CASE REPORT

Churg Strauss Syndrome: A Rare Case With Common Presentation

Surendra Singh Bhakal[1], Akhil D. Goel[2]

ABSTRACT

Introduction: In 1951, Churg & Strauss first described this rare disorder. Also known as eosinophilic granulomatosis with polyangitis (EGPA), is characterised by presence of asthma, eosinophilia and small to medium vessels vasculitis. Our aim of reporting this case is to ensure high index of suspicion required for diagnosis of CSS.

Case Report: We report a case of 26 year male that presented with classical clinical and histopathological characteristic of Churg Strauss Syndrome and discuss briefly about this rare disorder.

Conclusion: CSS should be suspected in patients with asthma, eosinophilia and systemic clinical features suggestive of vasculitis.

Keywords: Churg Strauss Syndrome, vasculitis, purpura, eosinophilia

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[1]Senior Resident, Department of Medicine, [2]Senior Resident, Centre for Community Medicine, All India Institute of Medical Sciences (AIIMS), New Delhi

Corresponding author: Dr. Surendra Singh Bhakal, 3/73, Hanuman Bagh Colony, Nagaur, Rajasthan, 341001

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INTRODUCTION

In 1951, Churg & Strauss first described this rare disorder.[1] Also known as eosinophilic granulomatosis with polyangitis (EGPA), is characterised by presence of asthma, eosinophilia and small to medium vessels vasculitis.[2] Our aim of reporting this case is to ensure high index of suspicion required for diagnosis of CSS.

CASE REPORT

A 26 year male presented to the Medicine OPD, Sawai Man Singh Hospital, Jaipur with chief complaints of low grade intermittent fever for 1 month, tingling and numbness of both lower limbs for 15 days, productive cough for 10 days, bloody diarrhea for 8 days and reddish lesions on legs for 2 days. He was a known case of Bronchial asthma on intermittent steroid inhaler use and Montelukast for last three years. He underwent polypectomy for nasal polyp 2 years back.

On physical examination, pulse 98/minute, BP 130/90 mm of Hg, Respiratory Rate 28/minute, pallor present. On legs, reddish multiple non blanching lesions, elevated from skin (palpable purpura) present (figure-1) Respiratory system examination revealed bilateral wheeze and crepts in left infrascapular region. Other systems were normal.

Lab Investigations showed microcytic hypochromic anemia (Hb 6.9gm/dl, MCV 71.7 fl, Serum Iron 21 µg/dl), Eosinophilic leukocytosis (TLC 12700/µl; E40%), Total Eosinophil Count 3800/ µl, Elevated ESR (155 mm/1st hour) and CRP (69.6 mg/l) Bleeding time and Clotting time were normal. 24 hour urinary protein was 0.54 gm. Peripheral smear for MP was negative. MP card test, dengue serology, Widal was negative. Sputum AFB was negative.

Bilateral maxillary sinus haziness was evident on paranasal sinus radiograph. On chest X-ray left lower zone opacity present (Figure-2). HRCT chest confirmed left lower zone consolidation with air bronchogram (Figure-3). This steered our differential diagnosis towards eosinophilic pneumonias or vasculitis.

Nerve conduction velocity done in view of tingling and numbness, was suggestive of sensory motor axonal involvement of bilateral peroneal nerve and right median nerve indicating Mononeuritis Multiplex. This was not consistent with eosinophilic pneumonias and prompted us to investigate further for vasculitis. C-ANCA was negative but P-ANCA was positive with 1:640 dilutions by ELISA.

Skin biopsy of palpable purpura on legs revealed necrosed area infiltrated with eosinophils and neutrophils around and in the walls of vessels, suggestive of necrotising eosinophilic granulomatous vasculitis. On the basis of clinical findings, investigations and skin biopsy, diagnosis of Churg Strauss syndrome was made. He was treated with intravenous methyl prednisolone 500mg once a day for three days, there after maintained on oral prednisolone in tapering dose and

azathioprine 100 mg OD. Patient ~~well~~ responded well to treatment and was discharged with stable vitals.

DISCUSSION

American College of Rheumatology (ACR) selected 6 criteria for diagnosis of Churg Strauss Syndrome (CSS). The presence of 4 or more of these 6 criteria yield a sensitivity of 85% and specificity of 99.7%.[3] This patient satisfied all the 6 diagnostic criteria with Bronchial asthma, Eosinophilia, Paranasal sinusitis, Non-fixed Pulmonary infiltrates, Mononeuritis multiplex and vasculitis on skin biopsy.

Natural history is characterized by three phases- Prodromal phase: asthma and allergic rhinitis; Eosinophilic phase: eosinophilic tissue infiltration, with or without granuloma formation; Vasculitis Phase: necrotizing small vessel vasculitis in the peripheral nerves, skin, kidneys, lungs, heart, and gastrointestinal tract.[4]

Diagnosis of CSS is typically preceded by history of bronchial asthma by many years. In our patient, asthma was diagnosed three years back which was controlled on treatment. Lung involvement is radiologically characterised by waxing and waning parenchymal infiltrates which was also seen in our case.

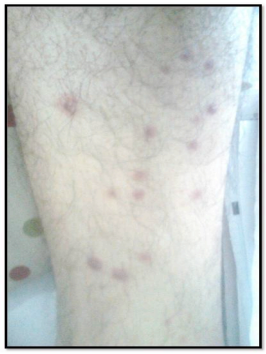
Neurological involvement manifests as mononeuropathy or polyneuropathy. Mononeuritis multiplex which was present in our case is considered most common neurological manifestation of CSS. Presence of Mononeuritis multiplex indicates poor outcome.[5]

Recognition of cutaneus manifestation like palpable purpura is important for early diagnosis as in our case. Frequent skin lesions seen in CSS are purpura, petechiae, papules, urticaria and livedo reticularis.[6,7]

Differential diagnosis includes eosinophilic pneumonias, allergic bronchopulmonary aspergillosis, hypereosinophilic syndrome, tropical pulmonary eosinophilia, and granulomatosis polyangitis. Only CSS and hypereosinophilic syndrome have a multisystem presentation, while tropical pulmonary eosinophilia and allergic bronchopulmonary aspergillosis show predominant involvement of lungs. Asthma or eosinophilia are not seen in granulomatosis polyangitis.[8]

The treatment is corticosteroid for limited disease. Pulse doses of intravenous corticosteroids combined with immunosuppressive agents like cyclophosphamide, azathioprine, and methotrexate is required in fulminant multisystem disease or relapse.[9]

The prognosis is good. Ten-year survival is observed in 80% of cases. Poor outcome is determined by the Five-Factor Score: azotemia (creatinine>1.5 mg/dl), proteinuria (>1g/dl), gastrointestinal tract involvement, cardiomyopathy, and central nervous system involvement.[5] The most common cause of death are myocardial infarction and myocarditis secondary to coronary vasculitis.[10]



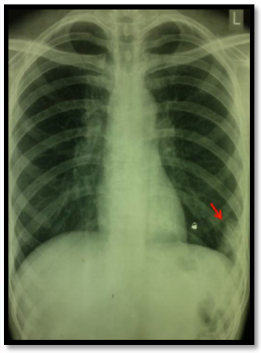


Figure-1: Purpura on lower limb; **Figure-2:** Left lower zone opacity on Xray Chest PA View

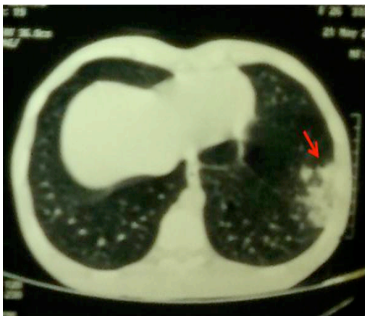


Figure-3: Left lower zone consolidation on HRCT Chest

CONCLUSION

The etiology of CSS is as yet unknown. CSS should be suspected in patients with asthma, eosinophilia and systemic clinical features suggestive of vasculitis. Background history of bronchial asthma usually presents before the diagnosis. Systemic manifestations include sinusitis, non-fixed pulmonary infiltrates, mononeuritis multiplex, palpable purpura and cardiac involvement. Cutaneus manifestations may prove pivotal role in establishment of diagnosis. Distinguishing CSS from other vasculitis is important as early intervention leads to better prognosis.

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