Eosinophilic granulomatosis with polyangitis (Churg-Strauss) : A Rare Diagnosis

Introduction: Eosinophilic granulomatosis with polyangitis (Churg-Strauss Disease) is characterised by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems.

Case History: We describe a case of a 40 year old male, who presented with complain of bilateral lower limb tingling numbness and foot drop, sudden onset diminution of vision of left eye, acute onset breathlessness at rest and fever with chills.

Discussion & Conclusion: All necessary investigations were done. Patient was found to have peripheral eosinophilia, mononeuritis multiplex, left Central retinal artery occlusion and multiple pulmonary infiltrates with pericardial effusion. A diagnosis of Churg-Strauss Disease (CSD) was established. The patient was treated with steroids and cyclophosphamide. A high degree of suspicion is necessary to arrive at the diagnosis.

KEYWORDS
Eosinophilic granulomatosis with polyangitis, peripheral eosinophilia, asthma, systemic vasculitis, cyclophosphamide.

INTRODUCTION:
Eosinophilic granulomatosis with polyangitis was described in 1951 by Churg and Staruss and is characterised by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems.

It is one of the three important fibrinoid, necrotising, inflammatory leukocytoclastic systemic small vessel vasculitides that are associated with ANCAs. Of the three conditions, CSD is the least commonly encountered. The others are Wegener's Granulomatosis and Microscopic Polyangitis.

CSD is an uncommon disease with an estimated annual incidence of 1-3 per million. The disease can occur at any age, with the mean age of onset at 48 years, with a female-to-male ratio of 1.2:1.

The necrotising vasculitis of CSD involves small and medium sized muscular arteries, capillaries, veins and venules. A characteristic histopathologic feature of CSD is granulomatous reaction that may be present in the tissues or even within the walls of the vessels themselves. These are usually associated with infiltration of tissues with eosinophils. This process can occur in any organ of the body; lung involvement is predominant. Skin, cardiovascular system, kidney, peripheral nervous system and gastrointestinal system are most commonly involved. Although the precise pathogenesis of this disease is uncertain, its strong association with asthma and its clinicopathologic manifestations, including eosinophilia, granuloma and vasculitis, point to aberrant immunologic phenomena.

Patient with CSD often exhibit nonspecific manifestations such as fever, malaise, anorexia and weight loss. The pulmonary findings in CSD predominate with severe asthmatic attacks and pulmonary infiltrates. Mononeuritis multiplex is seen in 72% patients. Allergic rhinitis and sinusitis is seen in ~61%. Skin lesions that include purpura with cutaneous and subcutaneous nodules is seen in 51% patients. Heart disease is seen in ~14%. Renal disease is less common and generally less severe than that of granulomatosis with polyangitis and microscopic polyangitis. Striking eosinophilia is seen in CSD with Absolute Eosinophil Count >1000/µl. ESR is elevated. p-ANCA is positive in only 48% of the patients.

A thorough history and clinical examination can raise suspicion of CSD. According to the American College of Rheumatology Criteria, clinical diagnosis is established when 4 of the following 8 manifestations are documented:

1. Allergic history
2. Asthma
3. Eosinophilia
4. Migratory pulmonary infiltrates
Patient was started on higher IV antibiotics, furosemide, cardioselective beta blockers and IV methylprednisone.

On further questioning, the patient gave a past history of recurrent episodes of rhinitis and sinusitis.

Eventually, we arrived at the diagnosis of Eosinophilic Granulomatosis with Polyangitis (Churg-Strauss Disease). Pulse therapy of IV Cyclophosphamide was given.

The patient was discharged on oral prednisolone with a rare diagnosis of CSD.

DISCUSSION AND CONCLUSION:

Patient presented primarily with complain of bilateral lower limb tingling numbness, foot weakness, fever with chills, malaise and weight loss, acute breathlessness at rest. With a past history of recurrent rhinitis and sinusitis.

The investigations revealed peripheral eosinophilia, elevated ESR, CRP, LV dysfunction (EF-20%), multiple pulmonary ground glass opacities with interstitial septal thickening. Fundus examination revealed left Central Retinal Artery Occlusion and EMG-NCV was suggestive of mononeuritis multiplex. Skin biopsy revealed eosinophilic vasculitis.

Other differential diagnoses like pulmonary tuberculosis and granulomatosis with polyangitis were satisfactorily ruled out as 2 sputum AFB was negative, p-ANCA & c-ANCA was negative and there was an absence of renal involvement.

The above positive clinical findings sufficiently fit the diagnostic criteria of CSD.

The patient improved with high dose IV and oral prednisolone and IV cyclophosphamide.

A thorough history, meticulous clinical examination, laboratory investigations, imaging studies and a high level of clinical suspicion is necessary to diagnose CSD. Timely diagnosis and treatment are necessary for the patient’s well being.

REFERENCES:
3. Fishman’s Pulmonary Diseases and Disorders, (5th Edition)