Multimodality imaging of Churg–Strauss myocarditis

Amit Sud, Roberta L. Brum, Kelly Victor, Panagiotis Koudounis, Gerald S. Carr-White and Ronak Rajani
Department of Cardiology, Guy’s and St Thomas’ NHS Foundation Trust, London, UK

Correspondence: Dr Ronak Rajani, Department of Cardiology, St Thomas’ Hospital, Westminster Bridge Road, London SE1 7EH, UK
Tel: +44 207 718 81004, fax: +44 208 399 4699, e-mail: Ronak.Rajani@gstt.nhs.uk

Abstract
A 26-year-old man presented with a 1-month history of hemoptysis, fever and chest pain. Investigations revealed a peripheral blood eosinophilia along with focal areas of lung consolidation on radiographic imaging. Subsequent echocardiography confirmed cardiac involvement, with a reduction in systolic function and a speckled appearance of the myocardium. The patient was diagnosed as having eosinophilic granulomatosis with polyangiitis and was treated with intravenous cyclophosphamide and corticosteroids. He made an excellent recovery and was subsequently discharged 4 weeks later. We present the radiographic and cardiac imaging findings of this condition along with a review of the value on cardiac screening in this patient population. Heart Metab; 2014;62:27–30

Keywords: Cardiac magnetic resonance; Churg–Strauss syndrome; echocardiography; eosinophilic granulomatosis polyangiitis; myocarditis.

Introduction
A 26-year-old man with a history of asthma presented to our institute with a 1-month history of intermittent hemoptysis, pleuritic chest pain and night sweats. Clinical examination revealed a tachycardia of 112 beats per minute and pyrexia of 37.9°C. The full blood count demonstrated an elevated total white cell count (16.4 × 10^9) with an abnormally elevated eosinophil count of 8.2 × 10^9. Serum biochemistry was normal apart from elevated serum troponin T measurement of 1366 ng/L. Given the clinical findings, the patient underwent an urgent 12-lead ECG and a chest X-ray. The ECG showed saddle-shaped ST-segment elevation suggestive of pericarditis, while the chest X-ray showed patchy consolidation in the left mid zone and coarse reticular marking in the mid zones bilaterally (Figure 1).

A computed tomographic pulmonary angiogram excluded the presence of pulmonary emboli but confirmed the presence of extensive lung parenchymal changes. The proximal airways were dilated with some opacification and distal dilatation. In addition, there was associated thickening of the bronchi and multifocal areas of ground glass opacification (Figure 2). These findings were suggestive of pulmonary hemorrhage along with eosinophilic pneumonia.

A subsequent transthoracic echocardiogram confirmed concomitant pericardial and myocardial involvement (Figure 3). There was a mild pericardial effusion and the myocardium was thickened (14 mm) with a heterogeneous speckled appearance indicative of edema. Left ventricular systolic function was mildly reduced at 40% and there was evidence of reduced long axis function.
Immunology demonstrated an elevated serum IgE level of 902 KU/L (normal range 0–81). There were no myeloperoxidase, anti proteinase-3, glomerular basement membrane, extractable nuclear antigens or antineutrophil cytoplasmic antibodies (ANCA). Given the elevated eosinophil count, elevated serum IgE level and imaging findings, a diagnosis of Churg–Strauss syndrome with myocardial involvement was made. The patient was promptly started on corticosteroids, an angiotensin converting enzyme (ACE) inhibitor and a β-blocker. Unfortunately, shortly thereafter he developed a right foot drop secondary to partial right common peroneal nerve damage, which initiated the start of intravenous cyclophosphamide. A cardiac magnetic resonance (CMR) scan performed 1 week later showed an improvement in systolic function to 55% and confirmed the myocardial involvement suggested by the transthoracic echocardiogram. On T2-weighted imaging there was evidence of circumferential enhancement indicative of myocardial edema. Following the administration of gadolinium, patchy areas of late enhancement were noted within the apex, anterior, septal and inferior walls with a transmurality of 25%. In addition, there were patches of intramyocardial late gadolinium enhancement within the mid ventricular septal and inferior walls (Figure 4).

The patient remained in hospital for a further 4 weeks during which his symptoms gradually improved. He was discharged home 5 weeks after his initial presentation. At follow-up 3 months later, he had received six infusions of cyclophosphamide, was on a reducing dose of prednisolone and was being maintained on an ACE inhibitor and β-blocker. His chest X-ray (Figure 5) and echocardiographic findings (Figure 6) had resolved and his foot drop had improved.

**Abbreviations**

ACE: angiotensin converting enzyme; ANCA: anti-neutrophil cytoplasmic antibodies; CMR: cardiac magnetic resonance; EGPA: eosinophilic granulomatosis with polyangiitis; FFS: five-factor score
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Discussion

In 1951, Churg and Strauss described a syndrome characterized by asthma and a "strikingly uniform clinical picture of fever and eosinophilia, and symptoms of cardiac failure, renal damage and peripheral neuropathy resulting from vascular embarrassment in various systems and organs" [1]. This entity was known as "Churg–Strauss syndrome" for many years but this has now been replaced by "eosinophilic granulomatosis with polyangiitis" (EGPA) [2].

The American College of Rheumatology has identified six criteria for the disease [3]. When at least four of these six criteria are met (asthma, eosinophilia >10%, neuropathy, non-fixed pulmonary infiltrates, paranasal sinus abnormalities and extravascular eosinophil infiltration) a vasculitis can be classified as being EGPA with a sensitivity of 85% and specificity of 99.7% [3]. EGPA is classically described as having three main sequential phases [4]. The first phase, or prodromal phase, usually consists of allergic rhinitis, nasal polyposis and airway irritability. The second phase, or eosinophilic phase, is characterized by peripheral blood eosinophilia organ involvement. The third phase, or vasculitic phase, is accompanied by clinical manifestations as a result of systemic necrotizing vasculitis.

Cardiac involvement

Cardiac involvement can occur in up to 62% of patients with EGPA [5] and is more common in ANCA-negative patients [6, 7]. Cardiac manifestations include restrictive or dilated cardiomyopathy, myocarditis, arrhythmias, valvar abnormalities, arrhythmias and sudden death. Dennert et al studied
32 consecutive patients in remission and found that 62% of patients had evidence of cardiac involvement. Clinical symptoms were present in 25%, major ECG abnormalities in 13%, echocardiographic abnormalities in 50% and CMR abnormalities in 62% of patients. The commonest ECG abnormality was the presence of T-wave changes (50%), while on echocardiography wall motion abnormalities (41%). CMR imaging was concordant with the echocardiographic findings in demonstrating wall motion abnormalities in 47% of patients. In addition, there was evidence of regional fibrosis in 22% and global fibrosis in 25% of patients. Fibrosis was only found in patients who had other cardiac abnormalities, and there was a good agreement between echocardiography and CMR imaging when fibrosis was excluded (sensitivity 88% and specificity 81%, respectively).

In general, the prognosis of EGPA is considered to be good, with an overall 10-year survival rate of 81–92% [8]. Up to 50% of EGPA mortality is caused by cardiac involvement, with up to 39% of patients dying during the acute phase of the disease [8]. In a recent study of 383 patients with EGPA followed up for a mean duration of 66.8 months, cardiac involvement determined by clinical evaluation, an ECG and echocardiography remained the greatest predictor of death with a hazard ratio of 4.11 (95% CI 1.96–8.60) [6]. Although there remains considerable interest in the detection of subclinical cardiac involvement with CMR and positron emission tomography, further longitudinal studies are required to determine their prognostic value [9]. Until these data are available, early clinical, ECG and echocardiographic screening is indicated for all patients with EGPA to detect early cardiac involvement.

Treatment
The choice of initial therapy is usually determined by the patient’s prognostic profile, as defined by the five-factor score (FFS) [10]. This includes cardiac, gastrointestinal and central nervous system involvement with proteinuria greater than 1 g/24 hours and creatinine greater than 140 μM/L. Patients with an FFS score of 1 or greater usually have a worse prognosis and are treated with corticosteroids and immunosuppressants. Corticosteroids are usually used in isolation for patients with an FFS score of 0. Although remission rates remain high with this approach, controversy still exists as to how many pulses of intravenous cyclophosphamide should be used, and relapse rates remain high for those patients receiving reducing doses of corticosteroids alone. There are no systematic trials evaluating the use of ACE inhibitors and β-blockers in EGPA-associated cardiac involvement.

Conclusions
The current case reports the multimodality imaging findings of a patient who presented with EGPA and demonstrates the value of early echocardiographic screening in these patients to identify early cardiac involvement to guide treatment.

REFERENCES