AN ATYPICAL PRESENTATION OF CHURG STRAUSS SYNDROME- A CASE REPORT.

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Abstract:
Churg strauss syndrome is a form of small vessel vasculitis characterised by bronchial asthma, peripheral and tissue eosinophilia, extravascular granulomas and vasculitis affecting multiple organ systems. It is an uncommon disorder with an estimated incidence of 1-3 per million per year. We report a case of churg strauss syndrome in a 16 yr old male, who presented with PUO and generalized lymphadenopathy.

Keyword: Churg strauss syndrome, PUO, generalized lymphadenopathy.

Case history
17 year old male was admitted with complaints of fever, breathlessness, abdomen pain and vomiting for 2 weeks. He was a known case of bronchial asthma on intermittent treatment since childhood on bronchodilators. He underwent a cardio thoracic surgery at the age of 1 year, details of which are not available. Clinical examination was unremarkable except for pyrexia and bilateral scattered wheeze on auscultation. Fever investigations were inconclusive. Patient was treated with bronchodilators and empirical antibiotics to cover respiratory pathogens. Patient improved symptomatically and became afebrile. He was discharged and advised to continue bronchodilators. Two weeks later patient came again with the complaints of intermittent fever and breathlessness. Clinical examination revealed generalized lymphadenopathy involving multiple cervical (1.5 cm), axillary (2 cm), epitrochlear (1 cm) and inguinal lymph nodes (2 cm), hepatomegaly apart from bilateral diffuse wheeze on chest auscultation. Examination of cardiovascular and central nervous system were normal. His complete blood count showed eosinophilic leukocytosis (absolute eosinophil count 1440/cubic mm) and elevated ESR. Blood urea, creatinine, serum electrolytes, liver function tests, urine routine and stool routine analyses were within normal limits. Chest x ray, ECG, echocardiogram were normal. Ultrasound abdomen showed hepatomegaly. Blood, urine and sputum culture were negative. Since the patient had fever with generalized lymphadenopathy we had a differential diagnosis of infections such as disseminated tuberculosis, toxoplasmosis, brucellosis, infectious mononucleosis, HIV infection, tropical pulmonary eosinophilia, lymphoproliferative disorders and connective tissue disorders. Excision biopsy of cervical lymph node was done and sent for histopathologic examination. Keeping the above differential diagnosis in mind we proceeded with further investigations. HIV elisa, sputum smear for acid fast bacilli, mantoux test, serum antibodies for toxoplasma, brucella, anti filarial antibody and serum antinuclear antibodies were negative. CT abdomen was normal with no evidence of intra abdominal lymphadenopathy. CT chest showed volume loss in left lung with compensatory hypertrophy of right lung and few ground glass opacities with parenchymal infiltrates. CT PNS showed features of maxillary sinusitis, which was done as patient had sinus tenderness.

CT PNS showing b/l maxillary sinusitis

CE CT ABDOMEN - NORMAL
CT CHEST showing b/l ground glass opacity with airspace infiltration
Cervical lymph node biopsy showed patchy areas of necrosis with focal palisading histiocytic aggregates, moderate eosinophilic infiltrates and evidence of vasculitis involving small blood vessels with destructive inflammation of vessel walls with lymphocytes, neutrophilic infiltrates and fibrinoid necrosis. AFB, TB and fungal stains of the biopsy specimen were negative.

BIOPSY REPORT
Other investigations were done subsequently. Anti ds dna, ANCA, rheumatoid factor, anti CCP were negative. Serum ferritin, C3, C4 levels were normal. Serum IgE levels were significantly elevated >2500. Bone marrow aspiration and biopsy were normal. A young male, known asthmatic presented with recurrent episodes of fever, generalized lymphadenopathy and sinusitis. Lymph node biopsy showed evidence of necrotizing small vessel vasculitis with extravascular eosinophil infiltrates. Peripheral smear study showed eosinophilia. CT chest showed transient ground glass infiltrates. The above clinical features were highly suggestive of churg strauss syndrome. Rheumatologist opinion was obtained, who also concluded that the clinical features were compatible with the diagnosis. our patient satisfied 5 out of the 6 criteria (proposed by ACR, cited below) for the diagnosis of churg strauss syndrome, which has specificity of 99%.

Patient was started on T Prednisolone 1 mg/kg. Patient responded dramatically with striking resolution of fever, lymphadenopathy and other constitutional symptoms. On discharge steroid dose was tapered to T Prednisolone 0.5 mg/kg. Patient was also started on cotrimoxazole for prophylaxis against pneumocystis carinii pneumonia and nocardiasis. Patient is on periodic follow up and doing fine.

Discussion:
Churg strauss syndrome was first described by churg and strauss in 1951, when they reviewed autopsy reports of patients who were previously diagnosed as polyarteritis nodosa (PAN) but had atypical features like asthma and extravascular granulomas. It is also called as allergic angiitis and granulomatosis. It is characterised by bronchial asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis affecting multiple organ systems. It is an uncommon disease with an estimated annual incidence of 1-3 per million. Vasculitis in CSS can affect any organ. Lung involvement is predominant. Other commonly involved organ systems include peripheral nervous system, skin, cardiovascular system, kidneys and gastro intestinal tract. The precise pathogenesis of this disease is unknown. Its association with asthma, peripheral and tissue eosinophilia, granuloma and vasculitis point to aberrant immunologic phenomena. Patients often exhibit non specific features such as fever, malaise, anorexia and weight loss. Pulmonary involvement is the most common clinical feature with severe asthmatic attacks and pulmonary infiltrates. Mononeuritis multiplex is the second most common manifestation (72%). Allergic rhinitis and sinusitis develop in 61%. Skin lesions include purpura and subcutaneous nodules (51%). Myocardial involvement (14%) is the most common cause of death. Generalised lymphadenopathy in churg strauss syndrome is uncommon but described in literature. The characteristic lab findings include striking eosinophilia, elevated ESR. 48% of patients have circulating anti neutrophil cytoplasmic antibody (ANCA) which is usually anti myeloperoxidase. The histologic features include necrotizing vasculitis involving small and medium sized muscular arteries, capillaries, veins and venules. A characteristic HPE feature is the granulomatous reaction in the tissues or vessel walls and infiltration of tissues with eosinophils. ACR criteria published in 1990 is most commonly used in the diagnosis.It has a sensitivity of 85% and specificity of 99%.

1. Bronchial asthma.
2. Eosinophilia >10% of the differential count.
3. Mononeuropathy or polyneuropathy attributable to vasculitis.
4. Transient pulmonary infiltrates attributable to vasculitis.
5. Paranasal sinus abnormality.
6. Extravascular eosinophils. If 4 out of 6 criteria are present, patient can be classified as a case of churg strauss syndrome.
Treatment
Glucocorticoids alone are effective in many patients. In patients with fulminant disease unresponsive to steroids alone a combination of steroids and cyclophosphamide can be used. Prognosis is favourable with treatment. Myocardial involvement is the most common cause of death.

Differential diagnosis
Diseases associated with pulmonary infiltrates and eosinophilia should be considered in the differential diagnosis such as Wegeners granulomatosis, drug allergy, fungal and parasitic infections, malignancy, eosinophilic granuloma, bronchocentric granulomatosis, allergic bronchopulmonary aspergillosis. Each of the above listed conditions can satisfy the ACR criteria for churg strauss syndrome but treatment differs for each of them. Hence it is extremely important to obtain tissue diagnosis whenever possible before commencing therapy. Our patient had biopsy evidence of vasculitis as well.

Conclusion
Churg strauss syndrome is an uncommon disease. Generalised lymphadenopathy in this disease is rare but described in literature. ACR criteria for the diagnosis has to be applied with caution as many diseases causing pulmonary infiltrates with eosinophilia can satisfy the criteria and mimic CSS but treatment differs markedly for each of them. Hence it is imperative to obtain tissue diagnosis whenever possible before labeling a patient as a case of CSS and commencing therapy.

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