

Churg Strauss Syndrome: A Case Report

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I. Introduction

Churg–Strauss syndrome (CSS) is a small and medium vessel vasculitis characterized by eosinophilic infiltration of organs with necrotizing vasculitis and interstitial and perivascular granulomas. Three phases have been described in the natural history of the disease (prodromal, eosinophilic, and vasculitic phases) although they do not always occur successively. Initial records show CSS is a condition highly responsive to steroids.

American College of Rheumatology (ACR) has proposed six criteria for CSS—four being necessary for CSS to be diagnosed with 85% sensitivity and 99.7% specificity. Even though allergic asthma, rhinosinusitis and eosinophilia is a part of CSS, most reports consider this vasculitis a disease by itself or a variant of asthma which occurs from immune system interference such as with the use of medications such as leukotrienes or inhaled corticosteroids—both conditions resulting from sudden withdrawal of oral steroids in chronic severe asthma. The condition must be distinguished from aspirin-induced asthma (AIA), mould-induced allergy allergic bronchopulmonary aspergillosis (ABPA), allergy to drugs (such as minocycline) and parasitic infections.

II. Case Report

31 year old male, presented with history of difficulty in breathing off and on even at rest. Presently having low grade fever, cough with expectoration since 10 days. Patient was evaluated earlier and was given symptomatic treatment comprising of antibiotics, analgesics and steroids. Patient responded but was not relieved with the course of time patient developed rashes on lower abdomen along with bilateral upper limbs. Patient also developed paraesthesia in bilateral lower limb.

On admission, patient's general condition was stable with BP: 120/80, PULSE : 88bpm regular, bilateral lung field showed coarse crackles and rhonchi. Other systemic examination were normal.

On investigation, X-RAY CHEST revealed bilateral pneumonitis, CBC [WBC- 20.2×10^3 , HB-12.8, EOSINOPHILS- 49.2%, PLATELETS 2.18×10^5]. Kidney, Liver function tests and serum electrolytes were in normal range. Urinalysis showed haematuria (RBC =12-13), Stool for occult blood was positive. Coagulation profile was within normal range. HRCT CHEST suggestive of EOSINOPHILLIC PNEUMONITIS. Total Eosinophil Count (TEC)= 7812 cells/mm³ , Absolute Eosinophil Count(AEC) = 3906 cells/mm³. IGE Levels were markedly raised (>7418 IU/ml). NCV of bilateral lower limb revealed motor axonal mononeuropathy of right tibial nerve. Sputum C/S was sterile. ANA, p-ANCA, c-ANCA, C3 AND C4 were all negative. Skin biopsy was done which revealed mononuclear cell infiltration and at places blood vessels showing neutrophilic infiltration in their walls along with increased collagen in dermis.

Patient was given symptomatic treatment that is oral steroids and inhaled steroids. Patients general condition improved, had no respiratory problems and was discharged in satisfactory condition.

III. Discussion

Churg–Strauss syndrome was first described in 1951 by Churg and Strauss.

It is a rare systemic vasculitis (2.5 cases/100 000 adults/year) occurring exclusively in people with asthma and is associated with blood and tissue eosinophilia. The most commonly involved organ is the lung followed by the skin. CSS, however, can affect any organ system of the body.

The clinical features develop in several sequential phases:

1. Prodromal phase: Characterized by atopic disease, allergic rhinitis and asthma. Occurs in 2nd and 3rd decades.
2. Eosinophilic phase: Peripheral blood eosinophilia and eosinophilic infiltration of many organs and commonly lung, seen.
3. Vasculitic phase: Can have life-threatening sequelae and heralded by constitutional symptoms. Skin involvement common.

Asthma is the cardinal feature of CSS and precedes vasculitic phase. It presents as a chronic severe form and requires frequent or long-term courses of systemic steroids. Upper airway abnormality in the form of

allergic rhinitis, recurrent sinusitis, and nasal polyposis is fairly common. Involvement of skin is a frequent feature of the vasculitic phase and presents as tender subcutaneous nodules, palpable purpura and hemorrhagic lesions. Cardiac and neurological involvement is often seen; cardiac complications in the form of infarction and arrhythmias is responsible for 50% of deaths. Early diagnosis and treatment prevents organ damage and mortality. However confirming the diagnosis is difficult as individual manifestations occur in isolation and lung parenchymal involvement is not universal. Moreover, although classified as vasculitis, ANCA positivity is seen in only 40%-60% of patients. To add to the problem there is no laboratory tests specific for CSS.

The laboratory abnormalities are nonspecific and includes eosinophilia, high IgE, raised acute phase reactants, hypergammaglobulinemia. Therefore, a diagnostic criteria specified by ACR is most commonly used for diagnosis. A minimum of four criteria is required for a confident diagnosis of CSS. Our patient satisfies four.

The major histopathology findings of CSS are as follows:

- a) eosinophilic infiltration of tissue.
- b) extensive areas of necrosis;
- c) eosinophilic/giant cell vasculitis; and
- d) interstitial/pervascular necrotizing granuloma.

Skin biopsy typically reveals leukocytoclastic vasculitis with eosinophilic infiltrates.

Untreated CSS has a very dismal prognosis.

Treatment regimen is based on FFS (five factor score) highlighting organ involvement.

FFS includes following five factors:

1. Cardiac involvement
2. Gastrointestinal (GI) disease (bleeding, perforation, pancreatitis)
3. Renal insufficiency (Creatinine > 1.6 mg/dl)
4. Proteinuria (> 1gm/day)
5. Central nervous system (CNS) involvement (mononeuritis, polyneuropathy)

The duration of treatment is prolonged with induction of remission (steroids alone or in combination with cyclophosphamide) followed by maintenance treatment sometimes lasting for 12–18 months or longer (azathioprine being the preferred agent with steroids).

Prognosis with current series is encouraging with survival rate of 70% at 5 years. Of the five factors cardiac and GI involvement appears to have the worst prognosis.

IV. Conclusion

Churg–Strauss syndrome is a rare disorder characterized by hypereosinophilia and systemic vasculitis occurring in patients with asthma and allergic rhinitis. Vasculitis commonly affects the lungs, skin and peripheral nervous system. Outcome and long-term survival is usually good with steroids alone or in combination with immunosuppressive agents. The syndrome has a low mortality rate compared with other systemic vasculitides.