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# EOSINOPHILIC GRANULOMATOSIS WITH POLYANGITIS (CHURG-STRAUSS) : A RARE DIAGNOSIS

Medicine

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## ABSTRACT

**Introduction-** Eosinophilic granulomatosis withpolyangitis (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 polyangits was described in 1951 by Churg and Staruss and is charaterised 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 48years, 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 exibit 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 Eosonphil Count>1000/microlitre. 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 dignosis is established when 4 of the following 8 manifestations are documented:

1. Allergic history
2. Asthma
3. Eosinophilia
4. Migratory pulmonary infiltrates
5. Paranasal sinus abnormality
6. Mononeuropathy or polyneuropathy
7. Clinical features consistent with vasculitis
8. Demonstration of extravascular infiltration of tissues on biopsy.

The prognosis of untreated CSD is poor, with a reported 5- year survival of 25%.With treatment, prognosis is favourable. Myocardial involvement is the most frequent cause of death in patients of CSD.

Glucocorticoids alone appear to be effective in many patients. Dosage tapering is often limited by asthma, and many patients require low dose prednisone for persistent asthma. In glucocorticoid failure or in patients who present with fulminant multisystem disease, particularly with cardiac involvement, the treatment of choice is a combined regimen of daily cyclophosphamide and prednisolone. Recent studies with Mepolizumab(anti- IL-5 antibody) in CSD have been encouraging, but this drug is still in its experimental stages.

## CASE HISTORY:

A 40 year old Hindu male, farmer by profession, presented with complain of tingling numbness in both lower limbs and decreased power in both feet, fever with chills, malaise and weight loss, since 2 months. He was admitted in the hospital, where clinical examination revealed pulmonary rhonchi on auscultation, power at both ankle joints of 2/5, decreased proprioception in toes and normal deep tendon reflexes. Rest of the neurological examination was unremarkable. Routine investigations were done, CBC showed leukocytosis with eosinophilia, with 52% eosinophils in differential count and absolute eosinophil count of 13936/microlitre. CRP was 56.6 IU. ESR was 40 mm at the end of first hour. Ferritin was elevated, 951microgram/litre. EMG-NCV was suggestive of mononeuritis multiplex (right sural, bilateral peroneal and tibial nerve). During the course of his stay, he developed complain of sudden onset painless loss of vision of left eye, fundus examination revealed Central Retinal Artery Occlusion in left eye.

After a few days he developed an episode of acute breathlessness at rest, a transthoracic echocardiogram was done that revealed anterior, lateral and inferior wall hypokinesia with an ejection fraction of 20% and pericardial effusion. Chest xray showed multiple pulmonary infiltrates, HRCT Thorax was done which showed soft tissue nodules with ground glass opacities and interstitial septal thickening of posterior basal segment of left lung and mild pericardial effusion.

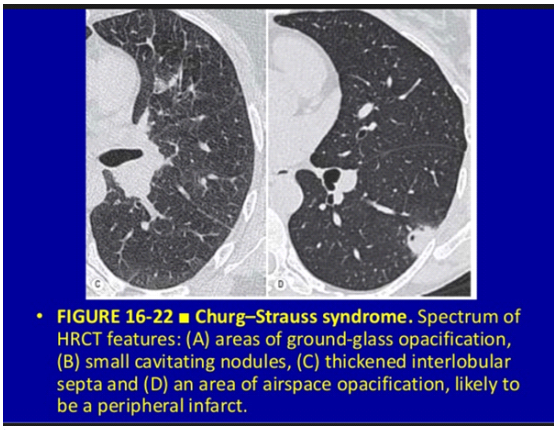
2 sputum AFB was negative. Renal function tests and urine R/M were within normal limits. HIV,HBsAg and HCV was negative. A provisional diagnosis of systemic vasculitides was kept and p-ANCA and c-ANCA tests were ordered that turned out to be negative. Skin biopsy revealed eosinophilic vasculitis.

Patient was started on higher IV antibiotics, furosemide, cardioselective beta blockers and IV methylprednisone.

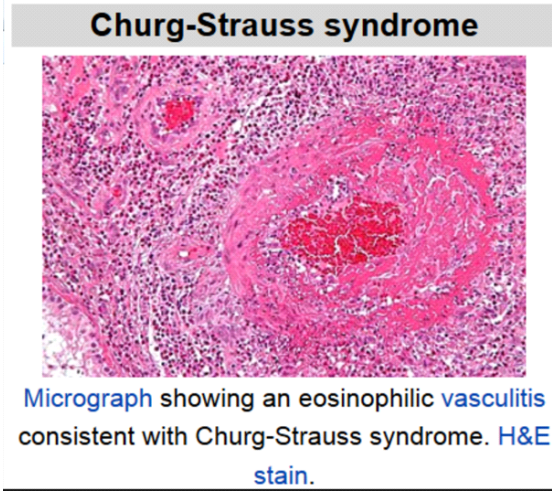
On further questioning, the patient gave a past history of recurrent episodes of rhinitis and sinusitis.

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.



- **FIGURE 16-22 ■ Churg–Strauss syndrome.** Spectrum of HRCT features: (A) areas of ground-glass opacification, (B) small cavitating nodules, (C) thickened interlobular septa and (D) an area of airspace opacification, likely to be a peripheral infarct.




Micrograph showing an eosinophilic vasculitis consistent with Churg-Strauss syndrome. H&E stain.

**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 necessry to diagnose CSD. Timely diagnosis and treatment are necessary for the patient's well being.

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