CASE REPORT

Cardiac involvement as the main presenting feature in eosinophilic granulomatosis with polyangiitis

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SUMMARY

Eosinophilic granulomatosis with polyangiitis is usually characterised by asthma, allergic rhinitis and peripheral eosinophilia. Presentations can vary greatly especially when there is cardiac involvement as demonstrated in these two case reports. Patient A initially presented to casualty with severe sinus pain and was diagnosed with severe sinonasal polyposis. After routine nasal polypectomy he had a cardiac arrest and was transferred to intensive care. Patient B presented to his general practitioner with a 4-week history of breathlessness, joint pain and a rash resulting in admission to hospital. Both patients had significant eosinophilia on routine bloods. High-sensitivity troponin T levels were raised in both; however, patient B's was significantly higher. Patient A had a large pericardial effusion on echo, the aspirate of which revealed numerous eosinophils. Patient B's echo was normal. Patient A's cardiac MRI was normal while Patient B's revealed myocarditis. Both were successfully treated with intravenous methylprednisolone and cyclophosphamide.

BACKGROUND

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome is a multisystem disorder characterised by asthma, allergic rhinitis and prominent peripheral blood eosinophilia.¹ The most commonly affected organs are the lungs and skin. It can however affect many other organs including the heart. Cardiac involvement is one of the more serious manifestations of EGPA accounting for approximately 50% of deaths from the condition.² Despite this, the diagnosis of EGPA can often be delayed when cardiac pathology is the major presenting symptom. Presentation can be very variable as demonstrated in these two case reports. They illustrate the importance of maintaining a high index of clinical suspicion of EGPA as when untreated it is potentially life threatening. Early aggressive treatment with corticosteroids and cyclophosphamide can result in improved outcomes.

CASE PRESENTATION

Patient A, a 33-year-old man with a previous history of childhood asthma, presented to ENT casualty with severe nasal pain. He had multiple failed treatments for sinusitis, including two courses of oral steroids which provided dramatic but temporary relief. Systematic questioning and medical history were otherwise unremarkable. He had an obvious mass in the right nasal passage and was admitted for intravenous antibiotics, analgesia and

CT scan of sinuses. The CT showed extensive soft tissue hypertrophy throughout all sinuses in keeping with severe sinonasal polyposis. A routine polypectomy was performed but during the procedure he became bradycardic, his QRS complexes widened and the rhythm degenerated into ventricular fibrillation. Sinus rhythm was restored with one synchronised DC shock and he was transferred to intensive care.

INVESTIGATIONS

Echocardiogram postarrest (figure 1) revealed a large circumferential pericardial effusion and 700 ml of straw coloured fluid was aspirated on pericardiocentesis. Coronary angiogram was negative for significant obstructive disease. Erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were 41 mm/h (Westergren) and 26 mg/dL, respectively. Troponin T high-sensitivity (TNT Hs) levels peaked at 60 ng/L. Antineutrophil cytoplasmic antibodies (ANCA), antinuclear antibodies, extractable nuclear antigens, serum ACE and antiglomerular basement membrane (anti-GBM) antibodies were negative. Urinalysis and urine microscopy were normal. Chest X-ray (CXR) and pulmonary function tests (PFTs) were normal. The pericardial fluid analysis revealed an exudate with mesothelial cells and numerous eosinophils. A differential white cell count revealed an eosinophil count of $2.34 \times 10^9/L$ which had been persistent since his initial presentation. Histological examination of the nasal biopsy specimens revealed vascular and perivascular inflammation with granulomatous matter, consistent with a vasculitis. He was diagnosed with EGPA.

TREATMENT

Disease remission was attained with pulsed intravenous methylprednisolone and intravenous

Figure 1 An echocardiogram showing a large circumferential pericardial effusion.

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cyclophosphamide with concomitant oral prednisolone following the CYCLOPS regime.³

OUTCOME AND FOLLOW-UP

Subsequent cardiac MRI showed no significant areas of myocardial involvement and normal ventricular function. He remains well on maintenance mycophenolate mofetil and a tapering course of oral prednisolone.

CASE PRESENTATION

Patient B, a 55-year-old man was admitted to the general medical take with a 4-week history of dyspnoea, nasal congestion, arthralgia and a well-circumscribed erythematous and blistering rash on his back and buttocks. Routine investigations by his general practitioner revealed an eosinophil count of $15.75 \times 10^9/L$ and elevated creatine kinase of 370 U/L and TNT Hs of 165 ng/L. He had a medical history of asthma, nasal polyps and over the previous year a non-specific skin rash for which he had previously attended a dermatologist.

INVESTIGATIONS

Admission bloods revealed a peripheral eosinophilia of $16.67 \times 10^9/L$. ESR and CRP were 26 mm/h (Westergren) and 49 mg/dL, respectively. Urea and electrolytes were normal. ECG was normal and echocardiogram revealed only a trace of pericardial fluid. On CXR he had left basal consolidation but CT chest showed patchy ground glass densities in both upper lobes.

Following rheumatological assessment a diagnosis of EGPA was suspected. Further investigation included MRI of the brain/sinuses which showed sinusitis. Tests for rheumatoid factor, ANCA and anti-GBM antibodies, complement and serum ACE were negative. PFTs revealed a mild obstructive defect. Skin biopsy was non-diagnostic.

Troponin levels continued to rise, and peaked at 1179 ng/L. He denied any chest pain. Given the clinical history and absence of abnormalities on echocardiogram, a cardiac MRI (figure 2) was performed which revealed oedema in the subendocardial region of the wall of the apex on T2 imaging consistent with myocarditis. There was persistence of gadolinium enhancement in the same region suggesting a degree of myocardial infiltration and possible scarring.

TREATMENT

He improved symptomatically and TNT Hs levels fell to <200 ng/L following pulsed intravenous methylprednisolone. He was also started on low-dose bisoprolol and ramipril while

continuing with methylprednisolone and cyclophosphamide using the CYCLOPS regime.

OUTCOME AND FOLLOW-UP

He currently remains well and subsequent cardiac MRIs have shown no persistent myocardial changes or persistent gadolinium enhancement.

DISCUSSION

These cases demonstrate the variability of cardiac involvement and subsequent presentation in patients with EGPA. In a multi-centre, cross-sectional analysis of patients with EGPA, 45% had cardiac involvement based on ECG, echo and MRI. Valvular insufficiency, pericardial effusions, heart failure and myocardial involvement were noted in 73%, 50%, 41% and 24%, respectively.⁴ Diagnosis can be challenging. Patients typically have a shorter duration of symptoms and are less likely to be ANCA-positive when cardiac disease is the main feature.² As stated previously cardiac involvement accounts for approximately 50% of deaths from EGPA. Early aggressive treatment with corticosteroids and cyclophosphamide can result in improved cardiac function and overall outcome, highlighting the importance of remembering this serious manifestation so that it can be recognised and treated promptly.⁵

Learning points

- ▶ Cardiac involvement is one of the more serious manifestations of eosinophilic granulomatosis with polyangiitis accounting for approximately 50% of deaths from the condition.
- ▶ Cardiac involvement can be very variable.
- ▶ Patients with cardiac involvement are less likely to be antineutrophil cytoplasmic antibodies positive.
- ▶ Early diagnosis and treatment may lead to improved outcomes, highlighting the importance of maintaining a high index of clinical suspicion.

Contributors The patients included in the case were managed by AP AM. The article was written by NM and was reviewed by AM and AP who gave final approval for the article.

Competing interests None.

Patient consent Obtained.

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Figure 2 A cardiac MRI showing high signal in the subendocardial wall on T2-weighted images consistent with active myocarditis.

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