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• NIB Awards 2016 for House Journal: Best Content

• Golden Peacock National Quality Award 2014 in Healthcare Sector.

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• Golden Peacock International Business Excellence Award for the year 2013 initiated by Institute of Directors, United Kingdom.

• Commendation Certificate of Kerala State Government for energy conservation for the year 2012.

• TRIMA CSR award 2012, for excellence in CSR Activities undertaken for the financial years 2010-2011 and 2011-2012.

• Dr.Prathap C. Reddy Safe Care award for Best Medication Safety Initiative 2011.

• Avaya Global Connect Customer responsiveness Award 2010.

• South Asian Federation of Accountants (SAFA) award for best presented accounts and corporate governance disclosure.

• A – stable rating by CRISIL for best financial reporting in the year 2008.

• Hospital Management Asia (HMA) Award for the Project Musculo skeletal injuries in 2009.

• AV Gandhi Memorial Award 2007 and 2008 for excellence in Cardiology.

• Award for transparency in financial reporting in the year 2005 and 2008.

• Best Power User Award by Cyber India Online for optimal power utilisation in the healthcare industry in India in 2004.

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Editorial

It has been a glorious year-end for KIMS as an academic institution. KIMS has been awarded the Scroll of Honour for Teaching and Clinical Excellence NBE accredited hospital-2018. This is a national award initiated by the Association of National Board Accredited Institutions (ANBAI) and the National Board of Examinations (NBE).

Prof. Dr. G Vijayaraghavan, Vice Chairman and Director Medical Services of KIMS has been awarded the highly prestigious title of Emeritus Regional Advisor to Royal College of Physicians (RCP), Edin. by the president of RCPE in October 2018.

Some of our doctors have also received accolades in recognition of the outstanding performance and excellence in their specialities. We congratulate them for their achievement.

The Government started the Kerala Antimicrobial Resistance Strategic Action Plan (KARSAP) and KIMS is a part of it to make optimum use of antibiotic and fight antibiotic resistance and make Kerala a role model state of India.

This is an era of online health information and the physician’s role is being redefined in the era of online health information. Patients will be accessing details of diseases and medication via the internet on computers, laptops, phones, or tablets. It is therefore essential that we discuss and understand how the internet and social media are used by patients to find health information, as well as the influence that the medical profession might have on such online information. Doctors increasingly work within a culture of litigation and blame, and sometimes they can’t provide the high value care patients expect. Correct documentation of details regarding a patient and communication are very important aspects of treatment.

This edition includes reports rare cases like “a curious case of ataxia, adenoma malignum of cervix”, “Unilateral thalamic infarct: A rare presentation of deep cerebral venous thrombosis” and so on. The case report “Ruptured left parietal AVM in a child” describes a life saving emergency procedure; it is also exceptional for the surgical management, medical team support and the rehabilitation by the
interdisciplinary team of doctors. The case report and review “Is Liver Transplantation the answer to Atypical HUS in the Indian scenario’ is a unique case presentation and discussion.

There is an interesting trip down the memory lane by Dr.Kesavan Nair, Professor & Senior Consultant in Respiratory medicine. He has recorded his own experiences in Ventilator management alongwith his view points as a chest specialist, a teacher and as a person who has had a good exposure to Critical care in a tertiary care centre. He emphasises the importance of team work in managing critically ill patients and the support it calls for.

The Chief Editor has penned his concerns about the rise of a new class of practitioners of surgery in India who have mostly commercial interests rather than passion and the harm it does to this noble proession.

CMEs initiated by the Academic division in the past few months and the forthcoming ones are also highlighted. Our medical quiz continues in this edition which would be interesting and informative.

We request your support and feedback for further improvement of content.

The Editorial Board
Itinerant Surgeons
Rise of Indian Medical Oligarchs

I script this essay principally to register my disquiet about the rise of a new class of practitioners of surgery in the province of Indian Medicine. In case I choose to describe them ‘Itinerant Surgeons’, that phrase will not be an absurd misnomer since these gentlemen like pub crawlers in the British Isles tenaciously hop from one nursing home to another for their daily sustenance. Through their imperious pomposity, neo-rich traits and indecorous conduct they felicitously attract yet another suitable descriptive phrase – medical oligarchs.

Let me pen a profile of the itinerant surgeons for you. A lot of commonalities seem to conglutinate them; the common denominator being their insatiable appetite for material wealth. Evidently hailing from affluent backgrounds they embraced their pecuniary tastes early in their lives and subsequently spent considerable lucre acquiring degrees and allied appurtenance to gain consultory perch. Despite being affluent, their cultural stock and pedigree are evidently profligate with meagre suffusion of liberality and egalitarian impulses and they remain mostly destitutes of any ingrained decent traits. The description ‘desiccated tissue mechanics’ suits them most agreeably.

They are wary of interacting with their patients directly; the nursing homes or hospitals they serve recruit and pigeonhole the patients for them. The treatment strategies are gerrymandered on minimal data transmitted electronically the previous evening.

They set out of their expansive abodes early in the morning after loading the dicky of their awesome limousines with equipments and seek out their first nursing home. By the time these stalwarts assail the precincts of the hospital their patients enfeebled by anaesthesia are already deposited on the operation table ready to be savaged.

Because of mutually salutary commercial transactions companies adore them. Drug companies seek them out, tend and sustain them with unwearied and unflinching constancy and devotion. At their behest the itinerant surgeons perambulate the exotic locales, shop at Harrods and Marks & Spencers without being parsimonious and frolic in the ethereal innards of the Star Cruise. They are of course,
not averse to behold scintillating cabaret sitting in the gilded balcony of Lido or Moulin Rouge.

There is no doubt that the commercial alignment of itinerant surgeons with dubious hospitals is very toxic and there is no visible limit to the harm it might do. It liquidates many of the ordinary guarantees to the patient which our civilized society proffers. It abridge and abrogate her rights of choice and other prerogatives.

It is disconcerting that even the magisterial MCI has not registered in such defilements and pass them over with careless inattention. Other professional groups including the formidable IMA blinded by their own agendas remain supine onlookers without providing any palliation. In fact all regulatory mechanisms seem to have given the itinerant groups carte blanche.

I suppose it is time that the general public take cognizance of these distortions and depurate the sullied Indian medical landscape. The consultant-patient interaction ought to represent a transparent human relationship. It endorses human values and above all the patients’ right to know. It furthers medical equity.

Prof. K Sasidharan
Contributions for KIMS Proceedings

All faculty members of Kerala Institute of Medical Sciences in India and abroad are invited to contribute to this medical journal. Since nursing service also play a crucial role in the healthcare delivery, they are also encouraged to contribute. We welcome purely medical articles either original or already published elsewhere, case reports, CPU reports and interesting topics of discussion. Materials from our sister concerns and invited guests will be entertained.

Instructions to Authors

• Original articles: Reports of original Clinical Research. The text should be limited to 1500 words with an abstract, maximum 3 tables and 15 references.

• Case reports: Reports of interesting clinical cases. The text should be limited to 2 tables and 10 references.

• Review articles: Evidence based reviews of topics relevant to practicing doctors. It should not be a personal interpretation of the topic but a critical evaluation of the topic with current evidence included. The text should be limited to 250 words with 5 tables or figures and 25 references.

• Articles require the full name of Author/Authors, Abstract, Keywords, Introduction, Case report, References and also Name of Corresponding Author, Designation with active email id.

• All abbreviations should be expanded at first use.

• References and Images to be marked at appropriate places in the text.

• Images used in article has to be good quality. Images also to be attached as (tiff/jpeg) alongwith article.

For contributions mail to:

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Late presentation of Vesico ureteric reflux

Dr. Renu Thomas
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Department of Urology

Abstract

Primary vesicoureteral reflux (VUR) is a common congenital urinary tract abnormality in children. There is considerable controversy regarding its management. Preservation of kidney function is the main goal of treatment, which necessitates identification of patients requiring early intervention. VUR usually manifests in childhood. Late presentation of VUR is rarely reported. We report the case of a 30 year old female who reported with history of recurrent episodes of urinary tract infections since 3 years. She was evaluated and found to have grade III-IV VUR left side which was treated endoscopically with Deflux injection. There is no consensus on the optimal management of VUR or on its diagnostic procedures, treatment options, or most effective timing of treatment. By defining risk factors (family history, gender, laterality, age at presentation, presenting symptoms, VUR grade, duplication, and other voiding dysfunctions), early stratification should allow identification of patients at high potential risk of renal scarring and urinary tract infections (UTIs).

Keywords: VUR, UTI, Renal scar, MCU/VCUG, Deflux.

Introduction

VUR, or the retrograde flow of urine from the bladder into the ureter, is an anatomic and/or functional disorder with potentially serious consequences such as renal scarring, hypertension, and renal failure. Patients with VUR demonstrate a wide range of severity, and a majority of reflux patients will not develop renal scars and probably will not need any intervention¹.

The main goal in the management of patients with VUR is the preservation of kidney function by minimizing the risk of pyelonephritis. By defining and analyzing the risk factors for each patient (age, sex, reflux grade, lower urinary tract dysfunction [LUTD], anatomic abnormalities, and kidney status), it is possible to identify those patients with a potential risk of upper urinary tract infection (UTI) and renal scarring. Controversy persists over the optimal management of VUR, particularly the choice of diagnostic procedures, the treatment (medical, endoscopic, or open surgical), and the timing of treatment.

Many children with VUR have no symptoms of UTI, and invasive diagnostic procedures are performed only when clinically indicated; therefore, the exact prevalence of VUR is unknown. However, the prevalence of VUR in normal children has been estimated at 0.4–1.8%². Among infants prenatally identified by ultrasonography to have hydronephrosis and who were screened for VUR, the prevalence was 16.2% (range: 7–35%)³. Siblings of children with VUR had a 27.4% (range: 3–51%) risk of also having VUR, whereas the offspring of parents with VUR had a higher incidence, 35.7% (range: 21.2–61.4%)³.
Case report

30 year old female P1L1 reported with history of dysuria, frequency and lower abdominal pain of 1 week duration. She gave history of mild fever-on and off fever which was low grade in nature, no history of hematuria/ flank pain / pyuria/ leucorrhoea. She had similar complaints in the past also- recurrent episodes since 3 years. Last child birth was 3 years back- full term LSCS. Pregnancy was uneventful, no history of UTI. First episode of UTI developed 3 days after child birth. Patient has high grade fever with lower abdominal pain and dysuria. Urine culture was positive, treated with oral antibiotics initially. Second episode was one week later. At that time she was admitted and treated with I V antibiotics. She was better and was discharged with oral antibiotics and afterwards uroprophylaxis was started. Whenever she stopped uroprophylaxis she used to get symptoms. No co-morbidities. No history of childhood UTI/ pulmonary TB in the past. Cycles were regular. Physical examination was unremarkable and routine lab investigations were normal. Urine microscopy showed plenty of pus cells and urine culture was positive for Klebsiella. She was treated with oral antibiotics and became symptomatically better. On follow up USG abdomen was done which showed diffuse changes in parenchymal echoes and prominence of renal sinus echoes in left kidney with dilated upper calyces and minimal parenchymal thinning. It also showed scar in the interpolar region – suggestive of sequelae of chronic pyelonephritis.

So micturating cystourethrogram was done, which showed grade III-IV reflux on left side.

Fig 1: USG showing scar in the interpolar region of left kidney with calyceal dilatation

Subsequently MR Urogram was done –showed smaller left kidney with scarring in poles and interpolar region.

Fig 2: MCU shows grade III-IV reflux on left side

Fig 3: MR Urogram showing small left kidney with scarred lower pole and interpolar region

With these investigations, a diagnosis of left VUR was made. She was admitted and cystoscopy was done.
Cystoscopy showed patulous left ureteric orifice, which was not lateralized. Sub ureteric injection of Deflux (dextranomer/hyaluronic acid) was done on left side. Uroprophylaxis was given and she was discharged. She was asymptomatic at 3 weeks follow up. Uroprophylaxis was stopped-no breakthrough UTI since 3 months.

Discussion

Vesicoureteral reflux is the abnormal backflow of urine from the bladder into the ureter and up to the kidney. It is the most common problem found in children with urinary tract infections. Reflux is found in 20-50% of children who have had a urinary tract infection. Reflux is dangerous because it allows bacteria that might be in the bladder to reach the kidney. This can cause a kidney infection (pyelonephritis) which potentially can lead to kidney scarring and/or damage.

Reflux is usually diagnosed in one of two ways.

1) Children who have a culture proven urinary tract infection when they undergo contrast x-ray evaluation called a voiding cystourethrogram (VCUG)

2) A prenatal ultrasound (ultrasound during pregnancy) may reveal a fetus with dilated kidneys.

-If this occurs, a VCUG is done soon after the birth of the baby.

Etiology

Mackie and Stephens proposed that abnormal ureteric budding and/or dysfunctional interactions between the ureteric bud and metanephric mesenchyme give rise to VUR and other renal abnormalities\(^4\). Many groups have studied VUR in the context of a familial disorder because sibling recurrence, parent-child transmission, and twin concordance (monozygotic 80% to 100%, dizygotic 35% to 40%) provide good evidence for heritability.

Primary VUR is caused by insufficient submucosal length of the ureter relative to its diameter. This is precipitated by congenital defect or lack of longitudinal muscle of the portion of the ureter within the bladder. Secondary VUR is caused by increased bladder pressures associated with obstruction, which distorts the ureterovesicular junction. The obstruction may be anatomical or functional. Anatomical causes include posterior urethral valves, urethral or meatal stenosis. Functional causes include bladder instability, neurogenic bladder and non neurogenic bladder. Bladder infections may cause reflux due to elevated pressures associated with inflammation.

Investigations

VCUG (MCU)

VCUG is recommended at age 0–2 yr after the first proven febrile UTI. If reflux is diagnosed, further evaluation traditionally consists of a DMSA scan. An alternative top-down approach is also an option, as suggested by several studies in the literature. This approach carries out a DMSA scan first, close to the time of a febrile UTI, to determine the presence of pyelonephritis, which is then followed by VCUG if the DMSA scan reveals kidney involvement.

Grading of reflux\(^5\)

Grade I - Reflux does not reach the renal pelvis; varying degrees of ureteral dilatation

Grade II - Reflux reaches the renal pelvis; no dilatation of the collecting system; normal fornices

Grade III - Mild or moderate dilatation of the ureter, with or without kinking; moderate dilatation of the collecting system; normal or minimally deformed fornices
Grade IV - Moderate dilatation of the ureter with or without kinking; moderate dilatation of the collecting system; blunt fornices, but impressions of the papillae still visible

Grade V - Gross dilatation and kinking of the ureter, marked dilatation of the collecting system; papillary impressions no longer visible; intraparenchymal reflux

The degree of reflux is used to assist in decision making with regard to treating the reflux and the ultimate prognosis of patients. More severe grades of reflux are associated with lower rates of spontaneous resolution and higher incidence of renal damage if not treated.

Other investigations

1) Kidney/Bladder Sonogram (Ultrasound): routinely recommended prior to the VCUG. This test is done to outline the kidneys, ureters and bladder. It looks for additional less common urinary tract defects that can be the cause of the urinary tract infection or kidney dilation.

2) Kidney (Renal) Scan: This test may be done if the above tests are abnormal or if repeated febrile infections have occurred. It is used to better demonstrate the actual function and/or drainage of the kidneys. A kidney scan can also show if there is kidney damage and/or scarring that may have resulted from a previous urinary tract infection.

3) Nuclear Cystogram: This test is very similar to the VCUG; however, it has less radiation and is very sensitive for reflux.

Treatment

There are two different management plans (conservative vs surgical) for reflux. These are based upon the degree of reflux, the age of the child at the time of diagnosis, the number and severity of urinary tract infections, and the amount of damage to the kidneys seen on X-ray studies.

Conservative treatment

Medical therapy is based on the knowledge that most reflux will resolve on its own as the child grows. It requires low dose daily antibiotics taken by mouth. An ultrasound and cystogram will be done on a yearly basis to assess the kidneys and to assess if the reflux has resolved. If the reflux persists for several years without change in the grade then surgery may be considered. If child continues to have febrile urinary tract infections despite being on antibiotics, then surgery should also be considered. Again, the goal is to prevent potential scarring and/or damage from each pyelonephritis episode. Resolution is nearly 80% in VUR grade I–II and 30–50% in VUR grade III–V within 4–5 yr of follow-up. Spontaneous resolution is low for bilateral high-grade reflux. The conservative approach includes watchful waiting, intermittent antibiotic prophylaxis or continuous antibiotic prophylaxis (CAP), and bladder rehabilitation in patients with LUTD (lower urinary tract dysfunction).

Surgical treatment

Surgical management is reserved for children with higher grades of reflux, febrile urinary tract infections despite being on antibiotics, and signs of renal damage on renal scans due to repeated infections. Surgery may also be discussed in cases when, after repeated VCUG’s and allowing for the growth of the child, the reflux does not appear improving. The goal of surgery is to cure reflux and avoid kidney damage. There are a few surgical options. These include an “open” operation, endoscopic injection and robotic/laparoscopic surgery.

1. Subureteral injection of bulking materials

- Injected bulking agent elevates the ureteral orifice and the distal ureter so that coaptation is increased.
- The lumen is thus narrowed, which prevents reflux of urine into the ureter while still allowing the urine’s antegrade flow.

2. Open surgical techniques

- Various intravesical and extravesical techniques have been described for the surgical correction of VUR i.e ureteric reimplantation
3. Laparoscopy

He or she will still need to take daily antibiotics following the surgery until the bladder and ureter are healed. An ultrasound will be performed approximately one month following surgery and depending on each case, a VCUG will be performed six months following surgery.

Resolution of secondary VUR will usually occur if the precipitating cause is treated or resolved.

Are there any risk factors for reflux?

- Specific modes of transmission are not known, but there is a very high incidence of reflux among siblings.
- Approximately 40% of siblings of those who have reflux will also have reflux.
- Younger siblings are at a much greater risk than older siblings.

In many of these affected siblings, there is no documented history of symptomatic urinary tract infections. Younger siblings of refluxers should be screened either with a VCUG or nuclear cystogram.

References


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A case of Hemophagocytic Lymphohistiocytosis

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Abstract

Hemophagocytic Lymphohistiocytosis is a disorder of cell mediated immunity. The disease can present with a wide range of symptoms and outcomes, with hyperinflammatory response leading to organ damage. HLH poses a diagnostic challenge among clinicians. The treatment of HLH is based on HLH 94 protocol – consisting of anti-inflammatory and cytotoxic agents. This is a case report of a young female presenting with a multitude of clinical features, who got diagnosed as having HLH.

Keywords: Hemophagocytic Lymphohistiocytosis, HLH, Cell mediated immunity.

Hemophagocytic Lymphohistiocytosis (HLH) is a disorder of cell mediated immunity. HLH can be genetic or acquired. Normally, an antigenic stimulation causes release of various cytokines (IF-γ, TNF-α, GM-CSF). These cytokines stimulates proliferation cytotoxic T-lymphocytes (CTLs), macrophages and NK cells. Cell mediated immunity functions in two pathways – Fas ligand Pathway and Perforin / Granzyme Pathway (Cytotoxic protein Pathway). The latter pathway is affected in HLH. The Perforin / Granzyme Pathway consists of five main steps:

1. Packaging of cytotoxic granules – Immature granule formation (perforin and granzyme)
2. Transport – Granule maturation and docking on microtubules
3. Fusion with the membrane - Granule exocytosis
4. In the Target cell:
   a. Formation of perforin-lined pore
   b. Granzyme enters the target cell through the pores
   c. Apoptosis

Apoptosis results in removal of antigenic stimulation and hence termination of inflammatory response. If any of the five steps in the perforin / granzyme pathway gets affected, apoptosis will not occur, and hence there will be persistence of antigenic stimulation, resulting in hyperinflammatory response and hypercytokininemia.

Hypercytokininemia results in:

1. Increased proliferation and tissue infiltration of macrophages (histiocytes) and Proliferation of Cytotoxic T-lymphocytes (Lymphohistiocytosis)
2. Chronic stimulation of Macrophages → Ingestion of non-physiologic quantities of cells, including blood cells (Hemophagocytosis)
3. Supress hematopoiesis and induce apoptosis in hematopoietic cells → Pancytopenia
4. Inhibit lipoprotein lipase → Elevated Triglycerides
5. Activated macrophages → secrete plasminogen activator → High plasmin levels, hyperfibrinolysis and Hypofibrinogenemia
6. Increased levels of heme-oxygenase → Elevated Ferritin levels
Infiltration of spleen by activated lymphocytes and histiocytes → Splenomegaly
Infiltration of liver by activated lymphocytes and histiocytes → Hepatomegaly with Transaminitis
IL-1 and IL-6 produced by activated macrophages → Fever

Various genetic variants of HLH are based on what step of perforin / granzyme pathway is affected:

- Packaging of cytotoxic granules - FHL 2
- Transport of granules – Hermansky-Pudlak Syndrome Type 2 & Griscelli Syndrome Type 2
- Granule exocytosis – FHL 3, FHL 4, FHL 5
- Perforin-lined pores on target cell – X-linked lymphoproliferative disorder
- Granzyme enters the target cell – FHL 2

The chromosomal involvement in various genetic variants of HLH are as follows:

- FHL 1 → Chr 9
- FHL 2 → PFR1 gene; Chr 10 (Most Common)
- FHL 3 → UNC13D gene; Chr 17
- FHL 4 → STX11 gene; Chr 6
- FHL 5 → STXB2 gene; Chr 19
- Griscelli Syndrome Type 2 → RAB27A gene; Chr 15
- Chediak-Higashi Syndrome → LYST gene; Chr 1
- Hermansky-Pudlak Syndrome Type 2 → AP3B1 gene; Chr 5
- XLP Type 1 → SHD2D1A gene; Chr X
- XLP Type 2 → BIRC4 gene; Chr X

The causes of acquired HLH are:

1. Infection associated
   1. Viral (esp EBV), Bacterial, Fungal, Parasitic
   2. Malignancy associated
   1. Hematologic → Lymphoma, Leukemia, Multiple Myeloma

2. Non-hematologic → Lung / Prostate / HCC
3. Autoimmune disease associated
   1. Systemic-onset juvenile idiopathic arthritis
   2. Kawasaki disease
   3. SLE
4. Seronegative spondyloarthropathies

Case report
A 25 years old female from Kanyakumari District, with history of on and off fever of six months duration, along with loss of appetite and loss of weight, and multiple hospitalizations for symptoms of fever, cough and breathlessness. The current admission complaints started as fever, associated with dry cough and breathlessness. Chest x-ray showed bilateral infiltrates. She was initially treated on OP basis from a local hospital with Ciprofloxacin, Doxycycline and anti-malarials. Five days later, since fever and breathlessness persisted, and chest x-ray showed features of ARDS, she got admitted to the local hospital. On evaluation at the local hospital, she was found to have low WBC counts and platelet counts along with mild transaminitis. She was treated with Inj Meropenam, Doxycycline and Methyl Prednisolone. All the routine fever work-ups were negative. After 2 days of admission, fever subsided, but breathlessness progressively worsened, and hence referred to KIMS Trivandrum for further management.

On Day 1 of admission to KIMS, she required NIV support, hemodynamically stable and physical examination revealed hepatosplenomegaly. Initial labs showed low WBC and platelet counts, elevated liver enzymes, and a normal renal function test. Procalcitonin was not significantly raised. Chest X-ray showed bilateral infiltrates, ABG showed type 1 respiratory failure, and Echocardiography showed normal cardiac function and no evidence of vegetations. On Day 2, the patient had worsening of respiratory function, and was intubated and
mechanically ventilated. WBC and platelet counts dropped further. Ferritin level was elevated.

The issues were prolonged febrile illness, ARDS, hepatosplenomegaly, bicytopenia and transaminitis.

The possibilities were a) Infection, b) Autoimmune disorder, c) Malignancy, d) Other rare disease like HLH.

Evaluation for infectious diseases did not yield any positive result, and the usual causative agents were ruled out. Autoimmune workup was also negative. The next step was to perform CT imaging of chest and abdomen, peripheral blood film and bone marrow aspiration study.

CT revealed bronchopneumonia / ARDS, hepatosplenomegaly, mild ascites and mild right pleural effusion. Peripheral blood film showed anisocytosis, leucopenia and thrombocytopenia.

Bone marrow aspiration study showed reactive changes with macrophage proliferation and evidence of hemophagocytosis. Thus the diagnosis of HLH was considered. The HLH diagnostic criteria (2004) needed 5 out of the 8 parameters to be present, for a diagnosis of HLH. In this case, 6 parameters were present – Fever, splenomegaly, cytopenias, hypertriglyceridemia, hemophagocytosis in bone marrow and hyperferritinemia – and hence the diagnosis of HLH was confirmed. EBV quantitative PCR showed a high EBV viral load – hence the causative association with HLH was established.

The patient was initiated on IV Dexamethasone according to HLH 94 protocol. The blood counts and respiratory function progressively improved, and the patient was extubated on Day 6 following admission. But, on Day 8, the patient had worsening of liver function and blood counts, and was initiated on IV Etoposide. On Day 9, the patient had worsening respiratory function, and was reintubated and mechanically ventilated. She had progressive hemodynamic worsening and a few hours later, the patient succumbed to death.

Discussion

HLH is a disorder with wide range of causes, symptoms and outcomes. It leads to hyper inflammatory response and organ damage. Most typical presentation is fever, hepatosplenomegaly and cytopenias. Other manifestations include hypertriglyceridemia, coagulopathy with hypofibrinogenemia, liver dysfunction, elevated levels of ferritin and transaminases, neurological symptoms – may be associated with a CSF hyperproteinemia and a moderate pleocytosis, pulmonary involvement and gastrointestinal symptoms.

Diagnosis of HLH requires at least one of the following:\^2:

- Molecular diagnosis consistent with HLH – PRF1, UNC13D, STXBP1, RAB27A, STX11, SHD1A or XIAP
- Diagnostic criteria for HLH fulfilled (atleast 5 out of 8)

HLH Diagnostic Criteria 2004 (atleast 5 out of 8)\^2

1. Fever
2. Splenomegaly
3. Cytopenias affecting at least two of the three lineages in peripheral blood (Hb < 90 g/L; Platelets < 100 x 10^9; Neutrophils < 1 x 10^9/L)
4. Hypertriglyceridemia (\(\geq 265\) mg/dl) and/or hypofibrinogenemia (\(\leq 1.5\) g/L)
5. Hemophagocytosis in bone marrow, spleen or lymph nodes
6. Low or absent NK-cell activity
7. Hyperferritinemia (\(\geq 500\) mcg/L)
8. High levels of sIL-2r (sCD25) (\(\geq 2400\) U/ml)

The components of HLH management includes:\^2:

- Supportive measures for life threatening presentation
- Elimination of triggers (mainly infection) to remove the stimuli that initiate abnormal immune system activation
- Suppression of inflammatory response by
immunosuppressives; suppression of cell proliferation (neoplasia) by cytotoxic drugs

Start therapy in genetically verified disease, Familial Hemophagocytic Lymphohistiocytosis (FHL), or if the disease is severe, persistent or recurrent.

The treatment of HLH is based on the HLH 94 protocol6,7,8:

Initial Therapy:
- Dexamethasone 10 mg/m²/day for 2 weeks, followed by 5 mg/m²/day for 2 weeks, 2.5 mg/m²/day for 2 weeks, & 1.25 mg/m²/day for 2 weeks
- Etoposide 150 mg/m² BSA for twice weekly for 2 weeks, and then weekly

Continuation Therapy:
- Dexamethasone pulses 10mg/m³ for 3 days every 2nd week
- Etoposide 150 mg/m³ every alternating 2nd week
- Cyclosporin A daily

Other modalities:
- Intrathecal Methotrexate – maximum 4 doses (neurological symptoms)
- Bone Marrow Transplantation

References

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Unilateral thalamic infarct: A rare presentation of deep cerebral venous thrombosis

Dr. Deepak Menon
Dr. Manoj K S

Introduction
Deep cerebral venous thrombosis (DCVT) remains a very rare entity among the spectrum of cerebral venous thrombosis (CVT). Due to the bilateral draining territories, DCVT nearly invariably causes bilateral infarction with predictably dismal prognosis. However, rare instances of DCVT with unilateral infarction having favourable prognosis have been described, but pose wide range of differentials to the clinician, and require careful interpretation of clinical and radiological features for accurate diagnosis. Here, we report two unusual cases of DCVT with unilateral thalamic infarction with excellent outcome. We also report a rare case of CVT, with simultaneous deep and cortical vein thrombosis. Through a relevant review of literature, we also examine the clinical presentations of unilateral infarction due to DCVT and their outcomes.

Case 1
A 43 year old lady presented to the ER with three days symptoms of diffuse mild to moderate headache which subsided by next day with paracetamol. Since then, she was noted to have dragging of right leg and clumsiness of right hand in the form of fumbling and occasional dropping of objects. She became withdrawn, tending to remain in bed with monosyllabic but appropriate responses and needed coaxing for activities including self care. Her history was also prominent for previous menorrhagia. At admission she remained apathetic and dull but had no aphasia or dysarthria. She had right UMN facial palsy with subtle right sided pyramidal signs and mild weakness of right upper limb and lower limbs. The clinical localisation was over left frontal cortical or subcortical region with likely etiologies including ADEM, cerebral cortical venous thrombosis or intracranial space occupying lesions.

Her MRI revealed left thalamic hyperintensity with mild mass effect along with left caudate and left lentiform nucleus hyperintensities sparing internal capsule. There were separate left superior frontal gyrus and left centrum semiovale hyperintensities. MIP and MRV images showed evidence for internal cerebral vein, vein of Galen and cortical vein thrombosis over the left frontal convexity. Diffusion and ADC sequences revealed patchy restriction for both thalamic and frontal cortical infarct suggesting mixed vasogenic and cytotoxic edema.

Her labs meanwhile showed severe iron deficiency anemia with hemoglobin of 5.8mg/dl and ferritin of 5.6ng/ml. Ultrasonogram of pelvis revealed multiple subserous fibroid. Evaluation for a vasculitic, infective or another procoagulant process was negative. Meanwhile lumbar puncture was done which revealed an opening pressure of 150mm of CSF with protein of 106mg/dl, with no cells, normal glucose levels and absent oligoclonal bands. She was initiated on anticoagulation and her anemia was corrected by parental iron and packed red cell transfusion with plan for elective hysterectomy. At one month review patient has become completely asymptomatic and has resumed her normal routine.
Case 2

A 22 year old lady presented with rather abrupt onset of holocranial predominantly back of scalp severe headache since past two days with bouts of vomiting. The pain persisted with only temporary relief with analgesics and was worse on lying down. There were no other symptoms at presentation and her examination did not reveal any neurological abnormality. She had in fact presented with nearly similar throbbing holocranial headache along with neck pain and bouts of vomiting and had been diagnosed with extensive thrombosis involving superior and left transverse sinuses but with no parenchymal changes. She had been on anticoagulation for six months since and her subsequent procoagulant work up had been negative. Her follow-up MRI with MRV had revealed near complete resolution with establishment of flow in superior and left transverse sinus.

Her current MRI showed left medial thalamic hyperintensity with no diffusion restriction with contrast MRV showing complete nonvisualisation of straight sinus, vein of gallein and deep cerebral veins as well as partial occlusion of posterior part of superior sagittal sinus. She was re-initiated on anticoagulation with plan for indefinite continuation of anticoagulation. Extensive evaluation including repeat procoagulant work up before initiation of anticoagulation failed to reveal an etiology. By discharge patient was completely asymptomatic with no focal neurological deficits.

Discussion

The deep venous system comprises of 1: paired internal cerebral veins which are midline structures, the main tributary being thalamostriate veins, choroidal veins and anterior septal veins 2: The basal vein of Rosenthal formed from tributaries on the medial surface and temporal horn of the temporal lobe and 3: Great vein of Galen formed by confluence of the two internal cerebral veins and the two basal veins just posterior and superior to the pineal gland. Anatomic variations from this classic confluence are however common.

In the international multicentric study on CVT, deep venous system thrombosis constituted 10.9% of all cases of CVT. Yousay had noted the rarity of unilateral internal cerebral vein thrombosis and the asymmetrical thalamic and basal ganglia changes thereof. Due to the symmetrical draining territories, infarctions associated with deep venous system infarction is usually bilateral, often affecting the thalami, corpus striati and adjacent white matter structures and upper midbrain. The presence of deep venous system thrombosis predicted a poor outcome in terms with hazard ratio of 2.9. However the degree of damage depends on the extent of thrombosis and efficiency of draining collaterals. Accordingly, partial DCVT syndrome, with remarkably good prognosis has been described rarely in literature. Despite deep venous system thrombosis, against the norm, asymmetrical involvement of unilateral thalamus and deep structures are seen in these cases. The details of clinical presentations, radiological features, differentials and outcome of the reported cases of unilateral DCVT infarction is summarised Table 1. In nearly all the reported cases in literature the infarction has been on the left side, the reason for which remains unclear. It may be that a more common anatomic variant predisposes the left sided system, especially the left internal cerebral vein with insufficient collateral venous drainage of thalamus, leading to left thalamic infarction. However this conjecture has not been proven in any of the reports and DSA evidence has been lacking. As has been previously proposed, the left sided infarction may be clinically more manifest as opposed to the nondominant side, again this remains in the realm of speculations. The current two cases capture the spectrum of DCVT highlighting the clinical features. As has been reported in previous reports, both the cases in our series had left sided infarction.
The diagnosis of unilateral deep venous infarct is not easy considering its clinical rarity and wide range radiological possibilities. In most cases the differential considered was that of a thalamic glioma or an abscess. In our first case, the possibility of acute demyelinating encephalomyelitis was also considered among other differentials and lumbar puncture was done before anticoagulation which showed elevated protein. In fact nearly a third of the patients in the multicentric series of CVT had undergone lumbar puncture. It may also have been employed for documenting the elevated CSF pressures which can be an isolated presentation of CVT. The second case in our series was easier to suspect with history of previous cortical vein thrombosis, though she had presented with isolated headache alone during both her episodes.

In DCVT, the triad of subacute onset of headache, mild to marked alteration of sensorium with hemiparesis have been the predominant clinical feature. It requires a careful scrutiny for absence of normal flow voids of deep cerebral veins along with use of venogram would confirm the diagnosis.

Of the very few reported cases in literature the outcome of unilateral deep venous system infarction has been good as opposed to deep CVT in general. It may refer to the that end of the spectrum were by the disease has been captured in the initial stages and initiation of anticoagulation has been done prior to further propagation of thrombosis and more extensive infarction leading to further downhill cascade of events.

The first case the current series is even more unique in that in addition to the deep venous system there was simultaneous ipsilateral superficial cortical vein thrombosis over left superior frontal gyrus. Combined superficial and deep cortical venous infarct has been reported in a single case series by Kumral et al however the imaging characteristics were not highlighted. All the patients with DCVT in addition to superficial venous thrombosis had headache and alteration of consciousness and the outcome remained poor compared to isolated superficial cortical vein thrombosis. Etiologically there were no differences between the groups vis a vis isolated superficial venous thrombosis and combined deep and superficial thrombosis. Though deep venous system infarction and combined superficial and deep system infarction has been a pointer for poor prognosis, our patient had an excellent outcome.

The etiological evaluation of CVT in the first case revealed no other cause than severe iron deficiency anemia. Coutinho reported a linear, inverse association between the risk of CVT and hemoglobin concentration. In their case control study after adjusting for all confounders anemia was associated with a fourfold increased risk of CVT and the risk was most for microcytic anemia. The possible pathogenic mechanisms have been postulated to be associated thrombocytosis, which we found in our patient, as well as elevated concentrations of factor VIII.

Both our patients could be started on anticoagulation within twenty four hours of presentation. With unilateral thalamic lesion, close attention has to be given to the radiological findings. Venous thrombosis leads to ischemia which appears as hyperintensities on T2 images with corresponding hypointensities unless hemorrhagic transformation of infarct has occurred which can be readily picked up in susceptibility weighted or gradient images or by CT scan. As opposed to arterial infarcts, these lesions, having a combination of cytotoxic and vasogenic edema will have patchy diffusion restriction in diffusion weighted images. While phase contrast or time-of-flight MR angiogram can highlight the nonvisualisation of vessels T1 and T2 images can be reveal the absence of normal venous flow voids and can aid in diagnosis.

The current case series highlights the varied presentation of DCVT. The presence of unilateral infarct does not rule out deep venous system involvement. The clinical presentation of subacute onset headache, alteration of sensorium and hemiparesis in the presence of unilateral
Table 1: Summary of the reported cases of unilateral infarct with deep cerebral venous thrombosis

<table>
<thead>
<tr>
<th>Author</th>
<th>No:</th>
<th>Age/sex</th>
<th>Presentation</th>
<th>Duration</th>
<th>Site</th>
<th>Vessel involved</th>
<th>DD</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yousry(3) (2002)</td>
<td>1</td>
<td>47/F</td>
<td>Headache, hemiparesis, retrograde amnesia</td>
<td>2 weeks</td>
<td>Left thalamus</td>
<td>Left ICV</td>
<td>-</td>
<td>Complete recovery</td>
</tr>
<tr>
<td>Van den Bergh(7) (2005)</td>
<td>4</td>
<td>30/F</td>
<td>Headache, hemianopia, drowsiness, seizure</td>
<td>2 weeks</td>
<td>Left thalamus</td>
<td>ICV, vein of Galen, Left TS and SS</td>
<td>Thalamic tumour</td>
<td>Residual headache, fatigue</td>
</tr>
<tr>
<td></td>
<td></td>
<td>28/F</td>
<td>Right hemiparesis</td>
<td>1 day</td>
<td>Left thalamus</td>
<td>Thalamic tumour</td>
<td>-</td>
<td>Mild hemiparesis, dysphasia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>53/F</td>
<td>Headache, aphasia, right hemiparesis</td>
<td>3 days</td>
<td>Left thalamus</td>
<td>Partial thrombosis of the straight sinus and the left internal cerebral vein</td>
<td>Thalamic tumour</td>
<td>-</td>
</tr>
<tr>
<td>Weishmann (8) (2008)</td>
<td>14/F</td>
<td>14/F</td>
<td>Headache, vomiting, drowsiness, aphasia, right sided weakness</td>
<td>3 days</td>
<td>Left thalamus</td>
<td>ICV, straight sinus, vein of galen</td>
<td>Tumour, biopsy done</td>
<td>Mild weakness, memory impairment</td>
</tr>
<tr>
<td>Hwang(9) (2012)</td>
<td>36/F</td>
<td>36/F</td>
<td>Headache, dysarthria, right hemiparesis</td>
<td>1 week</td>
<td>Left thalamus</td>
<td>Left ICV and thalamostriate vein</td>
<td>-</td>
<td>Mild headache</td>
</tr>
<tr>
<td>Deshpande (10) (2014)</td>
<td>31/M</td>
<td>31/M</td>
<td>Headache, vomiting, status epilepticus</td>
<td>4 days</td>
<td>Right thalamus</td>
<td>ICV, vein of Galen, SS</td>
<td>None</td>
<td>Memory impairment</td>
</tr>
<tr>
<td>Current series</td>
<td>2</td>
<td>43/F</td>
<td>Headache, apathy, right hemiparesis</td>
<td>3 days</td>
<td>Left thalamus</td>
<td>Left ICV, vein of Galen and cortical vein</td>
<td>ADEM</td>
<td>Complete recovery</td>
</tr>
<tr>
<td></td>
<td></td>
<td>23/F</td>
<td>Headache</td>
<td>2 days</td>
<td>Left thalamus</td>
<td>ICV, Vein of Galen, straight sinus, vein of gallen, SSS</td>
<td>-</td>
<td>Complete recovery</td>
</tr>
</tbody>
</table>

Thalamic lesion should raise the possibility of venous infarct. Delay in institution of treatment has far reaching consequences, as further extension of thrombosis can lead to bilateral infarction with significantly higher risk of mortality and long term morbidity.
Table 2: Relevant investigation results of patient in case 1

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>5.8mg/dl</td>
</tr>
<tr>
<td>MCV</td>
<td>76.4ul</td>
</tr>
<tr>
<td>Serum Ferritin</td>
<td>5.6ng/dl</td>
</tr>
<tr>
<td>Serum Iron</td>
<td>7.4ng/dl</td>
</tr>
<tr>
<td>Peripheral smear</td>
<td>Severe hypochromic microcytic anemia</td>
</tr>
<tr>
<td>Reticulocyte count</td>
<td>1.12</td>
</tr>
<tr>
<td>Vitamin B12</td>
<td>&gt;2000pg/ml</td>
</tr>
<tr>
<td>D dimer</td>
<td>884</td>
</tr>
<tr>
<td>ANA</td>
<td>Negative</td>
</tr>
<tr>
<td>APLA IgG and IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>LDH</td>
<td>112mg/dl</td>
</tr>
</tbody>
</table>

References


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Abstract

Arteriovenous malformations (AVMs) in children are relatively rare and represent 3% of all AVMs\textsuperscript{1,2}. The rate of rupture of paediatric AVM is high compared to adults and most of them are detected only after rupture. The AVMs incidentally diagnosed during evaluation in children, carry lifelong risk of bleeding and development of deficits. The microsurgical resection remains gold standard treatment for all AVMs, especially in cases of acute intracranial haemorrhage. The management of intracranial AVMs is still controversial. There is a possibility of re-rupture from the smallest remnant of an AVM even after surgical resection. Apart from conservative treatment and follow up in children without clinical symptoms, a multimodality approach is preferred in treating unruptured AVMs in recent days like Endovascular embolization, Open microsurgical resection and Gamma knife radiosurgery. Here, we present a successfully treated ruptured left parietal AVM in an 8 year old girl.

Keywords: Arteriovenous malformations, Microsurgery, Endovascular embolization, Gamma knife radiosurgery.

Introduction

In normal course, the arteries carry oxygenated blood from the heart to brain, supplies oxygen to brain parenchyma through capillary system and the veins carry blood with less oxygen from the brain to the heart. In AVMs, instead of a capillary system, a tangle of blood vessels bypasses normal brain tissue and diverts blood from arteries directly to the veins(Fig 1). An AVM has 3 components: the feeding arteries, nidus and draining veins. The presence of a single or multiple direct arteriovenous connections permit high flow arteriovenous shunting through small feeding arteries that lack a muscular layer and the absence of a capillary bed. The ‘weakened’ blood vessels dilate over time and may burst from the high pressure of blood flow from the arteries, causing bleeding into the brain which may be fatal depending upon the size, location and mass effect of the haematoma. The synonyms used previously to describe the arteriovenous malformations include Angioma arteriale racemosum (Virchow, 1863), Varix aneurysmaticus (Steinheil, 1895), Arteriovenous angiomas and anomalies.
Case report

An 8 year old girl was brought to the emergency room with history of sudden onset of headache followed by right sided weakness and unresponsiveness. She was initially treated at a local hospital where the MRI brain (Fig.2,3) showed acute left frontoparietal haematoma with intraventricular extension, mass effect and midline shift, suspecting an aneurysmal bleed. She was intubated for low GCS and referred for further treatment.

On examination: Intubated patient on ventilator with sedation and relaxant. Pupils: Right 2 mm, reacting to light and Left 4.5 mm, Not reacting. The heart rate: 52/minute, regular and the BP: 120 systolic. GRBS: 178 mg/dL. The cardiovascular and respiratory systems were clinically normal and no significant intra-abdominal pathology. A Digital Subtraction Angiography (DSA) could not be done due to pupillary asymmetry and bradycardia. The bedside ECHO showed normal study. Hb: 8.9 gms% and INR: 1.12.

The CT Brain and Cerebral angiogram (Fig.4,5,6,7,8,9,10,11) showed acute intraparenchymal haematoma in left high frontoparietal region extending into centrum semiovale and corona radiata measuring 4.1 x 3.6 x 4.8 cms with perilesional oedema. Mass effect in the form of effaced left lateral ventricle and midline shift of 6.8 mms towards right. Dependent blood densities in bilateral occipital horns of lateral ventricles extending to third ventricle. Diffuse effacement of sulci and cisterns suggestive of cerebral oedema. A nodular (Fig.12) intensely enhancing space occupying lesion seen superior and
Fig. 7  CT Brain Coronal view

Fig. 8  CTA

Fig. 9  CTA

Fig. 10  CTA

Fig. 11  CTA

Fig. 12  CTA
lateral to the intraparenchymal haematoma measuring 6.6 x 5.8 x 5.6 mms with arterial feeder coming from superior division of left middle cerebral artery. Small cortical draining vein arising from the superior border, seen joining the superior sagittal sinus. No obvious nidus seen.

She was admitted in Paediatric Intensive Care Unit (PICU) and continued on ventilator. Following evaluation and consent from the parents, she underwent an emergency left frontoparietal craniotomy, excision of left parietal arteriovenous malformation (Spetzler-Martin Grade II) and evacuation of haematoma.

**Surgical procedure**

Under GA, following craniotomy, the AVM was seen with prominent vein surfacing on the left parietal lobe. Circumferential excision of AVM (Fig.13) was done with coagulation of feeding arteries followed by evacuation of haematoma. Haemostasis achieved and re-confirmed. The brain was lax and pulsating well on closure. A lax duroplasty was done and bone flap was replaced.

The Histopathological examination reported as brain tissue with thick walled and dilated blood vessels of varying calibre filled with RBCs. Arterialisation of vessels noted with areas of haemorrhage in between - consistent with arteriovenous malformation.

She was reviewed in the OPD after one month, with normal speech and totally improved left ptosis, right hemiparesis 4+/5, ambulant without support (Fig.15) and on oral feeds.

A DSA was done which showed no evidence of residual arteriovenous malformation or aneurysm(Fig.16,17).
Discussion

Eventhough the developing nervous system in children is vulnerable to injury, it has the ability to rapidly adapt by

assigning functions to alternate areas of the brain. This phenomenon is known as 'Neural Plasticity' and allows children with AVMs to recover more quickly than adults after surgery.

Pathogenesis:

The cerebral AVMs are primarily congenital and most malformations occur during the third week of embryogenesis (Fig.18), either the persistence of primitive AV connection or development of a new connection after normal closure process.

Another concept focuses at a physiological trigger on susceptible vascular cellular elements which could be mechanical, hormonal, haemodynamic, thrombotic, thermal, ischaemic or inflammatory.

Vascular Endothelial Growth Factor (VEGF) is significantly high in patients with AVMs.

Patients with Hereditary Haemorrhagic Telangiectasia (HHT) have an increased chance of having AVMs and also elevated levels of VEGF. Release of VEGF also occurs from platelets in hypoxia, haemorrhage and thrombogenesis and infections.

Periventricular nodular hyperplasia in filamin 1 gene mutation have a role in neuronal migrational disorders and vascular malformations.

Clinical presentation:

The average age of initial diagnosis of AVM is about 30 to 40 years, more common in males and cases were reported in families.

The common clinical presentation in children with ruptured AVM include seizures, severe headache, muscular weakness or numbness in one part of the body depending upon the location, impairment of vision, slurring speech, altered sensorium and unsteadiness.
The complications in cases of AVMs are haemorrhage, reduced oxygen to brain tissue, thin or weak blood vessels developing into an aneurysm-susceptible to rupture, brain damage and hydrocephalus.

The AVMs are the most common cause of non-traumatic intracerebral haemorrhage in children. Computed Tomography (CT) with angiography is helpful in evaluating the location and size of the haematoma and the AVM. Magnetic Resonance Imaging (MRI) with angiography is useful in better localization of an AVM and planning of treatment. However, the early post rupture MR angiography, due to the compression by haematoma, might miss some important components of the AVM.

Treatment:

**Conservative management**

The consideration of conservative management and long term follow up in incidentally diagnosed, unruptured and smaller size AVMs in children have the risk of increase in size, rupture and development of neurological deficits. The technological improvement in microsurgical resection, endovascular embolization and radiosurgery has made the multimodality approach and complete cure easier.

**Surgical resection**

The complete microsurgical resection remains the gold standard of AVM treatment, especially in ruptured AVMs with haematoma and the AVMs in superficial and non-eloquent areas. The surgery has the advantages of immediate cure and evacuation of haematoma. The surgical resection alone can be used primarily in treating AVMs or combined with pre-operative endovascular embolization or radiosurgery, if needed, as a multimodality approach with complete cure and minimal morbidity. Kiris et al., reported 20 paediatric AVMs of Spetzler-Martin grade 1-3 treated surgically with an 89% radiographic obliteration rate, 5% morbidity and 5% mortality. The complications of microsurgical resection of AVM include haemorrhage which produces hypovolemic shock in children, that can be prevented by preoperative embolization and brain tissue damage during surgery can be prevented by preoperative functional MRI with intra-operative stereotactic guidance. The surgical microscope-integrated intraoperative angiography with intra-arterial injection of Indo Cyanine Green (ICG) (Fig.19) has a better phase contrast and spatial resolution than DSA and helpful in cerebral AVM surgery. A careful postoperative monitoring prevents complications like hyperperfusion, seizures, vasospasm, vascular thrombosis and stroke.

**Gamma knife Radiosurgery**

Altschular et al. in 1989, first used stereotactic radiosurgery in paediatric AVMs. Exposing the paediatric brain to radiation therapy was the reason behind the delay in using radiosurgery. The indications for using radiosurgery in children are the deep seated AVMs-not easily accessible to surgery and the AVMs in eloquent cortex. Gamma knife radiosurgery provides a non-invasive option that delivers high dose radiation directly to the abnormality while sparing the surrounding brain tissue from unnecessary injury. Several studies were done regarding safety and efficiency of radiosurgery in children. Yen et al., reported a series of 139 children with 80 months follow up after radiosurgery, in children of the age group of 6 to 222 months which showed an obliteration rate of 59%, which was increased to 69% who underwent multiple sessions.
Endovascular embolization

The Endovascular embolization is useful in complete obliteration in smaller AVMs and as an adjunctive therapy with microsurgical resection and radiosurgery in children. The faster technological developments in the field of endovascular treatment led to more utilization to cure smaller AVMs and staged embolization in the treatment of larger AVMs. A study conducted in a series of 1246 patients of cerebral AVMs showed complete obliteration after embolization in only 5%. The Endovascular embolization plays a significant role in the multimodality treatment of cerebral AVMs as an adjunctive therapy with other treatment modalities.

History

1853 / 1854: Arteriovenous malformation described by Luschka and Virchow.

1889: Giordano performed the first surgical exposure of an AVM and Krause was the first to attempt surgical elimination of an AVM by ligating its arterial feeders, without great success.

1932: First successful resection of a cerebellar AVM was done by Olivecrona (Fig. 20) and Riives.

1960: First description of embolization reported by Luessenhop and Spence.

1976: Yasargil et al published their first series of 10 AVM patients, successfully treated surgically with no mortality and minimal morbidity.

1986: Spetzler and Martin classification was published and the grading included the size, location and venous drainage of an AVM. The Spetzler-Martin grading system is used to assess the risk of neurological deficits after surgical resection. Eventhough there are many classifications and modifications seen throughout the literature, the Spetzler-Martin 5 point grading system is used widely till date.

1987: Yasargil was the first to suggest that AVMs are dynamic in nature (The initial description of an AVM is a simple static abnormal connection between the arterial and venous systems).

Conclusion

The main aim in the treatment of AVMs is to achieve a complete angiographic obliteration with minimal morbidity. A multidisciplinary approach involving a vascular neurosurgeon, an interventional neuroradiologist and a radiation oncologist is considered useful in the treatment of AVMs of all age groups. The effects of long standing ionizing radiation on the developing nervous system, chances of intracranial malignancy and neuropsychological retardation need detailed studies in future.

Acknowledgement

Thanks to the following teams for the excellent cooperation in the treatment of this child with Ruptured Left parietal Arterio Venous Malformation.

Paediatric ICU team:
Dr. Prameela Joji (Deputy Medical Superintendent and Consultant-PICU)
Dr. Neetu Gupta (Consultant-PICU)
Dr. Hridya (Paediatric surgery)
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3. De Novo cerebral arteriovenous malformations: case report and literature review

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Medical Quiz

Patient was on treatment for allergic contact dermatitis. The rash flared-up on discontinuing the topical application. What is the diagnosis?

Check your diagnosis > P 67 <
Abstract

Atypical HUS is disorder of complement dysregulation causing complement mediated damage resulting in haemolytic anaemia, thrombocytopenia and renal failure in susceptible individuals. With the advent of eculizumab, this disease seemed to have been cured. However, the duration of therapy with eculizumab is not established and the cost and availability of the drug a major deterrent in clinical practice. Liver transplant offers an alternative by replacing the offending organ by a healthy liver capable of synthesizing normal complement proteins and thereby effecting a complete cure to the disease. Here we present the case of a 26 yr old lady with complement factor H mutation mediated atypical HUS who underwent liver transplant for atypical HUS. Post transplant she is being followed up and has no evidence of disease activity during follow up for the last 6 months.

Keywords: Atypical HUS, Liver transplantation, Complement dysregulation

Introduction

Atypical HUS is a disease characterized by haemolytic anaemia, thrombocytopenia and renal failure. Haematological manifestations can lead to life threatening bleeding manifestations and severe anaemia. If untreated, this disease leads to End stage renal disease. Renal transplantation in this setting is not advocated as recurrence rates are very high and is recommended only in patients with genetic mutations proven to have low rates of recurrence such as MCP mutation. Plasmapheresis has been the therapy of choice for many years in atypical HUS as it filters out the defective complement and replaces fresh normal complement. Guidelines issued in 2009 recommended initial daily plasma exchange (50–70 ml/kg) with further titration of frequency according to clinical response. However for many patients, lifelong plasma exchange is difficult as vascular access may be a problem and patients may not tolerate the process of plasmapheresis. The introduction of eculizumab, a C5a blocker was a magical drug effecting a near cure of this disease, however the drug has to be taken lifelong which financially is not an option for most patients. Liver transplantation is an alternative and can effectively cure the disease.

Case report

27 yr old lady, with no pre morbidities conceived spontaneously and was undergoing regular ANCs at her local hospital. She had no complications till term and then underwent LSCS for antepartum haemorrhage. She was discharged on antibiotics and NSAIDs. On 8th post partum day, she developed altered sensorium, loose watery stools, no dysentery. Subsequently she developed anasarca and oliguria. She was evaluated at her local hospital. She was found to have a drop in haemoglobin from 12gm/dl at discharge to 4.3gm/dl. Platelet count was
1.5 lakh and serum creatinine Cr 11.2 mg/dl. On further evaluation, peripheral smear was suggestive of microangiopathic haemolytic anaemia, haptoglobin was low, stool culture was negative for Shigella and E Coli. She was transfused packed RBC with Haemodialysis support. On a clinical suspicion of atypical HUS, blood sent for Complement factor H Antibody and she was started on therapeutic plasma exchange. She required alternate day plasma exchange and haemodialysis with fluid removal as well as Packed RBC transfusions. Despite aggressive ultrafiltration, she had recurrent pulmonary edema and one episode of cardiopulmonary arrest requiring mechanical ventilation. Cardiac evaluation including ECHO showed fair LV function at the time. She continued to have recurrent Pulmonary edema. Repeat ECHO showed LV systolic and diastolic dysfunction and she was started on digoxin and hydralazine. LVEF improved to 40%. As she was not tolerating plasma exchange, she was started on Injection Eculizumab. Three weekly doses of eculizumab 900mg each were given. With this drug, she rapidly improved and was able to be maintained off haemodialysis with good urinary output at a creatinine of around 3 mg/dl. Genetic analysis showed mutation in CF H variant c.1792A>Cp (Lys598Gln). Antibody to Complement factor H was negative. One month after the last dose of eculizumab, she was again re admitted with acute pancreatitis possibly due to recurrence of the primary disease. With the subsequent fourth dose of eculizumab, all her symptoms again subsided. However she was requiring the same drug every month for disease flare evidenced by rising creatinine and schistocyte count. As she was not able to afford this treatment, she was planned for liver transplantation as an alternative cost effective treatment. She underwent liver transplantation with her husband as non blood related donor. She was given induction with methylprednisolone. Initially ductal anastamosis was tried however cholangiogram failed to demonstrate post sectoral duct and so hepato jejunostomy was done. She was transfused plasma during the procedure as per the consensus group protocol. On table renal biopsy showed TMA with 10% IFTA. Post operatively she underwent one prophylactic plasmapheresis on Post OP day 2. She however had oliguria and fluid overload for which she required Haemodialysis and fluid removal. She was started on triple drug immunosuppression with steroids, tacrolimus and mycophenolate mofetil. She had episodes of LFT derangement in the immediate post operative period however repeated liver biopsies did not show any evidence of rejection. CMV done was negative. She had one episode of GTCS with clinical and imaging suggestive of PRES. With Strict Blood pressure control and optimum diuretic prescription, she was asymptomatic and discharged at serum creatinine of 1.4mg/dl. She has been on follow up for the past 6 months. She had one episode of allograft rejection with low tacrolimus levels which was successfully managed with steroids. She also had one episode of E Coli sepsis treated with antibiotics during which creatinine transiently increased to 1.5mg/dl. There has not been any evidence of disease activity during follow up as evidenced by normal haemoglobin, renal function and platelet count.

**Discussion**

Although Eculizumab seems to be a magical solution for atypical HUS, it is an expensive drug and is not so easy to procure. In India till date there is only one case report of eculizumab being used. Also there is no data as to when the drug can be stopped. Pediatric guidelines have recommended stopping eculizumab once titre of anti CFH antibody Ab are less than 1000AU/ml in patients with complement factor H antibody. But others require lifelong eculizumab which effectively works at suppression of manifestation of the disease. On the other hand liver transplantation results in replacement of complement as opposed to blockage of complement action. Hence it offers a more complete solution in principle. Of course the morbidity and mortality of a liver transplantation is far not always so simple. But in centers geared for
liver transplantation, the risks may be more acceptable and definitely more affordable then eculizumab. The concept of liver transplantation for this disease is as old as the early 2000 where early desperate attempts were not successful.6,7 However these attempts showed that the concept can cure the disease and based on these early attempts and further successful transplantations6,9,10, the Consensus group came out with guidelines regarding the peri operative care for these patients.6 The main modification emphasized in the guidelines was large plasma exchange prior to transplantation and further plasma supplementation during transplantation. The rationale behind this method being an increase the amount of functional factor H until the graft begins normal function as well as removal of defunct factor H. With these modifications, successful liver transplantations have been done for atypical HUS and the follow up series by Saland et al shows a favourable outcome of 16 surviving graft out of 20 transplantations with most deaths related to vascular failure rather than disease recurrence.11 Kidney transplantation alone on the other hand is known to have a high rate of recurrence especially some genetic variants.12 Only one cohort was published showing good results with patients undergoing live renal transplantations without use of eculizumab or liver transplantation. Another controversial decision is whether to do combined liver-kidney transplantation or liver transplantation alone. In this case, since on table biopsy showed only 10% IFTA, the renal prognosis was predictable good. On follow up the patient has a serum creatinine of 1mg/dl and has normal urine output. Hence liver transplantation prior to development of ESRD is optimum and prevents the requirement of a renal transplantation.

Conclusion: Liver transplantation is an effective and complete cure for complement mediated atypical HUS and should be considered in patients with vascular complications not able to continue plasmapheresis and who are not able to afford complement 5 blocker eculizumab.

References


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Abstract
Adenoma malignum of the cervix is a rare variant of adenocarcinoma of uterine cervix. Its overall incidence rate is low. We report a case of 57 yr old multipara, postmenopausal, who presented with 6 month history of profuse vaginal discharge. Pelvic examination shows uterus 8-10 weeks size, Cervix hard, flushed up with walls and Scarring of lateral fornices
Transvaginal Sonography shows Uteres bulky 10.4 X 7.3 cm with homogenous myometrial echoes. Endometrial cavity shows fluid collection measuring 5.8 X 6.4 cm. Small hyperechoic lesion seen, just above the internal Os, measures 5mm. Cervical punch biopsy done- HPE suspicious of Adenoma Malignum, confirmed with IHC
Keywords: Adenoma Malignum, Minimal Deviation Adenocarcinoma, CA Cervix

Case Report
Jaya Sheela, 57 yr old, Married for 37 yrs, postmenopausal since 7 years. c/o profuse discharge per vaginum with dull abdominal pain since 6 months. Consulted @ GG hospital - said to have hydrometra -D and C done (Feb 2018) - said to be normal histopathology report. After that discharge decreased. Now c/o increased discharge pv. brownish in colour, mucoid, since 2 weeks, No foul smell, no itching Obstetric history- P2L2,1ST–FTND… (33 yrs back), 2ND–CS-sterilised LCB- 28 Years back. Past H/o-DM on OHA since 6 months, hypothyroidism on thyronorm 25mcg since 4 months. Family H-Mother CA thyroid
On examination, No pallor, no oedema, no lymphadenopathy, no clubbing
P/A–Obese, soft P/S-cervix not visualised properly, pulled up (Recent pap smear made available – negative hence not repeated again)
on pervaginal examination--uterus 8-10 weeks size, Cervix flushed up with walls - felt hard, Scarring of lateral fornices felt
Transvaginal Sonography-Uteres bulky 10.4 X 7.3 cm with homogenous myometrial echoes. Endometrial cavity shows fluid collection measuring 5.8 X 6.4 cm. Small hyperechoic lesion seen, just above the internal Os, measures 5mm.
Posted for FC Biopsy – Even though recent Biopsy report available. Pre anaesthetic panel done – it was normal.
examination shows multiple whitish tissue fragments aggregate, measuring 2 X 2 X 0.8 cm. Microscopic examination shows fragments of cervix partially lined by squamous epithelium with Koilocytosis. Endocervix is ulcerated and shows few irregular glands lined by low columnar epithelium with regular nuclei. These glands seen deep in cervical stroma. Diagnosis – suspicious of Adenoma Malignum, advised IHC to confirm. IHC stains done – results consistent with adenoma malignum

Referred to surgical oncology for 2nd opinion. Diagnosis – carcinoma cervix- minimal deviation adenocarcinoma/adenoma malignum. Advice - MRI Abdomen and pelvis, it was done on 5/8/2018. Entire cervical stroma shows altered signal with loss of normal T2 hypo intensity, replaced by poorly marginated heterogenous T2 mild hyperintense signals causing marked cervical canal stenosis. Outer cervical contour is irregular, nodular with circumferential stranding in parametral fat – likely early parametrial invasion. Circumferential thickening in vaginal fornix, thickening in upper third of anterior vaginal wall contiguous with abnormal signal intensity in cervix – invasion. A few bilateral mildly enlarged external iliac lymph


Biopsy sample sent for histopathology-Macroscopis
nodes - Stage T2B N1 Mx. Markedly distended endometrial cavity with altered signals, hyperintense in T2, iso to hyperintense in T1 – blood /mucin. Small T1 hypo intense nodular focus in inferior aspect of endometrial cavity, no enhancement- clot.

**Surgical oncology findings** - Since it is clinical evaluation (p/v-bulky and nodular cervical mass felt with bilateral
parametrial involvement not reaching to pelvic side walls. Anterior vaginal fornix involvement +). Diagnosis is stage II B – Unlikely to be suitable for radical surgery. Hence referred to radiation oncologist. Patient wants to take further treatment from RCC hence opted out to RCC.

Discussion

Essential features of adenoma malignum- 1 to 3 % of endocervical adenocarcinoma. It is very rare and easily misdiagnosed. Diagnostic challenge due to benign appearing histological characteristics. Consider in patients presenting with heavy vaginal discharge, cystic lesion on imaging, and atypical glandular cells on cytologic smear. Associated with Peutz-Jeghers syndrome with mutations in STK11 gene.

Pathology- Multiple irregular lobulations of distorted glands demonstrating a ‘Hairpin’ shape - pathological feature. Microscopy – Glands irregular in size and shape, lined predominantly by mucin containing columnar epithelial cells with basal nuclei.

Radiology description- Diagnosis with MRI and ultrasonography is often difficult due to benign appearance. Transvaginal sonography can detect presence of multilocular cystic masses in the cervix and may aid in diagnosis. Ultrasound imaging with Doppler examination is more efficient and accurate. MDA has increased intralesional vascularity. MRI: multiple irregular cystic lesions, cysts arranged in floret-like manner with aggregates of small cysts resulting in a “cosmos pattern”

Differential diagnosis- 1-Deep Nabothian cysts. 2. Tunnel cluster-multicystic dilatation of endocervical glands occasionally with mucoid discharge found exclusively during pregnancy can persist for variable period of time. 3- Endocervical glandular hyperplasia in OC pills users. It is seen in 10% of cases with Peutz Jeghers Syndrome. Literature search has suggested this variant is difficult to diagnose pre-operatively. cytology is insufficient to make diagnosis. Hence a deep incision biopsy or conisation is needed to make a preoperative diagnosis. Here we succeed in making pre operative diagnosis.

Management- Surgery is considered to be standard treatment when possible. Though there is no guidelines for what surgery is appropriate, it is treated as based on pathological stage as with other cervical carcinomas. It is essential to make appropriate diagnosis of adenoma malignum before posting for definitive surgery as suboptimal surgery could increase chance of recurrence and residual disease.
Conclusion

In this case we managed to diagnose case preoperatively, and with multidisciplinary efforts we were able to reach diagnosis and we have framed definitive treatment for this case. Persistent gynecological symptoms of unexplained origin should be referred to tertiary hospital with gynecological oncology service before symptom relief treatment is delivered. Since Adenoma Malignum is a rare variant and awareness can help to diagnose more cases, we need to be aware that there are several benign conditions which may have similar features and obtaining diagnosis of adenoma malignum by fine aspiration and cytology may be inconclusive. Since gynecological oncology services are continually developing new diagnostic tests and treatment technologies while improving existing ones, multiple treatment options can be offered even in case of incidental findings in a short frame time.

References


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A curious case of ataxia

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Abstract

We present the case of a 1 year 9-month old child who presented with acute onset of ataxia, which later progressed to Opsoclonus Myoclonus Ataxia Syndrome. This is presented in order to reinforce the fact that neuroblastoma has to be considered in the differential diagnosis of a child with any unusual presentation. It can present with symptoms pertaining to any system. If recognized early and in a favourable stage, outcome can be favourable.

Keywords: Ataxia, Opsoclonus, Myoclonus, Paraneoplastic, Neuroblastoma, Ganglioneuroblastoma

Introduction

Ataxia can be defined as a disturbance in the smooth, accurate coordination of movements. It is most commonly manifested as an unsteady gait.1

Ataxia is usually the result of cerebellar dysfunction. However, disturbances at many levels of the nervous system can also affect coordination.2 As an example, ataxia that develops as the result of loss of sensory function (such as proprioception) would be described as a sensory ataxia.

Acute ataxia is not a frequent problem in young children. Majority of cases with acute ataxia have a benign self-limited process. Though serious life threatening conditions like mass lesions and central nervous system (CNS) infection can occur. A focused history, specific physical findings and selected investigations can identify most causes of ataxia, especially those requiring immediate attention, stabilization and intervention.

Conditions that cause acute ataxia include acute infections, post-infectious inflammatory conditions, toxins, tumors, and trauma. Acute ataxia in association with opsoclonus-myoclonus (rapid, dancing eye movements and rhythmic jerking) may be the presenting manifestation of an occult neuroblastoma. As many as half of children with opsoclonus-myoclonus syndrome have a neuroblastoma.3

Opsoclonus-myoclonus syndrome (OMS) is a rare disorder that affects the nervous system. Symptoms include rapid, multi-directional eye movements (opsoclonus), quick, involuntary muscle jerks (myoclonus), uncoordinated movement (ataxia), irritability, and sleep disturbance. The onset of OMS is usually abrupt and often severe. The disease may become chronic. OMS typically occurs in association with tumors (neuroblastomas), or following a viral or bacterial infection. Treatment may include corticosteroids or ACTH (adrenocorticotropic hormone). When there is a tumor present, treatment may include chemotherapy, surgery, and/or radiation. In some cases, when the underlying cause of OMS is treated, symptoms improve.

Case Report

Ours is the case of a 1 year and 9-month old child who was developmentally normal, immunized up to age,
thrusting well born to a non-consanguineous couple who
developed acute onset of limb ataxia, later progressing
to involve the trunk over a period of 3 days. Child had
un URI 2-3 weeks ago. There was no history of trauma,
 inadvertent ingestion of drugs/toxins, no headache,
vomiting, symptoms s/o muscle weakness/cranial nerve
involvement/bowel or bladder symptoms.

Considering the possibility of a post infectious
cerbellitis/demelination child was managed with IVlg/
Methylprednisolone. MRI (plain with contrast) of brain
with a screening of spine was taken to look for any
evidence of demyelination. Imaging was normal except for
an incidentally detected presacral mass. By next day child
was noted to have spontaneous, arrhythmic, conjugate
saccades occurring in all directions of gaze without a
saccadic interval – opsoclonus. MRI plain with contrast
of pelvis was taken after consulting with neurologist
and pediatric surgeon. It revealed a contrast enhancing
presacral mass -? neuroblastoma. After explaining the
nature of the illness and the possible complications during
and post-surgery, child was operated through a posterior
approach. The mass was excised in toto, biopsy of
which revealed ganglioneuroblastoma. A CT chest/bone
marrow aspiration with biopsy was done for staging the
disease which were normal. Child was initiated on oral
prednisolone therapy (2 mg/kg/day). Irritability decreased,
truncal and limb ataxia and opsoclonus reduced in 2 days,
post-surgery. Child is on oral prednisolone therapy, under
regular follow up, opsoclonus and ataxia subsided.

Discussion

Opsoclonus-myoclonus syndrome (OMS), also known
as opsoclonus myoclonus ataxia, is a syndrome that
includes opsoclonus along with diffuse or focal body
myoclonus and truncal titubation with or without ataxia
and other cerebellar signs.

- Opsoclonus is a disorder of ocular motility
  characterized by spontaneous, arrhythmic, conjugate
  saccades occurring in all directions of gaze without a
  saccadic interval.

- Myoclonus is a clinical sign that is characterized by
  brief, shock-like, involuntary movements caused by
  muscular contractions or inhibitions.

OMS affects mainly young children with a mean age of
1.5 to 2 years. Some series, but not others, have found
a higher prevalence in girls. A female predominance is
characteristic of most, but not all, autoimmune disorders.

The most common malignancy associated with OMS in
children is neuroblastoma. Almost 50 percent of children
with OMS have an underlying neuroblastoma, and in turn,
around 2 percent of children with neuroblastoma
develop paraneoplastic OMS. While neuroblastoma is
somewhat more common among boys than girls, the
associated OMS is more common in girls.

Nonparaneoplastic OMS in children is believed to have a
parainfectious origin in some cases. Implicated pathogens
include hepatitis C, Lyme disease, Epstein-Barr virus,
post-streptococcal infection, HIV (possibly with immune
reconstitution inflammatory syndrome), Coxsackie B3,
Mycoplasma pneumoniae, and Rotavirus. In idiopathic
cases of OMS, a post-viral origin is sometimes inferred
based upon a suggestive history of a viral prodrome. One
case of OMS has been reported in the setting of celiac
disease (an immune-mediated inflammatory disorder of
the small intestine).

There are no controlled studies of treatment in pediatric
OMS. Removal of the neuroblastoma, when present, does
not appear to improve neurologic symptoms.

Observational studies suggest that OMS in
children may respond to immunologic treatment (eg, glucocorticoids, ACTH, plasma exchange,
intravenous immunoglobulin, azathioprine,
cyclophosphamide, mycophenolate mofetil often
used in combination. Examples of treatment regimens
include prednisone 2 mg/kg per day with a prolonged taper
over several months and pulsed dexamethasone therapy
using 20 mg/m² for three days, given monthly.
One protocol describes using ACTH for 40 weeks, beginning with 75 international units/m\(^2\) twice a day for one week and subsequent tapers based upon clinical response\(^9\). Rituximab added to other immunotherapies has been reported effective in small case series.\(^9\)

In one nonrandomized comparison of immunotherapies, blinded rater assessments suggested that corticotropin therapy was more efficacious than glucocorticoid therapy and that combination therapies (corticotropin with rituximab and/or cyclophosphamide) appeared to be more effective than corticotropin alone, although also associated with a higher rate of complications.\(^10\) Multicenter clinical trials of immunotherapies in pediatric OMS are in progress.\(^3\)

The neurologic prognosis is guarded. Among different case series, motor symptoms appear to improve or resolve in approximately 60 percent during initial treatment.\(^11\) Neurologic relapses may occur during steroid withdrawal or with intercurrent infections requiring prolonged treatment in some cases.\(^8\) However, regardless of pathogenesis, approximately 60 to 80 percent of patients have residual behavioral abnormalities or psychomotor retardation that sometimes become increasingly problematic later in life.\(^6\) Early and more intensive treatment does not clearly improve neuropsychiatric disability, which can occur even while motor symptoms have resolved.\(^12\) Sleep problems and associated rage attacks may respond to trazodone.\(^13\)

Tumors in children with paraneoplastic OMS appear to have a better prognosis than tumors in patients without paraneoplastic symptoms.\(^14\)

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Medical Quiz

This child was on long term treatment for recurrent itchy rash on the face. What is the diagnosis?

Check your diagnosis > P 67 <
Atraumatic clostridial myonecrosis in an immunocompetent host

Dr. Harikrishnan S
Dr. Deepak V
Dr. Suresh Kumar V K

Abstract

Background: Necrotizing fasciitis is usually associated with a surgical or traumatic wound. Clostridial myonecrosis is an uncommon but deadly infection that can develop in the absence of a wound and is often associated with occult gastrointestinal cancer or immunocompromise, or both.

We report a case of catastrophic atraumatic Clostridium septicum infection in an immunocompetent host. This case reminds physicians that patients can present with spontaneous gas gangrene due to C. septicum. Providers should consider this diagnosis in immunocompromised patients who present with acute onset of severe atraumatic limb pain.

Keywords: Infections in immunocompromised patients, Limb pain, Necrotizing fasciitis, Occult gastrointestinal cancer.

Introduction

Necrotizing fasciitis is an uncommon but frequently considered diagnosis in the emergency department (ED). It typically presents with severe pain, skin changes, and signs of sepsis in a patient with an open wound. Among patients with immune compromise, colonic carcinoma, diverticulitis, gastrointestinal operation, or irradiation, clostridial myonecrosis must also be considered. We present a case of catastrophic atraumatic Clostridium septicum infection.

Case report

A 62 YR old male, alcoholic, reformed smoker with no other known comorbidities presented to kims emergency room at 9 30 am with complaints of pain Lt gluteal region and upper part of Left thigh since morning. On initial examination, the patient was clinically stable with stable hemodynamics and an unremarkable general and physical examination. Local examination revealed no external signs of inflammation viz redness/tenderness. A hard woody induration could be felt on the affected limb up to thigh. On physical examination, the patient appeared to be in severe distress, rolling from side to side on the emergency room cot. He was alert and able to provide his own history but deferred most details to his wife. His blood pressure was 140/94 mm Hg, heart rate was 106 beats/min, respiratory rate was 18 breaths/min, oxygen saturation was 97% on room air, and temperature was 37.6°C orally. His skin was warm but diaphoretic and pale. His neck was supple, and heart and lung examinations were normal, with the exception of tachycardia. The patient’s abdominal examination was unremarkable, including a normal genital examination. He had no scrotal swelling, erythema, or lesions. No costovertebral angle tenderness and no particular point tenderness were found over the spinous processes or paraspinal musculature of the back. Examination of the right hip and leg was essentially normal. The pain could not be exacerbated by
particular movements or palpation. The skin was normal in appearance with no lesions, rash, or erythema. It was not hot to the touch and the compartments were soft. He had full range of motion in the hip, knee, and ankle joints. Toes were warm with brisk capillary refill, and his dorsalis pedis pulse was palpable. Laboratory evaluation included a negative urine analysis and normal electrolytes and creatinine levels. The complete blood cell count was notable for hemoglobin of 11.2 g/dL and white blood cell count of 1100, with an absolute neutrophil count of 400. The severity of pain with known hip and lumbar spine metastases raised concern for pathologic fracture or neuropathic pain. A plain radiograph study of the pelvis showed right iliac wing sclerosis that appeared more prominent than in previous images, but no fracture was seen. Plain radiograph studies of the lumbar spine showed sclerotic changes at L4 and L5. After discussion with the orthopaedic and internal medicine teams, the patient was admitted for pain control with a plan for advanced imaging of his spine and pelvis.

By 11 30 am, patient was still in severe pain. Internal medicine advised to rule out peripheral occlusive arterial disease/aortic dissection/gluteal/perianal access and advised to do further imaging and decide pain persisting and escalated; another shot of inj fentanyl 100mcg given and decided to expedite MRI. By 3 45 pm, the patient was in severe pain, was restless, agitated in the MRI suite, unable to lie supine and still and only screening MRI could be taken. By 6 pm, in v/o increased WOB-tachypnoea, O2 req, it was decided to shift to MDICU care. Patient received in MDICU 2 by about 6 30 pm, MRI prov report suggestive of myositis involving hamstring muscles. ABG showed severe metabolic acidosis, and in v/o severe met acidosis, inc WOB, mechanically intubated after securing IT radial artery access-pc mode piperacillin/tazobactam, doxycycline started by 7 30 pm. Patient was in hypotension, central venous access obtained, inotropes initiated and by now wound was showing blebs, meropenam loaded citing hypotension and worsening hemodynamics. By 8 pm, patient was on triple inotropic support went into bradycardia, arrest, code blue was initiated and in spite of best resuscitative efforts, patient was pronounced dead by 8 40 pm.

Discussion

Clostridial myonecrosis (also referred to as gas gangrene) is a fulminant skeletal muscle infection caused by toxin-producing clostridia, associated with gas production in the muscle, not subcutaneously, and crepitus. Gas gangrene is historically associated with battlefield wounds, but now it is most often seen in civilian traumatic or surgical wounds. The most common cause of clostridial myonecrosis is Clostridium perfringens. C. septicum infection is a rare and dramatic disease and accounts for only approximately 1% of all reported clostridial infections. Predisposing host factors include colonic carcinoma, diverticulitis, gastrointestinal surgery, leukemia, lymphoproliferative disorders, cancer chemotherapy, radiation therapy, and acquired immune deficiencies syndrome. Stevens et al. cite that “acquired neutropenia is also strongly associated with an increased incidence of spontaneous gas gangrene due to C. septicum, and in such cases, necrotizing enterocolitis, cecitis, or distal ileitis are commonly found.” Clostridium species manufacture exotoxins that lead to rapid breakdown of connective tissue, hemolysis, and platelet aggregation. They are believed to also inhibit local inflammatory responses. C. septicum is 300 times more aerotolerant than C. perfringens, which is potentially why it is able to cause tissue destruction in healthy, nontraumatized tissues. It is thought to be an opportunistic invader with a short incubation of between 6 h and 3 days. The first symptom of infection may be confusion, followed by abrupt onset of excruciating pain. Initial presentation may be subtle because patients often do not have fever and are usually normotensive until late in the infection course. Tachycardia is common, but fever and hypotension are late findings. The classic sign of crepitus most often associated with gas gangrene is commonly

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a late finding as well. Skin examination often appears normal initially, but progresses rapidly to reddish-purple discoloration with serosanguinous bullae formation. The mortality rate of spontaneous gangrene ranges from 67% to 100%, with most deaths occurring within 24 h of onset. Treatment of clostridial myonecrosis includes aggressive resuscitation, broad surgical debridement, and prompt antibiotic administration. Penicillin is still recommended as first-line therapy, in combination with a protein synthesis–inhibiting antibiotic, such as clindamycin. In some medical institutions, hyperbaric oxygen therapy can be considered in geographic locations where it is immediately available as an adjunct to usual therapy. We theorize that he might have had an occult gastrointestinal malignancy and that combined with his more than occasional indulgences in alcohol with resultant compromised immunity might have provided the right environment for gastrointestinal invasion and hematologic spread of C. septicum. As with most previously reported cases, this unusual infection spread rapidly and led to death. Our case underscores the unusual presentation of this deadly disease.

References

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Abstract

Subacute combined degeneration (SCD) refers to the gradually progressive myelopathic or myeloneuropathic presentation of vitamin B12 deficiency. While polyneuropathy has been well recognised with vitamin B12 deficiency, it has been debated whether neuropathy can manifest in isolation without myelopathy. SCD would seldom pose as a clinical or electrophysiological mimicker of Guillain-Barre syndrome (GBS). We describe the case of a middle age gentleman who presented with two weeks of rapidly progressive sensory predominant neuropathic syndrome with conduction findings consistent with a demyelinating process. In view of certain clinical-electrical discrepancies patient was further imaged with MRI revealing features classical of SCD with nearly undetectable Vitamin B12 levels. Upper gastrointestinal endoscopy as part of evaluation revealed a polyp in body of stomach with histopathological diagnosis of WHO II neurogastroendocrine tumour (carcinoid tumour) along with chronic atrophic gastritis. As far as to our knowledge, the association between gastric carcinoid and SCD have not been reported. We also discuss the relevant clinical points of this GBS-like presentation of SCD and possible pathomechanisms of the association with carcinoid. With parenteral Vitamin B12 supplementation patient had near complete resolution of symptoms and improvement in conduction parameters on follow-up.

Keywords: Guillain Barre Syndrome; Subacute combined degeneration; Carcinoid tumour; Myelopathy

Introduction

The neurological presentation of vitamin B12 deficiency can be varied, but most identifiable is as subacute progressive posterolateral myelopathic syndrome aptly named sub acute combined degeneration of spinal cord (SCD) or as a myeloneuropathy. While polyneuropathy has been well recognised with Vitamin B12 deficiency, it has been debated whether neuropathy can manifest in isolation without myelopathy1. With a gradually progressive course, subacute combined degeneration would seldom pose a clinical or electrophysiological differential to Guillain Barre Syndrome (GBS). Through the current report, we describe a case of SCD, mimicking GBS, with further workup revealing a hitherto unreported association with gastric carcinoid tumour.

Case report

This 52 year old gentleman was referred to us with rapidly progressing neurological symptoms and presented thirteen days after symptom onset. He initially noted tingling paresthesia which he likens to “electric shock” and “pins and needles” over tips of all fingers bilaterally. The next day he noticed similar sensation over bilateral toes as well but of lesser intensity. The progression and severity
were more for upper limb and the paresthesia ascended up to elbow and followed by the same sensations over legs till knee. One week later he noted distal hand clumsiness in the form of difficulty in picking notes from pocket, dialing mobile phones, buttoning shirts and using fingers for handling food items. At the same time he also noted loosening of footwear without awareness and difficulty insinuating feet into footwear unless guided with hand. Two days prior to presentation he felt unsteadiness with swaying to either side while walking. There was no nocturnal worsening of gait or wash-basin phenomenon. There were no other constitutional complaints, preceding infections or vaccinations. He was on a non-vegetarian diet. His examination was notable for impairment of vibration and joint position sense in bilateral upper and lower extremities, pseudoathetosis of both hands and mild proximal lower limb weakness. His deep tendon reflexes in upper limb was sluggish and was absent in lower limb with flexor plantar response.

The clinical possibility of sensory ganglionopathy or a sensory predominant AIDP was considered. His NCV on day of presentation showed a symmetric demyelinating sensori-motor neuropathy predominantly affecting lower limbs. However since the upper limb symptoms and signs were more severe in the face of preserved upper limb SNAPs, a possible preganglionic or posterior column localisation was considered and MRI was done. His MRI revealed changes typical for SCD (Figure 1). Meanwhile his blood routine showed florid features of macrocytic anemia with extremely low serum Vitamin B12 levels (<50pg/ml). Serological study for antiparietal cell antibody was negative. Since dietary deficiency was unlikely a possibility of pernicious anemia was considered and OGDscopy was done which revealed antral gastritis, evidence of H.pylori infection along with a 10mm polyp in the body of stomach which was excised. The histopathological diagnosis was of WHO grade II neurogastroendocrine tumour(carcinoid) along with severe chronic atrophic gastritis. CT scan of abdomen did not reveal other tumours of gastrointestinal tract. He was subsequently initiated on parenteral Vitamin B12 supplementation. At 2 months follow-up he had significant improvement in all his symptoms, except for mild paresthesia of both hands. His repeat nerve conduction study showed improvement in conduction parameters with near normal F wave latencies and reappearance of lower limb SNAPs.

Discussion

Deficiency in s-adenosyl cobalamine vital for succinyl CoA synthesis and reduced tetrahydrofolate reductase impairing cell turnover have been theorized as the key factors leading to SCD. Recently there has been evidence for interplay between increasing levels of myelinotoxic cytokines and decreasing levels of myelinotrophic IL-6 and EGF in CSF in SCD pathogenesis. Traditionally SCD have been recognised as gradually progressive neurological manifestation of chronic Vitamin B12 deficiency. In most studies the mean duration of symptoms ranges from 4 months to 11 months. Acute presentations of SCD have been seldom reported in literature. Shukla describes a series of five patients with SCD all of whom had presented acutely, with duration of symptoms ranging from 6-15 days. However in all of them the presentation was as acute posterior or posterolateral myelopathy with no mention of neuropathy.

In the absence of Lhermitte phenomenon and girdle sensation, brisk DTR and other pyramidal signs the clinical localisation for a glove and stocking sensory impairment would be more in favour of peripheral neuropathy. As exemplified in this case, the duration of symptoms, rapidity of progression, and absence of deep tendon reflexes and presence of abdominal reflex would raise the clinical possibility of ganglionopathy or sensory predominant AIDP. The nerve conduction study was consistent with a demyelinating sensori-motor neuropathy. Vitamin B12 deficiency has been known to manifest as polyneuropathy, however the electrophysiological pathology has been a point of contention. Early studies
by McCombe employing sural nerve biopsy argued in favour of axonal degeneration\textsuperscript{8}. More recently Saperstein examining patients with cobalamin deficient neuropathies noted none of the patients to have evidence of demyelination in nerve conduction studies\textsuperscript{1}. On the other hand Steiner and recently Puri have clearly highlighted the conduction parameters consistent with demyelination in patients with low Vitamin B12 levels\textsuperscript{9,10}. It could be argued that the pathology depends at what stage the disease is captured, with demyelination being the predominant feature early in the course and secondary axonal changes setting in later. The present case could provide evidence for this hypothesis.

In the current case MRI was ordered due to the clinical-electrical discordance wherein the sensory deafferented upper limbs had essentially normal SNAPs. A predominant LMN involvement due to neuropathy must have masked the clinical feature of pyramidal/posterolateral cord involvement from manifesting.

The American Society for Gastrointestinal Endoscopy recommends a single endoscopic evaluation at the diagnosis of pernicious anemia. This is largely to confirm gastritis and since patients with pernicious anemia have a 2-3 fold increased in risk in incidence of gastric cancer\textsuperscript{11}. Neurogastroendocrine tumours are associated with chronic atrophic gastritis and often pernicious anemia\textsuperscript{12}. The incidence of gastric carcinoid tumours in patients with PA is as high as 10\%\textsuperscript{13}. Endoscopically tumours appear as polypoidal lesions with central ulceration. Prevailing theories of etiopathogenesis point to high gastrin load which is seen in atrophic gastritis driving enterochromaffin cells proliferating into carcinoid tumours. These are usually diagnosed on routine OGD related to chronic gastritis and anemia. The consensus in the management of gastric carcinoids is that WHO grade III carcinoids are treated surgically with extensive resections as for adenocarcinoma while options for type I and II include surveillance, endoscopic polypectomy or resection.

While the association between gastric carcinoid and pernicious anemia is rare but well documented there have been no reports of association with neurological manifestations of Vitamin B12 deficiency. It is interesting to note that while presence of gastric carcinoid and atrophic gastritis points to a chronic process, the neurological manifestation was acute. This may point to an unidentified mechanism which has abruptly tipped the balance triggering off the neuropathogenic process, which leads to clinical manifestation. Whether the association with gastric carcinoid is of relevance as far as neurological manifestations is concerned, remains presently undetermined and future reports are necessary to clarify the significance. In their series, Misra et al had not noted any difference in the clinical picture of SCD between patients with and without antiparietal cell antibodies\textsuperscript{6}. However in the current case the etiology of SCD might have been chronic atrophic gastritis related to H. pylori infection rather than pernicious anemia especially with absence of evidence for autoimmune gastritis and positive serological and rapid urease test. In fact it has been argued that intrinsic factor negative atrophic gastritis could have been initiated by H. pylori infection\textsuperscript{14}.

Without a careful scrutiny our patient could well have received immunoglobulins and had in fact been referred as GBS. However the haematological parameters could
alert the clinician to an alternate disease process. The association with carcinoid with SCD needs to be explored further but an upper gastrointestinal endoscopy should be considered mandatory in any middle aged person with Vitamin B12 deficiency.

MRI T2 weighted images axial(A) and sagittal(B) sections showing the intrinsic cervical cord hyperintense signal changes predominantly affecting the pos-terior columns with the characteristic inverted ‘V’ appearance in axial section.

H&E stains of biopsy from gastric polyp(A(magnification x4) and B(magnification x10)) and gastric mucosa (B(magnification x4) and (C(magnification x 10)). A and B showing nests of tumour cells with features of neuroendocrine neoplasm in the lamina propria of gastric mucosa. C and D showing atrophy of gastric glands, moderate inflammation and pseudopyloric metaplasia suggesting chronic atrophic gastritis.

Table 1 Summary of relevant blood investigations

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Result</th>
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<tr>
<td>Hb</td>
<td>10.1 mg/dl</td>
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<tr>
<td>MCV</td>
<td>107.5fL</td>
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<tr>
<td>MCHC</td>
<td>34.3</td>
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<tr>
<td>Serum Ferritin</td>
<td>96.6(30-400ng/ml)</td>
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<tr>
<td>Serum Iron</td>
<td>128(59-158microgm/dl)</td>
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<tr>
<td>Serum Vitamin B12</td>
<td>&lt;50 (197-866 picogm/ml)</td>
</tr>
<tr>
<td>Peripheral Smear</td>
<td>Macrocytic anemia, hypersegmented neutrophils and evidence of mild hemolysis</td>
</tr>
<tr>
<td>Morning serum cortisol</td>
<td>(7AM- 10AM) - 14.8 (6.2-19.4 microgm/dl)</td>
</tr>
</tbody>
</table>

References


9. Steiner I, Kidron D, Soffer D, Wirguin I, Abramsky O.


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**Emergency Medical Service**

Major services offered:
- Management of acute medical, surgical, cardiac, paediatric and neonatal emergencies.
- Care to all trauma victims including poly trauma following RTAs, fall or assault in concordance with international Level 1 Trauma center standards.
- Acute burns & multidisciplinary poisoning management
- Disaster management
- Well trained clinicians & nurses.
- Emergency Ambulance service for patients
Rett syndrome like presentation of NMDA Encephalitis: A case report

Abstract

The clinical spectrum of autoimmune encephalitis is expanding and presentations as acquired epileptic encephalopathy are being more frequently recognised. Presentations mimicking autism spectrum disorders have been rarely reported in the literature. Through this case report, we highlight the significance of early recognition and early institution of treatment with which a child with clinical features of autism spectrum disorder had an excellent outcome.

Keywords: Autism Spectrum disorder, Rett syndrome, Autoimmune encephalitis, NMDA encephalitis

Introduction

The etiopathogenesis and neurophysiology behind autism spectrum disorder remains largely obscure. In less than ten percent of children with Autism Spectrum Disorder (ASD), a medical condition or a genetic syndrome has been associated as the causative factor\textsuperscript{1}. Aberrant immune mechanism has previously received attention as putative causative agents in ASD\textsuperscript{2} and population based studies have noted a weak association of parental autoimmune disorders with ASD in off-springs. With the ever expanding spectrum of autoimmune encephalitides a strong case can be made for screening children with ASD for potentially treatable autoimmune process. Here we present the case of a five year old boy with a subacute, rapidly progressing autistic features having clinical phenotype of Rett- syndrome, in whom evaluation revealed a diagnosis of NMDA encephalitis. With initiation of immunomodulation, child had mild improvement in autistic features and is being followed up.

Case report

A 5 year old boy with normal birth and developmental and no antecedent illness presented with 3 weeks history of change in behaviour. Normally a docile and well behaved child he started throwing temper tantrums and became irritable at the slightest provocation. There were prolonged crying spells for no apparent reasons and physical aggressiveness towards his parents. His communication dropped off rapidly initially with bisyllabic responses, not answering calls and at presentation was nearly mute with angry vocalisation. Parents noted repetitive wringing hand movements and facial contortions during awake state. He was tending to be aloof, preoccupied with observing his hands and constant hand movements.

His examination revealed a poorly interactive child with very limited eye contact. Visual and auditory regards were well preserved. No meaningful communication could be established. There was ongoing perioral chewing movements and bimanual motor stereotypies in the form of hand wringing and frequent mouthing (Video 1). Thus his clinical features were concluded as a rapidly progressive autistic regression syndrome, most typical of Rett-like phenotype.
His MRI and routine CSF examination was normal and video-EEG revealed no electrical correlate for the dyskinesias. His CSF and serum NMDA antibody titre was positive by direct immunofluoresence. He was initiated on intravenous steroids and immunoglobulins as pulse dose with no improvement. At one month, he was initiated weekly dose of Rituximab. He was also on symptomatic treatment with Risperidone. At the end of 2 months child shows signs of improvement with good eye contact, reciprocates emotions and started smiling at parents.

**Discussion**

Our case had the clinical features satisfying autistic regression syndrome, more specifically Rett-phenotype. Despite essentially normal MRI and nonspecific EEG changes, the rapidity of progression and the predominant perioral stereotyp es made us suspect NMDA encephalitis. Lim and colleagues had recently reported a case of NMD encephalitis with striking similarities to our case. Both cases showed favourable clinical response to immunosuppression but our patient responded to Rituximab. While the autoimmune nature of ASD has remained speculative, a recent study has demonstrated elevated levels of IgM in CSF significantly elevated in Rett syndrome, compared to PDD and normal controls. A parallel can be drawn with certain epileptic encephalopathies such as Landau-Klefner syndrome where the etiology is presumed to be autoimmune and is effectively treated with immunomodulation. Such speculations aside, possibility of an unusual presentation of potentially treatable autoimmune encephalitis needs to be given priority. Despite absence of seizures, suggestive MRI or EEG features and normal routine CSF, recent onset of autistic traits with or without dyskinesia warrants exhaustive evaluation. A strong recommendation need to be made for screening even though the ASD has not been recognised as a presenting syndrome of Autoimmune Encephalitis.

**References**


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A case of extreme thrombocytosis

Dr. Said Jabir
Dr. Mathew Thomas

Abstract

Increased platelet count or thrombocytosis, defined as a platelet count greater than or equal to 350 x 10(9)/litre, is a common hematologic aberration. Several aetiologies are documented for thrombocytosis. Extreme thrombocytosis, defined as a platelet count greater than or equal to 1,000 x 10(9)/litre, is rarely seen in general practice.

Thrombocytosis is a laboratory abnormality which may be encountered on routine evaluation of an unrelated medical problem. The degree of elevation of platelet count was once considered to be an indicator whether the thrombocytosis was reactive or due to a primary haematological disorder. But, current evidence seems to indicate that the degree of elevation cannot be used as a criteria to differentiate between the two. We report a case of reactive thrombocytosis with an unusually high platelet count.

Introduction

Thrombocytosis is quite often discovered as an incidental finding during the laboratory evaluation of an unrelated medical problem. However, when it is detected, it constitutes an important diagnostic challenge. Thrombocytosis can either be due to a reactive process (secondary thrombocytosis) or a clonal bone marrow disorder (primary thrombocytosis or clonal thrombocytosis). It is often exceedingly difficult to differentiate between the reactive and clonal types of thrombocytosis on the basis of clinical findings or laboratory test results. Yet there are fundamental differences between them in terms of cause, pathophysiological features, and clinical implications. We report a case of reactive thrombocytosis with an unusually high platelet count.

Case report

A 52 year old male patient admitted in our hospital on 14th April 2018 with chest pain of 1 week duration, ECG was suggestive of ACS, coronary angiogram done showing TVD underwent CABG. He was discharged on dual antiplatelets and statins. He came for follow up on 27th April 2018.

On examination

General and systemic examinations were normal. On investigation, he was found to have platelet counts of 15 lakhs/cu.mm. Based on this initial report, our initial differential diagnosis were primary and secondary thrombocytosis.

Primary causes include myeloproliferative neoplasm and essential thrombocytosis. Secondary causes include anaemia, infection, inflammation trauma, post splenectomy and functional asplenia.

We proceeded with further evaluation to identify the cause. Peripheral blood film—showed normocytic normochromic anaemia with thrombocytosis.
On imaging

US abdomen–spleen was not seen in its normal anatomical position
CT abdomen–spleen not visualised in the expected location or anywhere else in the abdomen
So our initial provisional diagnosis was probable asplenia.
Workup done
for asplenia, sickling test, ANA, APLA workup, HB electrophoresis, homocystine were negative.

Final diagnosis
Probable congenital asplenia causing extreme thrombocytosis precipitated by stress during the period of surgery.

On follow up gradually his platelet count decreased.

The patient was vaccinated for Pneumococci, H.influenza and Meningococcal coverage.
Now he is on follow up – platelet count is in decreasing trend. Last platelet count was 6 lakh/cumm.

Discussion
The most striking feature of this case is the gross elevation in the platelet count. The degree of elevation of platelet count in this patient was such that further work up for a primary haematological problem was warranted. However, further investigations in that direction did not reveal any evidence of a primary hematological disorder. The cause of reactive thrombocytosis has been ascribed to cytokine mediated increased synthesis of thrombopoietin, which in turn leads to increased synthesis of thrombocytes.

The various causes of extreme thrombocytosis that deserve due consideration in this particular regard include. Primary–myeloproliferative disorders. Secondary–Anaemia, Infections, Trauma, Postsplenectomy, Post-operative and asplenia.

Unanswered questions in this case
- This patient has remained asymptomatic without any serious infections till now without a spleen.
- Is this asplenia really congenital or was it acquired later in his life?
- If it has occurred later, what could be the cause??

References

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I took voluntary retirement from Government service after 22 years and joined Private Medical College and KIMS. I have been working in KIMS, Thiruvananthapuram for more than 16 years and when we got Dip. NB Chest here, I got relieved from SUT Medical College, Thiruvananthapuram as Professor and HOD, Dept of T.B. & Chest Diseases. Because of my experience in both Government and private sectors for many years, I thought I will share my experiences with you, especially the budding physicians, chest physicians and intensivists.

I am aware of the fact that the Critical care management has come a long way ahead in Government Medical Colleges but the Private sector shows an improvement in leaps and bounds. The following is my story inside the ICUs in the Kerala Institute of Medical Sciences (KIMS), Thiruvananthapuram almost exclusively for about 5 years. The ventilated patients were being managed by the Anaesthesia Department, when I joined KIMS and I went for rounds with them. They were not familiar with Non-invasive ventilation and most of the chronic patients in the medical ICU were COPD with exacerbation. With the introduction of Non invasive ventilation by me, these patients could be weaned off easily. Then the anesthesia people left the ICU ventilator management to me.

When I was asked to look after the ventilated patients in the ICUs, Medical, coronary, surgical and paediatric, I approached the idea with some trepidation but the very fact that I had joined KIMS for ‘learning about ventilator management’ and the confidence that chest physicians know about the lung better than any anaesthetist egged me on! It was a ‘dream come true’ for me as I always felt that I lacked sufficient knowledge in managing ventilated cases and it was a ‘hands on experience’. Intensivists were unheard of at that time at least in this part of the country.

I familiarized myself with the ventilator gadgetry, learned about Invasive and Non invasive ventilation from the best of books on the subject and immersed myself in the management of ventilated patients in the ICUs. I spent a lot of time in the ICUs and got a good exposure to the complicated cases needing ventilator. Knowledge about the clinical conditions affecting the lungs helped me judge the cases better. Extubating or liberating a patient from mechanical ventilation is an art which you learn by experience. I stuck to the old adage ‘the very fact that you are putting a patient on ventilator is to get him/her out of it as soon as possible’. I was adhering to the ‘ventilator bundles’ before the same were published and so we did not have Ventilator Associated Pneumonia (VAP) in the MICU for nearly 2 years! The ID specialist who joined later could not believe it. I had to work with 16 ventilators (Drager) which could be used both invasively and non-invasively and 4 Non invasive ventilators. I had assistance from an anaesthetist, and a few dedicated staff nurses.

All the 16 ventilators were put into use at the time of the
first outbreak of Dengue fever in 2003 when we had many complicated cases with respiratory failure. We lost 7 cases of haemorrhagic shock, myocarditis and massive pleural effusion etc. out of 272 cases but in 2004 we could save 206 cases out of 207.

During the SARS outbreak we had an interesting case, a Malayalee working in Abu Dhabi. He developed viral pneumonia while in Kerala and came to the MICU with a saturation of 30%. He was intubated and ventilated. The other patients left the ICU and he had to be shifted to a room and ventilated. The other patients got discharged from the floor. But his pneumonia cleared thanks to a great team effort and he went back happily.

Saving cases of ARDS is always mentally satisfying. ARDS in the early stages may respond to proper mechanical ventilation. I shall cite some anecdotal cases.

A middle aged lady with features of Viral fever- with Pneumonia.

Admitted in local hospital and then transferred here due to deterioration and as swab sent from there was +ve for H1N1.

At the time of admission, febrile, tachypnoeic, bilateral crackles, cyanosed with the saturation 54% on room air, CXR bilateral infiltrates. Saturation picked up to 80% only with 10 L/min flow of oxygen.

Put on Non-Invasive ventilation (NIV) immediately using Savina ventilator - BIPAP mode.

ABG showing paO2 of 50mm Hg on 100% at initiation.

Added broad spectrum antibiotic, methyl prednisolone and supports, continued mechanical ventilation with a PEEP of 12, and PIP of 20mm, slowly bringing down the FiO2. The PEEP was also reduced paralleling the clinical and radiological improvement.

Weaned off ventilator on the 4th day with oxygen and NIV support at night.

Developed post viral IED which responded to oral steroids with full recovery.

CXR at admission

CXR at discharge

A school teacher from Maldives who had a road traffic accident, was given multiple blood transfusions and developed ARDS. He was airlifted, came to MICU directly from airport, was ventilated, made a full recovery and flew back.

The ‘Blue lady’ is a case which became famous in KIMS. The lady was referred from a Kollam hospital with a ‘difficult to wean’ label. She asked me the next day to get the ‘ET tube’ out and I obliged. I usually wait for about 15 minutes after extubation with NIV to see whether there is any post extubation problem. She literally turned blue from the finger tips to the torso in 5 minutes. Luckily I
kept the NIV mask on and she slowly came round again when we removed the NIV she fumed blue again. Then it dawned on me that she may have a thyroid nodule or retrosternal extension. There was indeed a nodule on CT thorax which was removed after tracheostomy and she made a complete recovery. She visited me 2 years back and I could recognize her only after her brother told me that this is the ‘blue lady’ of old.

Another case is that of a young man from Kollam who was brought to the casualty and MICU on 100% on oxygen. He was written off by the local doctors. He was ventilated using high PEEP, low tidal volume, antibiotics and high dose steroids and other supportives. He was on ventilator for a month, but could be successfully weaned off with a mild residual fibrosis.

A few words about Drotrecogin Alfa (XIGRIS). I used it in about 6 cases. I was hesitant in using it as the side effect like bleeding was highlighted by many. I will just cite one or two examples. One was the only son of a Medical shop owner who was brought from Emakulam with severe sepsis following high dose IV steroids for Multiple sclerosis. He had severe ARDS and when I suggested to the father that XIGRIS is a last option he told me to go ahead as the boy was his only son. I started him on Xigris as everything else was failing and he could be weaned off ventilator within a week’s time and he walked out of the hospital in 2 weeks. It was a miracle! Another lady with post partum sepsis and ARDS, a priest with severe sepsis also had the same result. But I had to stop the drug due to bleeding in other cases. Later reports showed that Xigris had only placebo effect only and was withdrawn. How did these patients improve? +God’s hand?

Another lady from Maldives who had severe kyphoscoliosis, with respiratory failure, was airlifted, and accompanied by an Anaesthetist who wanted to take the patient back by return flight. She was weaned off ventilator after a week but she did not want to go back and stayed in Trivandrum for 10 years.

ARDS is not a common condition and most of the cases labeled as ARDS are Viral or aspiration pneumonia which can be managed with appropriate anti-viral/antibiotic and Non Invasive ventilation and supportives.

Neurological problems require prolonged ventilation with tracheostomy otherwise tracheostomy was a rarity in our ICU. Good results are obtained with Guillain-Barre syndrome.

I also remember an Ayurveda student who was brought unconscious. The cause was not known but Prof. Vijayaraghavan Sir after ECHO said she has only 20% EF and she is in cardiogenic shock. She was given cardiac support, ventilated and she made a full recovery in 7 days. I also had cases referred to me with full invasive ventilatory support from other hospitals as “difficult to wean”. One of them with severe COPD and Eventration diaphragm who was weaned off 12 years back still comes to see me off and on with mild exacerbations.

COPD is a condition which poses problems for weaning after Type 2 respiratory failure. I have found that good use of Non Invasive Ventilator (NIV) will help weaning these patients early. The no. of patients requiring invasive ventilation has come down drastically with the introduction of NIV. I would say that Non invasive ventilator is a ‘wonder gadget’ in the chest physician’s therapeutic armamentarium. It can be used in any type of respiratory failure, pneumonias, or when there is increased work of breathing due to restrictive lung diseases. Its effectiveness in pulmonary oedema, & OSA have also been wonderful. It requires more attention than that for invasive ventilation.

Though I have been doing FOB from the early 1990s I had to resort to doing bronchoscopy through the ET tube only rarely. Collapse-consolidation of lobes or segments may occur in mechanically ventilated patients but can usually be easily relieved by N. Acetyl cysteine or N. Saline nebulisation and good chest physiotherapy. FOB through the ET tube spoils the scope.
Managing severe sepsis, ARDS etc are difficult and taxing and I have also had failures and setbacks. I have a very good no. of patients who come to see me yearly for years after I have liberated them from ventilator. That is the dearest award to me for working in ICUs.

One of the social aspects regarding Critical care management in Kerala is keeping the anxious relatives posted on the latest developments in the ICU. The lucidity with which I explain about inhaler therapy in asthmatics I have inculcated in Critical care also. Even then some relatives produce problems which have to be dealt with in the most pleasing manner. One other problem is when the children come to the hospital from abroad and want things to happen either way, that is the patient should either come round or die.

I am now looking after my cases in ICU when they are admitted with respiratory failure as we have Intensivists managing the Multi-Disciplinary ICUs.

**Take home points for the young specialists**

1. Ventilator management is an art which comes by experience. There is no substitute for hard work inside the ICUs.

2. Make good use of Non-Invasive Ventilator. Any patient who comes with a pCO2 upto 120 mm Hg can be given a trial of Non-Invasive Ventilation if you are sure that the patients’ unconsciousness is due to Type II respiratory failure!

3. In my experience Viral pneumonia responds to parenteral steroids with early clearance of the lung shadows when ventilated for 3-5 days.

4. Treat the “patient” and not lab values or ABG. Do not go for frequent ABGs, which is not necessary and will drain only the purse of patients.

5. The good old antibiotics still do not fail in most of the cases. You can escalate if needed, but there is no hard and fast rule that you should be using high-end antibiotics only, in the ICUs.

6. Severe COPD patients do well on home oxygen therapy.

And last but not the least! Discharge patients as soon as possible from the ICUs, rooms or wards. Most of them do better at home and come back happier than ever before.

What I have tried to put in above with all honesty have been my own experiences, my own view points as a chest specialist, a teacher, and as a person who has had a good exposure to Critical care in a tertiary care centre. Managing critically ill patients is essentially team work. You should have the support from the authorities, and dedicated supporting staff. I was lucky in having both these aspects to my favour. I wish to thank Dr. M I Sahadulla, CMD, KIMS, & Prof. Dr. G Vijayaraghavan, Vice Chairman for giving me a free hand to manage ICUs and to the dedicated staff of ICUs for their kind-hearted co-operation.
### Programs from September 2018 to January 2019

<table>
<thead>
<tr>
<th>No.</th>
<th>Event Description</th>
<th>Date</th>
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<tbody>
<tr>
<td>1</td>
<td>Research Methodology Class on “Systematic Review”.</td>
<td>4th Sep 2018</td>
</tr>
<tr>
<td>2</td>
<td>Research Methodology Class on “Test of Significance”.</td>
<td>11th Sep 2018</td>
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<tr>
<td>3</td>
<td>CME on “Endocrine problems in Neonates”.</td>
<td>15th Sep 2018</td>
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<td>4</td>
<td>Clinical Club Meeting.</td>
<td>19th Sep 2018</td>
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<tr>
<td>5</td>
<td>Webcast on “High Intensity Statins in established CVD”.</td>
<td>8th Oct 2018</td>
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<td>6</td>
<td>Surgical Skill Training Workshop on “Bown Anastomosis, Episiotomy &amp; Perineal repair, Knotting, Suturing &amp; abdominal wall closure, basic lap &amp; intra corporeal suturing”.</td>
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<tr>
<td>7</td>
<td>Clinicopathological Meeting.</td>
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<td>8</td>
<td>External Clinical Club Meeting on “Clinicopathological correlation”.</td>
<td>15th Oct 2018</td>
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<td>9</td>
<td>Clinical Club meeting.</td>
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<td>10</td>
<td>Research Methodology on “Test of Significance”.</td>
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<td>11</td>
<td>Internal CME on “In-Hospital Heart Failure”.</td>
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<td>12</td>
<td>Teaching Programme on Neonatology case discussion.</td>
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<td>13</td>
<td>CME on “Cardiac arrest in pregnancy”.</td>
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<td>14</td>
<td>Research Methodology Class on “Diagnostic Study”.</td>
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<td>15</td>
<td>World Prematurity Week 2018.</td>
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<td>Internal CME on “Exclusive Human Milk Diet”.</td>
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<td>Research Methodology Class on “Journal Club Diagnostic Study”.</td>
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<td>CME on “Hyponatremia: the Physiological basis of management – an interactive discussion”.</td>
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<tr>
<td>21</td>
<td>State level Neonatology workshop: STABLE program.</td>
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<td>22</td>
<td>Clinical Club Meeting.</td>
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<tr>
<td>23</td>
<td>Webcast of Asia-Pacific Association of Echocardiography on “Constrictive Pericarditis”.</td>
<td>20th Dec 2018</td>
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<td>24</td>
<td>Clinicopathological Meeting.</td>
<td>20th Dec 2018</td>
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<td>25</td>
<td>Internal CME on “In-Hospital Heart Failure”.</td>
<td>26th Oct 2018</td>
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<td>27</td>
<td>Clinical Club Meeting.</td>
<td>16th Jan 2019</td>
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<td>28</td>
<td>Basic Laparoscopy course.</td>
<td>19th &amp; 20th Jan 2019</td>
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<td>29</td>
<td>Workshop on Neurology for Neonatologists.</td>
<td>19th &amp; 20th Jan 2019</td>
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### AHA Programmes

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<th>No.</th>
<th>Course</th>
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<tr>
<td>1</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>5th to 7th July 2018</td>
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<td>2</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>19th to 21st July 2018</td>
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<td>3</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>2nd to 4th Aug 2018</td>
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<td>4</td>
<td>AHA PALS Provider Course.</td>
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<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>6th to 8th Sep 2018</td>
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<td>6</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>26th to 28th Oct 2018</td>
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<td>7</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>1st to 3rd Nov 2018</td>
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<td>8</td>
<td>AHA BLS &amp; ACLS Instructor Courses.</td>
<td>30th Nov to 1st Dec 2018</td>
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<td>9</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>6th to 8th Dec 2018</td>
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<td>10</td>
<td>AHA PALS Provider Course.</td>
<td>12th and 13th Dec 2018</td>
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<td>11</td>
<td>AHA BLS &amp; ACLS Provider Courses.</td>
<td>3rd to 5th Jan 2019</td>
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</table>

### Forthcoming Programme

“Neonatal Videolaryngoscopy & Difficult Airway Course” on 26th February 2019 at the Osler Hall.
SCROLL OF HONOUR for Teaching and Clinical Excellence NBE accredited hospital-2018

National Award from the Association of National Board Accredited Institutions (ANBAI) & National Board of Examinations (NBE)

Dr. M I Sahadulla, Chairman & Managing Director, KIMS Group and Dr. P M Saffia, Vice Dean Academics, KIMS receiving the National award - Scroll of Honour for Teaching and Clinical Excellence, National Board accredited Hospitals from Shri. Vajubhai Vala Saheb, Honourable Governor of Karnataka, Dr. Alexander Thomas, President, Association of National Board Accredited Institutions (ANBAI), Dr. Jayshree Mehta, President, Medical Council of India (MCI) and Dr. Abhijat Sheth, President, National Board of Examinations (NBE).

The Scroll of Honour Award instituted by National Board Accredited Institutions and NBE is given to NBE accredited institutions. The Awards committee was chaired by Shri. V K Gupta, former Chief Justice of Jharkand-Himachal High Court. The award function was held at J N Tata Auditorium, IISC Bengaluru.
### Academics

<table>
<thead>
<tr>
<th>Sl. No.</th>
<th>Authors</th>
<th>Title of the article/chapter</th>
<th>Name of the Journal or Publication</th>
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<tr>
<td>1</td>
<td>Dr. Jayakumar Menon et al (Editors) Dr. Arya R Raj &amp; Dr. Jayakumar Menon Dr. Manju Issac &amp; Dr. Jayakumar Menon Dr. Ragitha Binu Krishnan &amp; Dr. Jayakumar Menon Dr. Jayakumar Menon</td>
<td>Dysphagia Management in Head &amp; Neck Cancers 2018 Setting up a swallowing clinic Fluoroscopic Evaluation of dysphagia TNE &amp; Pharyngeal Manometry Phagosurgery</td>
<td>Springer Publication</td>
</tr>
<tr>
<td>2</td>
<td>Dr. Jayakumar Menon et al</td>
<td>Interdisciplinary Telemedicine in the Management of Dysphagia in Head and Neck</td>
<td>Dysphagia</td>
</tr>
<tr>
<td>3</td>
<td>Dr. Suman O S., Prof. G Vijayaraghavan, Dr. Muneer A R</td>
<td>Aneurysm of the Saphenous Vein Graft after Coronary Artery Bypass surgery</td>
<td>Journal of the Indian Academy of Echocardiography and Cardiovascular Imaging</td>
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<td>4</td>
<td>Prof. G Vijayaraghavan and Dr. S Sivasankaran (SCITMST)</td>
<td>Restrictive Cardiomyopathy: The Indian Face</td>
<td>CSI Jaypee</td>
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<tr>
<td>5</td>
<td>Prof. G Vijayaraghavan</td>
<td>Myocarditis in hospitalized patients with dengue fever</td>
<td>Proceedings of 2018 Healthcare and Cardiology conference and 2018 Mental Health and Neurology conference, Bangkok</td>
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<tr>
<td>6</td>
<td>Dr. Sitanshu Bank (KIMS Alumni), Dr. Muhammad Nazeer, Dr. Babloo Thomas Mani (KIMS Alumni).</td>
<td>Accelerated Ponseti Technique in the Management of CTEV</td>
<td>European Journal of Orthopaedic Surgery and Traumatology</td>
</tr>
<tr>
<td>7</td>
<td>Dr. Mathew Thomas Dr. Manish Kumar Yadav Dr. Elsa George</td>
<td>Partial Splenic Artery Embolization for the treatment of ITP: A Case Series-Pilot Study</td>
<td>Journal of Clinical Interventional Radiology ISVIR</td>
</tr>
</tbody>
</table>

### Appreciations

- Prof. G. Vijayaraghavan has been awarded the highly prestigious title of **Emeritus Regional Adviser** to Royal College of Physicians (RCP), Edin. by the president of RCPE in October 2018

**Achievement at the National Board of Examinations**

- Dr. Rubeesh Hassan, All India First Rank and Gold Medal - DNB Orthopaedic Surgery (2015)
- Dr. Dipin Kumar, All India First Rank and Gold Medal - DNB ENT (2015)
- Dr. Chitra Gopal, All India First Rank and Gold Medal - DNB Obstetrics & Gynaecology (2016)

**Winners received the award from Shri. Ashwini Kumar Choubey, Honourable Minister for Health and Family Welfare.**

**Other Achievements**

- Dr. Anoop Sugunan, Dept of Neurology, secured 2nd prize in scientific poster presentation on “Clinical Spectrum of Enterovirus meningitis-A Single centre retrospective case series” at the conference of Annual Mid Term Meet of Kerala Association of Neurologists held on 21st October 2018 at Kochi.
- Dr. Sreejith (3rd Yr MEM resident) & Dr. Waseem (2nd Yr DNB resident) won 2nd place for the quiz competition and Dr. Bhadra Menon (3rd yr MEM resident) secured 2nd place for the young faculty presentation competition at the 10th Annual National CME by Emergency Physicians Association: PACE 2018 on 22nd and 23rd at Trivandrum.
Patient was on treatment for allergic contact dermatitis. The rash flared-up on discontinuing the topical application. What is the diagnosis?

**Answer:** Perioral dermatitis (seen in long-term use of topical steroid)

Name one of the commonest side effects of fairness cream?

**Answer:** Hypertrichosis (prolonged use of topical steroid combinations)

This child was on long term treatment for recurrent itchy rash on the face. What is the diagnosis?

**Answer:** Tinea incognito (fungal infection exacerbated the application of topical steroid)
The Art of Chest X-ray Diagnosis

This book has an extensive collection of radiographs and CT images of various chest-lesions ranging from simple or incidental issues to complex clinical problems. This would be very useful for clinicians, budding pulmonologists, radiologists, general practitioners and also to Post graduate students of Internal Medicine as a ready reckoner in their practice and also to gain a lot of information in chest-radiology.

Author
Dr. Kesavan Nair MBBS M.D. (Resp. Medicine), M.Phil, FCCP
Sr. Consultant Respiratory Medicine
Former Professor in Respiratory Medicine, Govt Medical College
Worked in Govt. & Private Medical Colleges and retired as Professor of Respiratory Medicine

Address for communication
email: veekeyen@gmail.com
9446478615

The book is available in Notion Press showroom in Chennai and through Amazon and Flipkart only

Egg shell calcification of Mediastinal L.N. - Silicosis - CT Thorax

Cysts in Azygos lobe - CT Thorax

Bamboo spine in Ankylosing Spondylitis with severe kyphosis associated with Cavities in upper lobes sometimes

Mucocele following lobectomy - a rare occurrence
Accreditations

• ACHSI (Australian Council on Healthcare Standards International)
  KIMS got ACHSI accreditation in the year 2006 for demonstrating continuous improvements in patient safety and delivery of quality healthcare that is at par with international standards.
• NABH (National Accreditation Board for Hospitals & Healthcare Providers - India)
  KIMS received NABH in the year 2006 as a recognition of its commitment to ensure safe healthcare practices and infection control measures.
• NABL (National Accreditation Board for Testing & Calibration Laboratories)
  The Laboratory at KIMS is accredited by NABL in the year 2008, for ensuring precise diagnosis and following safe practices.
• NABH (National Accreditation Board for Hospitals & Healthcare Providers - India)
  KIMS Blood Bank is accredited by NABH in the year 2011, as recognition of its commitment to make safe blood and blood products easily available at the hour of need by adhering to modern techniques and quality standards.
• KIMS is certified with nursing excellence by NABH in the year 2015, as a recognition of its commitment towards safe and ethical nursing care.
• NABH Medical imaging services is awarded in the year 2016 for its outstanding contribution to sound and ethical radio diagnostics practices.

Recognitions

• Scroll of Honour for Teaching and Clinical Excellence NBE accredited hospital 2018.
  National Award from the Association of National Board Accredited Institutions(ANBAI) & National Board of Examinations (NBE)
• Best Hospital IT Project Award 2017.
• Australian Council on Healthcare Standards International Medal for outstanding contribution at an international level to improving quality and safety in health service.
• NIB Awards 2016 for House Journal: Best Content
• Golden Peacock National Quality Award 2014 in Healthcare Sector.
• Best Service Provider Award 2014 from Star Health and Allied Insurance Company Ltd.
• Golden Peacock International Business Excellence Award for the year 2013 initiated by Institute of Directors, United Kingdom.
• Commendation Certificate of Kerala State Government for energy conservation for the year 2012.
• TRIMA CSR award 2012, for excellence in CSR Activities undertaken for the financial years 2010-2011 and 2011-2012.
• Dr.Prathap C. Reddy Safe Care award for Best Medication Safety Initiative 2011.
• Avaya Global Connect Customer responsiveness Award 2010.
• South Asian Federation of Accountants (SAFA) award for best presented accounts and corporate governance disclosure.
• A – stable rating by CRISIL for best financial reporting in the year 2008.
• Hospital Management Asia (HMA) Award for the Project Musculo skeletal injuries in 2009.
• AV Gandhi Memorial Award 2007 and 2008 for excellence in Cardiology.
• Award for transparency in financial reporting in the year 2005 and 2008.
• Best Power User Award by Cyber India Online for optimal power utilisation in the healthcare industry in India in 2004.
• Kerala State Pollution Control Board Award for biomedical waste management in 2004 & 2006.
• Health Tourism Award 2005 for maximum foreign exchange earnings.
• Best Customer Site Award from HCL Infosystems Ltd.
• Regional ACLS Training Center by American Heart Association.

KIMS CENTRES OF EXCELLENCE

• Cardiac Science
• Neuro Science
• Urology
• Orthopaedics
• Gynaecology & IVF
• Oncology
• Gastroenterology & Liver Transplant
• General & Minimally Invasive Surgery
• Nephrology & Renal Transplant
• Dermatology & Cosmetology
• Emergency Medical Services
• Endocrinology & Diabetes
• Obstetrics & Gynecology
• Psychiatry & Behavioral Medicine
• Physical Medicine & Rehabilitation
• Pediatric Gastroenterology
• Respiratory Medicine
• Plastic & Cosmetic Surgery
• Interventional Radiology
• Radiodiagnosis & Imaging Sciences

Thank you. With your trust, our legacy continues.
NOTHING SHOULD LIMIT US FROM WALKING OUR PATHS

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Middle East: Oman | Qatar | Dubai | KSA (Jubail, Riyadh) | Bahrain (RBH, KBMC)