

# Eosinophilic Granulomatosis with Polyangitis Presenting as Cardiac Tamponade

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## Abstract

Eosinophilic granulomatosis with polyangitis (EGPA; earlier called Churg-Strauss syndrome) is a small-vessel necrotising vasculitis typically characterised by asthma, lung infiltrates, extra-vascular necrotising granulomas and hyper-eosinophilia. Cardiac disease is a major contributor to disease-related deaths in EGPA. We describe the case of a 39-year-old woman with late onset asthma, allergic rhinosinusitis, and high extra-vascular and peripheral blood eosinophilia, presenting with peripheral neuropathy and pericardial effusion. Early therapy with intravenous corticosteroids led to resolution of the pericardial effusion and significant clinical improvement. The present case also highlights the importance of being vigilant about potentially fatal cardiac complications in patients with EGPA. [Indian J Chest Dis Allied Sci 2015;57:121-123]

**Key words:** EGPA, Pericardial tamponade.

## Introduction

Eosinophilic granulomatosis with polyangitis (EGPA; earlier called Churg-Strauss syndrome) is characterised by asthma, eosinophilia and vasculitis that chiefly affects the skin, peripheral nerves and lungs. Although the skin and the lungs are among the most commonly affected organs, pathological findings of necrotising eosinophilic vasculitis can be demonstrated in nearly all major organ systems. Cardiac involvement is an important, though an under-recognised feature in patients with EGPA.

## Case Report

A 39-year-old female was admitted with a 2-week history of diffuse chest pain, progressive dyspnoea on exertion and orthopnoea. She also complained of severe pain in the left calf and sole associated with paraesthesias. She was suffering from bronchial asthma for the past 10 years and was on regular treatment with inhaled corticosteroid, long-acting beta-2 agonist combination and leukotriene antagonist. She also gave a history of allergic rhinitis, nasal polyposis and sinusitis. On general physical examination, jugular venous pressure (JVP) was elevated. Respiratory system examination revealed scattered wheeze. On neurological examination reduced pinprick and vibration sense were noted in both lower limbs suggestive of peripheral neuropathy.

Laboratory examination revealed total leucocyte count  $14,000/\text{mm}^3$  with 44% eosinophils on differential leucocyte count; serum creatinine was 0.8 mg/dL.

Erythrocyte sedimentation rate (49 mm at the end of the first hour) and C-reactive protein (48 mg/L) were elevated. Urine analysis was normal. Chest radiograph revealed bilateral small pleural effusions (Figure 1) and echocardiography (Figure 2) revealed moderate pericardial effusion with the evidence of tamponade. Pericardiocentesis was carried out under echocardiographic guidance and 360 mL of pericardial fluid was drained.

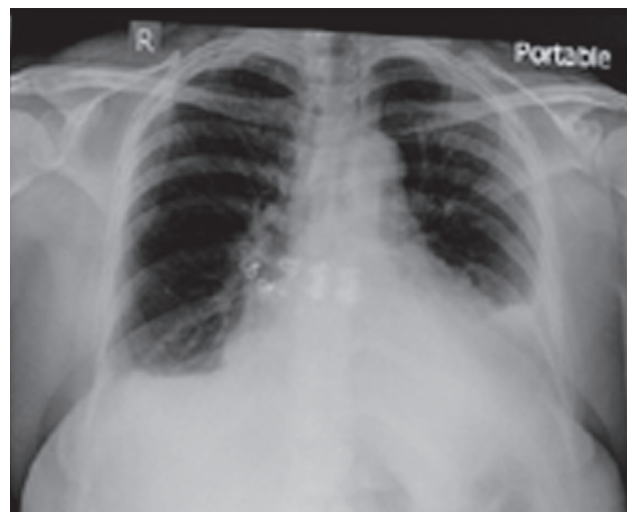


Figure 1. Chest radiograph (portable bedside view) showing bilateral pleural effusions.

Laboratory testing of pericardial fluid revealed protein 4.24 mg/dL; total leucocyte count  $3200/\text{mm}^3$  with 80% eosinophils, absolute eosinophil count  $3200/\text{mm}^3$ , adenosine deaminase (ADA) 35 IU/L,

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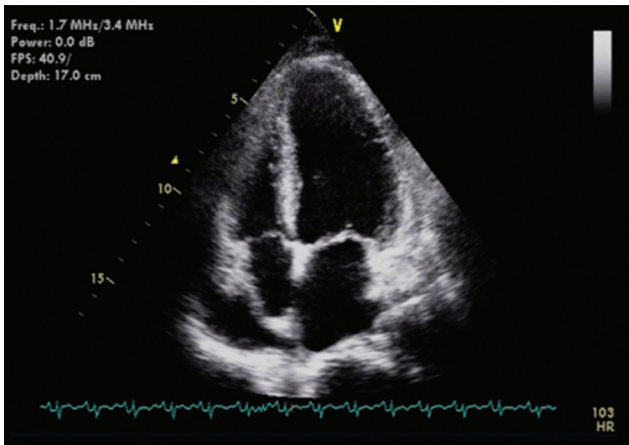


Figure 2. 2D-Echocardiography showing moderate pericardial effusion.

lactate dehydrogenase (LDH) 208 IU/L, cytology was negative for malignant cells and microbiology revealed that the fluid was sterile, Collagen vascular screen including rheumatoid factor (RF), anti-nuclear antibody (ANA) and anti-double stranded deoxyribonucleic acid (anti-dsDNA) antibody were all negative. Anti-neutrophil cytoplasmic antibody (ANCA) was also negative. Serum immunoglobulin E (IgE) levels were 470 U/mL (normal range <180 U/mL). Pulmonary function testing done after pericardiocentesis showed a moderate degree of airway obstruction with reversibility. Nerve conduction study revealed a mild motor axonal neuropathy involving the left tibial nerve.

On the basis of the background history of bronchial asthma, pericardial and pleural effusions, evidence of peripheral neuropathy and pericardial fluid eosinophilia the patient was diagnosed to have EGPA. She was started on intravenous methylprednisolone 40 mg 8 hourly for 5 days. This resulted in a dramatic clinical response with complete resolution of pericardial fluid and pain in the left calf and sole associated with paraesthesias. Her peripheral blood eosinophil count dropped to zero when re-checked after a week. She was discharged on oral prednisone (40 mg/day), azathioprine (100 mg/day) along with regular inhaled corticosteroids; the oral corticosteroid dosage was gradually tapered to 10 mg. She remains well at one year of follow-up with no recurrence of her pericardial effusion and has a normal eosinophil count.

## Discussion

First described in 1951 as an allergic and granulomatous angiitis, EGPA is a small-vessel vasculitis. Mean age at the time of diagnosis is approximately 50 years, with both genders being affected. Bronchial asthma is the central feature of EGPA and precedes the systemic manifestations in almost all cases, whereas 70% of the patients have

maxillary sinusitis, allergic rhinitis, and/or sinus polyposis. General symptoms are frequent, and associated with pulmonary infiltrates in 38% to 77% of the patients; peripheral neuropathy, usually mononeuritis multiplex, in 64% to 75%; skin involvement in 40% to 70%; and gastrointestinal tract symptoms in 37% to 62%.<sup>1</sup>

Cardiac involvement occurs in 50%, more frequently than with other vasculitides and is the major cause of early death, in the absence of any previous cardiac or pulmonary disease.<sup>2</sup> The pathogenesis is caused by vasculitis of myocardial or coronary vessels, eosinophilic myocardial infiltration or rarely extra-vascular granuloma formation. In contrast to eosinophilic cardiomyopathy (Loeffler's endocarditis), eosinophil infiltration and fibrosis is less intense and focal necrosis and vasculitis is seen. Cardiac disease is more common in ANCA-negative EGPA patients and in those with a higher eosinophil count which is consistent with the observation that such involvement is rare in ANCA associated granulomatosis with vasculitis and microscopic polyangitis. Endomyocarditis leads to cardiomyopathy with restrictive, congestive, or dilated patterns and myocardial infarction. Left ventricular failure (LVF) is common and both supra-ventricular and ventricular dysarrhythmias occur and can result in sudden death. Valvular abnormalities arise from sub-endocardial inflammation or fibrosis in particular mitral regurgitation and can be associated with pulmonary hypertension. Pericardial effusion secondary to pericarditis occurs in 20% and may progress to tamponade.<sup>3,4</sup> Constrictive pericarditis may occur over time. EGPA may present as isolated cardiac tamponade. Whereas pericarditis with myocardial injury warrants immunosuppressive therapy, isolated pericarditis without other visceral involvement only requires corticosteroid therapy.

Cardiac manifestations may be sub-clinical and may go unrecognized unless the patient with EGPA is actively worked up and investigated. In a recent study<sup>5</sup>, 32 consecutive patients with EGPA in remission who were previously unaware of cardiac involvement were compared with 32 randomly selected age- and gender-matched control subjects, using clinical evaluation, electrocardiography, echocardiography, and cardiac magnetic resonance imaging. Detailed cardiac evaluation revealed a 62% prevalence of cardiac involvement in EGPA patients compared with 3% in controls ( $p < 0.001$ ), with clinical symptoms in 26% and 3%, respectively ( $p = 0.009$ ).<sup>5</sup> A similar case with pericardial tamponade and acute myocarditis being the presenting manifestations of EGPA has been reported.<sup>5</sup> An interesting case report even documented oculomotor nerve palsy following cardiac tamponade in a patient with EGPA.<sup>6</sup>

In conclusion, cardiac involvement in EGPA occurs more commonly than suspected and may be, as in our

patient, the initial clinical manifestation of this generalised vasculitis. It may be potentially fatal or clinical suspicion is the key as steroids if promptly started can be life-saving.

## References

1. Guillevin L, Pagnoux C, Mouthon L. Churg-Strauss syndrome. *Semin Respir Crit Care Med* 2004;25:535-45.
2. Bourgarit A, Le Toumelin P, Pagnoux C, Cohen P, Mahr A, Le Guern V, et al. Deaths occurring during the first year after treatment onset for polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome: a retrospective analysis of causes and factors predictive of mortality based on 595 patients. *Medicine* 2005;84:323-30.
3. Neumann T, Manger B, Schmid M, Kroegel C, Hansch A, Kaiser WA, et al. Cardiac involvement in Churg-Strauss syndrome: impact of endomyocarditis. *Medicine* 2009;88:236-43.
4. Agard C, Rendu E, Leguern V, Ponge T, Masseur A, Barrier JH, et al. Churg-Strauss syndrome revealed by granulomatous acute pericarditis: two case reports and a review of the literature. *Semin Arthritis Rheum* 2007;36:386-91.
5. Dennert RM, Van Paassen P, Schalla S, Kuznetsova T, Apzand BS, Staessen JA, et al. Cardiac involvement in Churg-Strauss syndrome. *Arthritis Rheum* 2010;62:627-34.
6. Baili L, Aydi Z, Soussi G, Ben Dhaou BB, Zidi A, Berraies A, et al. Cardiac tamponade and myocarditis in Churg-Strauss syndrome. *Ann Cardiol Angeiol (Paris)* 2014;63:271-5.
7. Suganuma K, Hashimoto T, Sato H, Suzuki T, Sakurai S. Oculomotor nerve palsy following cardiac tamponade with Churg-Strauss syndrome: a case report. *Case Rep Neurol* 2011;3:274-7.