Editorial Board

Patron : Mr. Michael S. Potter
        Chief Executive Officer

Editor in Chief : Prof. Dr. Md. Shahid Karim

Executive Editors : Prof. Dr. Anisur Rahman
                   Dr. Chandra Prakash Dokwal
                   Dr. Mohammed Naseem Siddiq
                   Prof. Dr. Tareak Al Nasir

Business Editor : Dr. Shagufa Anwar

Correspondent Editor : Dr. Simeen M Akhtar
Correspondent Executive : Mr. Iqbal Hossain Howlader

Editorial Board (alphabetical) : Dr. Abu Sayeed Mohammad Iqbal
                                Dr. Abdul Mannan Sarker
                                Dr. Alim Akhter Bhuian
                                Dr. A. K. M. Fazlul Haque
                                Dr. A. K. Gupta
                                Dr. Azizul Hasan
                                Dr. Borhan Uddin Ahmed
                                Prof. Dr. Jahangir Alam
                                Dr. Jasmin Manzoor
                                Prof. Dr. Kazi Mesbahuddin Iqbal
                                Dr. Krishna Mohan Sahu
                                Dr. Lutful Latif Chowdhury
                                Dr. M. Quamrul Hassan
                                Prof. Dr. Mathew J. Chandy
                                Prof. Dr. Md. Shahab Uddin Talukder
                                Prof. Dr. Md. Zilur Rahman
                                Dr. Monowara Begum
                                Dr. Mohiuddin Araf
                                Dr. Mrinal Kumar Sarker
                                Dr. Nandkumar Katakhand
                                Dr. Prabhat Dutta
                                Dr. Sandeep Attawar G.
                                Dr. Sheeba Khan
                                Dr. Wahed Zaman

Office of the Editor : Department of Paediatric Surgery & Paediatric Urology
OPD Level 5; Apollo Hospitals Dhaka
Phone: 880-2-8401661; Ext. 2525
Contents

Volume 4  Number 1  January 2010

Editorial

Original Article

- Morbidities of preterm VLBW neonates and the bacteriological profile of sepsis cases
  Dr. Md. Mahbubul Hoque, Dr. ASM Nawshad Uddin Ahmed, Dr. Swapan Kumar Halder, Dr. Md. Faizul Haque Khan, Dr. M A K Azad Chowdhury

- Study of blood lead and semen lead concentrations in male infertility
  Dr. Bidhan Chandra Debnath, Dr. Mohammad Ibrahim, Dr. Parvin Fatima

- Avulsion fracture of tibial insertion of PCL—operative management and outcome
  Dr. Md. Muztaba Ali, Dr. Prashant Agrawal, Dr. Nandkumar Katakhond, Dr. Mohammad Arshadullah, Dr. Abu Mohammad Sayem, Dr. A.T.M. Zulfiquar Rahman

Review Article

- Pulmonary Manifestations of Collagen Vascular Diseases
  Dr. Chandra Prakash Dokwal

Case Report

- Regression of plaque burden after primary percutaneous coronary intervention (PCI) in a patient with TVD: 4 years follow-up
  Dr. AHM Waliul Islam, Dr. Shams Munwar, Prof. (Dr.) Md. Shahab Uddin Talukder, Dr. Syed Sakib Nazir

- Primary malignant CNS lymphoma in an immunocompetent patient
  Dr. Ahmad Khaled, Dr. S A Salauddin, Dr. SMQ Zaman, Prof. (Dr.) Tareak Al Nasir

- Sarcomatoid carcinoma of the urinary bladder
  Dr. Ashraf Uddin Malik, Dr. Md. Ziaur Rahman, Dr. Mohammad Mahbub Rashid Sarker

- Fibrosarcoma of the chest wall
  Major (Retd.) Dr. Sheikh Firoj Kabir

- Transcatheter technique now standard for secundum ASD closure
  Dr. A.Q.M Reza, Dr. Atique Bin Siddique, Prof. (Dr.) Shahab Uddin Talukder, Dr. Shams Munwar, Dr. AHM Waliul Islam, Dr. Dr. Shaifur Rahman Shohel, Dr. Md Abdul Ghani

- A Fibrinogenemia
  Dr. Tahera Naznin, Dr. Pinkoo Attawar, Prof. (Dr.) Md. Moniruzzaman, Major (Retd.) Dr. Ikramul Islam

- Bilateral renal angiomyolipoma not associated with tuberous sclerosis
  Dr. Ahmad Khaled, Dr. Naznin Uddin Md. Arif, Dr. Wahed Zaman, Prof. (Dr.) Tareak Al Nasir

- Astroblastoma
  Dr. Ahmad Khaled, Dr. Md. Aliuzzaman Joarder, Prof. (Dr.) Mathew J. Chandy, Prof. (Dr.) Tareak Al-Nasir

- Fibrolipomatous hamartoma of the digital branches of the median nerve
  Dr. Md. Mozaffor Hossain, Dr. Md. Mahfuzul Momen, Dr. A. K. M. Fazul Haque

- Dysembryoplastic neuroepithelial tumour
  Dr. Ahmad Khaled, Dr. Md. Aliuzzaman Joarder, Prof. (Dr.) Mathew J. Chandy, Prof. (Dr.) Tareak Al-Nasir

Apollo Hospitals Dhaka

- CME (July 09 to Dec 09)
- Instruction to Authors
- Message from CEO
The very word "digital" hospital brings images of an impersonal facility, where patients may think they will consult a machine or a robot. But the reality is just the opposite. Digital hospitals - where information, images and clinical know-how all exist and are all connected wirelessly, where the interpersonal interactions and quality of care are much superior. It is the hospital of the future.

The skyrocketing costs, reports of medical errors, consumers demand for better care, and the ever-growing need to move away from paper, files, charts are all slowly leading to a restructuring of the health system. The shifting from paper to paper less, from wire to wire less, from big shelf full of record rooms to space saving electronic recording has already begun in many hospitals in USA & Europe1 and to a lesser extent even in Bangladesh. By the end of 2010, the European Commission predicts that five percent of national health budgets will be invested in e-health systems and services.2

In digital hospitals expensive, hard-to-share and easily lost traditional films (X-rays, for example) are replaced with digital images. It means moving from scribbled notes and paperwork to wireless networked software that accurately registers and quickly transmits patient records. Everything is linked into the system, from the automated pharmacy to the X-ray room, to laboratory to doctors office eliminating the need for most faxes, phone calls, and other administrative hassles. According to a 2005 study by PriceWaterhouse Coopers approximately 90 percent of the more than 30 billion healthcare communications that occur in the US each year are currently by fax, paper, mail or phone.

With a few clicks of index finger, doctors and nurses will have immediate access to high-resolution digital images, laboratory results and medication histories and an automated system of warning of medicine interactions and abuses or overuses through wireless networking system. Doctors moving around the hospital will receive new data about their patients. Trauma calls, alarms and ordinary messages - all go via the wireless IP network to a single handheld device. Doctors can access updated health records instantaneously and make more informed decisions about diagnoses and treatment, it saves time, money and most importantly, lives.

Internet access will be available in each patient bed. The hospital’s Nurse Call system will operate through the patient’s bedside terminal and connects the patient to a call handler. The system is built with a predetermined order of call routing, sending the call to a preassigned nurse. If the nurse does not respond within a certain time, the call is routed to an alternative nurse and onwards until the call has been responded to. For the patient, it leads to more responsive care from the right person at the right time.

One of the barriers may be reluctance on the part of doctors, nurses and others to use computers, to type rather than write, to look at monitors rather than reading in black and white. The other may be concerns about the security of computer systems. Wireless networks use shared radio frequencies to move data, so security concerns about this method of information transmission have always been high. Hospitals have already made patient records available to doctors and patients via the Internet on the HealthSouth website and hasn't had any security or privacy problems.

Technology suppliers must not only be state of the art, but state of the future art. The hospital ICT infrastructure is going to be tuned to unlock the benefits of modern healthcare communication for the coming decades.

Finally, I would like to emphasize that to be a top hospital it may not be sufficient to recruit top doctors, nurses, administrators and other personnel, the hospital will have to be also top in technology use as well. I would end the editorial with a quote from Swaid N. Swaid, a neurosurgeon working as a consultant to HealthSouth, “To marry technology with medicine is exciting...... it's going to be a tremendous way to provide patient care that is superior to anything we have seen.”

Prof Md Shahid Karim
Paediatric Surgery and Paediatric Urology
Editor in Chief
PULSE
Apollo Hospitals Dhaka

Reference:
Morbidities of preterm VLBW neonates and the bacteriological profile of sepsis cases

M M Hoque1, A S M N U Ahmed2, S K Halder3, M F H Khan4, M A K A Chowdhury5

Abstract

Background: Preterm very low birth weight babies are at increased risk of perinatal, neonatal and postnatal mortality and morbidity, mainly due to infections and complications of prematurity. Mortality of VLBW neonates is 30 times more than that of normal weight. Outcomes of such infants have been reported extensively from developed countries, but less is known from developing countries like Bangladesh though prematurity is very common.

Objective: To determine the morbidities associated with preterm VLBW neonates with particular emphasis on sepsis.

Methods: A prospective cohort study was done in Special Care Baby Unit of a tertiary care teaching hospital from July 2009 to December 2009. Preterm VLBW neonates admitted within 7 days of age comprised the cohort for the study. Detailed physical findings and information on pregnancy, delivery and immediate postnatal period were recorded on enrolment. Sepsis workup was done whenever sepsis was suspected clinically. Daily follow-up was given till discharge/death and relevant clinical findings recorded. Data were analyzed using SPSS 12.

Results: A total of 738 neonates were admitted during study period, 92 were preterm VLBW and enrolled in the cohort. Fifty-two (56.5%) of the enrolled cases were male and 40 (43.5%) female, male female ratio of 1.3:1. Mean gestational age was 30.8±2.4 weeks and mean birth weight 1,320±133 grams. Eight cases (8.7%) had features of septicaemia on admission and 49 (53.2%) neonates subsequently developed nosocomial infections. Other morbidities were jaundice (34.8%), RDS (8.7%), NEC (4.3%), TTN (3.3%), IVH (2.2%) and PDA (1.1%). Blood culture was positive in 29.8% among 57 suspected sepsis cases; Acinetobacter (41.2%) was the most common organism, followed by Klebsiella pneumoniae (23.5%), Escherichia coli (23.5%) and Pseudomonas sp. (11.8%). Sixty (62.5%) preterm VLBW neonates were improved and discharged, 3 (3.3%) cases were discharged on risk bond and 29 (31.5%) died.

Conclusions: Preterm VLBW neonates are at increased risk of morbidity and mortality. Septicaemia is the most common and devastating morbidity, most infections are hospital acquired. Therefore strict protocol for asepsis in neonatal units must be adhered to when handling these high risk infants.

Key words: Preterm, Very low birth weight, Septicaemia, Morbidity, Mortality.

Introduction: Between 1992 and 2008, globally under-five mortality rate have declined by two thirds, falling from 85 to 28 deaths per 1,000 live births; neonatal mortality dropped by one half during this period. In an article entitled ‘Why are 4 million newborn babies dying each year?’ Lawn et al. recognized that the greater part of the impressive reduction in infant mortality observed throughout the twentieth century was the result of reductions in post-neonatal mortality, neonatal mortality rates remained static. Among the neonates, very low birth weight (VLBW) preterm babies are at increased risk of perinatal, neonatal and postnatal mortality and morbidity, mainly due to infections and complications of prematurity. Mortality of VLBW neonates is 30 times more than that of newborns of normal weight. Despite constituting a small portion of LBW newborns, VLBW infants have a large impact on both neonatal mortality and morbidity. They demand high technology health care delivery and consume a great amount of resources.

Although outcomes of VLBW infants have been reported extensively from industrialized countries, less is known from the developing countries. Its prevalence is directly correlated with the developmental state of a country and is associated with poverty. Bangladesh is a developing country and an estimated 22% of babies are born prematurely and have low birth weight (LBW), but no such data is available for VLBW cases. Like any other part of the developing world, in Bangladesh also there has been a substantial reduction in under-five and infant mortality, but still having high neonatal mortality rates. In Bangladesh, survey in 2007 reported that the neonatal mortality (37/1,000 live births) accounts for about two-thirds of infant deaths (52/1,000 live births) and about half of deaths among children aged under 5 years (65/1,000 live births). The Bangladesh Demographic and Health Surveys indicate that the neonatal mortality rate declined in the early 1990s, but remained at 41-42 between 1995-1999 and 1999-2003. Preterm VLBW babies are especially vulnerable because of immaturity of structures and functions of various systems. Well thermal control, monitoring of heart rate and respiratory rate, oxygen therapy, maintenance of fluid and electrolyte balance, special attention to nutritional support and safeguard against infection are the corner stone of management of these neonates. The use of intensive care facilities has been successful in decreasing the mortality rates of premature infants. However, with increasing survival, an increase in morbidities is encountered. Preterm VLBW neonates are vulnerable to develop respiratory distress syndrome (RDS), septicaemia, necrotizing enterocolitis (NEC), patent ductus arteriosus (PDA), intraventricular haemorrhage (IVH), and long-term sequelae such as threshold retinopathy of prematurity (ROP), chronic lung disease (CLD) and...
Morbidities of preterm developmental disabilities. Among the morbidities, sepsis is the most common in developing countries compared to the developed world. Smaller infants with lower gestational ages are also prone to invasive clinical interventions which result in higher incidence of nosocomial infections. Gram negative bacteria are mostly responsible for sepsis in this age group in developing countries. However, the organism pattern is different in developed and developing countries, and within a geographical location also may change with time.

The purpose of the present study was to report the morbidities associated with preterm VLBW infants with particular emphasis on sepsis. Identification of bacterial etiology of sepsis in a tertiary care hospital will enable the physicians to treat these babies more rationally and thereby to reduce the morbidity and mortality of VLBW babies.

Materials and methods:
Study site: This prospective cohort study was conducted in the Special Care Baby Unit (SCABU) of Dhaka Shishu Hospital (DSH) from 1st July 2009 to 30th December 2009. DSH is the largest pediatric hospital in Bangladesh for primary and tertiary care. The SCABU has 30 neonatal beds and operates at full capacity at nearly all times.

Study Population: Newborn infants weighing <1,500 grams, gestational age<35 weeks and age <7 days, admitted during this period in SCABU were eligible for enrolment. Weight was measured on admission using a baby scale of 50 gm variation. Gestational age was determined on the basis of maternal dates (time from the first day of the last menstrual period) and further confirmed by New Ballard score.

Patient evaluation: On admission, detailed history was taken about pregnancy, delivery and immediate postnatal period for each subject by interviewing the parents and thorough clinical examination was conducted by one of the investigators and findings were recorded in a structured questionnaire. The parents of the neonates were explained about the study and then witnessed verbal consent was taken. After enrolment, all neonates with suspected septicaemia underwent the following diagnostic procedures: complete blood count (CBC), C - reactive protein (CRP) and blood culture. To diagnose other morbidities, CSF study (cytology, biochemistry and culture), serum bilirubin, x-ray chest, ultrasonography of brain, echocardiography and other relevant investigations were done as indicated. All neonates included into the cohort were closely followed during their hospital stay for clinical signs of infection. Sepsis workup was done whenever sepsis was suspected clinically. A patient was leveled as having septicaemia when, any two of the following signs and symptoms were present: lethargy, fever, hypothermia, recurrent apnea, bradycardia, abdominal distension, bleeding per rectal or gastric aspirate; plus any two of the following investigations were positive: blood culture, leukocytosis (total leukocyte count >20,000/cu mm of blood), leucopenia (total leukocyte count <5,000/cu mm of blood), thrombocytopenia (platelet count <100,000/cu mm of blood), positive C-reacting protein (>10 mg/L).

Patient management: All babies were managed by the unit consultant with their own protocol. Other than antibiotic therapy for sepsis cases, supportive therapy such as incubator care, correction of acidosis, maintenance of fluid and electrolyte balance, ventilatory assistance, phototherapy and blood transfusion was given as required. Investigators followed up all neonates daily till their discharge/death and recorded relevant clinical findings. The day of development of septicaemia was recorded.

Statistical analysis: The data were subjected to statistical analysis according to standard procedure. SPSS version 12.0 for Windows (SPSS Inc, Chicago, IL, USA) software was used for data recording and analysis.

Results: A total of 738 neonates were admitted in the SCBU during 1st July to 30th December 2009, 92 among them were preterm very low birth weight babies and enrolled in this study. Among the VLBW neonates, 60 (65%) were admitted within 24 hours of birth and rest admitted between 24 hours and 7 days of birth. Fifty-two (56.5%) of the enrolled cases were male and 40 (43.5%) were female, male female ratio of 1.3:1. Their mean gestational age was 30.8±2.4 weeks (ranged from 27-34 weeks) and mean birth weight of 1,320±133 grams (ranged from 1,030-1,500 grams).

Out of 92 cases, 51 (55.4%) VLBW babies were admitted only due to prematurity, rest 41 (44.6%) had other complications along with prematurity, like suspected RDS (20.7%), perinatal asphyxia (10.9%), sepsis (8.7%) and jaundice (4.3%).

Table I: Causes of admission of the VLBW neonates (n=92)

<table>
<thead>
<tr>
<th>Diseases on admission</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suspected RDS</td>
<td>19</td>
<td>20.7</td>
</tr>
<tr>
<td>Perinatal asphyxia</td>
<td>10</td>
<td>10.9</td>
</tr>
<tr>
<td>Neonatal sepsis</td>
<td>8</td>
<td>8.7</td>
</tr>
<tr>
<td>Neonatal jaundice</td>
<td>4</td>
<td>4.3</td>
</tr>
<tr>
<td>Prematurity only</td>
<td>51</td>
<td>55.4</td>
</tr>
<tr>
<td>Total</td>
<td>92</td>
<td>100</td>
</tr>
</tbody>
</table>
Morbidity of preterm

Other than 8 cases (8.7%) that had features of septicaemia on admission, 49 (53.2%) neonates subsequently developed septicaemia during hospital stay (nosocomial infections).

So incidence of septicaemia in preterm VLBW neonates in this study was 62%. In addition to increased incidence of septicaemia among the enrolled VLBW neonates, there were other morbidities including jaundice (34.8%), RDS (8.7%), NEC (4.3%), TTN (3.3%), IVH (2.2%) and PDA (1.1%).
Morbidities of preterm

The above complications were present alone or in combination with septicaemia.
Among the laboratory investigations, CRP was positive in 86% cases and thrombocytopenia was found in 80.7% cases. Leukopenia (52.6%) was more common than leukocytosis (8.8%).

<table>
<thead>
<tr>
<th>Name of investigations</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood culture positive</td>
<td>17</td>
<td>29.8</td>
</tr>
<tr>
<td>Leukocytosis (TLC &gt;20,000)</td>
<td>5</td>
<td>8.8</td>
</tr>
<tr>
<td>Leukopenia (TLC &lt;5,000)</td>
<td>30</td>
<td>52.6</td>
</tr>
<tr>
<td>Thrombocytopenia (Platelets &lt;100,000)</td>
<td>46</td>
<td>80.7</td>
</tr>
<tr>
<td>CRP positive (≥10mg/L)</td>
<td>49</td>
<td>86.0</td>
</tr>
</tbody>
</table>

Sixty (65.2%) preterm very low birth weight neonates were improved and discharged, 3 (3.3%) patients were discharged on risk bond and 29 (31.5%) died.

**Discussion:** A total 92 patients were studied during the study period. Sixty (65.2%) neonates were admitted within 24 hours and 32 (34.8%) were admitted after 24 hours up to 7 days of age. There is no obstetric unit attached to this hospital and cases are referred from different centers, so babies coming from a distance are admitted late. Fifty-seven percent were male and 43% female with male female ratio of 1.3:1. Male babies are more likely than females to develop septicaemia and this male preponderance is similar to other studies. Males are cared for more in our society, this may also be a cause of male preponderance.

Septicaemia occurred in 57 (62%) cases of VLBW neonates in this series. Very low birth weight infants develop 2.7 times more sepsis than other infants since their immune system and skin barrier are immature and they are exposed to many invasive diagnostic and therapeutic procedures but it was much higher than other similar studies. Banu et al. found septicaemia in 16.4% of preterm VLBW neonates. Nagar et al. in their study of small for gestational age babies found the incidence of neonatal infection is about 26.2%. In a multi center cohort study done by Stoll et al. showed that the incidence of culture positive early onset neonatal sepsis was 1.9% though 50% of the cohort had clinical sepsis and 25% had culture positive late onset neonatal sepsis.

Majority (49/57, 86%) of the septicaemia were hospital acquired. Nosocomial infections are a common problem in neonatal wards and an important cause of mortality in developing countries. The reported incidence of nosocomial sepsis in neonates from India ranges from 15.4% to 37%. Risk factors for nosocomial sepsis include preterm very low birth weight neonates, overcrowding in nurseries, assisted ventilation and use of peripheral venous and umbilical vascular catheters. Bed capacity of the SCABU at DSH has been increased from 20 to 30 without improvement of manpower specially nurses and cleaning staffs and other auxiliary supports. In our series we studied only the preterm VLBW babies and overcrowding at the nursery was responsible for this high nosocomial sepsis.

Among the 57 neonates diagnosed as septicaemia, 17 (29.8%) had positive blood culture. Culture positivity rate was little lower than another of Ahmed ASMNU et al. in 1998 at the same SCABU, that showed 35% culture positive among the suspected septicaemia cases. Like other studies from developing countries, gram negative organisms were predominant in this study. Acinetobacter (41.2%) was the most common organism found followed by Klebsiella pneumoniae (23.5%) and Escherichia coli (23.5%), Pseudomonas sp. was found in 2 (11.8%) cases.

In this series the mean age of development of septicaemia was 6.1±2.4 days indicating late-onset septicaemia was more common than early-onset disease, in contrast to other reports in which early-onset septicaemia generally has been more
Morbidity of preterm

common.27 Predominantly nosocomial origin of infection was the reason for late-onset sepsis. The mean duration of hospital stay in neonates with septicaemia was 13.57±5.39 days compared to 10.88±4.02 days neonates without septicaemia. It demonstrates that septicaemia increases duration of hospital stay, increase workload and morbidity of VLBW neonates.

Other than septicaemia, common morbidities in this series were jaundice, RDS, NEC, IVH and PDA in 34.8%, 8.7%, 4.3%, 2.2% and 1.1% cases respectively. Jaundice was mostly physiological, aggravated by sepsis. Several studies have reported almost similar findings.14,28 Almost one-third (31.5%) of the enrolled cases in this study died. Mortality is high in preterm VLBW neonates and this finding is comparable to other studies.13,29

Conclusions: Preterm VLBW neonates are at risk for several morbidities, of which septicaemia is the most common and devastating morbidity leading to high mortality. Most cases of septicaemia are hospital acquired. Therefore to keep the infection rates low, strict protocol for asepsis in neonatal units must be adhered to when handling these high risk infants. There is a need for continuous neonatal infection surveillance to generate information on neonatal nosocomial infection rates and risk factors in our setting, so that appropriate preventive strategies can be adopted.

References:
Study of blood lead and semen lead concentrations in male infertility

B C Debnath1, M Ibrahim2, P Fatima1

Abstract
Objective: To evaluate the association of blood and semen lead with male infertility.

Design: It was a case control study carried out in the department of Biochemistry, BSMMU, from January, 2004 to December, 2004.

Subjects: Of 52 male subjects, 26 were infertile with oligospermia and / or asthenospermia. Age matched 26 normospermic subjects with normal standard semen parameters (volume, count, motility and morphology) were selected as controls. None had the history of occupational exposure to lead.

Results: Median values of blood lead were 19.96 µg/dl and 17.68 µg/dl (normal <25 µg/dl); semen lead were 29.56µg/dl and 28.17 µg/dl (normal 15.37 ± 3.92 µg/dl) in cases and controls respectively. Neither blood lead nor semen lead showed significant difference between cases and controls. There was significant positive correlation between blood lead and semen lead concentrations. There was no significant correlation of semen and blood lead concentrations with other semen parameters & serum hormone concentrations (FSH, LH, & Testosterone) in cases.

Conclusion: Though the association of blood or semen lead with male infertility was not established in our study, yet, increased seminal lead concentration found in our study subjects may be indicative of subtle exposure of lead in our environment. So, further study with larger sample size is needed. The clinicians should consider the lead measurements when evaluating male partners of couples with unexplained infertility and for assessment of lead status, semen lead measurement can be recommended.

Key words: Male infertility, blood lead, semen lead.

Introduction: Lead poisoning is one of the important occupational and environmental diseases in the world. Despite its recognized hazards, lead continues to have widespread commercial application. Though the incidence of serious overt lead poisoning is decreased due to diminished use of lead in gasoline and public health measures, there remains considerable concern over the effects of low level lead exposure. Lead may have adverse effects - at lead concentrations once considered "normal" or "safe". Lead serves no useful purpose in the human body. Lead exerts multi-systemic toxic effects by inhibiting enzyme activity, (sometimes as a consequence of binding to sulfhydryl groups), interfering with the action of essential cations, (particularly Ca, Iron and Zinc) and by altering the structure of cell membranes and receptors. Lead petrol, lead pipes in household water supplies, newspaper, Xerox copies, cigarette smoking are sources of lead exposure. Battery repair, radiators repair, soldering, painting and printing are occupations prone to get lead poisoning. It is not biodegradable. Lead is a cumulative poison and accumulated in tissues over years. There is no 'safe' level in blood, although about 10 µg/dl can be tolerated. >10 µg/dl in children & >25 µg/dl in adults may cause toxic manifestations. Pollution level of lead in Bangladesh are among the world's highest during dry season, falling during periods of medium and heavy rainfall & the current blood level of lead concentration for adults in Dhaka city is about 50 µg/dl. Lead is a notorious reproductive toxin. Blood lead > 40 µg/dl have been associated with diminished or aberrant sperm production. Reports of declining human sperm concentration and male fertility have renewed interest about the role of environmental exposures in the etiology of human male infertility. The role of lead in male factor sub fertility is of particular current interest. WHO estimates that there are 60-80 million infertile couple worldwide. About 40-50% of infertility is totally or in part due to a male factor. Unexplained infertility varies from 10 - 20%. Evaluation of male partner of infertile couples is to identify - potentially correctable conditions and treatment of which may improve the male's fertility. In order to fertilize an egg, a sperm has to bind with it. A sugar called mannose on the outer coating of the egg is crucial to binding. Mannose receptors located on the head of human sperm recognize the mannose on the coating of the egg and regulate the binding process. Then the sperm has to penetrate the egg. Successful binding induces an event called mannose induced acrosome reaction (MIAR) that help to release digestive enzymes from the sperm to ease its passage through the egg coating to its nucleus? To study the effects of lead upon the human male reproductive tract, a systematic analysis of lead levels & correlation of it with other trace metals, semen parameters, sperm fertilization potential and biomarkers of human sperm function in men without occupational exposure to lead was done. Analysis of lead in seminal plasma as a biomarker to predict IVF success was performed examining parameters that could affect IVF outcome. The mean lead value was significantly higher. A significant negative relationship was detected between semen lead levels and the fertilization rate in IVF. Threshold
value for semen lead was analyzed to be 42.29 µg/dl. Threshold values for blood lead, >40 µg/dl/ above which the semen parameters are affected. Above this threshold value for the lead in semen, less than 63% of oocytes could be predicted to be fertilized. Below this threshold value of lead in semen more than 63% of oocytes could be predicted to be fertilized by IVF. So, simple measurement of semen lead level might be a useful addition in the evaluation of the male partner prior to an IVF attempt. Blood lead levels determined by the National Institute for Occupational safety and Health (1994) protocol, > 40.0 µg/dl require medical intervention. However, studies by several groups now suggest that lead intake levels even below this threshold are associated with a variety of adverse health effects in somatic tissues. Ascorbic acid supplementation 1000 mg daily for 4 weeks may significantly reduce lead toxicity. Calcium deficiency potentially may result in elevated lead accumulation. Maintenance of adequate dietary calcium levels appeared extremely important toward minimizing susceptibility to lead toxicity. An alteration of Zinc and Copper homoeostasis is done by lead poisoning and Zinc and Copper supplementation may decrease the elevated lead levels.

In Bangladesh no work has been done on lead status of general population of the country outside Dhaka city. Moreover no work has yet been done on infertile males of unexplained infertility. In this study, whole blood and semen lead concentrations were measured and correlated with fertility status. In our country IVF is now becoming popular for infertile couples but it is of huge cost with many unexplained failures. It is of great concern to find out the possible causes of the failures including the role of lead exposure. In this context, this study might be helpful for lead status evaluation and to find out its association with male infertility. Therapeutic intervention may be done by clinicians to decrease lead level and preventive measures may be taken nationally to reduce or eliminate even the low level lead exposure. Above all, by treating this reversible, treatable cause for infertility, some persons may procreate in natural ways without undergoing huge costly attempts of artificial methods of high technology.

Materials and methods

This case control study was carried out in the department of Biochemistry of Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbag, Dhaka, from January 2004 to December 2004. Informed written consent was taken from each study subject. 52 subjects were selected from the Centre for Assisted Reproduction, Shamoli, Dhaka. The subjects were male partners of infertile couples where there was no abnormality in female partners that may affect fertility. The subjects were divided into two groups: Group-I (Cases): 26 males with oligospermia and or asthenospermia, Group-II (Controls): 26 healthy age matched males with normospermia. Semen was collected in a previously treated metal free wide mouthed plastic container following three days abstinence. Semen analysis was done within 60 minutes after liquefaction of the semen. Blood samples of the subjects were collected (with EDTA for lead, without anticoagulant for hormone measurement). The serum was taken into micro-centrifuge tube. The serum and whole blood with anticoagulant were preserved at -35°C. The clean supernatant of seminal fluid (after centrifugation) was taken into cryo-tubes and preserved at -70°C till analysis was completed. Blood and semen lead concentration was measured by Graphite furnace atomic absorption spectrophotometry. Serum FSH & LH were measured by-Abbott AxSYM system, based on MEIA principle and Serum Testosterone by ELISA. Semen analysis was done by ordinary light microscope using Makler chamber. All plastic wares including test tubes, semen collection cups, micro-centrifuge tubes, containers and pipettes were cleaned and made metal free, following a standard protocol. All statistical analysis was done by SPSS software package, 10.0 for windows. Values were presented as mean ± SD and also as median. 95% confidence limit was taken as level of significance. Mann Whitney U test was done to see the significant difference of blood and semen lead between the case and controls and spearman’s correlation test was done to see any significant correlation between the relevant values.

Results: Grouping of study subjects was shown in Table-I. No significant difference of age distribution was found between two groups. Table-II shows the serum FSH, LH & Testosterone concentrations in cases and controls. No significant difference of serum hormones between cases and controls was found. Whole blood lead & semen lead concentrations were compared between cases and controls by Mann-Whitney U test but no significant difference was observed (Table-III). Spearman’s correlation test was done between whole blood lead concentration and semen lead concentrations in cases & significant positive correlation was found (P<0.05) (Table-I) & (Fig-1).

<table>
<thead>
<tr>
<th>Group</th>
<th>Number</th>
<th>Range</th>
<th>Age(years) (mean±SD)</th>
<th>t-value</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group I-cases (Oligospermic&amp;/or Asthenospermic)</td>
<td>26</td>
<td>27-45</td>
<td>37.04± 4.57</td>
<td>0.643</td>
<td>&gt;0.05</td>
</tr>
<tr>
<td>Group II-controls (Normospermic)</td>
<td>26</td>
<td>26-45</td>
<td>37.92± 5.31</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table I: Grouping of study subjects and age distribution
The aim of this case-control study was to measure the lead level in Blood and Semen of infertile Bangladeshi males and to correlate these values with other semen parameters like semen volume, sperm count, motility and morphology to see any effect of lead on fertility.

The strategy of the study was to examine the interaction between two main variables: first, the degree of environmental exposure of lead & second, the fertility status. Chemical biomarkers of exposure (lead in blood and semen) were correlated with endocrinologic /biologic biomarkers (total testosterone, LH & FSH) and semen parameters were correlated with other semen parameters (semen volume, total count, motility and morphology of sperms) were performed. No significant correlation was found. Spearman's correlation of blood and semen lead concentrations with serum FSH, LH & Testosterone concentrations were done, but no significant correlation was observed.

Table II: Comparison of Serum FSH, LH and Testosterone between cases and controls

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group I (Case) n=26</th>
<th>Group II (Control) n=26</th>
<th>t-value</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>FSH (mIU/mL)</td>
<td>5.24±1.38</td>
<td>5.35±1.43</td>
<td>0.108</td>
<td>&gt;0.05</td>
</tr>
<tr>
<td>LH (mIU/mL)</td>
<td>5.86±1.84</td>
<td>6.62±2.09</td>
<td>1.083</td>
<td>&gt;0.05</td>
</tr>
<tr>
<td>Testosterone (ng/mL)</td>
<td>4.65±1.44</td>
<td>5.40±1.92</td>
<td>1.450</td>
<td>&gt;0.05</td>
</tr>
</tbody>
</table>

Table III: Comparison of lead concentration between cases and controls

<table>
<thead>
<tr>
<th>Variable (Lead) (median)</th>
<th>Group I (Case) n=26</th>
<th>Group II (Control) n=26</th>
<th>Mann-Whitney U</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood (µg/dl)</td>
<td>19.96</td>
<td>17.68</td>
<td>271.00</td>
<td>&gt;0.08</td>
</tr>
<tr>
<td>Semen (µg/dl)</td>
<td>29.56</td>
<td>28.17</td>
<td>331.00</td>
<td>&gt;0.05</td>
</tr>
</tbody>
</table>

Discussion: The aim of this case-control study was to measure the lead level in Blood and Semen of infertile Bangladeshi males and to correlate these values with other semen parameters like semen volume, sperm count, motility and morphology to see any effect of lead on fertility.

The strategy of the study was to examine the interaction between two main variables: first, the degree of environmental exposure of lead & second, the fertility status. Blood lead level was used as a biologic index for recent exposure and semen lead level was used as an indicator for the direct exposure of reproductive tissue. Subtle changes in fertility status were evaluated through an assessment of semen parameters and reproductive hormones. Thus, chemical biomarkers of exposure (lead in blood and semen) were correlated with endocrinologic /biologic biomarkers (total testosterone, LH & FSH) and semen parameters were correlated with other semen parameters (semen volume, total count, motility and morphology of sperms) were performed. No significant correlation was found. Spearman's correlation of blood and semen lead concentrations with serum FSH, LH & Testosterone concentrations were done, but no significant correlation was observed.
Study of blood

(volume, count, motility and abnormal forms). The median values of blood lead and semen lead concentrations were slightly higher in oligospermic population (cases) than in the normospermic (controls) but the difference was not significant. This finding was similar to El-Zohairy et al (1996). No difference of lead between cases and controls may be due to the fact that both were exposed to the same environment, none was occupationally exposed to lead. Serum FSH, LH & Testosterone concentrations in cases and controls showed no significant difference (within normal reference ranges). The phenomenon of this normal hormone concentration in cases is very much implied because of the fact that 90% of sub fertile males are due to non-endocrine abnormalities (Bhatla 2001). Significant positive correlation was observed between the lead concentration of whole blood and semen in cases. This finding is similar to Aribarg & Sukcharoen (1996) and Benoff et al (2003). This might be due to any mechanism operating to transfer lead from blood to the male reproductive tract readily. This finding suggests that seminal lead concentration would be a useful biomarker for evaluation of male infertility.

Blood and semen lead concentrations showed no significant correlation with semen parameters. This is in contrast to Benoff et al (2003) who found weak negative correlations between seminal lead levels and semen parameters and stronger negative correlation between seminal lead values and sperm function tests like - manrose receptor expression and manrose-stimulated acrosome loss. The probable reason of our finding may be due to the fact that threshold levels for semen and blood lead concentrations were not exceeded above which semen parameters could be affected. Moreover, we did not perform sperm function tests which could be more informative in relation to effect of lead on semen parameters. No significant correlation of blood and semen lead concentrations with serum hormones (FSH, LH & Testosterone) was observed. This is consistent to the finding of Benoff et al (2003) and Lancranjan et al (1975). This may be explained in light of low blood lead concentration of our population consequential of low environmental lead level that failed to affect hypothalamic pituitary testicular axis.

In our study, we could not prove the effect of environmental lead exposure on male infertility, yet we cannot deny the association of lead with male infertility because of some limitations of our study. Sample size was small. We used hot plate heating block for semen sample digestion. Use of microwave oven for semen sample digestion could improve detection of semen lead. Moreover, infertility in our study subjects might be due to some causes other than the lead exposure effect, like vitamins and trace elements deficiency and free radical assault etc. It is a matter of great concern to find out the causes of unexplained infertility. So, further broad based study with large sample size is needed to explore the association of lead status with male infertility in a more comprehensive and convincing manner.

References:
5. BIRPERHT. Assessment of reproductive health care needs and review of services provided at the Levels of Thana, Union and Village. Dhaka Bangladesh: Institute of Research for Promotion of Essential & Reproductive Health and Technologies; 1997.
Avulsion fracture of tibial insertion of PCL-operative management and outcome

M Ali, P Agrawal, N Katakdhond, M Arshadullah

Abstract
There are controversies about the management of the PCL injuries among the Orthopaedic surgeons. We present our result of 14 cases that underwent surgical management for the avulsion fracture of the tibial insertion of the PCL. Open reduction and internal fixation of the avulsion fracture of the tibial insertion of the PCL were done and results were analyzed. The study group consisted of 13 males and 1 female; the mean age was 27 years. The avulsion fractures of more than 3 weeks old were excluded. The result showed that there were no instability in any of the patients and each patient obtained pain free, full range of movement for the affected knee within a mean follow-up period of 11 months. Therefore it is recommended to take up surgical management for avulsed PCL if presented within 3 weeks of injury.

Introduction: PCL is the stronger of the two cruciate ligaments of the knee. It is the primary constraint to the posterior tibial translation at 90º of knee flexion. PCL avulsion injuries are not uncommon in our country. Though there are controversies, most authors have recommended operative management of a displaced bony avulsion of the tibial insertion of the PCL. Torisu, Trikey, Lee, Myers are sought to be giants in PCL surgery & their researches stated that excellent result can be achieved by fixation of the avulsed fragment. However the result of surgery is less satisfactory when performed beyond 11 weeks from the time of injury. To identify the results and gain further confidence about the surgical management of the avulsed PCL we have conducted this study and found that OR & IF (Open Reduction & Internal Fixation) can provide excellent outcome in the management of the avulsed PCL from tibial attachment. On the contrary without surgery there would have been early OA of the knee & sometime instability would cause significant difficulty in activities in daily living (ADL).

Material & Methods: In this series there were 14 cases. The average age of the patient was 27 years, extending from 19 to 35 years of age. Male female ratio was 13:1. In all the cases the injuries were less than 3 weeks old. In each case the injury was isolated and there was no history of surgery of the affected knee before. It is to be mentioned here that the fracture fragment has to be large enough to be fixed by an at least 3.5mm screw with washer. In this regards real image by digital x-ray can provide necessary information. Even though, in half of the cases where fracture fragment needed further evaluation, we did CT scan. From the history it was evident that the motor bike accidents were the main cause of the injury (78%), then dash board (14%) and also fall from rickshaw on a hard object in a flexed knee (7%).

Technique: Surgeries were performed under spinal anesthesia in 10 cases, and spinal - epidural anesthesia in 4 cases. In all cases tourniquet was applied. Diagnostic arthroscopy for all cases was done to identify any associated injuries of the knee. The patient was then shifted to prone position and the surgery was done through posterior approach of the knee.

Results: Between 2005 to 2009, 13 men and one woman aged from 19 to 35 years underwent OR & IF by 3.5 mm screw and washer after diagnostic arthroscopy. At 11 months of mean follow-up it has been seen that the tests for instability (posterior sag sign and posterior drawer test) were negative in 100% of cases, 98% achieved full ROM within 5 months post-op. It was also found that 91% patients were pain free by 11 months and equal percentage of patients were satisfied with the management.

Discussion: Treatment is indicated in patients when there is posterior instability on physical examination and a bony fragment is seen on x-ray. We have not done any MRI in this study as we know that the chance of intra-substance injury is a remote possibility in avulsion injury of any ligament. We performed the surgeries in acute phase of the injury.
Avulsion fracture

injury; and fixed the fragments with 3.5mm cannulated cancellous screws with washer. Though the surgeries were performed at acute phase, there were no arthrofibrosis, might be due to arthroscopic washout. These patients had been immobilized for 6 weeks by pop and another 6 weeks with knee braces. The early removal of the pop and application of the knee brace is to achieve early ROM and rehabilitation. Study shows there were no, or minimum instability following the procedure in this series and obtained solid bony union and functional capacity along with satisfaction. It is evident that the associated intra-substance injury if any has not affected the postoperative posterior instability of the knee.

Conclusion: In conclusion, open reduction and internal fixation for avulsed PCL from tibial attachment after diagnostic arthroscopy is a reliable and effective method of management for the avulsed PCL from tibial attachment. So, it should be practiced.

References:
Pulmonary Manifestations of Collagen Vascular Diseases

C. P. Dokwal

The collagen vascular diseases (CVDs) include a heterogeneous group of chronic inflammatory immunologically-mediated systemic diseases, such as rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), systemic sclerosis (SSc), Sjogren's syndrome (SS), polymyositis (PM)/dermatomyositis (DM), and mixed connective tissue disease (MCTD). They present with a wide range of clinical manifestations. The clinical features in CVDs frequently overlap causing much clinical confusion.

The lung is frequently affected in CVDs and is the cause of significant morbidity and mortality. The common pulmonary manifestations include pleural disease, pulmonary fibrosis, bronchiolitis obliterans, obliteration, organizing pneumonia, bronchiectasis, aspiration pneumonia, and diaphragmatic weakness. In the following paragraphs, the pulmonary manifestations in various collagen vascular diseases are discussed.

**SYSTEMIC LUPUS ERYTHEMATOSUS**

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease which shows a great variability in its presentation and course. Most patients have at least four of the American College of Rheumatology Criteria for SLE.1 Patients frequently present with systemic constitutional symptoms, such as fever, malaise, weight loss, fatigue, lymphadenopathy and hepatosplenomegaly, and these manifestations are seen throughout the course of SLE. Skin, musculoskeletal, lung, and renal systems are also commonly involved in SLE.

The pleuro-pulmonary involvement occurs in about 50-60% of patients, especially later in the course of the disease when other organs are already affected. Pleuritis and pleural effusion are the most common pulmonary manifestation, although, lung parenchymal disease, pulmonary vascular disease, upper airway involvement, and diaphragmatic dysfunction also occur.

**Pleuritis and pleural effusion**

The pleural effusion is seen in 50% of patients with SLE during the course of their illness, and is the presenting feature in 5-10%.2, 3 The autopsy series have revealed pleural involvement in up to two thirds of patients.4 Plural effusion is typically bilateral, often associated with pericarditis, and frequently recurs. Pleural fluid is exudative in nature with either lymphocytic or polymorphonuclear cell predominance. Small pleural effusion usually resolves spontaneously, but severe and persistent one requires drug therapy.

**Interstitial lung disease**

Interstitial lung disease (ILD) has been described in 4% of patients with long-standing SLE in the past.5 However, recent HRCT series revealed that about 1/3rd of patients with SLE have ILD, most having early sub clinical disease.6 It usually develops insidiously and is associated with recurrent pleural effusions.

Chest radiographs typically show diffuse alveolar opacities. HRCT of chest often reveals a cellular, fibrotic, or mixed non-specific interstitial pneumonia (NSIP) pattern.7 Usual interstitial pneumonia (UIP) and lymphoid pneumonia (LIP), particularly in those with associated secondary Sjogren's syndrome, have also been described.7, 8 Rarely, organizing pneumonia has also been reported.

**Diffuse Alveolar Haemorrhage**

Diffuse alveolar haemorrhage (DAH) is a rare complication seen in 4% of patients with SLE. Pulmonary capillaritis is often the underlying cause in most patients with DAH.9 It is often fatal in 50% of cases despite therapy, although mortality is declining with aggressive treatment with corticosteroids and cyclophosphamide.9, 10 Glomerulonephritis is also present in most patients with DAH.9 DAH should be differentiated from acute lupus pneumonitis which also presents similarly, however, significant systemic symptoms suggestive of a SLE flare are typically absent in DAH.11 DAH may also occur secondary to uraemia or thrombocytopenia.

Chest radiograph in DAH commonly reveals bilateral, often extensive, airspace consolidation. Histopathology examination of lung tissue reveals a non-specific alveolitis, although in some patients the infiltration of small arteries, arterioles, and capillaries by polymorphonuclear cells may be seen.

**Acute Lupus Pneumonitis**

Acute lupus pneumonitis (ALP) has been described in 1% to 12% of patients, and often has a serious outcome 2, 12, it often presents acutely with fever, cough, dyspnea, pleuritic pain, and hypoxia, and sometimes haemoptysis.13 Occasionally it may be the presenting feature of SLE.11 The chest roentgenogram typically shows bilateral areas of airspace consolidation particularly in lower lung zones, and is often associated with bilateral pleural effusions.

ALP is a diagnosis of exclusion. It should be differentiated from diffuse alveolar haemorrhage and infective pneumonias. Surgical biopsies often reveal a histology pattern consistent with diffuse alveolar damage with or without alveolar haemorrhage 14, 15 ALP requires therapy with high dosage of systemic corticosteroids, and in refractory cases with additional immunosuppressive therapy, such as cyclophosphamide.
Pulmonary Manifestations

**Pulmonary vascular disease**

Acute reversible hypoxemia is extremely rare, and has been described in acutely ill patients. It presents clinically as a sudden onset of hypoxemia with normal radiologic findings, and often responds rapidly to corticosteroids. Patients with this syndrome have diffusion abnormality which is believed to be due to occlusive vasculopathy as a result of neutrophil aggregation within pulmonary capillaries.17

The prevalence of pulmonary arterial hypertension (PAH) in SLE is about 14%, however, it increases with the progression of the disease. Raynaud's phenomenon is seen in 75% of SLE patients who also have PAH, compared with 20%-30% of those without clinically evident PAH. Although, 50% of SLE patients with PAH have been reported to have underlying vasculitis, it has been found much less common in recent biopsy studies. The prognosis of patients with vasculitis is poor.

Antiphospholipid antibodies are present in up to two thirds of patients with SLE, and it poses more than six fold risk of venous thrombo-embolism in this patient subgroup.22

**RHEUMATOID ARTHRITIS**

Rheumatoid arthritis (RA) is the most common autoimmune disease affecting 1-2% of general population. The involvement of heart, skin, lung, and eye is seen in 50% patients with RA. Pulmonary complications are common and often occur within 5 years of initial diagnosis of RA. They are also the presenting manifestations in 10% of patients. The commonest pulmonary manifestations are pleuritis and interstitial lung disease (ILD). Lung involvement is second to infection as the most common cause of death in patients with RA (18% vs 27%).

**Pleural disease**

It is seen clinically in about 3-5% of patients with RA, although post mortem studies reveal a prevalence of 40-80%. It is mainly seen in patients who have already developed extra-articular manifestations. Pleural thickening is more common than pleural effusions. Pleural effusions are usually unilateral and may be loculated. They commonly occur late in the course of the disease, and may be associated with pericarditis and subcutaneous nodules. They are usually asymptomatic, but tend to recur after aspiration. The Pleural fluid is exudative with high protein component; however its glucose content is characteristically very low. Occasionally, pleural fluid may appear chylos due to high cholesterol content. Small pleural effusions do not require any specific therapy, and usually resolve spontaneously, but large effusions require aspiration. The recurrent pleural effusion may be treated with corticosteroid or pleurodesis.

**Interstitial Lung Disease**

ILD is the commonest and most serious complication seen in 15-20% of patients with RA. The usual interstitial pneumonia (UIP) is the commonest histological pattern followed by non-specific interstitial pneumonia (NSIP). Cryptogenic organizing pneumonia (COP) is less frequent. ILD typically occurs a few years after the onset of joint symptoms, however, it may be the first manifestation in 15% to 20% of RA patients. Chest radiograph typically shows reticular or reticulo nodular opacities in the lower lung fields. HRCT shows a reticular pattern and irregular septal thickening, mainly in the peripheral and lower lung zones. Honeycombing is a late manifestation of ILD. Rarely, some patients present with progressive upper lobe fibrosis and cavitations.

**Pulmonary Nodules**

Pulmonary nodules occur in up to 1% of patients. They are usually associated with advanced disease and subcutaneous nodules. They may be single or multiple, varying from 1-3 cm in diameter. They tend to be subpleural, often cavitate leaving thick-walled cavities. Occasionally, such cavities may be thin walled, which may disappear gradually. Rarely, rheumatoid nodules are seen in patients without RA, although some patients have developed RA disease later. Pulmonary nodules are rare in coal worker's pneumoconiosis, however, they may cause progressive massive fibrosis in some patients.

**Bronchiolitis Obliterans/Bronchiolitis Obliterans Organizing Pneumonia (BOOP)**

Occasionally, a severe rapidly progressive form of airway obstruction develops in patients with RA who present with widespread inspiratory crackles and often a mid inspiratory squeak. Chest radiograph is usually normal, but may show over inflation. HRCT of chest may show a characteristic mosaic pattern of attenuation and perfusion due to redistribution of blood flow away from the areas of abnormal ventilation. In early cases, the end-expiratory HRCT scan may show air-trapping when end-inspiratory scan is normal. Lung function studies show a severe obstructive deficit with low transfer factor for carbon monoxide (TLC). The autopsy studies reveal a widespread fibrous obliteration of bronchioles. Corticosteroid is not effective and most patients die in 2-3 yrs, although some patients have survived up to a decade.

**Other Pulmonary Manifestations in RA**

Pulmonary infections are frequent in RA patients. Rarely, cosinophilic pneumonia, bronchocentric granulomatosis, pulmonary hypertension, and progressive loss of lung volume due to diaphragmatic dysfunction may also occur. Small airway disease has been described in up to 30% of patients. Bronchiectasis is also common. The involvement of crico-arytenoid joint of larynx may cause stridor, which in severe case may require tracheostomy.

**SYSTEMIC SCLEROSIS (SCLERODERMA)**

Systemic sclerosis (SSc) is a rare, chronic, multisystem disease of unknown cause that is characterized by fibrosis and
Pulmonary Manifestations

Vasculopathy of the skin and visceral organs. There are 2 forms of SSc, the limited SSc and the diffuse SSc disease. The limited SSc is seen in 70% of patients, and it manifests with subcutaneous calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia. It tends to involve the distal limbs and face and has a lower risk of internal-organ involvement. However, PAH is more common in limited SSc disease. About 70% of patients have circulating anticientromere antibodies. The diffuse disease often involves both distal and proximal limbs, the face, trunk and internal organs and has an increased associated mortality rate compared with the limited form. 38 Although patients without skin changes (systemic sclerosis sine scleroderma) do not meet established criteria for a diagnosis of systemic sclerosis, they may have many features of the disease. 29, 30

The lung is commonly involved in SSc. Most patients with SSC have pulmonary symptoms, such as exertional dyspnea and dry cough. The interstitial lung disease and pulmonary vascular disease are the most common pulmonary complications. Aspiration pneumonia is also frequent due to oesophageal dysfunction, however, pleural disease is relatively rare, and is usually accompanied by lung parenchymal involvement.

ILD is seen in 20-60% of patients. It can occur in both limited and diffuse SSc. It rarely precedes scleroderma. Chest radiograph typically shows a bibasilar fine reticular pattern in the initial stages which progresses to coarse reticulation and honeycombing. 31 NSIP is the most common histologic pattern, followed by UIP. 24, 32, 33

HRCT scan of chest is more sensitive in diagnosing early ILD. It typically reveals lower lobes and peripheral involvement with areas of ground-glass attenuation, reticular shadowing, and poorly defined sub pleural nodules. Honeycombing and traction bronchiectasis predominate in long standing cases.

MIXED CONNECTIVE TISSUE DISEASE

Mixed connective tissue disease (MTCDD) is a syndrome characterized by overlapping features of SSc, SLE, and PM/DM. The presence of high titres of antibodies against U1 small nuclear ribonucleoprotein (RNP) is a must for the diagnosis. About 80% of patients with MTCDD have respiratory system involvement. 34 Many patients with MTCDD present with Raynaud's phenomenon, swollen hands, sclerodactyly, arthritis, PM and pulmonary involvement. 35

The common lung manifestations include ILD, pleural effusion and pulmonary arterial hypertension. ILD occurs in up to 65% of patients. 36 It usually occurs within first 2 to 4 years of onset of the disease. Rarely, ILD may be the first or predominant manifestation of MTCDD. 37 NSIP is the most common histologic pattern on lung biopsy. 24, 38

Other pulmonary disorders include pulmonary vasculitis, pulmonary thrombo embolism, aspiration pneumonia, alveolar haemorrhage, pulmonary nodules, cysts, mediastinal lymphadenopathy, and respiratory muscle dysfunction.

SJOGRÉN'S SYNDROME

Sjogren's syndrome (SS) is characterized by lymphocytic infiltration of exocrine glands and other numerous extra glandular sites with resultant sicca syndrome. 39 It can occur alone as primary Sjogren syndrome (pSS) or in association with other autoimmune diseases, such as RA, SLE, and progressive SS known as secondary Sjogren's syndrome (SSS). It is predominantly seen in middle aged women with a female/male ratio of 9:1. 40 It has recently been proposed that, the diagnosis of SS should include four of the following features: ocular symptoms of inadequate tear production, corneal damage due to inadequate tear production, dryness of mouth due to inadequate salivary production, salivary gland histopathology showing presence of foci of lymphocytes, impaired salivary gland function on clinical testing, and the presence of antibodies (anti-SS-A/Ro or anti-SS-B/La, or both), however, the compatible histologic findings or presence of auto antibodies has to be one of the four criteria. 41

Lung involvement is common extra glandular manifestation of SS, and is seen in 9% to 75% patients. 42, 43 However, a recent study reported lung involvement in 11% of patients with primary Sjogren's syndrome. 44 The common pulmonary manifestations in SS include small airway disease, large airway obstruction or desiccation of the tracheobronchial tree, and interstitial lung disease.

AIRWAY INVOLVEMENT

Upper airway is commonly involved in SS. The dryness of the oral and nasal mucosa affects smell and taste functions. Upper airway bleeding, sinusitis, and septal perforation may also occur. 45

Dry cough is frequently present due to dryness of mucosa of trachea (xerotrachea) and large airways. Bronchial glands in large airway reveal changes similar to those in salivary exocrine glands, and mucosa shows lymphocytic infiltration predominantly with CD4 T lymphocytes.

Small airways are commonly involved in SS, but severe symptoms due to this are rare. With the progression of the disease, air trapping, mosaic attenuation, and bronchiectasis may manifest on CT scan of chest.

FOLLICULAR BRONCHIOLITIS

Follicular bronchiolitis is commonly associated with lymphocytic bronchitis, bronchiolitis, or LIP in patients with primary SS. Histological examination often reveals nodules of lymphocytic infiltrates with reactive germinal centres surrounding the terminal and respiratory bronchioles.

DIFFUSE INTERSTITIAL LUNG DISEASE (ILD)
**Pulmonary Manifestations**

Diffuse ILD is seen in 5% of patients with SS. Rarely, ILD may present without the sicca syndrome. NSIP is the commonest Interstitial lung disease in primary SS, which commonly manifests as increased reticular markings, ground-glass opacities, and traction bronchiectasis on HRCT scan of chest.\(^46, 47, 48\) Honeycombing is a late feature of NSIP. LIP is seen in 0.9% of patients with SS, whereas 25% of adults with LIP suffer from SS. A pattern of ground-glass opacities with thin-walled cysts are found in 50% of patients with LIP with other frequent radiologic findings being centrilobular nodules and interlobular septal thickening. Rarely, pleural effusion may also be present.\(^49\) UIP is rare in SS. HRCT of chest often reveals fibrosis, honeycombing, and traction bronchiectasis in lower lobes. Organizing pneumonia (OP) is extremely rare in SS with only a handful of cases reported. Recently, a patient with OP has been described, who subsequently developed SS 3 years later.\(^50\)

**Lymphoma**

The risk of lymphoma is 44 times higher in Sjogren's syndrome than that in a healthy population.\(^51\) Lymphoma develops in 4-8% of patients with SS with NHL being the commonest type.\(^52, 53\) Primary pulmonary lymphoma is also seen in 1%-2% of patients which can present as a solitary or multifocal nodules, bilateral alveolar infiltrates, or interstitial markings.\(^43, 54\)

**Other Manifestations**

Despite lung being commonly involved in SS, pulmonary hypertension is rare.\(^55, 56\) Pleural effusions and pleural thickening are also extremely rare conditions in SS.

**POLYMYOSITIS/DERMATOMYOSITIS**

Polymyositis (PM) and dermatomyositis (DM) affect skeletal muscles and various other organs. About 5% of patients with PM/DM develop lung manifestations with ILD and aspiration pneumonia being the commonest. Pulmonary vascular disease is rare. As PM/DM also occurs in malignancy, an active search for malignancy should be made in those who present with PM/DM.

The most common radiographic finding is aspiration pneumonia due to pharyngeal muscle weakness. The diaphragmatic weakness leads to high diaphragmatic elevation, reduced lung volumes, and basilar atelectasis.

ILD is seen in 50% of patients with PM/DM.\(^57, 58\) ILD commonly follows the musculoskeletal involvement, but may precede these in 1/3rd of cases.\(^59, 60\) ILD may be the sole manifestation in 20-30% of patients with DM/PM.\(^59, 60\) and in 10%-20% patients with amyopathic form of DM.

The lung bases are predominantly involved in ILD. Chest radiography reveals a reticulo-nodular pattern or honeycombing. HRCT of chest may reveal predominant linear abnormalities and ground-glass attenuation. Airspace consolidation is also often present in the lower lung zones with a peri bronchial and sub pleural distribution usually due to organizing pneumonia which is more common in PM/DM compared to other collagen vascular diseases.\(^61\) Although NSIP is the most common histological pattern, COP (cryptogenic organizing pneumonia), diffuse alveolar damage (DAD) and UIP are also frequently found.\(^59\)

**RELAPSING POLYCHONDritis**

Relapsing polychondritis (RP) is a rare disease characterized by recurrent episodes of cartilaginous inflammation with degeneration, loss of structure, and fibrosis throughout the body. The ears, nose, and airways are the most frequently affected organs in RP. About 1/3rd of patients will also suffer from another rheumatologic disorder, such as vasculitis, RA, Sjogren's syndrome, and ankylosing spondylitis. Rarely RP is seen in patients with inflammatory bowel disease, primary biliary cirrhosis, and myelodysplastic syndrome.

Airway manifestations are seen in 50% of patients with RP during the course of their illness. These are also the most serious complications presenting a diagnostic and therapeutic challenge.\(^62\) It may be the presenting feature in 10% of patients. The common airway involvement in RP includes subglottic stenosis, tracheal stenosis, tracheal wall thickening and calcification, and tracheobronchomalacia.

Respiratory symptoms are common in patients with RP, such as cough, dyspnea, hoarseness of voice, and stridor. Bronchoscopy is performed in patients having respiratory symptoms to assess for mucosal inflammation as a marker for active disease. The common bronchoscopic features are generalized redness of mucosa, mucosal oedema, tracheobronchomalacia with airway collapse, fixed narrowing and stenosis from granulation tissue and peribronchial fibrosis. However, tracheobronchomalacia and focal stenosis in the tracheobronchial tree are the commonest bronchoscopic findings.\(^63\) The sparing of posterior tracheal mucosa is an important diagnostic clue.

Most patients with respiratory symptoms reveal computed tomography (CT) scan abnormalities including tracheobronchomalacia and air trapping during expiratory scan which may not be apparent in inspiratory CT scan of the chest. Hence, the dynamic CT imaging is recommended in patients with suspected airway involvement.\(^64\) Recent developments in the imaging modalities such as three-dimensional imaging will allow early and improved detection of airway involvement in RP.

**ANKYLOSING SPONDYLITIS**

Ankylosing spondylitis (AK) is a multisystem disorder of unknown pathogenesis which primarily affects the joints of the axial skeleton. It commonly affects young males, 90% of them being HLA-B27 positive. The progressive destruction of the joints results in stiffness of spine, chest, and pelvis.

The lung is involved in up to 30% of patients with chest wall restriction and upper lobe fibrosis being the commonest.\(^6, 65\)
Pulmonary Manifestations

The restrictive ventilator defect may develop as a result of either fusion of costovertebral joints and ankylosis of thoracic spine or anterior chest wall involvement.

The lung parenchymal involvement generally occurs many years after the onset of arthritic changes. Upper lobe fibroblastic disease is the most common lung manifestation described in 1.3% to 30% of patients with ankylosing spondylitis. 66, 67 Bilateral apical scarring is believed to be the earliest pulmonary manifestation (54). 67 HRCT of chest may reveal peripheral interstitial lung disease, apical fibrosis, bronchiectasis, paraseptal emphysema, tracheobronchomegaly. Cavitation is uncommon, and was reported in only 0.45% of patients in one series. 68 There is a higher risk of pneumothorax in patients with AK, and it may be recurrent in some.

CONCLUSIONS

Patients with CVDs present with a wide range of pulmonary manifestations which cause a significant morbidity and mortality in this patient population. In addition, the frequent use of immunosuppressive therapy often makes them vulnerable to a wide range of infections which may create a diagnostic dilemma in many patients. To make the matter worse, some patients may present with lung involvement as a sole or predominant feature of an underlying VCD. Thus, a presence of a CVD should be suspected in a proper clinical context, particularly in patients with interstitial lung diseases, so that unnecessary delay in arriving at a diagnosis is avoided, and outcome in this patient group is improved.

References:

Pulmonary Manifestations


Case Report

Regression of plaque burden after primary percutaneous coronary intervention (PCI) in a patient with TVD: 4 years follow-up

AHM W Islam¹, S Munwar², S Talukder³, A Q M Reza², T Ahmed²

Abstract
Coronary Artery disease (CAD) is an important cause of mortality and morbidity in the developed world as well as in Bangladesh. Treatment of Acute Myocardial Infarction (AMI) patient either by Streptokinase (STK) or Primary Percutaneous Coronary Intervention (pPCI) has increased the survival outcome and reduced the mortality. Several studies have documented the significant beneficial role pPCI in terms of in-hospital survival outcome over thrombolysis. Our patient, who had Anterior MI in 2004 and his CAG revealed TVD. pPCI of the culprit mid LAD lesion with Bare Metal Stent (BMS) was done immediately after hospitalization. He was later referred for CABG, but decision was postponed because of asymptomatic status. His re-look CAG on 20-02-2008 (i.e., 4 yrs after the original procedure), revealed patent LAD stent with the regression of atherosclerotic plaque in Ostio-Proximal LAD and proximal LCX. Our findings indicated that pPCI with rigid control of CAD risk factors and modification of lifestyle plays a key role in the regression of atherosclerotic plaque and maintenance of stent patency.

Correspondence and Reprint request: Dr. A H M Waliul Islam, Dept of Interventional and Invasive cardiology, Apollo Hospitals Dhaka, Bangladesh. Ahmwislam@apollodhaka.com

Introduction: Acute coronary syndrome (ACS) is one of the important causes of death in the developing as well as developed world. Managing the patient either by STK or pPCI can reduce the mortality and improve 30 days in-hospital survival outcome. Primary angioplasty is the preferred treatment modality for AMI in centers with available cath lab facilities and well-trained personnel. The main advantage of pPCI is the achievement of a higher rate of coronary recanalization with a lower risk of intracranial bleeding. Now a days pPCI has become the choice of revascularization procedure compared to thrombolysis, for establishing TIMI III distal flow. It has already been established that pPCI is superior to thrombolytic therapy for the treatment of patient with STEMI. Many studies have demonstrated that pPCI within 2hrs of onset of symptoms has lower mortality and higher rate of complete reperfusion. Our present case shows the survival outcome in patient with acute Ant MI with TVD who had pPCI in Mid LAD in the year 2004.

Case: 61 yr Bangladeshi gentleman admitted to a local hospital with the onset of sudden chest pain for one and half hrs. His admission ECG revealed acute ST elevation in anterior chest leads (Fig.1).

He is a known hypertensive on medication and active smoker with no known positive family history for CAD. He was taken to the cardiac cath lab and CAG revealed TVD (Ostial + mid LAD, tight distal LCX and 100% RCA) with the culprit lesion in the mid LAD (Fig. 2).

Figure 1: ECG before Primary PCI in 2004 during chest pain showing Acute ST elevation in Anterior Chest leads

Figure 2a: Culprit lesion in Mid LAD

He was later referred for CABG, but decision was postponed because of asymptomatic status. His re-look CAG on 20-02-2008 (i.e., 4 yrs after the original procedure), revealed patent LAD stent with the regression of atherosclerotic plaque in Ostio-Proximal LAD and proximal LCX. Our findings indicated that pPCI with rigid control of CAD risk factors and modification of lifestyle plays a key role in the regression of atherosclerotic plaque and maintenance of stent patency.

Correspondence and Reprint request: Dr. A H M Waliul Islam, Dept of Interventional and Invasive cardiology, Apollo Hospitals Dhaka, Bangladesh. Ahmwislam@apollodhaka.com
Regression of plaque

He was discharged in a stable condition and given beta blocker, losartan plus HTZ, atorvastatin with nicotinic acid, Clopidogrel plus aspirin. He was followed-up in the OPD with the continuation of medication, regular exercise, and diet control. He stopped smoking. He remained asymptomatic throughout. He did not attend OPD regularly, but otherwise remained compliant. In 2007, he was advised for SPECT perfusion scan for routine follow-up. SPECT perfusion revealed MI involving apical wall with a strip of peri-infarct hibernation and also showed myocardial hibernation in septum (Fig.4.).

Emergency pPCI to culprit lesion was done with a BMS of 3 x 19 mm size deployed at 14ATM and post dilated with 3.5 x 12 mm balloon at 12ATM. Final CAG showed no residual stenosis with achievement of TIMI III distal flow. Post procedural ECG showed almost normalization of Elevated ST in the anterior chest leads (Fig.3).

He was planned for a re-look CAG to evaluate his coronary status. This revealed, a patent LAD stent and regression of Ostio-proximal LAD plaque (Fig.5)(Fig.6).
Slow flow in the circ and diffuse small vessel disease was noted, explaining SPECT perfusion findings. He was discharged in a stable condition with the advice of continuing medication, lifestyle modification and regular follow-up.

**Discussion:** It has been well established that the survival outcome of patients with Acute Myocardial Infarction by primary PCI is superior to conventional thrombolytic therapy. Weaver et al., demonstrated that pPCI results in reduced rates of mortality, reinfarction, and stroke. It has been well documented that PTCA alone in AMI, recurrence of ischaemia or re-infarction occurs in 37-49% and late infarct related artery re-occlusion in 9-14%. Stone has demonstrated that pPCI (stenting) has low rates of in-hospital death (0.8%), re-infarction (1.7%), recurrent Ischaemia 3.8% and pre-discharge target vessel revascularization for ischaemia (3.8%). In patient undergoing pPCI, procedural success provides significant prognostic values. Patient selection, lesion size, vessel caliber and plaque burden, overall door-to balloon time are the key factors in the procedural success and in-hospital 30 days survival outcome. Many have demonstrated that pPCI reduces the risk of left ventricular free wall rupture. Many have demonstrated that the better survival outcome among patients treated with pPCI, even when need to be transported to a center with cath lab facilities. Considering the better survival outcome and less procedural complications and decreasing morbidity, the UK government is considering to establish a national primary angioplasty service for patients with acute myocardial infarction. Our present patient demonstrated that the rigid control of CAD risk factors by regular exercise, complete withdrawal of smoking and anti-platelet and lipid lowering agents are not only the key factors in the maintenance of stent patency, but can reduce plaque burden. Nicholls et al. has described male gender, diabetes, and a history of prior revascularization are strong independent predictors of atherosclerotic burden in coronary disease patients. Birgelen CV et al. documented that positive linear relation between LDL cholesterol and annual changes in plaque size, with an LDL value of 75 mg/dL predicting, on average, no plaque progression. HDL cholesterol shows an inverse relation with annual changes in plaque size. In the landmark COURAGE study of patient with Stable Angina, revealed that PCI in addition to optimal medical therapy reduced the prevalence of angina. Therefore, our present patient's strong compliance to lifestyle modification and rigid cholesterol control by lipid lowering agent, may have caused regression of plaque burden.

**References:**
2. Aros F, Loma-Osorio AA, Aloso JJ. Guías de la sociedad espala de
Primary malignant CNS lymphoma in an immunocompetent patient: A case report

Khaled A¹, Salauddin  SA², Zaman SMQ³, Nasir TA⁴

Abstract

Aim and Objective: Primary malignant CNS lymphoma in otherwise healthy immunocompetent subject is relatively uncommon. They occur more frequently in immunocompromised patients. In this case report, we presented a report of an immunocompetent 48 year old male with primary CNS lymphoma

Clinical presentation: A 48 year old male was admitted in the neurosurgery department of Apollo hospital Dhaka because of continuous headache and vomiting for 15 days. There was no history of any illness, medication or head trauma. He was tested negative for HIV test. MRI revealed a periventricular mass which was confirmed as primary CNS lymphoma, diffuse large cell type by histopathology and immunohistochemical examination.

Conclusion: We presented this case because of relatively uncommon occurrence and raising incidence of primary CNS lymphoma in immunocompetent subjects in last one or two decades.

Key Word: Brain tumor, CNS lymphoma, immunocompetent

Introduction: Primary CNS lymphoma is extranodal malignant lymphomas arising in the CNS in the absence of obvious lymphoma outside the nervous system at the time of diagnosis.¹ Primary CNS lymphomas arise sporadically in otherwise healthy subjects but occur more frequently in the immunologically compromised particularly those with HIV infection and post transplant patient.² Primary CNS lymphoma affects all ages with a peak incidence in immunocompetent subjects during the 6th and 7th decades and usually 3rd and 4th decade in immunocompromised patient. About 60% of primary CNS lymphoma involve the supratentorial space including the frontal (15%), temporal (8%), parietal (17%), occipital (3%), basal ganglia/periventricular region (10%), corpus callosum (5%), posterior fossa (13%) and the spinal cord (1%).¹

Case report: A 48 year old male was admitted in the neurosurgery department of Apollo hospital Dhaka because of continuous headache with associated vomiting for 15 days. There was no history of febrile illness, head trauma, recent diagnosis of cancer, medication or any other illness. He also did not give any history of loss of consciousness or seizures. His physical examination revealed signs of raised intracranial pressure and both pupil were non reactive to light with decreased Glasgow coma scale (E4V4M6). Muscle power and other vitals were within normal limit. All laboratory examination was within normal range and he was negative for HIV test. MRI with contrast showed multiple well enhancing periventricular mass lesions with edema and mass effect suggestive of lymphoma. Left occipital craniotomy followed by debulking of tumor was done and tissue was sent for histopathological examination. Grossly, there were multiple grayish white pieces of tissue measuring between 3x2.5x2cm and 1x0.8x0.5cm. Microscopic examination showed diffuse infiltration of atypical large lymphoid appearing cells in the brain parenchyma with marked perivascular cuffing (angiocentric pattern) (Fig:1).

Immunohistochemically, the neoplastic cells were strongly positive for leukocyte common antigen (LCA) (Fig:2) and a diagnosis of primary malignant CNS lymphoma, diffuse large cell type were made.

Figure 1: Diffuse infiltration of atypical large lymphoid cells. In the brain parenchyma with angiocentric pattern

Figure 2: Tumor cells showing strong immureactivity for Leucocyte Common Antigen(LCA)
Primary malignant CNS lymphoma

Discussion: It is not known whether primary CNS lymphoma arise within or outside the brain and why they manifest in an organ that lacks a regular lymphatic system. Three hypothesis have been put forward (1) B cells may be transformed at a site elsewhere and then develop adhesion molecules specific for cerebral endothelia (2) lymphoma cells may be systemically eradicated by an intact immune system but may be relatively protected within the CNS (3) A polyclonal intracerebral inflammatory lesion may expand clonally and may progress to the monoclonal neoplastic state. Epstein-Barr virus play's a major role in the etiology of CNS lymphoma in case of immunocompromised patients but it has minor role in immunocompetent cases.

In an extensive study by Paul and his team on 56 Indian populations for a period of 20 years, they found median age of onset was 41 years, one patient was HIV positive with rest of them immunocompetent and histopathologically all the cases were diffuse large cell lymphoma. Their findings are almost similar to this presenting case. Over the last decade, the incidence of primary CNS lymphoma reported in most studies has risen threefold, which cannot be entirely explained by the increased prevalence of AIDS, or the growing number of organ transplantation. Incidence grew significantly not only in immunosuppressed patients, but also in immunocompetent patients. Mi and Mittal reported a case of 77 year old immunocompetent women with cognitive decline in whom a tentative diagnosis of primary CNS lymphoma was made. Brunnstrom et al also reported an immunocompetent 76 year old patient with cognitive and neurological symptoms.

The treatment of primary CNS lymphoma includes chemotherapy and radiotherapy. With this therapy immunocompetent patients show response rate of 85% with a median survival of 17- 45 months and immunocompromised patients 2-6 months. Some authors have also found that histological subtypes have a direct correlation with survival. In conclusion, we presented herein a report of an immunocompetent 48 year old male with primary CNS lymphoma. Pathological examination and immunohistochemistry provide the diagnosis of diffuse large cell lymphoma.

References:
Sarcomatoid carcinoma of the urinary bladder: A case report

A U Mallik, M Z Rahman, M M R Sarker

Abstract

Background: Sarcomatoid carcinoma of the urinary bladder is a very rare variety of transitional cell carcinoma (TCC) with a prominent component of spindle-shaped cell. The incidence of this tumor is 0.5% of all bladder tumors.

Case report: Herein, we report a case of sarcomatoid carcinoma of urinary bladder of a farmer of 70 years, a smoker with the complaints of lower abdominal pain, mild burning during urination and 3 episodes of painless gross haematuria. USG of KUB revealed a cystic lesion in front of the anterior bladder wall. Partial cystectomy was carried out. Diagnosis was confirmed by histopathological study. Patient refused radiotherapy or chemotherapy and died after 7 months of surgery due to metastasis.

Key Words: TCC, Sarcomatoid carcinoma. Partial cystectomy

Introduction: Sarcomatoid carcinoma (SC) is an aggressive type of malignant tumor that begins in tissues which line or cover internal organs and frequently a poorly differentiated urothelial carcinoma that may be difficult to distinguish from a sarcoma.

SC is a very rare variety. The incidence rate of this tumor is 0.5% of all bladder tumors and has an aggressive behavior, yielding a poor prognosis despite different treatment modalities.

It has a variety of names such as sarcomatoid carcinoma, pseudo sarcoma, malignant mixed mesodermal/mullerian tumor, metaplastic carcinoma and spindle cell carcinoma. Herein, we report a case of sarcomatoid carcinoma of the urinary bladder that was treated by partial cystectomy and diagnosed with histopathological study. Since the guardian of the patient refused total cystectomy with chemotherapy and radiotherapy, the outcome was found to be poor.

Case report: A 70 year old male farmer with a habit of smoking came with the complaints of 3 episodes of painless gross hematuria within 1 and half months. He also complained of mild burning during urination and lower abdominal pain intermittent in nature. His past history and family history was unremarkable. On physical examination, no abnormality was detected. The blood chemistry was within normal limit except for mild anemia. Urine microscopic examination revealed pyuria. USG of abdomen revealed a cystic lesion in front of the anterior bladder wall the first time. In cystoscopy, a hemorrhagic area in the anterior bladder wall was found. Patient was discharged. After 1 month he again came to us with one episode of hematuria. Time, USG of lower abdomen was unremarkable. It was decided to explore the urinary bladder for the cystic lesion in front of the anterior bladder wall. A hard mass was found in the anterior bladder wall. A total cystectomy was planned, but patient's guardian refused, so partial cystectomy with removal of 1 cm bladder wall surrounding the mass was undergone. Specimen was sent for histopathological study.

Macroscopically 5x 4.5x 3 cm a tumor infiltrating the adipose tissue at the anterior wall of bladder was seen. Microscopically, this tumor infiltrated all layers of the bladder wall. Histopathological findings show a malignant tumor of sarcomatoid carcinoma. It is composed of malignant epithelial component with glandular differentiation in a background of areas having sarcoma like appearance. The features are consistent with area of sarcomatoid carcinoma

Discussion: SC is an aggressive form of locally advanced carcinoma and has high rates of recurrence. Biopsy for histopathological and immunohistochemical study is required for accurate diagnosis. The treatment options for SC will vary depending upon many factors having to do with the patient's over-all condition. SC is composed predominantly of a sarcomatoid component and an obviously carcinomatous component. The sarcomatoid component is composed of a mixture of spindle cells, round cells, and pleomorphic giant cells. The carcinomatous component consisted of papillary or non-papillary high-grade TCC. In our case the sections of tissues shows a malignant epithelial component with glandular differentiation in a background of areas having sarcoma like appearance. The features are consistent with...
Sarcomatoid carcinoma

area of sarcomatoid carcinoma.1 One reported that of 4191 patients with bladder carcinoma, the incidence of SC was 0.3% and 1.06% in another report. As SC is a very rare tumor, data regarding etiology and outcome is also limited. Although, no definite risk factors for SC are identified to date, an association with radiation and cyclophosphamide therapy has been noted. The male female ratio is approximate 3:1.4 In general, SC presents a diagnostic difficulty if any definite abnormalities are not found in investigation, as was the case in our patient. We reported such a case of SC of urinary bladder treated with partial cystectomy. The median age at diagnosis is 60 to 70 years and most of the patients with SC of urinary bladder presents with painless hematuria as was in our case. Although the term sarcomatoid carcinoma and carcinosarcoma have been used together, the term sarcomatoid carcinoma has been used to describe malignant spindle cell type tumor with epithelial differentiation, while carcinosarcoma describes the tumor composed of both malignant epithelial and malignant soft tissue elements.9 In sarcomatoid carcinoma, the sarcomatous elements are chondrosarcoma, leiomysarcoma, fibrosarcoma and rhabdomyosarcoma and epithelial elements consist of transitional cell carcinoma, small cell carcinoma, squamous carcinoma and other mixed carcinoma.

To differentiate sarcomatoid carcinoma and carcinosarcoma, immunohistochemical study is very important. In our case report, IHC demonstrated that the tumor was composed of malignant epithelial component with glandular differentiation with areas of sarcoma like appearance.7 This type of tumor has a very aggressive behavior, local invasion are the striking features of this cancer. Total cystectomy is the preferred treatment modality with radiotherapy or chemotherapy, although different treatment has different consequences.10,12

Our case was in preliminary stage, no positive findings were in our investigations, except a cystic lesion found in front of the anterior bladder wall by USG. With this suspicion, we explored the urinary bladder with a lower midline incision and found a hard mass in the anterior bladder wall. We planned for total cystectomy but because the patient’s guardian refused, so partial cystectomy was done. Post operative period was uneventful. Survival time was 7 months after surgery. Some authors found median survival time 10 months and others found 2 or 3 months after treatment.11,12 Regarding clinical outcome of sarcomatoid carcinoma of the urinary bladder very few data have been published, so it is difficult to assess due to vague diagnostic standards used in each group. Clinical behavior of such a rare tumor is difficult, but very aggressive and usually loco-regional extension is the main feature of such tumors. Total cystectomy is the preferred method of treatment of SC of urinary bladder. For achievement of better local control, adjuvant radiotherapy and chemotherapy may show prospective result.13,15 Froehner et al reported a case of a metastatic SC remission of the bladder with cisplatin and gemcitabine, although in the literature, adjuvant radiotherapy or adjuvant chemotherapy are apparently of no effect.2,4 Our patient refused chemotherapy and radiotherapy, but the patient survived for 7 months after partial cystectomy due to early diagnosis and early treatment.

Conclusion: Routine investigations may not give any clue for early diagnosis of sarcomatoid carcinoma of the bladder. Total cystectomy is the choice of treatment for SC but postoperative quality of life should be considered before any aggressive decision is taken. Further study with more cases and experience may provide great value, clinically and pathologically. More study with more cases is necessary to see the survival time.

References:

Fibrosarcoma of the chest wall: A case report

SF Kabir¹

Abstract

An 85 year old man presented with a large swelling over the right side of the chest wall of 3 years duration. Patient had no complaints other than a swelling which looked aggressive. On the 1st instance it looked malignant. Histopathology of the specimen confirmed that it is a case of fibrosarcoma. Excision of the tumor done with 2cm healthy margin and the patient was referred to radiotherapist for possible adjuvant therapy.

Introduction: Fibrosarcoma is the most common primary soft tissue cancer of the chest wall. It occurs most frequently in young adults. The incidence of fibrosarcoma is about 0.05 cases per 100,000 population per year.² Male and female are equally affected. More than half of the cases occur in the long bones, the distal femur and proximal tibia are the commonest sites. The humerus and scapula together have been reported as representing perhaps the second or third most common site. These tumors have been reported in every decade but are most common in the third and fourth decades. The signs and symptoms are those of pain and, at times, local swelling.

Case report: An 85 yrs Bangladeshi farmer from Noakhali, weighing 55 Kg, non smoker, non diabetic was admitted in a private clinic at Dhaka with the complaints of a large swelling on the right side of the anterior chest wall for 3 years. The swelling was gradually increasing in size but in the last three months the swelling had increased two fold according to the patient's statement. Other than the swelling he had no complaints. On examination, he was mildly anaemic, non ecteric, no lymph node enlargement, both axillary and neck regions were normal. Skin was free from the swelling. It is about 12.5cm x 10cm in size, mobile from side to side and above downwards, not fixed to the underling muscle, the upper part of the swelling was ulcerated.

Laboratory investigations are within normal limit, X-ray chest and ECG were normal. Fine needle aspiration was positive for sarcomatous lesion.

Under general anesthesia the swelling was resected with 2cm healthy margin. Wound was closed keeping a drain insitu. The histopathology report confirmed fibrosarcoma of moderate grading.

Discussion: Soft tissue sarcomas account for approximately 1% of all new cancer diagnosed. Slightly more than half of all patients diagnosed with the disease eventually die as a result of the cancer. Classically, soft tissue sarcomas present as asymptomatic large masses in the extremities or retroperitoneum, but they also develop with some frequency in the neck and within the abdominal viscera. Fifty percent of these tumors occur in the lower extremity, most commonly in the thigh. Soft tissue sarcomas originate from a wide variety of mesenchymal cell types and include liposarcoma, fibrosarcoma, rhabdomyosarcoma, and desmoids tumors. While the histopathology of these tumors is highly variable, with some exceptions they tend to behave in a fashion directed by tumor grade rather than the cell of origin.

Most soft tissue sarcomas arise de novo, and rarely do they result from malignant degeneration of a benign lesion. There are some familial syndromes in which patients are genetically predisposed to the formation of soft tissue sarcomas. Any mass suspected of harboring a soft tissue sarcoma should be imaged prior to operation. Both CT and MRI can be used to image these tumors, but MRI is often more accurate in defining the extent of the tumor and invasion of surrounding structures. A chest x-ray or chest CT should be obtained in order to evaluate for pulmonary metastases in patients with high-grade tumors. In imaging a retroperitoneal tumor, the liver should be examined in the same radiographic study.

The most important prognostic variables for patients with soft tissue sarcoma are the size and grade of the primary tumor.³ As grading is based on the cellular architecture and invasive nature of the tumor, FNA is not a typically useful biopsy technique for the initial diagnosis of a sarcoma. Core needle biopsies may be performed for large, palpable superficial tumors. For large deep tumors or those adjacent to vital structures, incisional biopsy is usually the diagnostic method of choice. If a tumor is small (<3cm) and superficial, excisional biopsy should be performed.

Treatment: Surgery is the mainstay of therapy for sarcoma, but these tumors are also relatively sensitive to radiation.⁴ A pseudocapsule composed of tumor cells surrounds sarcomas, and local invasion along fascial planes and neurovascular structures is common. Local capsular excision is inadequate for cure, as local recurrence rates for such procedures are greater than 90%.

Generally, the margin for wide local excision should be at least 2cm. The dissection should be carried out at least one fascial plane away from the tumor and may necessitate a compartmental resection. The surgeon should take care to avoid direct visualization of the tumor during the procedure in order to prevent tumor from seeding into the wound. Surgical excision of the tumor may involve resection of important neurovascular structure.

Large soft tissue defects often require the construction of myocutaneous flaps to improve function and cosmesis.⁵ Soft tissue sarcomas rarely invade bone or skin, and wide

¹ Registrar, Department of General and Laparoscopic Surgery, Apollo Hospitals Dhaka
Fibrosarcoma of the chest wall

resections of these structures are infrequently necessary. Following wide local excision, metal clips should be placed at all margins of the resection in order to guide subsequent radiotherapy.

Radiation therapy is a necessary adjunct in the local control of sarcomas. Based on the location and size of the tumor, several modes of delivery are used, including EBRT, intraoperative radiation, and brachytherapy. In many centers, small (<2cm) low-grade extremity tumors are treated with wide local excision alone. Larger tumors should be treated by wide excision and postoperative radiation therapy, either by EBRT or brachytherapy catheters. Preoperative radiotherapy appears to have distinct advantages in patients with tumors larger than 10cm, improving local control rates and sometimes shrinking tumors sufficiently to allow for limb sparing procedures.

Conclusion: The natural history is one of progressive growth, frequent pathologic fracture, and distant metastases usually to the lung, with death if appropriate treatment is not given. Treatment consists of local removal of the primary tumor, which in most situations requires amputation. Neither radiation therapy nor chemotherapy has been helpful. The 5 years survival rate is 30-35%.

Reference:
Case Report

Transcatheter technique now standard for Secundum ASD Closure, A case report of our first experience.

A Q M Reza¹, A B Siddique², M S U Talukder³, S Munwar⁴, A H M W Islam⁵, S R Shohel⁶, M A Ghani⁷

Introduction: Atrial septal defect (ASD) is one of the more commonly recognized congenital cardiac anomalies presenting in adulthood. Atrial septal defect is characterized by a defect in the interatrial septum allowing pulmonary venous return from the left atrium to pass directly to the right atrium. Depending on the size of the defect, size of the shunt, and associated anomalies, this can result in a spectrum of disease from no significant cardiac sequelae to right-sided volume overload, pulmonary arterial hypertension, and even atrial arrhythmias.

Surgical closure of atrial septal defects has been practiced successfully for 50 years, nowadays reaching the expectation of zero mortality and an optimal functional result. In the attempt to minimise operative morbidity, practice has moved to a less invasive surgical approach, including minimal skin incisions, reduced opening of the thoracic cage and alternative vascular access sites. More recently, the development of catheter-based technology has advanced device occlusion as the first option for closure of ostium secundum atrial septal defects. This mode of therapy appears now to be widely accepted, based on the obvious advantage of saving the patient from open heart surgery, allowing subsequently faster rehabilitation. Catheter-based closure is now considered the first-line treatment strategy for secundum atrial septal defects (ASD). Although in some cases, surgery remains the only option (eg, for those with an insufficient rim to secure a device, or those with coincidental anomalous pulmonary venous return), a percutaneous approach is suitable for the vast majority of patients with secundum ASD. As skill and experience grow, defects as large as 40 mm in size have been successfully closed in this manner. We will report a case of Secundum ASD Device Closure which was first done in Apollo Hospitals Dhaka.

Case Report: This 50 years old hypertensive (newly diagnosed), diabetic (newly diagnosed) Bangladeshi lady from Chittagong with a positive family history of heart disease was referred from outside to OPD, Apollo Hospitals Dhaka as a case of Secundum ASD for further evaluation & treatment. Since last year she was having shortness of breath and unusual tiredness. For this reason she underwent routine checkup (OGTT, ECG & Blood pressure monitoring). ECG showed incomplete RBBB with flattening of T waves from V1-V6 and for this reason she was further advised for Transthoracic Echocardiogram. Echo was done on 27th May 2009 which showed ASD(Secundum) with left to right shunt - about 23 mm approximately, Dilated RA(40mm), RV(30 mm), PA(23 mm) with mild pulmonary hypertension(PASP 55 mmHg) & Mild Tricuspid Regurgitation. Pulmonary flow was 1.73 m/sec & PPG 12.03 mmHg.

On examination in Apollo Hospitals Dhaka, at OPD revealed Pulse - 80 bpm regular, B.P 140/90 mmHg, Lungs - Clear, Heart - Ejection Systolic murmur with a wide fixed splitting of the second heart sound. She was started with antihypertensives & was put on strict diabetic diet and was advised for TEE, CAG & Cardiac catheterization. CAG and Cardiac catheterization was done on 04.06.09. CAG revealed Normal Epicardial Coronary Arteries and Cardiac catheterization revealed Qp : Qs = 2.73: 1, PVR = 5 wood unit, L-R shunt 63 %, significant step up of O₂ saturation in mid RA and she was advised for ASD Device closure.

On 08.06.09 TEE was done which revealed Moderate size ASD (Secundum) about 25 mm with left to right shunt and good rim size of IAS.

On 19.07.09 ASD device closure was done successfully through right femoral approach under local anaesthesia without any sedatives and without Trans Esophageal Echo (TEE) with a 26 mm device. Transthoracic Echo was used per operatively & post operatively and device was implanted with no residual shunt. Patient was discharged home on 22.07.09 with Dual Antiplatlet, antihypertensives and advice to come for a follow up 1, 3 and 6 month. The follow-up scheme after device occlusion comprises a physical examination, routine blood (CBC), CXR and transthoracic echocardiography. Her first follow up after 1 month, patient had no complaints. Physical examination revealed Pulse-68 bpm regular, B.P 125/80 mm Hg, Lungs-Clear, Heart-Normal, 1st & 2nd heart sound, Echo was done which showed device normaly in situ with no residual shunt. At 3rd month CBC & CXR were done which were within normal limit. CXR showed device in position. At 6th months routine blood tests were done which showed no abnormality. Echo showed device in situ with no residual shunt and chamber dimensions and other parameters are within normal limit - RA (30), RV (27.8) and PA (19.5), Pulmonary Flow 0.99 m/sec, PPG-3.9 mmHg, PASP 29 mmHg. Antiplatelets were discontinued from 6th month after the procedure.

Figure 1: Apical 4 chamber view showing ASD.
Transcatheter technique

**Discussion:** Transcatheter closure is becoming an accepted technique for closure of patent foramen ovales and some ASDs. Because it can be performed without general anesthesia, without cardiopulmonary bypass, and without an incision, the technique must be considered an attractive alternative to conventional surgical closure. Although the cost of the device and the implantation facilities may be similar to those of surgical closure, hospitalization is shortened or eliminated and time off from work is greatly reduced. A close working relationship between the surgeon and the interventionist will permit successful and safe application of this technique. The surgeon's role continues to be that of patient advocate, recognizing favorable and unfavorable anatomy, and participating in the selection of patients for the transcatheter approach.

In our first experience we have successfully closed a Secundum ASD in a middle aged women without any minor or major complications by Transcatheter device closure. Our achievement was, we did it without sedations and we used per procedural Transthoracic Echocardiography instead of Transesophageal Echocardiography for positioning of the device during implantation and to check whether there is any residual shunt present or not and ensured better comfort of the patient.

**Conclusion:** In view of the high success rate of complete closure, safety of device closure, and growing experience in use of this device, it seems likely that device closure of ASD will become the preferred mode for closure of ASD with suitable anatomy.

**References:**
1. Markham LW, Cribbs MG. Atrial Septal Defect. eMedicine from WebMD. Updated: Jan 21, 2010.
Case Report

Afibrinogenemia

Tahera Nazrin1, Pinkoo Attawar2, Md. Moniruzzaman3, I Islam4

Abstract

Afibrinogenemia is a rare bleeding disorder with an estimated prevalence of 1:10,000,000.1 It is an autosomal recessive disease resulting from mutations in any of the 3 genes that encode the 3 polypeptide chains of fibrinogen and are located on the long arm of chromosome 4.2 Spontaneous bleeding, bleeding after minor trauma, and excessive bleeding during interventional procedures are the principal manifestations.3,4 Here we have reviewed the process of diagnosing a case of such rare disorder in Apollo Hospitals Dhaka. We have also highlighted the treatment and management plan of such a case.

Introduction: Afibrinogenemia is a very rare inherited disorder in which factor I or fibrinogen deficiency occurs.5 Afibrinogenemia is an autosomal recessive disease and most of the patients are commonly descendents of consanguineous marriage.5,6 It was first discovered in 1920 by two German physicians, Fritz Rabe and Eugene Salomon. People affected by this disorder and those close to them have very little written information about it. This literature therefore seeks to provide information for people trying to cope with this health problem. Here we present a case with afibrinogenemia, the process of diagnosis and explain the causes of the disorder and currently available treatments.

Case summary: Muna, a 3 years old girl, weighing 11.5 kg having 0.93 meter height, only issue of a 1st degree consanguineous parents, was admitted in Paediatric department, Apollo Hospitals Dhaka with the complaints of multiple bruises and echymoses all over the body. She had h/o same type of bleeding spots on limbs during crawling or after falling on ground while walking or running. Her mother stated that there was bloody discharge from the umbilical stump even at 2 weeks of her age. Her milestones of development were slightly delayed. She had no h/o hospital admission for this illness and there was no h/o such type of illness in her family.

Physical examinations on OPD visit revealed temperature 98.4°F, H.R-130 b/m, R.R-20/m. She was conscious, cooperative, having mild pallor, multiple bruises and echymoses all over the body. She had h/o falling on ground while walking or running. Her mother stated that there was bloody discharge from the umbilical stump even at 2 weeks of her age. Her milestones of development were slightly delayed. She had no h/o hospital admission for this illness and there was no h/o such type of illness in her family.

Physical examinations on OPD visit revealed temperature 98.4°F, H.R-130 b/m, R.R-20/m. She was conscious, cooperative, having mild pallor, multiple bruises and echymoses all over the body. She had h/o falling on ground while walking or running. Her mother stated that there was bloody discharge from the umbilical stump even at 2 weeks of her age. Her milestones of development were slightly delayed. She had no h/o hospital admission for this illness and there was no h/o such type of illness in her family.

Relevant investigations were done as stated below:

HB%: 11.4gm/dl, PCV: 0.33L/L, MCV: 78.0fl, MCH: 27.0pg, MCHC: 34.0%, Platelets: 362 10^5/c mm, TLC: 10.7 10^9/L, N: 27%, L: 62%, E: 09%, B: 00%, M: 02%, BT: 7 min, CT: Clotting was not seen within 30 minutes, PT: >118 sec (contr:12-14sec), INR: >12, APTT: >180sec (contr:32-38sec), TT: >60 sec(constr:17.0sec), Urea solubility test (2% acetic acid): result unstable.

Investigation reports with prolonged CT, PT, APTT & TT suggests clotting disorder. This picture could be found in liver disease, fibrinogen deficiency and inhibition of fibrin polymerization or Hyperfibrinolysis.6 Serum fibrinogen level of this girl was less than 15 mg /dl (ref:200-400mg/dl). Liver disease was also excluded by normal liver function tests: total S. Bilirubin 0.5 mg/dl (normal range- 0.5-1 mg/dl), S. SGPT: 27 IU/L (norm: 5-40 IU/L), Alkaline Phosphatase; 229 IU/L (32-385 IU/L).Total protein and A/G ratio were also within normal limit. Hence, this is a case of fibrinogen deficiency disorder, Afibrinogenemia.

Counseling to the parents was done about the disease, treatment and protection of the baby.

Discussion: Fibrinogen/factor I, a plasma protein produced by the liver plays an important role in blood coagulation, missing or functional problems of which results in hemorrhage or thrombosis.7 These fibrinogen disorders can present as afibrinogenemia or hypofibrinogenemia (quantitative defects) or dysfibrinogenemia (qualitative defects).7 The normal volume of fibrinogen in the blood is from 2 to 4 g/l.8

Afibrinogenemia is a very rare inherited bleeding disorder that affects both male and female of all races and ethnic origin.9 The first clinical report of congenital afibrinogenemia dates back to 1920 when a 9 year old boy suffering from recurrent bleeding episodes since birth and lacking of fibrinogen in blood was described and shown subsequently to be autosomal recessive in inheritance with variable penetrance.10,11 The estimated prevalence of afibrinogenemia which is most severe form of the disorder is around 1 in 10 lacs2 and recent registries from Italy, Iran and North America have greatly improved understanding of the clinical spectrum of presentation.12 In fact, a seven fold higher incidence of fibrinogen disorders was observed in the Iranian Registry (where consanguinity is high) for rare bleeding disorders in comparison with similar registries in Italy and United Kingdom.8

In congenital afibrinogenemia (fibrinogen level <0.2 g/l), bleeding can vary, from slight to severe. Bleeding can start in the neonatal period with 85% presenting with umbilical cord bleeding10 or bleeding into the skin, gastrointestinal tract, genitourinary tract or central nervous system bleeding.

1. Registrar, Department of Paediatrics, Apollo Hospital Dhaka, 2. Consultant, Department of Paediatrics, Apollo Hospital Dhaka, 3. Consultant, Department of Hematology, Apollo Hospital Dhaka, 4. Specialist, Department of Hematology & Clinical Pathology, Apollo Hospital Dhaka
Afibrinogenemia

(5%). Musculoskeletal bleeding (including hemarthroses) occurs in 54% of the patients. In afibrinogenemia unusual manifestations such as spontaneous rupture of the spleen and presence of bone cysts are also observed. In females menorrhagia (7%) have been noted. Further, impaired wound healing and wound dehiscence post surgery have been reported because of the non tensile clot and inadequate deposition of healing proteins delaying wound healing.

It is strongly recommended that people who suffer from afibrinogenemia learn to recognize the signs and symptoms of bleeding that could threaten their lives or the integrity of a limb, so that they can react adequately and in a reasonable time. Bleeding that affects the head, neck, chest or abdomen can be life-threatening and may require immediate medical attention. This kind of bleeding can occur either following an injury or spontaneously. Intra cranial hemorrhage is very serious, and may be manifested as headache, visual problem, nausea, vomiting, change of personality, somnolence, loss of balance, loss of fine motor skills, fainting or convulsion. Intra thoracic bleeding may present with chest pain, breathing difficulty, cough or bloody sputum or swallowing difficulty. Intra abdominal hemorrhage causes pain in abdomen or low back pain or blood in urine or stool. If one of these symptoms occurs, immediate consultation with a physician is necessary.

There are other kinds of bleeding that are not necessarily life-threatening, but for which treatment is necessary. These are soft tissue bleeding, bleeding in the joints manifested by reduced joint mobility, or swelling or heat in the joint with or without bruising. Replacement therapy is the mainstay of treatment of bleeding episodes in patients with afibrinogenemia and includes plasma-derived fibrinogen concentrate, cryoprecipitate and fresh frozen plasma. The aim of the treatment is to increase the fibrinogen level to 1 g/L when there is minor bleeding, and 2 g/L for serious bleeding or for surgery. The girl in our hospital was treated with fresh frozen plasma due to lack of availability of other options. At present, the most frequently used treatment abroad is fibrinogen concentrate. The concentrate is obtained from human plasma and contains fibrinogen only. The quantity of fibrinogen required can be calculated as follows:

\[ \text{Dose (g) = Desired increment (g/l) \times plasma volume \times patient weight (kg)} \]

Here, plasma volume = 0.07 \times (1- hematocrit).

There are five fibrinogen concentrates currently available: Hemoocompletan P (CSL Behring, Marburg, Germany), Clottagen and FIBRINOGENE T1 (LFB, Les Ullis, France), Fibrinogen HT (Benesis,Osaka, Japan) and FibroRAAS (Shangai RAAS, Shangai,China).

Prevention is very important for this disease. The patient was advised never to take aspirin, and to prevent dental problem she needs to visit dentist every six months. The patient should always contact with hematologist if she needs surgery or a tooth extraction in order to plan adequate preventive treatment. She also advised to carry a card explaining her coagulation problem. Contact sports were also asked to avoid due to the significant risk of bleeding. She is also recommended to receive vaccine against hepatitis A and B by fine needle.

Conclusion: Even though afibrinogenemia is a rare disorder, it might acquire greater importance in future when this disorder might be more prevalent due to increasing consanguineous marriages. Patients with afibrinogenemia should be referred to and registered with a center of bleeding disorders guided by hematologist. The development of new tests to identify higher risk patients and safer replacement therapy will improve the management of these cases in future.

References:

Bilateral renal angiomyolipoma not associated with tuberous sclerosis: A case report

Khaled A, Arif NUM, Zaman W, Nasir TA

Abstract

Angiomyolipoma is a tumour composed of varying admixtures of blood vessels, smooth muscles and adipose tissue. Renal angiomyolipoma can be unilateral or bilateral. Bilateral angiomyolipoma has a very strong association with tuberous sclerosis. In non tuberous sclerosis patient, bilateral renal angiomyolipoma is relatively rare. We have described a 49 year old non tuberous sclerosis female presented with bilateral renal angiomyolipoma. Renal angiomyolipoma commonly behaved in a benign way and resection is curative in most of the cases.

Case Report

Introduction: Angiomyolipoma is a tumor composed of varying admixtures of blood vessels, smooth muscle cells and adipose tissue. The lesion should be considered a choriostoma, a disordered arrangement of mature tissue appearing at a site where that tissue does not normally reside. The frequency of angiomyolipoma of kidney varies from 0.7 to 2% of all renal tumors depending upon whether cases were discovered as incidental findings or as symptomatic tumor with or without associated tuberous sclerosis. Patients are predominantly female. The average age at diagnosis is 41 years. Among symptomatic patients, flank pain related to intratumoral hemorrhage is the most common complaint. Angiomyolipoma have been observed in patients with several hereditary disorders including Von Recklinghausen disease, Von Hippel Lindau syndrome, and autosomal dominant polycystic kidney disease. The association is particularly strong with tuberous sclerosis (mental retardation, epilepsy, cutaneous hamartoma, depigmented spots and subungual fibromas of fingers). Angiomyolipoma occurs in 80% of individual with tuberous sclerosis and in most cases it is bilateral. The association is so close that all patient with multiple renal angiomyolipomas should be evaluated for tuberous sclerosis.

Case report: A 49 year old non diabetic, hypertensive female came with the complaint of long standing left lumber pain in the urology outpatient department of Apollo hospital Dhaka. Her physical examination revealed lumps on both side of abdomen extending from hypochondrium to lumber region, non tender, firm and bimanually palpable. Laboratory examination showed raised serum creatinine, neutrophilia but urine examination was negative for haematuria. Ultrasonogram of whole abdomen showed bilateral renal mass and suggested CT/MRI correlation. MRI revealed bilateral enlarged kidneys with lobulations, loss of architecture and replacement by fat containing mass lesion very much suggestive of bilateral diffuse angiomyolipomatosis of the kidneys. She was screened for other signs of tuberous sclerosis and found to be negative. Her bone scan was also negative. Left sided nephrectomy was done and specimen was sent for histopathological examination. Grossly, the total specimen measured 26x14x7 cm. Cut surface showed a huge mass with yellowish and hemorrhagic areas replacing almost whole of kidney tissue. Sections from the growth revealed the feature of angiomyolipomas characterized by areas of mature adipose tissue, tortuous thick walled blood vessels, bundles of smooth muscles and perivascular epithelioid cells (Fig: 1).

Discussion: Angiomyolipomas of the kidneys are hamartomas that may be sporadically found. They are usually asymptomatic and more common than previously appreciated, approaching 13 per 10.000 adults. Moreover, they may be found either in one or in both kidneys. Multiple lesions are found in about one third of the cases and bilateral tumors in 15%. Such tumors may occur in other organs (e.g. liver), in association with renal angiomyolipoma. Our patient had bilateral disease. Angiomyolipomas are much more prevalent in patients with tuberous sclerosis in a percentage as high as 50-80%, accompanied by cysts and occasionally by renal cell carcinoma. Tuberous sclerosis is a multisystem syndrome characterized by neurological symptoms and tumors in multiple organs including kidneys, brain, skin, eyes, heart and lungs. In symptomatic patients, the most common manifestations are pain, retroperitoneal hemorrhage or hypovolaemic shock, haematuria, hypertension, palpable mass, anemia, acute pyelonephritis and fever. The size of the tumor varies and they are classified as small (< 4 cm), medium (4-8 cm) or large (> 8 cm). Renal angiomyolipoma of
Bilateral renal

less than 4 cm is generally asymptomatic. Angiomyolipomas greater than 8 cm, responsible for significant morbidity, require treatment, prior to the development of symptoms and potential complications. The large ones are more common in women than in men and their rapid growth during pregnancy suggest that hormones may be responsible for increasing size of angiomyolipoma. Imaging methods for the diagnosis of renal tumors include US, CT, angiography and MRI. US reveal a high acoustic renal mass. In CT, the excessive fat tissue of the tumor has a characteristic appearance and after intravenous bolus injection of contrast media, the pathological blood circulation of the tumor is evidenced. Typical angiomyolipomas are benign tumors. However, the smooth muscle component is troublesome because it may exhibit hypercellularity, marked pleomorphism and moderate mitotic activity. These features may prompt a mistaken diagnosis of leiomyosarcoma. In addition, the regional lymph nodes may be involved, but it is generally regarded as an expression of multicentricity rather than true metastasis. Furthermore, they may show resemblance to renal cell carcinoma because of their yellow color, intratumoral hemorrhage and frequent extra renal growth. The percutaneous needle aspiration biopsy under US, CT or MRI control, usually confirms the diagnosis of angiomyolipomas, allowing differentiation of this entity from other renal tumors.

The management approach of angiomyolipomas is surgical resection, which should be aimed at parenchyma preservation. This can be effectively accomplished by limited surgery, without postoperative dysfunction or need for dialysis, even in solitary kidney. Furthermore, complete or selective renal embolization is a viable alternative to surgery, mainly in patients with poorly functioning kidneys who present with pain or bleeding, and in those who are poor operative candidates. The embolization is safe, reliable and minimally invasive with few long-term sequelae, and it is well tolerated. In conclusion, current management options of renal angiomyolipomas include observation, embolization and partial or total nephrectomy. Recommendations of treatment are usually based on the symptoms observed and the size of the lesion.

In conclusion, we have described a bilateral renal angiomyolipoma in a non tuberous sclerosis patient along with general characteristics of renal angiomyolipoma.

References:

Astroblastoma

Khaled A', Joader A', Chandy M', Nasir TA'

**Introduction:** Astroblastoma is a rare glial neoplasm with preferential manifestation in young adults, histologically characterized by a typical perivascular pattern of astrocytic cells with broad non-tapering processes radiating towards a central blood vessels. The biological behavior of astroblastoma is variable. In the absence of sufficient clinicopathologic data, it has been decided not to establish a WHO grade and they are listed in the category of neuroepithelial tumor of unknown origin 1.

**Case report:** A 10 year old girl presented in the neurosurgery outpatient of Apollo hospital Dhaka with intermittent frontal headache and history of seizure two years back which persisted for six months and was controlled by anti-convulsiv drugs. Now for the last one month she has developed right sided weakness, unable to stand or walk with loss of appetite and occasional headache. On examination, her Glasgow coma scale was 15 (E4V5M6), both pupils were equally reactive to light with normal vision and other vitals. But there was right sided muscle power weakness. Her routine laboratory examination and chest x ray was normal. MRI scan of brain revealed a mass in the left parietal supratentorial region consistent with astrocytoma. Left fronto-parietal craniotomy with removal of the tumor mass was done and the specimen was sent for histo-pathological examination. Grossly, the specimen consisted of one irregular grey brown mass measuring 6x4x3 cm. Cut surface was solid and homogenous. Microscopically, the tumor was characterized by perivascular pseudorosettes pattern proliferation of glial cell like cells with broad, non tapering processes towards blood vessels. The stromal blood vessels show collagenous thickening and hylialinization of the wall with focal obliteration of the lumen (Fig:1).

![Figure 1: Magnetic resonance imaging showed a large well-defined mass in the left parietal region extending from the cortex to the periventricular region.](image)

The tumor was non-infiltrative and surrounded by normal brain tissue. Histo-morphologically, a diagnosis of astroblastoma was made.

**Discussion:** Astroblastoma, a rare tumor of cerebral hemispheres has been known to occur from infancy to fifth decade 2,3,4,5. The cell of origin of astroblastoma is still a debatable entity, but widely accepted to be the astroblast - an intermittent cell between spongialast and astrocytes. Till recently, the possible origin from tanyctye was reported 6,7, however, till date no consensus has been reached whether it arises from an immature astroblast or by process of differentiation of astrocytes 6. Clinically raised intracranial pressure and seizure episodes remain the common presentation. Others may present with signs and symptoms pertaining to anatomical structures involved like visual loss, memory disturbances, seizures, weakness, and altered sensorium 3,4,8. Cerebral hemispheres, mostly frontal and occipital lobes are the site favored by these tumors as in our case, but corpus callosum, cerebellar hemispheres, optic nerves, brainstem, and cauda equina tumors have been reported in literature 6,9.

On MRI, these tumors tend to be large lobulated masses which characteristically extend from the peripheral cortex to the periventricular region. On imaging, these tumors are well-demarcated lobular tumors. T1 and T2 weighted sequences of these tumors appear well demarcated, heterogeneous solid-cystic components with inhomogeneous contrast enhancement. Solid component give typical heterogeneous appearance on MRI, and peritumoral T2 hyperintensity is less compared to their large size 10. In spite of the characteristic imaging features, they are usually confused for glioma as in this case, primitive neuroectodermal tumors (PNET), or ependymoma 2,4.

Histologically, these tumors are characterized by
Astroblastoma

perivascular pseudossets throughout the tumor tissue more distinctly observed in solid areas of tumor. Most of the tumor cells are monotonous with pseudorosettes arrangement pattern. These rosettes were characterized by short and thick blunt ended foot plates of astroblastoma cells directed toward the central blood vessel. These pseudorosettes are also observed in glioblastoma and anaplastic astrocytoma, but appearance remains focal, while in astroblastoma these are spread all over the tumor tissue. Ependymoma also can have pseudorosettes, but arrangement of cells remains more compact and cytoplasmic processes are thin, tapering toward central vessel and often fibrillated compared to thick and blunt ended in astroblastoma cells. Blood vessels in tumor tissue show areas of hyalinization. Surgical excision of the tumor remains the mainstay of the treatment, which achieves decompression of tumor relieving raised intracranial pressure and decreases the cellular load. As these tumors generally present with raised intracranial pressure, role of surgery as a primary therapy is well justified2,3,4. In postoperative period, chemotherapy and radiotherapy have been tried in different series with limited success4,6,13. The overall prognosis of this tumor remains average with average survival being 4 years after the diagnosis in several series 4,6,8,13,14. In some patients, chemotherapy with methotrexate, vincristine, and leukovarit can have been tried successfully6. The present consensus is to do surgical excision of the patients with postoperative radiotherapy and chemotherapy with the grade of excision being the major determinant of prognosis14.

References:
Fibrolipomatous hamartoma of the digital branches of the median nerve

MM Hosain¹, MM Momen², AKM F Haque³

Abstract

Fibrolipomatous Hamartoma is an idiopathic disorder which may be related to hypertrophy of mature fat cells and fibroblasts in the epineurium. It is a rare condition which most commonly affects the median nerve. Presentation is usually before 30 years of age.

Some of them are presented with signs and symptoms of nerve entrapment and some are associated with macrodactyly (macrodystrophia lipomatosa) with gross cosmetic abnormality. MRI can be an important preoperative diagnostic tool. Biopsy and histopathology report can confirm the pathology.

This report describes a case of Fibrolipomatous Hamartoma of the digital branches of left index and middle fingers of median nerve associated with macrodactyly. A 25-years-old male patient having this problem came to Apollo Hospitals Dhaka with an abnormally large left index and middle finger. Debulking by excision of the lipomatous tissue along the digital branches of median nerve was performed. Three weeks postoperatively, the patient had no major complaints and the mass decreased in size without any motor or functional losses. Sensory function also found to be near normal with a small area of numbness at the tip of the left index finger.

Key Words: Fibrolipomatous Hamartoma, Rare benign tumour, Macrodactyly, Digital branches of Median nerve.

Introduction:

Fibrolipomatous Hamartoma is a rare benign tumour. The condition has also been designated as Fibrolipomatous nerve enlargement, Lipofibromatous Hamartoma, Lipofibroma, fibro fatty nerve enlargement and Neurolipoma. World Health Organization tumour classification describes Fibrolipomatous Hamartoma as lipomatosis of the nerves. A review of the literature showed only few cases of Fibrolipomatous Hamartoma. This rare condition most commonly affects the median nerve. Some times it may present with macrodactyly. MRI can be an important diagnostic tool. Treatment by surgery has some indications - multiple biopsies can be done to confirm the diagnosis, release of nerve entrapment if any, de-bulking - if gross cosmetic abnormality.

Patient, Methods and Results

A 25 years old man came to our plastic surgery out patient clinic with an enlarged left index and middle finger since his childhood. For the last 3 years it increased in size significantly. It was associated with paraesthesias of left index and middle finger. There was no history of trauma.

On examination, his general condition was good, all the vital parameters were normal. Local examination showed left index and middle fingers were exceptionally large and enormous. Skin color was normal. There was no visible vein or pulsation or scar mark on the surface. Local temperature was not raised with normal capillary refill. Patient could move the fingers but some restriction of movement was noted at the distal interphalangeal joint of the left index finger. Sensations of the affected fingers were less than normal. The finger swelling was soft, non tender and non compressible similar to neurofibromatosis.

Radiographic examination showed bony spur at the base of the distal phalanx of left index finger, the joints were normal. A soft tissue swelling involving the whole of the left index finger and some areas of left middle finger was noted.

Figure 1: Exceptionally large left index finger

Figure 2: Macrodactyly of left index and middle finger with normal fingers in the right side.
Fibrolipomatous hamartoma

Fine Needle Aspiration Cytology showed Benign Mesenchymal tumour.

Debulking of fingers was planned under tourniquet control. The proposed fingers were explored with Brunner's incisions with the aim to explore both the digital nerves of fingers.

After exploration, all the digital nerves of those two fingers were found enlarged and thickened with infiltrated lipomatous tissues. A diffuse yellowish lipomatous tissue was also found along and around the digital nerve fibers.

**Discussion:** Adipose tissue is a normal constituent of peripheral nerves which is usually located within the perineurium and epineurium. The intraneural lipomatous tumours are believed to arise from these normal adipose cells which are found infiltrating between the nerve bundles. It may also occur from nerve sheath. It most commonly occurs in the upper extremity of infants and children and usually affects the median nerve. In 1978, Terzis JK et al, classified benign fatty tumors of peripheral nerves into three types: well capsulated intraneuronal lipomas, diffusely infiltrating fibrofatty tumors or Lipofibromatous Hamartoma and macrodystrophia lipomatosa which is an infiltrating fibrofatty lesion with associated focal macrodactyly.

There are some significant differences between the well encapsulated and the diffusely infiltrative Fibrolipomatous Hamartoma types of intraneural lipomas. Average age at the time of appearance of well encapsulated type is 45 years with female predominance while the infiltrative type arises in a younger age group with no sexual predominance. Typically it presents in the 3rd to 4th decades of life with signs and symptoms of nerve compression in the distribution of the affected nerves. There is usually a long history of a painless mass since childhood. Patient may be aware of a painless mass for years. Patients may have signs and symptoms of nerve compression in the distribution of the affected nerve characterized by paraesthesia, motor deficit and pain. Patient may present with abnormally large finger(s) (macrodactyly). Bony abnormality may also occur.

In our patient, painless masses in the left index and middle fingers were present since childhood. He came to us in his 3rd decade, with macrodactyly (left index and middle finger) and paraesthesia (predominantly hypoesthesia) with bony deformity at the distal phalanx of left index finger.

As with all neoplasms, the exact etiology of lipoma is unknown. Some researchers consider Fibrolipomatous Hamartoma to be congenital tumour, while others believe that it is incited to grow by nerve irritation, inflammation, or prior trauma. There is no familial predisposition. About 78-96% of patients present with involvement of upper limb and there is a marked predilection for median nerve. More than 80% of Fibrolipomatous Hamartoma arises exclusively in the median nerve. Other nerves may also get involved - e.g. ulnar nerve, radial nerve, axillary nerve, musculocutaneous nerve, brachial plexus, and cranial nerve. Nerves in the lower extremity can be affected in 4-22% of patients. In 27-67% of cases Fibrolipomatous Hamartoma is associated with macrodactyly (Macrodystrophia lipomatosa) where it mostly affects the index and middle finger. Histologically, perineural and endoneural fibrosis cause thickening of the neural fascicles. The interfascicular connective tissue is infiltrated by mature fat cells. In our case, mature fat cells were found in the excised tissues. The differential diagnoses...
Fibrolipomatous hamartoma

of the digital nerves masses include intraneuronal lipoma, neurofibromatosis and vascular malformation. The presence of mature fat within the nerve virtually excludes all other diagnostic considerations except for intraneural lipoma. In case of intraneural lipoma, the fat content arises from fatty tissue within the epineurium, so this condition will present as a focal mass separate from the neural fascicles, instead of infiltrating in between and separating the neural fascicles. MRI should readily differentiate between the two entities. MRI finding e.g. coaxial-cable-like appearance on axial planes and a spaghetti-like appearance on coronal planes of the lesion is often pathognomonic and can allow a confident diagnosis, even without the need for biopsy. The treatment of Fibrolipomatous Hamartoma is controversial.

Conventional treatment involves carpal tunnel decompression (if needed) by excising the transverse carpal ligament, followed by biopsy of the enlarged nerve. This procedure has resulted in clinical improvement in 60% of patients in one study. Debulking of the tumor can also be done but may compromise the vascularity and that may affect neurological function. For complete resection, both satisfactory results and catastrophic motor and sensory deficit have been reported.

Conclusion: In conclusion, Fibrolipomatous Hamartoma is a rare benign tumour that most commonly affects the median nerve. It may present with macroactyly. In this case, it mainly affects the digital branches of left median nerve. Careful history and methodical examination is essential for its diagnosis. Local X-ray as well as MRI can help in accurate diagnosis. Final diagnosis can be achieved from histopathological reports. As the treatment is still controversial so, reports of such rare benign tumors need publication and the treatment protocol shared among the surgeons.

References:
WE INVITE ARTICLES FROM OUR ESTEEMED READERS

Please Refer to Instruction to Authors (Page 38)

Editorial Board - PULSE
Dysembryoplastic neuroepithelial tumour: A case report

Khaled A 1, Joader A 2, Chandy M 3, Nasir TA 4

Abstract

Aim and Objective: Dysembryoplastic neuroepithelial tumour is an unusual brain tumour with varied incidence commonly occurring in younger age groups. We report a 16 year old female diagnosed as a case of dysembryoplastic neuroepithelial tumour.

Clinical presentation: A 16 year old female was examined for headache and convulsion for two months. MRI revealed a brain tumor which was later confirmed as dysembryoplastic neuroepithelial tumor by histopathological examination.

Conclusion: DNET is a relatively rare brain tumor and needs differentiation from other closely resembling brain tumor because of its favorable prognosis.

Key Words: brain tumor, Dysembryoplastic neuroepithelial tumor, epilepsy.

Introduction: Dysembryoplastic neuroepithelial tumor was first proposed by Daumas-Duport in 1988. DNT is characterized as mixed neuronal-glial tumor in the current WHO classification of CNS tumors corresponding to WHO grade. This tumor demonstrates typical histological features such as glial nodules and the so-called glioneuronal element. DNTs are clinically associated with drug-resistant focal or secondary generalized seizures arising in childhood probably due to an up-regulation of several multi-drug transporters. The vast majority of these tumors have been reported in the cortex with the temporal lobe being most common. In up to one third of cases contrast enhancement in MRI is observed. Their favorable prognosis is also due to the fact that most lesions remain stable, yet rare cases with slow progression or hemorrhage due to hamartomatous vessels have been reported. Outcome after surgical resection has also been considered favorable, as the majority of children remain seizure-free. In contrast, a long history of epilepsy, older age at time of surgery and adult cases are associated with poor seizure control. Rarely, cases with multi-focal lesions or familial occurrence have been described. There are very few reports of DNTs with elevated proliferative activity that underwent transformation into malignant gliomas at a later stage.

As the lesion carries a favorable prognosis and these patients do not require radiation following surgery, it becomes very essential that this lesion should be accurately diagnosed by the surgical pathologist. Besides, there is no published report of DNET from our country so far. With this background knowledge, in this case report we describe the clinical and histo-morphological features in a patient with DNET.

Case History: A 16 year old non diabetic, normotensive female came in the neurosurgery out patient department of Apollo hospital Dhaka with the complaint of left sided headache for two months associated with decreased hearing for one month and history of convulsion 1.5 month back. She also gave history of convulsion for few times two years back. A family history of CNS malformation or neurofibromatosis was excluded. Additionally no family member had a positive history for intracranial tumor. Her Glasgow coma scale score was 15(E4V5M6). Her routine neurological examination and laboratory examination were within normal limit. MRI revealed a well defined lesion in the left temporal fossa posteriorly abutting the cranial vault and tentorium cerebelli. Brilliant enhancement noted on contrast study. After taking written informed consent, left occipito-temporal craniotomy and excision of the mass was done and specimen sent for histopathological examination. Grossly the specimen was one lobulated grayish-white piece of tissue measuring 3.5x2.5x3 cm having macrogyri like appearance. On sectioning it shows homogenosity with viscous consistency. On microscopic examination, it shows glial nodules in association with specific glioneuronal elements and foci of cortical dysplasia. The heterogeneous appearance is due to proliferation of pleomorphic cells composed of astrocytes, oligodendrocytes and neuronal elements. Areas of focal myxoid change, microcystic changes and vascular proliferation are also noted (Fig:1).

A diagnosis histomorphologically consistent with Dysembryoplastic neuroepithelial tumour was made.

1. Specialist, Department of Histopathology, Apollo Hospital Dhaka, 2. Registrar, Department of Neurosurgery, Apollo Hospital Dhaka, 3. Sr. Consultant, Department of Neurosurgery, Apollo Hospital Dhaka, 4. Sr. Consultant, Department of Histopathology, Apollo Hospital Dhaka
Dyssembryoplastic neuroepithelial

Discussion: The morphologic features of DNT consisting of a preferential cortical topography, multinodular architecture and the young age of onset, led to the hypothesis that DNT is an own tumor entity and may be derived from the secondary germlinal layers. Histopathological hallmarks are bundles of axons lined by oligodendroglia-like cells, forming columns in a pale mucoid matrix in which isolated neurons float. These so-called glioneuronal elements are observed both in simple and complex forms of DNT. The heterogeneous appearance of the latter is due to additional glial or neuronal cell populations which mimic low-grade gliomas. Our patient presented with a clinical history of focal epileptic seizures and histology showed the typical features of DNT including glioneuronal elements. Additionally, the tumor contained mainly oligodendrogial components and some astrocytic and neuronal areas in disarranged cortical layers. Thus, the diagnosis of a DNT, complex variant, WHO grade I was made.

A non-specific variant of DNT without typical glioneuronal elements has been proposed by Daumas-Dupont. These are histologically indistinguishable from certain low-grade gliomas. One case without typical glioneuronal elements but mature ganglion cells within a multinodular architecture has been reported in the literature. Because of this histopathological confusion with oligodendroglioma, low grade glioma and ganglion cell tumor, neuroradiological appearance together with clinical presentation always needs to be taken into consideration.

Most DNTs show no or a very low proliferative activity, with MIB-1 labeling indices usually being <1%. However, few cases with occasional mitotic activity and elevated MIB-1 index (28%) which is more typical for high-grade gliomas, have been reported. In addition, despite a traditionally benign course, in rare cases of elevated proliferative activity DNT might undergo transformation to malignant gliomas. One case without typical glioneuronal elements but mature ganglion cells within a multinodular architecture has been reported in the literature. Because of this histopathological confusion with oligodendroglioma, low grade glioma and ganglion cell tumor, neuroradiological appearance together with clinical presentation always needs to be taken into consideration. 21

References:

44

January 2010; Volume 4
Subscription Form – National Subscribers

No subscription fee required for doctors

I would like to subscribe regularly to PULSE the medical journal of Apollo Hospitals Dhaka. I understand no charge is required for the doctors. My particulars are:

Name: .......................................................... Designation: .............................................
Degree / Diploma:  ...
Postal Address: ..........................................................
E-mail: .......................................................... Mobile: .............................................
Signature: ..........................................................................................................................

(Photocopy Allowed)
# CME in Apollo Hospitals from July to December 2009

<table>
<thead>
<tr>
<th>SL.</th>
<th>Topic</th>
<th>Department</th>
<th>Date</th>
<th>Speaker</th>
<th>Discussant</th>
<th>Chairperson of the Session</th>
</tr>
</thead>
</table>
| 1   | a. The complexities of aortic surgery and why it is the most exciting of all adult cardiac surgeries.  
   b. Left ventricular aneurysm repair.  
   c. Surgery for ascending aortic aneurysm with aortic incompetence | Dept. of Cardiothoracic Surgery                      | 12th January-09       | Dr. Attawar Sandeep G. Sr. Consultant & Co-ordinator Dept. of CTS  
   Dr. Zulfiqur Haider. Specialist Dept. of CTS  
   Dr. Sohail Ahmed. Sr. Registrar Dept. of CTS | Dr. Syed Sakib Nazir Consultant & Coordinator Dept. of Cardiology | Prof. Dr. Md. Shahid Karim Sr. Consultant & Coordinator Dept. of Paediatric Surgery & Paediatric Urology |
| 2   | a. Advanced ENT surgeries in Bangladesh.  
   b. Cochlear Implantation, Apollo Experience | Dept of ENT                                          | 9th Feb-09            | Prof. (Dr.) Md. Zillur Rahman Sr. Consultant & Co-ordinator Dept. of ENT  
   Dr. Sanjeev Gupta. Consultant Dept. of ENT | Dr. Chandra Prakash Dokwal Sr. Consultant Dept. of Respiratory Medicine | Dr. S.M. Rezaul Islam Sr. Consultant Dept. of General & Laparoscopy Surgery |
| 3   | Management of Primary Headaches                                       | Dept. of Neurology                                   | 9th March-09          | Dr. Alim Akhter Bhuiyan Consultant & Co-ordinator Dept. of Neurology | Prof. Quazi Deen Mohammad Dept. of Neurology & Principal Dhaka Medical College | Prof. Dr. Md. Shahid Karim Sr. Consultant & Coordinator Dept. of Paediatric Surgery & Paediatric Urology |
| 4   | Polycystic Ovarian Diseases & Fertility Center                        | Dept. of Fertility Center                            | 13th April-09         | Dr. Mrinal Kumar Sarker Sr. Consultant & Co-ordinator Dept. of Obs/Gyn & IVF | Dr. A.S.M. Iqbal Sr. Consultant & Coordinator Dept. of Neonatology | Prof. Dr. Md. Shahid Karim Sr. Consultant & Coordinator Dept. of Paediatric Surgery & Paediatric Urology |
   b. Perianal Problems; AHD Experience                                | Dept. of General & Laparoscopic Surgery              | 11th May-09           | Dr. S. M Rezaul Islam Sr. Consultant, Dept. of General & Laparoscopy Surgery  
   Dr. Bidhan Chandra Das Specialist, Dept. of General & Laparoscopy Surgery | Prof. Dr. Anisur Rahman Sr. Consultant & Coordinator Dept. of General & Laparoscopy Surgery | Dr. Azizul Hasan Sr. Consultant & Coordinator Dept. of Internal Medicine |
| 6   | a. Leptospirosis.  
   b. Doctor patient communication in an Electronic Era               | Dept. of Internal Medicine                           | 8th June-09           | Dr. Md. Sadiqul Islam Consultant & Co-ordinator, Dr. Imran Rouf Assistant Professor of Medicine | Prof. Dr. Anisur Rahman Sr. Consultant & Coordinator Dept. of General & Laparoscopy Surgery | Dr. Sanjeev Gupta Consultant Dept. of ENT |

January 2010; Volume 4
Instruction to authors

Pulse is the official medical journal of Apollo Hospitals Dhaka. The aim is to ensure and maintain strong, up-to-date academic base and share updated medical knowledge, views and performances. We encourage all doctors from home and abroad to send articles to Pulse.

Papers written in English will be considered for publication provided these have not been published previously and are not under consideration for publication elsewhere.

Conditions for manuscript submission

• All manuscripts will be subjected to peer and editorial review.

• Accepted manuscripts become the property of the Pulse. Any reproduction in whole or part will require written permission from the editorial board of the journal.

• The author should obtain written permission from appropriate authority if the manuscript contains any table; data or illustration from previously published other journals. The letter of permission should be submitted with the manuscript.

• If the photographs are not disguised, the permission from the patient or parents/guardians to print should accompany the manuscript. Otherwise identity will be blackened out.

• Rejected manuscripts/electronic copies/illustrations/photographs will not be returned to the authors.

• Editors are not responsible for electronic/courier/postal failure.

Manuscript preparation

The editorial board has decided to comply with "Uniform Requirements for Manuscripts Submitted to Biomedical Journals“ published by the International Committee of Medical Journal Editors in Vancouver, British Columbia in 1979 (the widely accepted "Vancouver style") last updated in 2007 & published in Jan 2009. All scientific units should be expressed in System International (SI) units. Authors are referred to international committee of medical Journal Editors was p a g e (http://www.icmje.org/ manuscript_prepare.html) for guidance in the use of SI units. All drugs should be mentioned in their generic form.

• Original articles, reviews, special articles, case reports and any other articles of medical interest are welcome.

• Should be typed in English and on one side of A4 (290 x 210cm) size white paper, using Times New Roman font, size 12, with single space.

• There should be one original and two paper copies and one IBM compatible electronic copy.

• There should be a margin of 2.5 cm at top and bottom and at least 1 cm on either side.

• Pages should be numbered in English numerical at the upper right hand, consecutively, beginning with the title page.

Manuscripts should be submitted in the following order

• Title page

• Abstract (should include background, objective, methodology, results, conclusion in short) with key words

• Text (Introduction, Materials & Methods, Results, Discussion, Conclusion).

• Acknowledgements

• References

Photographs

• Unmounted glossy paper, 12.7x17.3 size

• Should be clipped to a white paper with appropriate labelling (number in English numerical, title of photographs and title of manuscripts.)

• May also be submitted in CD

Illustrations

• All illustrations should be cited in the text

• Illustration should be numbered in English numerical and labeled properly.

Tables

• Should not duplicate the text.

• Should be appropriately titled.

• Numbered with roman numerical in order of text.

• Abbreviations if used, should be explained in footnotes.

Placement

• All photographs, illustrations and tables should be placed in the text in their appropriate places where their description are given.

References

1. References should be indicated by superscript numbers consecutively in the text (e.g. "...has been reported1"; or as shown by Akbar2) in the order in which they are first mentioned and should be listed in numerical order on a separate sheet at the end of the article.

2. References cited only in tables or legends or illustrations should be numbered in accordance with a sequence established by the first mention in the text.

3. Titles of journals should be abbreviated according to Index Medicus or given in full.

4. References from journal must include:

(i) all authors, surnames and initials if there are 6 authors or fewer. If there are more than 6 authors, the first six
Instruction to authors

authors should be listed and "et al" (means "and others") to be added with comma and one space between each name. The last author must have a full stop after the name. Surname to be followed by 1 space and initials (no space or punctuation between initials);
(ii) the full title of the paper; no need to use italics or underlining; only the first word of title of articles are capitalised;
(iii) the abbreviated or full title of the journal followed by full stop no punctuation marks are used in the abbreviated journal name - just spaces;
(iv) the year of publication followed by semicolon;
(v) the volume number;
(vi) the issue number in brackets without any space immediately after vol no (again no space between vol no and issue no);

5. Reference from books must include:
(i) authors name as mentioned under journal above;
(ii) title of article, as under journal;
(iii) editors name/s, the word "editor" or "editors" in full after the names (avoid confusion with "ed" for edition);
(iv) Title of the book followed by full stop;
(v) edition if other than first edition. Abbreviation of word edition to "ed" followed by fullstop;
(vi) place of publication followed by colon and a space;
(vii) name of publisher, followed by semicolon and 1 space;
(viii) year of publication followed by full stop and;

6. Documents in electronic format/from internet must include:
(i) title;
(ii) authors name/s;
(iii) year of publication;
(iv) web site address;
(v) date of access. Example: What does a swollen testis mean for an 11 year old boy. Svanyo, 2009 (http://www…….), accessed on (dd/mm/year)

Manuscripts Submission: the manuscripts should be submitted to the editor / executive editors with a covering letter, mentioning that the work has not been published or submitted for publication anywhere else. (Both soft and hard copies). We are also operating to open a new pulse email address (pulse@apollodhaka.com).

Reprints for the authors: 2 copies of original journal and three copies of each article will be provided to the corresponding author free of cost.

Copyright: No part of the materials published in this journal may be reproduced, stored or transmitted without prior written permission of the editorial board.

Reference:
Apollo Hospitals Dhaka
leading the way in quality

Access to high quality health care is the mark of emerging and developed nations worldwide. Apollo Hospitals Dhaka has been one of the key leaders in the emergence and continued development of modern hospital care in Bangladesh.

STS Group recognized the need for private sector investment as a means of accelerating the development of modern health facilities. More than five years of planning, development and construction resulted in the opening of Apollo Hospitals Dhaka in 2005. The management, staff and Physicians at Apollo Hospitals Dhaka have continuously reviewed the needs of the community as a guide to the development and addition of services available in the facility. Their vision has resulted in the first and only internationally accredited super specialty hospital in Bangladesh. The Joint Commission International is an organization dedicated to ensuring rigorous standards are met by hospitals seeking accreditation.

The eleven story main hospital building is situated on 4 acres and has almost 450,000 square feet of floor space, in its 11 story structure. While designed to accommodate up to 450 inpatients as well as a wide range of outpatient services, the hospital has only opened additional beds as the need demands. The approved inpatient capacity is currently 360 beds, with additional beds planned for completion in 2011 and beyond. Plans are also being made to add a new building on the campus to accommodate rapid growth in the demand for a wide range of Tertiary Level Specialty services and treatments on an outpatient basis.

The group has also committed to developing an additional inpatient hospital. Located in Chittagong, this facility will be modeled after the successful Dhaka flagship and will serve the southeastern area of Bangladesh. While providing a broad range of services in Chittagong, the new facility will also be able to refer cases to its sister hospital in Dhaka if super-specialty care is required.

We are confident that our facilities and services are leading the way in the continued development of world class healthcare in Bangladesh.

Michael S. Potter, FACHE
Chief Executive Officer
Apollo Hospitals Dhaka
PULSE Committee Members

Chairman : Prof. Dr. Md. Shahid Karim
Co-Chairman : Dr. Mohammed Naseem Siddiq

Committee Members : Prof. Dr. Anisur Rahman
Dr. A.H.M. Waliul Islam
Dr. Chandra Prakash Dokwal
Dr. M. Quamrul Hasan
Dr. Shagufa Anwar
Dr. Simeen M Akhtar
Prof. Dr. Tareak Al Nasir

Committee Coordinator : Mr. Iqbal Hossain Howlader

Office of the Editor : Department of Paediatric Surgery & Paediatric Urology
OPD Level 5; Apollo Hospitals Dhaka
Phone: 880-2-8401661; Ext. 2525
Centers of AHD:
Apollo Bone & Joint Center
  Orthopaedics
  Rheumatology
  Physical Medicine
Apollo Heart Center
  Cardiothoracic Surgery
  Interventional Cardiology
  Pulmonology
  Cardio-Pulmonary Rehabilitation
  Cardiac ICU
Apollo Kidney & Urology Centre
  Nephrology
  Renal Transplant
  Dialysis
  Andrology
Apollo Mother & Child Health Center
  Obstetrics and Gynaecology
  Fertility Center
  Paediatrics
  Paediatric Surgery and Paediatric Urology
  Neonatology
  Neonatal ICU
Apollo Neuroscience Center
  Neurology
  Neurosurgery
  Neuro Rehabilitation Neuro ICU
Apollo Diagnostic Center
  (Imaging & Laboratory)
  Diagnostic Modalities
  Interventional Radiology
  Laboratory
  Reference Laboratory
  Blood Bank
Apollo Surgical Center
  General Surgery
  Laparoscopic Surgery
  ENT
  Plastic Surgery
  Anaesthesia Surgical ICU
  Daycare
Apollo Therapeutic Center
  Dermatology Medical Oncology
  Ophthalmology
  Dental
  Psychiatry
Apollo Emergency & Primary Care Centre
  Emergency Department
  Triage
  Master Health Check
  Internal Medicine
  Endocrinology
  Gastroenterology
  Vaccination

www.apollodhaka.com

Emergency Hotline: 10678
Appointment: (02) 8845242, 01841 APOLLO, 01729 APOLLO, 01195 APOLLO, 01612 APOLLO
Ambulance: 01714-090000 | Emergency: 01911-555555 | PABX: (02) 8401661
Plot 81, Block E, Bashundhara R/A, Dhaka-1229, Bangladesh
Info Center: Chittagong: 01713-064555
  Sylhet: 01713-047461
  Bogra: 01713-229988
  Khulna: 01713-489191
info@apollohdhaka.com
www.apollodhaka.com
Enriched Critical Care

Our critical care service is more enriched with more than 100 ICU beds

State of the art specialized personal care for every critical patient

Supervised by trained ICU doctors and nurses 24 hours a day, 7 days a week

- Central monitoring system
- Hi-tech vital sign monitor for every patient
- Invasive & non-invasive hemodynamic monitoring system
- Syringe & infusion pumps for metered medications and accurate volume infusions
- Ultramodern ventilator with monitoring facilities for patient's own breathing effort
- Instant Arterial Blood Gas analysis & electrolyte assessment
- Portable X-ray machine within ICU
- Bed side Echocardiogram and Ultra sonogram
- Intra Aortic Balloon Pump (IABP) facility for maintaining blood pressure in circulatory failure patients
- Total Parenteral Nutrition (TPN) for patients who are unable to have nutrition by mouth
- Bedside Continued Renal Replacement Therapy (CRRT) for moderate to severe kidney failure patients

The first and only Joint Commission International (JCI) Accredited hospital in Bangladesh
Simply dial and inject¹

Just a gentle push¹

30% lower dose force¹

Setting new standards with a more convenient injection

Accurate: Dose after dose, time after time²

Trust FlexPen® to deliver an accurate dose, time after time

Virtually painless NovoFine® needle ETW

Distinct colours to avoid mix ups


changing diabetes®
novo nordisk®