A case of Churg - Strauss syndrome

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Abstract

Churg-Strauss syndrome is an extremely rare disease belonging to a group of primary vasculitis syndrome, which is present in approximately 1 in 1 to 3 million people. One rarely comes across 1 or 2 cases in lifelong practice. According to Indian studies, its frequency is 1.78 in Indian population1 .Only 11 patients of this disease were seen during a period of 11 years at AIIMS, New Delhi.2 This disease was first described by Churg & Strauss in 1951. The more common diseases of this group are Wegener's granulomatosis, polyarteritis nodosa, Bechets syndrome, Takayasu 's arteritis. Present case was a 40 year old male, who was referred as a suspected case of pulmonary TB / infective complications of Bronchial asthma.

Keywords : *" Vasculitis, Asthma, Eosinophilia*

A 40 years old male patient presented with 5 years history of irregular wheezing episodes and cough. He was being treated with bronchodilators. He had recent development of low grade pyrexia with dry cough and painful swelling of joints (metatarsophalangeal, wrist and elbows) of 15 days duration. He had one chest x ray done with a private clinic which showed multiple bilateral lung shadows. He was suspected to be suffering from pulmonary Koch's disease (Fig. 1) and was put on Anti tubercular regimen.

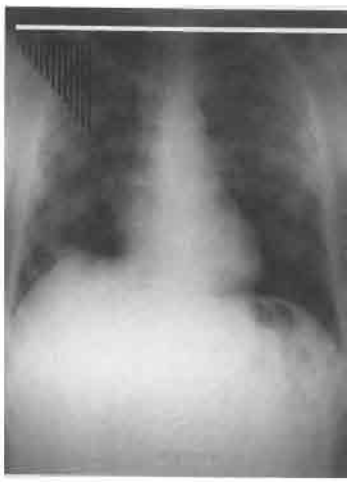

Fig 1: X-Ray chest on admission

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Except for 5 years of occasional history of asthmatic episodes, no other relevant past or family history was available. He did not suffer from weight loss, expectoration or haemoptysis, nor had he any joint stiffness in the past. He was a non smoker and non alcoholic. Relevant investigations on admission were: Hb-6.5 gm/dl, TLC: 11500/cumm, DLC: Eosinophilia 29%, ESR was 85 mm / hr (Wintrobe), Sputum AFB was negative, HIV screening test: Non reactive, HBsAg: negative. Renal and liver function tests were within normal limits. USG abdomen showed bilateral altered echogenecity of kidneys and mild hepatomegaly. Since a possibility of aspergillosis was also entertained, sputum for isolation of the fungi was undertaken but was negative. His X-ray chest showed increased lesions (fig 2) and his pyrexia and joint pains continued. Antibiotics were added to the regimen being followed (A combination of amoxycillin, metronidazole.

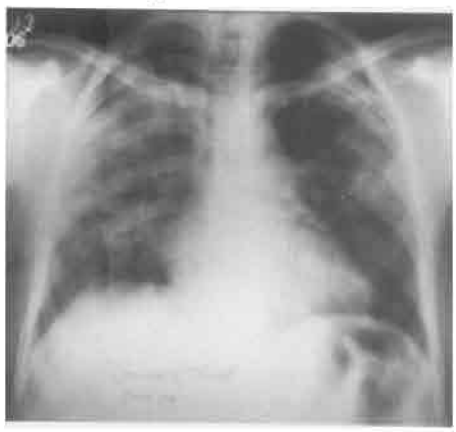

Fig 2: X-Ray Chest: Increase of Lung Lesions

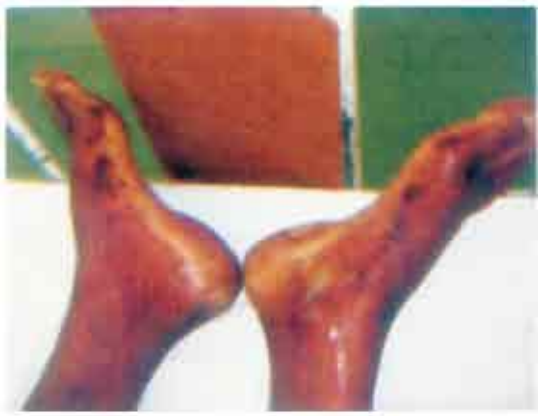

Fig 3 : Purpura Like Lesions Over Feet.

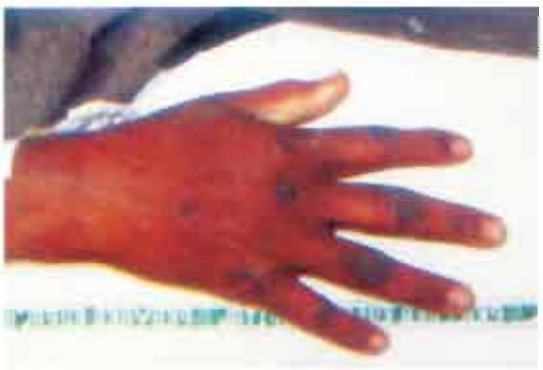

Fig 4 : Purpura Like Lesions over hand.

A bronchoscopy was planned but on the 15th day of hospitalization, he was found to have developed erythematous raised purpura like irregular lesions over feet and hands (Fig 3 and 4). A CT scan of the thorax revealed lesions suggestive of bilateral alveolar and interstitial type of opacities (Fig 5). By now he had developed raised BUN and creatinine level (blood urea: 123 mg/ dl and creatinine; 2.7 mg/ dl). Keeping in view the pulmonary and skin lesions suggestive of vasculitis, ATT and antibiotics were withdrawn and a skin biopsy carried out. As first report suggested "Non specific dermatitis," a repeat skin biopsy was sent together with other clinical details as there was a strong suspicion of vasculitis. This was reported as small vessel vasculitis with eosinophilic infiltration(Fig 6). C-ANCA and P-ANCA were negative. The overall picture of lung lesions, eosinophilia and evidence of vasculitis fitted clinically and, histopathologically into CHURG-STRAUSS syndrome. ECG and ECHO cardiography carried out were within normal limits.

High power view of skin biopsy from dorsum of right foot showing infiltration of inflammatory cells including eosinophils, around capillaries.

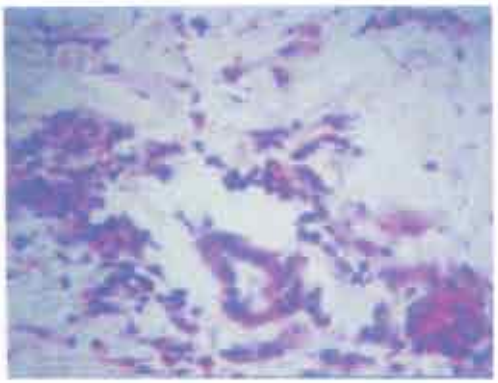

Fig 6: Histopathology

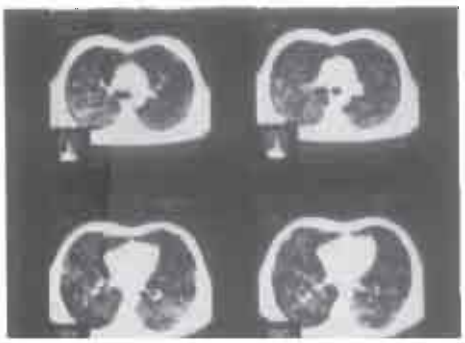

Fig 5 : CT Scan, Thorax : Bilateral alveloar & interstitial opacities.

Patient was first treated with high doses of steroids. There was a clinical response to this treatment but it was slow, hence Cyclophosphamide in doses of 100 mg once a day was added. A rapid clearance of lung and skin lesions was noted with disappearance of eosinophilia, joint swellings and pyrexia. On follow up, patient was asymptomatic after 3 weeks of therapy (Fig 7). Renal function test results were within normal limits.

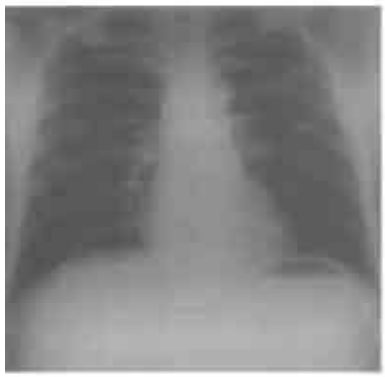

Fig 7: Disappearance of Lung Lesions following treatment.

Discussion

The vasculitis syndromes are a large group of clinical conditions some of which are very rare. The clinicopathological process in all these groups of

disorders is characterized by inflammation of blood vessels resulting in subsequent ischemic and necrotic changes in various organ systems of the body. These diseases are classified as : Primary and secondary vasculitis. Secondary causes are : Drug induced, allergic, infections, malignancies and miscellaneous autoimmune diseases. The vasculitis syndromes are also classified as large vessel, medium vessel, small vessel diseases.[1,2]

Some of the important primary vasculitis syndromes are Wegner's, Poly arteritis nodosa, giant cell arteritis, Bechets and Kawasaki syndrome and Churg-Strauss syndrome. Our case can be grouped as small vessel vasculitis. Churg-Strauss syndrome is a very rare disease (1 in 1 -3 million), described by Churg and Strauss in 1951 as a separate entity. Patients with this disease almost invariably present with respiratory symptoms.[3,4,5] It is characterized by asthmatic episodes, peripheral blood and tissue eosinophilia and vasculitis of various organs of which lung and skin are major target organs. Age of onset is usually in the 3rd to 4th decade and female to male ratio is 1.2:1. Other organs involved in order of frequency are: Kidneys, peripheral nervous system, gastro intestinal tract and heart. Pulmonary lesions and eosinophilia are dominant in over 61%, while skin lesions presenting as elevated purpura is seen in 70% of cases. Renal lesions are less common but when present usually show microscopic polyangitis.

Pathology

The disease is usually due to type 3 hypersensitivity reaction to an external or internal antigen. An alternative cell-mediated mechanism postulated, is that macrophages are recruited to the vessel wall by lymphokines released from sensitized lymphocytes. The macrophages may then be activated to release lysosomal enzymes, with resultant vessel wall damage, at the same time the macrophages have the potential to transform into epitheloid cells with the formation of granulomas.[6] Laboratory findings are eosinophilia, raised ESR, increased a2 globulins. Circulating ANCA (Anti myeloperoxidase) is present in only 45% of cases. Optimal diagnosis is made by a careful skin biopsy examination. Treatment options are cortico-steroids and other immunosuppressive drugs. Addition of cyclophosphamide or other immunosuppressive drugs with very satisfactory prognosis has been recorded. Without treatment prognosis is grave, and 5 years survival is only 25%.

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