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A Rare Cause of Asthma with Eosinophilia - Churg Strauss Syndrome

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***Abstract*— Churg strauss syndrome is a rare disorder mainly affecting small and medium sized systemic blood vessels. Diagnostic characteristics include asthma, peripheral eosinophilia >10%, mononeuropathy multiplex or polyneuropathy, pulmonary infiltrates, paranasal sinus abnormality, biopsy showing blood vessels with extravascular eosinophilia [4]. Here we present the case of a 64 year old female who presented with complaints of giddiness, intermittent cough, weakness and paresthesia of both upper limbs and lower limbs and bilateral pedal edema. Absolute eosinophilic count was 26900, peripheral blood smear showed leucocytosis with eosinophilia, immunoglobulin E >1000 and computed tomography scan showing bilateral left lower lobe bronchiectasis and left upper lobe bronchiectasis which pointed towards the diagnosis of Churg strauss syndrome. Patient was treated with immunosuppresants and corticosteroids and she responded well.**

***Index Terms*— Churg Strauss syndrome (CSS), Eosinophilic granulomatosis with polyangiitis (EGPA), Eosinophilia, Bronchiectasis, Asthma.**

I. INTRODUCTION

Churg-Strauss syndrome is characterized by inflammation of blood vessels. This inturn will restrict the blood flow to various tissues and often causes permanent damages. The other name for this disorder is eosinophilic granulomatosis with polyangiitis (EGPA). This is a relatively rare condition and has no documented cure. Symptomatic control is usually achieved with steroids and other immunosuppressant medications. The cause of Churg-Strauss syndrome is unknown. The classical histological findings and its association with asthma help to differentiate from other vasculitides. Drugs like anti- leukotrienes are suggested to be associated with the development of this disorder eventhough the clinical rationale is questionable [1]. Known cause of eosinophilic pneumonia include Parasitic infestation like Ascaris, Filaria, Schistosoma, Toxocara, Strongyloides, drugs like minocycline, non-steroidal antiinflammatory agents, captopril, penicillamine, L-tryptophan, cocaine, allergic bronchopulmonary aspergillosis. Unknown causes include Churg-Strauss syndrome, cryptogenic pulmonary eosinophilia and hypereosinophilic syndrome [2].

The disorder is separated into three distinct phases – prodromal, eosinophilic and vasculitic. These phases may or may not occur sequentially. Some affected individuals will not develop all three phases. With proper treatment, Churg-Strauss syndrome can be successfully managed. Without treatment, the disorder may progress to cause life-threatening complications.

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II. CASE REPORT

A 64 year old female presented with complaints of giddiness, cough on and off, weakness and paresthesia of both upper limbs and lower limbs and bilateral pedal edema.

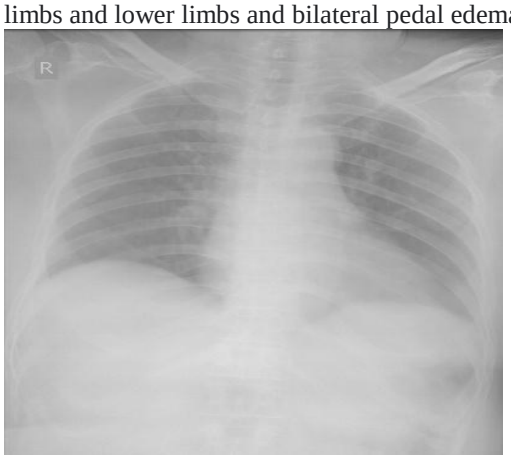
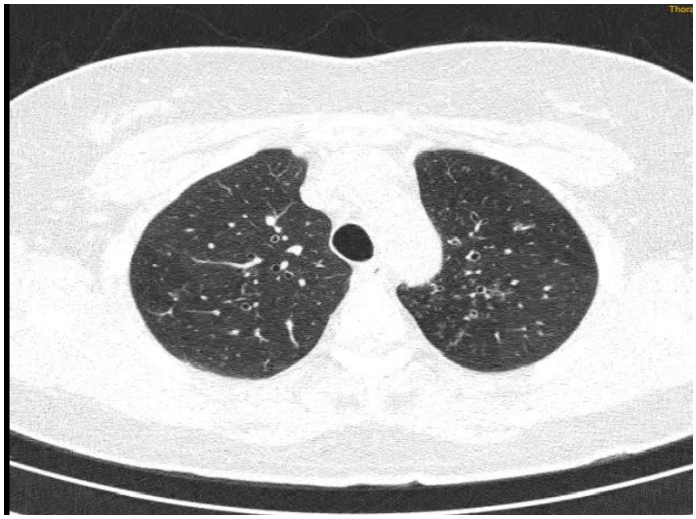


Figure 1- Chest x-ray at the time of initial presentation

Absolute eosinophilic count was 26900, Total count 43600, Peripheral blood smear showed leucocytosis with eosinophilia, Immunoglobulin E >1000 and raised erythrocyte sedimentation rate was 100. Serum electrophoresis showed hypergammaglobulinemia. CT scan showing bilateral left lower lobe bronchiectasis and left upper lobe bronchiectasis (Fig: 2 & 3).



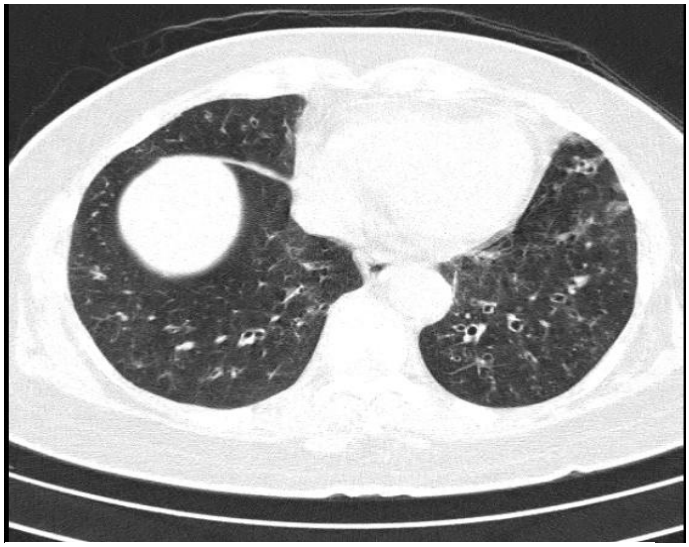


Figure 2 & 3- CT scan showing bilateral left lower lobe bronchiectasis and left upper lobe bronchiectasis.

She was diagnosed with pulmonary eosinophilia, ? Eosinophilic granulomatosis with polyangitis, Demyelinating polyneuropathy. Patient was treated with azathioprine, and tapering dose of oral prednisolone and inhaled corticosteroids.

Patient presented with complaints of nasal obstruction, mouth breathing, snoring, recurrent cold or headache after 3 years. Bilateral anterior middle turbinectomy, uncinectomy, bullotomy, anterior/ posterior ethmoidectomy was done. Nasal polyp occupying whole of nasal cavity was removed. Bilateral middle meatal antrostomy and bilateral sphenoidotomy was done. Bilateral sphenoid recess widened. Polypoidal nasal mucosa debrided completely. Polypoidal nasal mucosa from right frontal sinus removed completely. Biopsy showed tissue eosinophilia.

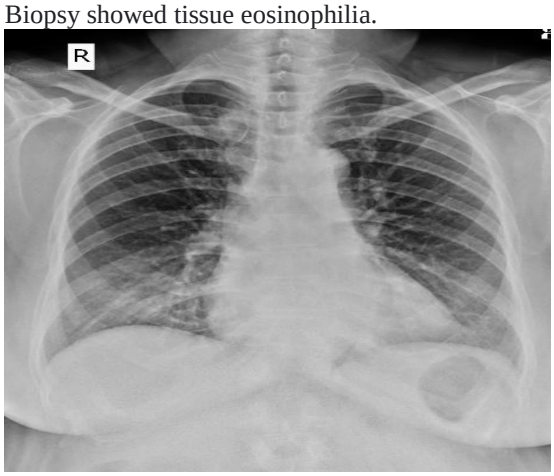


Figure 4- Chest x-ray after 3 years

III. DISCUSSION

The disorder can affect individuals of almost any age and has ranged from 15 to 70 years of age. Most cases occur in individuals between 30 and 50 years of age. The estimated mean annual incidence is 2.4 individuals per million.

As per Conron et.al, the estimated prevalence of Churg-Strauss syndrome (CSS), now known as eosinophilic granulomatosis with polyangiitis (EGPA), is 10.7 to 14 per million adults worldwide. There is no gender difference in the incidence of the disease [2,3].

Diagnostic criteria include asthma, peripheral eosinophilia >10%, mononeuropathy multiplex or polyneuropathy, pulmonary infiltrates, paranasal sinus abnormality, biopsy showing blood vessels with extravascular eosinophilia [4].

Most individuals with Churg-Strauss syndrome are first treated with corticosteroid medications such as prednisone or methylprednisonone, which suppress the activity of the immune system (immunosuppressive) and reduce inflammation. Individuals usually receive high-doses of corticosteroid initially and then after improvement is seen the dosage is slowly reduced (tapered off). Many affected individuals only need corticosteroids to achieve remission of their symptoms (monotherapy).

In 2017, Nucala (mepolizumab) was approved by the U.S. Food and Drug Administration (FDA) to treat adult patients with eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome). Nucala is manufactured by GlaxoSmithKline.

Complications of Churg-Strauss syndrome (CSS) include peripheral nerve damage, heart-related complications include inflammation of the membrane surrounding your heart, inflammation of the muscular layer of your heart wall, heart attack and heart failure and kidney damage [1].

Swietlik and Doboszynska et al suggested the use of inhaled corticosteroids as a recurrence preventing approach and this step was taken in our patient also [6]. In their most recent publication, Lamprecht et al summarized that anti-CD20 therapy and TNF-alpha-antagonists might represent strategies for refractory disease, but evidence from controlled trials is still lacking [7].

IV. CONCLUSION

Churg strauss disease is a rare pulmonary eosinophilic condition affecting lung, nervous system and blood vessels. Early identification of this condition is the key to achieve control and remission.

ABBREVIATIONS

CSD- Churg-Strauss disease.
CT- Computed tomography.
FDA- Food and Drug Administration.
EGPA- Eosinophilic granulomatosis with polyangiitis.

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