Case Report

531. Systemic Mastocytosis Masquerading as Recurrent Spells: A Diagnostic Challenge

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A 38 years man was referred to our institution for evaluation of ‘spells’ for 15 years. His spells typically began when he would go to sleep with a sudden throbbing in his head and palpitations often accompanied by vomiting and diarrhea. He would remain alert throughout though be completely incapacitated with weakness, which could last for several minutes. He would have approximately one spell a year and could think of no precipitating factors. Physical examination revealed a healthy appearing male with a macular rash on his trunk and limbs. Darier’s sign was positive. Complete blood counts, electrolytes, MRI brain, EEG were normal. Skin biopsy revealed high density of mast cells consistent with urticaria pigmentosa (UP). Serum tryptase and histamine levels were elevated, as were urinary histamine and beta prostaglandin F2 levels. Bone survey was negative. Bone marrow biopsy revealed 10% mast cells. Cytophenic studies of bone marrow demonstrated a normal karyotype. A diagnosis of Systemic mastocytosis Type 1B was made. He was started on H1 and H2 blockers, instructed to avoid alcohol, aspirin/NSAIDs, polymixin B, seafood, narcotics and contrast dye and always keep an Epipen available. Differential diagnosis such as carcinoid syndrome, pheochromocytoma, VIPoma, and anaphylaxis were evaluated and excluded in our case. The prognosis in our patient is good.

532. Visceral Leishmaniasis from South India

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Abstract: Kala azar is an endemic important public health problem in India, especially the Gangetic basin of Northern India. We saw a native of Kerala and another of Karnataka with kala azar, suggesting a possibility of its resurgence.

Case Reports and Methods: Case 1: Thirty years male, a tidy taper from Ernakulum district of Kerala, presented with five month history of high grade intermittent fever with chills and rigors, increasing fatigue and weight loss (8 kg). He had no localizing symptoms. Following a road traffic accident 2 years ago, he had been transfused one pint of blood. He is a non-smoker but consumes about 100-150 ml of arrack daily. He denies history of sexual exposure. His last travel dates back to five years ago, to Mangalore and to Coimbatore. Examination showed a thin ly built individual (43 kg), with temperature of 100°F and pallor. Abdominal examination revealed a soft hepatomegaly (3 cm) and splenomegaly (17 cm). Neurological and cardio respiratory system examination was normal. Laboratory investigation: Haemoglobin: 10.5 gm% with reticulocyte count: 3%, white blood cell count: 1300 cells/mm3 (normal differential) and platelets: 75,000/mm3. Peripheral smears (3) were negative for malaria. Liver function tests were normal except elevated globulins (5.9 gm/dl). Electrolytes and creatinine were normal. Two sets of blood cultures were sterile. Ultrasound abdomen showed hepatosplenomegaly. Bone marrow trephine smears showed hypercellular marrow and diffuse plasmacytosis. Leishman-Donovan (LD) bodies were seen on staining with Giemsa. One gram of Amphotericin B was administered (25 gm/kg) over 28 days, with resolution of fever over a month. On follow up 6 weeks later, he had gained 8 kg weight and spleen had regressed (4 cm).

Case 2: Fifty five years male from Kolar, Karnataka was admitted for evaluation of dry cough of 5 days duration. He had been diagnosed to have HIV 1 infection in 2000. In March 2001 he was evaluated for fatigue, pallor, and hepatosplenomegaly. He was started empirically on antituberculous therapy, with improvement of symptoms over the course of therapy, and weight gain of 5 kg. He has never had a blood transfusion in the past. On examination, he was pale, with no lymph adenopathy. Abdominal examination showed hepatomegaly (1 cm) and splenomegaly (5 cm), but no free fluid. Cardiorespiratory and neurological examination showed hepatomegaly (1 cm) and splenomegaly (5 cm), but no free fluid. Cardiorespiratory and neurological systems were normal. Investigation revealed: Haemoglobinin 9.4 gm/dl, reticulocyte count 0.8%, with total white cell count 2800 (myelocytes 3%, band forms 1%, neutrophils 60%, eosinophils 6%, lymphocytes 29%, monocytes 1%) and platelet count of 102000/mm3. Liver function tests were normal except for altered albumin globulin ratio (total protein 9.5 gm%, albumin 3.5 gm%) and an elevated alkaline phosphatase (136 IU/L). Acid fast bacilli were not seen on three sputum samples. Chest X-ray was normal. Ultrasound abdomen showed moderate splenomegaly. Bone marrow trephine smears showed cellular marrow with numerous intra and extracellular LD bodies suggestive of Leishmaniasis, on staining with Giemsa. The patient was started on Amphotericin B along with HAART (Stavudine, lamivudine and nevirapine). He recovered, with decrease in splenic size, and continues to do well on follow up.

Discussion: A century has elapsed since the description of the splenic amastigote of Leishmania donovani by Donovan in Madras. Subsequently, very few reports of visceral leishmaniasis from this region have been documented. Our patients have had no history of travel to an endemic area. The possibility of transfusion related kala azar was considered in our first patient. As the asymptomatic sub clinical period of Leishmanial infection post transfusion may be of variable duration, and as parasite viability in stored blood has been documented earlier, this could not be ruled out. Kerala and Karnataka have not had prior history of visceral leishmaniasis in adults. There has been prior documentation of cutaneous leishmaniasis, and most recently, there have been a report of two children with visceral leishmaniasis from Kerala. This could represent a re emergence of a disease in areas considered non endemic.

539. Lemierre’s Syndrome Caused by Community Acquired Methicillin-Resistant Staphylococcus aureus Infection

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Lemierre’s syndrome is septic thrombophlebitis of internal jugular veins arising as a rare complication of infection elsewhere
in the head and neck. Community acquired methicillin-resistant Staphylococcus aureus (CA-MRSA) infection is emerging as a real threat. We report a rare case of CA-MRSA infection presenting as orbital cellulitis with Lemierre’s syndrome. A 16 years girl presented with high grade fever of 2 weeks duration and swelling around the Rt. eye for 9 days. On examination, she was febrile with Rt. sided proptosis with painful restriction of movements of the globe in all directions and minimal ipsilateral papillary dilatation. Visual acuity was preserved. Palpation of the neck revealed firm, cord-like structures bilaterally and jugular venous waves were not apparent. Bilateral coarse crackles were present. She was started on parental antibiotics with anaerobic coverage, after drawing blood cultures. Chest radiograph showed bilateral alveolar infiltrates with breakdown. Contrast enhanced CT-scan showed thrombosis of bilateral internal jugular veins and multiple, bilateral, thin walled cavities in the lung parenchyma. Orbital ultrasound showed a 16 x 9 mm fluid collection in the superomedial orbit. Cavernous sinus thrombosis was excluded by magnetic resonance venography; Anti-cardiolipin and anti neutrophil cytoplasmic antibodies were negative. On the third hospital day she had massive haemoptysis and aspiration and was intubated and mechanically ventilated. Aerobic blood cultures grew MRSA and no anaerobes were grown. With antibiotic therapy and ventilatory support she improved and was successfully extubated. On follow up visit, she was fully functional with ventilatory support she improved and was successfully extubated. On follow up visit, she was fully functional with ventilatory support she improved and was successfully extubated.

562. Gujar Lung

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Gujar lung refers to a form of chronic bronchitis associated with radiographic evidence of diffuse reticular and nodular shadowing, thought to be caused by pine wood fuel burning in mud houses called “Kothas” in the gujjar community which is a social and ethnic group in the mountain regions of Kashmir valley.1,2 We describe the disease state in a 58 years old male gujar who got admitted with complaints of recurrent cough and progressive dyspnea of one year duration. There was no history of any significant illness in his past requiring hospitalization or prolonged treatment. Examination revealed the patient of average built with body mass index of 27.5 kg/m². His general physical examination was normal. Chest examination revealed few rhonchi bilaterally. Rest of the systemic examination was normal. On investigating the patient, hemoglobin was 10.8 g/dL and ESR of 12 mm in 1st hour (Wintrobes method). Chest radiograph showed bilateral reticulonodular shadows in mid and lower zones, sparing the upper zones. High resolution computed tomography of chest revealed multiple centrilobular nodulation, with reticulation. Closed lung biopsy showed evidence of deposits of carbon nodules interspersed with collagen fibres, with fibrogenic reaction in some areas. Investigation profile for tuberculosis including PCR and ELISA and culture was not suggestive of the disease and fungal studies were negative. Pulmonary function tests revealed mild obstruction of small airways. All possible hematological, biochemical, immunological, endochronological and serological studies were normal. The patient was given an empirical therapeutic trial using antitubercular drugs for nine months, in addition to bronchodilators and steroids. The radiological features persisted and he is under our regular follow up, for future course.


571. Extramedullary Blast Crisis

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We reviewed three cases of chronic myelocytic leukaemia (CML) in chronic phase. One of them (F/32 yrs) presented with moderate pleural effusion and ascites and two (M/35 yrs, M/40 yrs) presented with generalized lymphadenopathy with hepatosplenomegaly, clinical features suggestive of lymphoma/leukaemia. In all cases peripheral blood and bone marrow examination revealed chronic phase of CML with few blast cells. Pleural fluid cytology revealed cells with morphological feature of myeloblast and pleural biopsy showed undifferentiated monomorphic cellular infiltration. Lymphnode biopsy mimic lymphoma. Infiltrated tumour cell was diagnosed as lymphoblast as a part of extramedullary blast crisis of CML by histochemical an immunological test. Varied therapeutic responses observed in these patients.

Extramedullary blast crisis in chronic phase is relatively entity. Some may present in accelerated phase as first presentation. Frequently involved sites are lymphnode, serosal surface, skin, soft tissue, CNS, breast, GI tract, bone and manifest accordingly. Diagnosis mainly based on detection of myeloblast/lymphoblasts in involved organ by immunophenotyping and FISH (Fluoresence in situ hybridization) analysis. Appropriate diagnosis is essential to select the ideal therapy for early remission and longer survival.

Case Report : A 35 years male presented with weakness, exertional dyspnea and malena since two months. He had past history of episodic epistaxis and migraine since 4 years. There was no history of bleed from any other site. He had severe pallor, mild hepatosplenomegaly and a bruit was heard over his right chest. The chest X-ray showed a lobulated mass. CT scan and digital subtraction angiography (DSA) revealed a pulmonary arteriovenous malformation and an A-V fistula arising from the coeliac axis and going upto the pelvis. Transcatheter embolotherapy was performed to occlude the abdominal A-V fistula and hematinics were given, with which patient improved. Patient was well for a year when he again presented with worsening dyspnea and epistaxis. Repeat DSA showed occlusion of the abdominal fistula. ENT evaluation and endoscopy revealed multiple telengectatic lesions. He improved with blood transfusion and hematinics and also underwent surgery for pulmonary A-V fistula A final diagnosis of hereditary hemorrhagic telengiectasia was made, based on the criteria of International HHT foundation.

Conclusion : HHT is a rare vascular disorder that is often overlooked. The underlying abnormality is malformation of blood vessels affecting various organs of the body including nose, skin, lungs, liver, brain and GIT causing excessive bleeding. Treatment is usually conservative, but embolization, surgical excision or ligation is indicated for enlarging or symptomatic fistulas.
Previous studies have shown that with such an approach, mortality is less than 10%. Gene replacement therapy may be promising in the future.

588. Atypical Case of Congenital Ectodermal Dysplasia Presenting as PUO

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Introduction: Prolonged PUO is a clinical challenge to physicians. We present a case which illustrates how clinical acumen can do away with extensive, invasive and needless investigations.

Case Reports: Seventeen years Oriya male from Bhubaneswar presented with quotidian spikes of fever up to 105°F for 3 months. Temp. peaks during day time and remits during nights and morning. Patient gives past H/O similar cycles of fever for which he has been investigated extensively particularly for lymphoma and TB. He had visited referral premier hospitals and all routine specialised investigations like scans, cultures and biopsy have been non informative. Examined on 12th May at 3 pm, patient was having temp. of 104°F but didn’t appear toxic. He was sweating profusely from face but had no sweating from rest of the body at all and skin was dry and hot. There was no abnormality in teeth, nose or facial structure. Systemic exam and hemogram were normal, test for MP negative.

Patient admitted of having febrile episodes during summer every year. He avoids going out in the sun and always cools himself by using wet ‘Camuchha’ (towels) over body.

A diagnosis of CED was made. Absence sweating was demonstrated by 2% iodine alcohol soln. followed by starch in castor oil application. Biopsy of palmar skin showed almost absent sweat glands.

Conclusion: 1) CED is not rare but a case with facial area escaping the anomaly and without other stigmata (hair, teeth, nose) of the disease is not described in the literature. 2) Possibility of CED must be borne in mind while evaluating a case of PUO particularly during tropical summer months. 3) Simple clinical observation is sometimes far superior to elaborate and costly investigations.

590. Kikuchi Fujimoto Disease

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A female aged 16 years was admitted with fever of 2 years duration. She used to get high-grade fever with chills, on and off initially and became continuous of late. There was history of generalized body pains. There was loss of appetite and weight. She was moderately built and ill nourished. Her weight was 35 kgs. No abnormality was detected on examination except for the pyrexia.

She was investigated for the cause of PUO. Hemoglobin was 9.3 g/dl with MCV of 75.9 fl/cumm. Total leucocyte count was 7200 cells/cumm (polymorphs 81%), lymphocytes 15%, eosinophils 2% and monocytes 2%. Platelet count was 3.9 lakhs/cumm. Peripheral smear did not show any abnormal cells and negative for malarial parasite. ESR was 124 mm. Routine urine examination was normal. Random blood glucose was 93 mg/dl and blood urea 26 mg/dl. Total serum proteins were 8.2 g/dl with albumin of 3.5 g/dl. Serum bilirubin was 0.5 mg/dl, serum alkaline phosphatase 223 U/L, AST 36 U/L and ALT 30 U/L. Chest radiograph was normal and Mantoux test negative. 2D echocardiogram was normal. ANA and RF were negative. Ultrasonogram and CECT of abdomen showed normal spleen with lymph nodes at hilum and spleneculi. Bone marrow study revealed reactive hyperplasia with absent iron stores. Enterobacter species bacteria were isolated in blood. She was given antibiotics, iron supplementation and discharged on request.

She was readmitted after one month, as fever did not subside. Small cervical lymph node on left side was detected but biopsy was inconclusive. Blood and urine cultures were negative. LDH was 675 U/L and uric acid 1.8 mg/dl. CRP was positive. ELISA for HIV and HBsAg were negative. She was vaccinated for capsulated organisms before operation. Spleen was removed along with hilar lymph nodes and spleneculi. Biopsy of the liver was also done.

No pathogen was identified on staining or culturing the specimens. On histology, the splenic hilar lymph nodes revealed necrotizing lymphadenitis suggestive of Kikuchi Fujimoto disease. Liver and spleen were normal. No granulomas were noted. Spleneculi showed reactive histiocytosis. She was given prednesolone 10 mg/day for 6 weeks. Steriod was stopped after tapering doses. She was on follow up for more than 1 year. She became normal and fever did not recur. She steadily gained 16 kgs weight.

*635. Recurrent Bacterial Meningitis due to C2 Complement Deficiency

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A 34 years man presented to the emergency department with a one day history of increasing fever, chills and abdominal pain with some nauseaus and vomiting. At presentation, the patient was found to be febrile and hypotensive. Blood, urine and cerebrospinal fluid (CSF) cultures were obtained; intravenous fluids were given and the patient was admitted to the intensive care unit, intubated due to hypoxemic respiratory failure and started on intravenous ceftriaxone and vancomycin. Blood and CSF cultures grew Neisseria meningitidis. The disease course was complicated by severe disseminated intravascular coagulation (DIC), leading to necrosis and eventual amputation of the fingers, toes and tip of the patient’s nose. His recovery was delayed by complications such as MRSA bacteremia, multiple soft tissue infections and ulcers as well as osteomyelitis at the amputation sites, particularly of the lower extremities.

The patient’s medical history prior to this illness was significant for an episode of pneumococcal meningitis with associated septicemia and septic arthritis at age 21, pneumococcal septicemia at the age of 23, upper extremity cellulitis and a possible history of rheumatoid arthritis and Raynaud’s syndrome. HIV serology was negative. In light of the history of recurrent serious infections, immunologic evaluation was performed, demonstrating complement C2 deficiency (< 10 U/ml).

Although C3 deficiencies are more commonly associated with recurrent infections, complement C2 deficiency may result in vulnerability to pyogenic infections. This deficiency may result in a wide spectrum of disease, ranging from asymptomatic cases to severe disease. C2 deficiency is commonly associated with autoimmune disease but may also lead to severe recurrent pyogenic infections.

* Adjudged Best Papers and got an award of Rs. 1000/- each from Chairman Scientific Committee, Diamond APICON 2005.