

# Eosinophilic granulomatosis with polyangitis (Churg–Strauss syndrome): a diagnostic rarity with an atypical presentation

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We report a case of a 33-year-old woman who presented to us with symptoms of bronchial asthma and peripheral neuropathy. After investigations, the diagnosis of eosinophilic granulomatosis and polyangitis (Churg–Strauss syndrome) was made.

## Keywords:

antineutrophil cytoplasmic antibody, Churg–Strauss syndrome, revised nomenclature, systemic vasculitis

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## Introduction

Eosinophilic granulomatosis and polyangitis (EGPA) (Churg–Strauss syndrome) is a rare disorder characterized by the presence of asthma, eosinophilia, and small-to-medium-sized vessel vasculitis [1,2]. This case is presented to highlight the fact that EGPA is a diagnostic rarity among eosinophilic pneumonias and is difficult to diagnose particularly at an early stage. The rapid evolution of multisystem involvement is not the typical presentation in the natural history of Churg–Strauss syndrome. The patient also had a clinical syndrome that was antineutrophil cytoplasmic antibody (p-ANCA) positive. The literature is also reviewed in view of the recommendations of the 2012 revised international Chapel Hill Consensus Conference (CHCC2012) nomenclature of vasculitides.

## Case report

A 33-year-old woman presented with a history of cough for 5 months. Cough was intermittent, more at night, initially dry, and later with mucoid expectoration. A history of intermittent nocturnal wheeze was present for the last 5 months. The patient also had a history of hemoptysis for the last 2 weeks. There was no history of dyspnea. The patient had a history of pain in the right calf and ankle region for 2 months, followed by pain in the left ankle for the last 1 week. The pain was continuous and severe enough to disturb sleep. A history of paresthesia in the bilateral soles was present for last 1 month. A history of distal weakness in the left lower limb in the form of difficulty in walking and inability to clear the ground was present for the last 7 days. No history of radicular pains, claudication, arthritis, and swelling was present.

There was no history of fever, anorexia, malaise, weight loss, rash, and pleuritic chest pain. No similar complaints in the past were present. No significant family history was present. Review of other systems was normal. The patient was a housewife and had never gone out of the district. She had no significant history and had a normal menstrual history. There was no history of addictions. The general physical examination was normal. On chest examination, vesicular breath sounds with prolonged expiration and bilateral wheeze were observed. Neurological examination indicated asymmetrical distal sensory motor neuropathy in the form of left foot drop, absent ankle jerks, and sensory loss along the left sural nerve. The results of hemogram at admission are shown in Table 1.

On peripheral smear examination, red blood cell morphology was normocytic, normochromic, and eosinophilic leukocytosis was observed (Fig. 1). The biochemistry profile was normal and C-reactive protein was elevated. Urine examination indicated active urinary sediment in the form of leukocytes and erythrocytes and 24 h urine protein was 1065 mg. Stool examination for ova, parasites, and cysts was normal. Sputum examination showed intact and degenerated eosinophils, squamous epithelial cells, and alveolar macrophages and was negative for acid-fast bacillus. Serum immunoglobulin E level was 314 kUA/l (normal <64). Investigations for autoantibodies showed positive p-ANCA (antimyeloperoxidase). Antinuclear antibody and rheumatoid factor were negative. Pulmonary function tests documented a moderate degree of obstruction. Chest radiography showed bilateral pulmonary infiltrates (Fig. 2a). Frontal sinus haziness was observed in radiograph paranasal sinuses (Fig. 2b). High-resolution computed tomography chest showed areas of scattered ground glass opacities

in both lung fields with superimposed intralobular and interlobular thickening and yielded a crazy-paving pattern (Fig. 2c). On nerve conduction studies, axonal sensory motor polyneuropathy was observed. Sural nerve biopsy was suggestive of vasculitis. Kidney biopsy showed crescentric pauci-immune glomerulonephritis. On the basis of clinical findings and investigations, the diagnosis was EGPA. The patient was managed with oral prednisolone and pulse therapy of cyclophosphamide. The patient is under regular follow-up and the results of serial hemogram are shown in Table 1. Neurological deficit is improving and proteinuria has disappeared. Radiological clearance can be seen in the chest radiography taken during follow-up (Fig. 2d).

**Discussion**

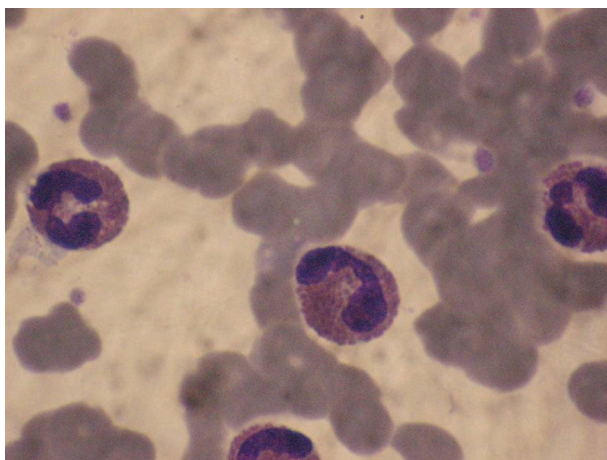
Medical knowledge is ever evolving as with advancements in the understanding of disease manifestations and pathogenesis. The disease names and definitions also

**Table 1 Serial hemogram before and after treatment**

Parameters	At admission	During follow-up		
		At discharge	At 1 month	At 3 months
Hemoglobin (g%)	11.3	11.4	12	11.6
TLC (cells/cm <sup>2</sup> )	41 000	15 600	11 800	7300
DLC (%)				
Polymorphs	28	53	62	61
Lymphocytes	9	32	31	33
Monocytes	1	1	1	1
Eosinophils	62	14	6	5
AEC (mm <sup>3</sup> )	25 420	2184	708	365
Platelets (10 <sup>3</sup> /mm <sup>3</sup> )	243	310	260	216
ESR (mm/h)	42	34	38	35

AEC, absolute eosinophil count; DLC, differential leukocyte count; ESR, erythrocyte sedimentation rate; TLC, total leukocyte count.

**Figure 1**



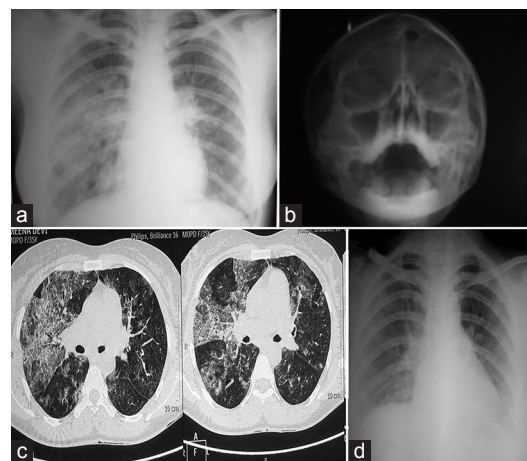
Photomicrograph showing eosinophils in the peripheral smear (Giemsa x1000).

evolve over time; thus, older nomenclature is replaced by a new one that is more relevant, useful, and updated. Advances in the understanding of vasculitis have led to the development of a revised nomenclature of vasculitides. The International Chapel Hill Consensus Conference (CHCC2012) adopted EGPA for Churg–Strauss syndrome. The eponym ‘Churg–Strauss syndrome’ was replaced by ‘EGPA’ in part to achieve nomenclature symmetry with microscopic polyangiitis and granulomatosis with polyangiitis (Wegener’s granulomatosis). Eosinophilia in the blood and tissue is an essential feature of EGPA and is thus highlighted in the name [1].

In natural history, the disease is characterized by a three-phase course, although not as a rule, and the phases may not occur successively. In the prodromal phase, asthma and/or allergic rhinitis with or without nasal polyposis precedes full development of the syndrome, usually by many years. In the next eosinophilic phase, tissue infiltration by eosinophils, with or without granuloma formation, occurs in various organs, particularly in the upper and lower respiratory tract, the gastrointestinal tract, and the myocardium. Finally, systemic vasculitis develops, with necrotizing small-vessel vasculitis clinically apparent in the peripheral nerves, skin, kidneys, lungs, heart, and gastrointestinal tract. The vasculitic phase distinguishes EGPA from other eosinophilic disorders [3].

Uniform diagnostic criteria are lacking for EGPA [2]. The clinical criteria of Lanham and colleagues [2,4–6], histopathology-based Chapel Hill criteria, and American College of Rheumatology-based classification criteria have been used as diagnostic tools.

**Figure 2**



(a) Chest radiograph showing bilateral pulmonary infiltrates. (b) Radiograph paranasal sinuses showing hazy frontal sinuses. (c) Computed tomographic chest showing areas of scattered ground glass opacities in both lung fields. (d) Chest radiograph showing radiological resolution during follow-up.

Antimyeloperoxidase p-ANCA is present in 30–40% of cases and appears to determine a subgroup of patients with a higher frequency of renal disease, alveolar hemorrhage, and central nervous system involvement [2]. Only 25% of patients with EGPA who have no renal disease are ANCA positive, whereas 75% with any renal disease and 100% with documented necrotizing glomerulonephritis have ANCA present [1]. ANCA-negative patients have cardiomyopathy, nonhemorrhagic pulmonary infiltrate, nasal polyposis, eosinophilic gastritis, or enteritis commonly [7]. Epidemiology of ANCA-associated vasculitis has shown geographical variation, ethnic, and sex differences. However, the prevalence of ANCA-associated vasculitis in India is unknown [8]. Immunoglobulin E levels are frequently elevated [3].

The differential diagnosis includes acute and chronic eosinophilic pneumonias, allergic bronchopulmonary aspergillosis, hypereosinophilic syndrome, tropical pulmonary eosinophilia, and granulomatosis polyangiitis (Wegener's granulomatosis). Only EGPA and hypereosinophilic syndrome have a multisystem presentation, whereas tropical pulmonary eosinophilia, acute and chronic eosinophilic pneumonia, and allergic bronchopulmonary aspergillosis show involvement of only the lungs in addition to blood eosinophilia. Granulomatosis polyangiitis does not present with either asthma or eosinophilia. In the absence of clinical or histologic evidence of vasculitis, it may be difficult to differentiate between EGPA and other systemic eosinophilic disorders [2].

The treatment is corticosteroid therapy alone for limited disease. Steroids are slowly tapered to a maintenance dose. Complete weaning off steroids should be done if possible. Maintenance dose is indefinitely required in some patients. An aggressive approach with pulse doses of intravenous corticosteroids combined with other immunosuppressive agents, such as cyclophosphamide, azathioprine, and methotrexate, is required in fulminant multisystem disease or during relapse. Regular monitoring of patients is performed even in the remission period [9].

The prognosis is good. Ten-year survival is observed in 80% of cases. Long-term remission occurs in 81–92% of patients. Relapses occur in 25% of patients that are more common within the first year. Poor outcome is determined by the Five-Factor Score: azotemia (creatinine >1.5 mg/dl), proteinuria (>1 g/dl), gastrointestinal tract involvement, cardiomyopathy, and central nervous system involvement [10].

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## Conclusion

EGPA should be suspected in patients with adult-onset asthma or worsening asthma, eosinophilia, and clinical features consistent with vasculitis.

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## Acknowledgements

### Conflicts of interest

There are no conflicts of interest.

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## References

- Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, *et al.* 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013; **65**:1–11.
- Comarmond C, Pagnoux C, Khellaf M, Cordier JF, Hamidou M, Viallard JF, *et al.* French Vasculitis Study Group Eosinophilic granulomatosis with polyangiitis (Churg–Strauss): clinical characteristics and long-term follow-up of the 383 patients enrolled in the French Vasculitis Study Group cohort. *Arthritis Rheum* 2013; **65**:270–281.
- Langford C. Clinical features and diagnosis of small-vessel vasculitis. *Cleve Clin J Med* 2012; 79 Suppl **3**:S3–S7.
- Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg–Strauss syndrome. *Medicine (Baltimore)* 1984; **63**:65–81.
- Jennette JC, Falk RJ, Andrassy K, Bacon PA, Churg J, Gross WL, *et al.* Nomenclature of systemic vasculitides. Proposal of an international consensus conference. *Arthritis Rheum* 1994; **37**:187–192.
- Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, *et al.* The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990; **33**:1094–1100.
- Kallenberg CG. Churg–Strauss syndrome: just one disease entity? *Arthritis Rheum* 2005; **52**:2589–2593.
- Koldingsnes W, Nossent HC. Epidemiology of ANCA associated vasculitis. *Norsk Epidemiologi* 2008; **18**:37–48.
- Campos LE, Pereira LF. Pulmonary eosinophilia. *J Bras Pneumol* 2009; **35**:561–573.
- Lhote F, Cohen P, Guilpain P, Guillevin L. Churg–Strauss syndrome. *Rev Prat* 2008; **58**:1165–1174.