A CASE REPORT OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS, ALSO KNOWN AS CHURG-Strauss SYNDROME

Ajit Kumar Pegu1, Priyam Goswami2, Anupam Dutta3, Denleena Paul4, Harsh Vardhan5

1Associate Professor, Department of General Medicine, Assam Medical College and Hospital, Dibrugarh, Assam.
2Assistant Professor, Department of General Medicine, Assam Medical College and Hospital, Dibrugarh, Assam.
3Assistant Professor, Department of General Medicine, Assam Medical College and Hospital, Dibrugarh, Assam.
4Postgraduate Trainee, Department of General Medicine, Assam Medical College and Hospital, Dibrugarh, Assam.
5Postgraduate Trainee, Department of General Medicine, Assam Medical College and Hospital, Dibrugarh, Assam.

ABSTRACT

BACKGROUND
Eosinophilic granulomatosis with polyangiitis (Churg-Strauss), described by Churg and Strauss in 1951, is a rare systemic disease characterised by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems. It is a necrotising vasculitis of small and medium blood vessels. Its estimated annual incidence is 1 - 3 per million. It can occur at any age with possible exception of infants. Mean age of onset is 48 years. Female-to-male ratio is 1.2:1. A 55-year-old male presented to the Department of Medicine with a 6-month history of intermittent dyspnoea and wheezing and 10 days history of productive cough, low-grade fever with several episodes of haemoptysis. Physical examination revealed tachypnoea, tachycardia with wheeze and crackles on auscultation of the chest. Chest x-ray showed right upper lobe consolidation. The patient was suspected to be a case of sputum negative pulmonary TB and started on ATT under DOTS. He was readmitted when he presented with swelling of both lower limbs and decreased urine output. Jaundice had subsided, but urine for albumin was positive (+), blood urea was 243 mg/dL and serum creatinine was 10.6 mg/dL and there was microscopic haematuria. ATT was discontinued and patient underwent 2 sittings of haemodialysis. He came back with palpable purpuric rash over the back and lower limbs, which was typical of vasculitis. On examination, there was anaemia. CBC showed WBC of 43,900 with 70% eosinophiles. Nerve Conduction Study showed mononeuritis and axonopathy. There was elevation in CRP and ESR, ANA was negative but pANCA was positive (+++). We conclude by reporting a rare case of Churg-Strauss syndrome presenting with skin, lung, renal as well as peripheral nervous system involvement. Diagnosis of Churg-Strauss syndrome is based on clinical, pathological and laboratory correlates. Asthma is pivotal to the diagnosis and late in age of onset. The ultimate diagnosis is by biopsy in a patient meeting the clinical criteria. The histopathological confirmation can be challenging as the pathognomonic features often do not occur simultaneously.

KEYWORDS
Eosinophilic Granulomatosis, Polyangiitis, Churg-Strauss Syndrome.


BACKGROUND
Eosinophilic granulomatosis with polyangiitis (Churg-Strauss), described by Churg and Strauss in 1951, is a rare systemic disease characterised by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems. It is a necrotising vasculitis of small and medium blood vessels. Its estimated annual incidence is 1 - 3 per million. It can occur at any age with possible exception of infants. Mean age of onset is 48 years. Female-to-male ratio is 1.2:1. Most commonly involved organ is lung followed by skin along with involvement of any organ system.

Here, we present a case of a 55-year-old male with asthma, hyper-eosinophilia, fleeting pulmonary infiltrates with renal involvement, who presented with dyspnoea and haemoptysis and subsequently developed palpable purpuric skin lesions.

CASE REPORT
A 55-year-old male presented to the Department of Medicine on 16th January 2016 with a 6-month history of intermittent dyspnoea and wheezing and 10 days history of productive cough, low-grade fever with several episodes of haemoptysis. Physical examination revealed tachypnoea, tachycardia with wheeze and crackles on auscultation of the chest. Chest x-ray showed right upper lobe consolidation. Sputum for AFB was negative. The patient was suspected to be a case of sputum negative pulmonary TB and started on ATT under DOTS (Cat-1). The fever subsided and patient was discharged. Two (2) weeks later, he presented with jaundice and mild hepatomegaly. His ATT regimen was modified with isoniazid, ethambutol only and streptomycin was added.

Another week later, he was readmitted when he presented with swelling of both lower limbs and decreased urine output. Jaundice had subsided but urine for albumin was positive (+), blood urea was 243 mg/dL and serum creatinine was 10.6 mg/dL and there was microscopic haematuria. ATT was discontinued and patient underwent 2 sittings of haemodialysis. Serum creatinine level dropped to 3.6 mg/dL. Patient improved symptomatically and was discharged.

He came back with palpable purpuric rash over the back and lower limbs, which was typical of vasculitis. He had pain and paraesthesia in both lower limbs. On examination, there was anaemia. CBC showed WBC of 43,900 with 70%
Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) or EGPA is an uncommon. Approximately 60% of patients with a major form of vasculitis are recognised to have EGPA. Among the three anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitides (EGPA, granulomatosis with polyangiitis (Wegener’s) and microscopic polyangiitis), EGPA is least common. The mean age at diagnosis of EGPA is 40 years. EGPA is an uncommon cause of vasculitis in people older than 65 years, accounting for 5 percent of histologically proven vasculitis among 38 elderly patients with various systemic forms of angitis. EGPA is also rare in children and adolescents. When it does occur in this age group, it appears to follow a more aggressive course with prominent pulmonary and cardiovascular manifestations. EGPA does not exhibit gender predominance. A cross-sectional nationwide survey in Japan estimated the prevalence of EGPA at 17.8/1,000,000. The mean age at onset was 55 ± 14 years (±SD). Among the patients tested for myeloperoxidase (MPO) anti-neutrophil cytoplasmic antibody (p-ANCA), 50 percent were positive; however, only 2.5 percent were positive for proteinase 3 (PR3) c-ANCA. There was female predominance (2:1).

The mainstay of treatment is systemic glucocorticoids, initiated at a dose of 0.5 to 1.5 mg/kg per day. Higher dose is used for patients with more severe vasculitis. With acute multiorgan disease, intravenous glucocorticoid (e.g. Methylprednisolone 1 g for 3 days) is used for initial therapy followed by oral glucocorticoid therapy. It is gradually tapered over approximately 12 to 18 months as tolerated. In a study of 72 patients without poor prognostic factors (cardiac, renal or CNS involvement) followed for five years, 93% achieved remission with glucocorticoids alone. However, patients with heart, kidney, GIT or CNS involvement will require additional immunosuppressive therapy.

**DISCUSSION**

Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) or EGPA is a multi-system disorder characterised by asthma, vasculitis and peripheral blood eosinophilia (> 1500 cells/microl and/or >10% eosinophils on DLC). The exact aetiology and pathogenesis is unclear. ANCA are detected in 40 - 60% patients and EGPA is classified among ANCA positive vasculitis.

**Clinical Features Develop in 3 Phases**

a. Prodromal phase - characterised by atopic disease, allergic rhinitis and asthma, which usually precedes the vasculitic phase for around 8 - 10 years.

b. Eosinophilic phase - includes peripheral blood eosinophilia and eosinophil infiltration of multiple organs, especially lung and GIT.

c. Vasculitic phase - in the 3rd and 4th decades of life, a life-threatening systemic vasculitis of medium and small vessels occur, frequently associated with vascular and extravascular granulomatosis.

The epidemiology of EGPA remains unclear because of the uncertainties related to diagnosis. Approximately, 10 percent of patients with a major form of vasculitis are recognised to