Cardiac eosinophilic granulomatosis with polyangiitis: rapid imaging with contrast CT and contrast echo aids early diagnosis

A 49-year-old male asthmatic (on Montelukast) with multiple sclerosis presented with shortness of breath and generalised weakness. On examination, he was in cardiac failure with a systolic murmur, rapid atrial fibrillation and weakness in all four limbs with slurred speech. His white cell count was $37 \times 10^9 / L$, eosinophils 25×10⁹/L, urea 13.6 mmol/L, creatinine 131 μmol/ L, troponin I 4448 ng/L, anti-neutrophil cytoplasmic antibodies (ANCA) weakly positive (negative myeloperoxidase (MPO) and proteinase 3 (PR3)). Differential diagnoses included stroke, myocarditis and eosinophilic granulomatosis with polyangiitis (EGPA). Contrast echocardiography showed massive hypertrophy of the right ventricular apex and basal segments of the left ventricle (figure 1A) with severe mid-cavity obstruction (95 mm Hg gradient). A contrast CT of the chest showed right lung nodules and hilar lymphadenopathy, and confirmed the myocardial infiltration (figure 2). Brain MRI revealed multiple ischaemic foci throughout both cerebral hemispheres in keeping with a vasculitic aetiology. A clinical diagnosis of EGPA (formally Churg-Strauss syndrome) with myocardial eosinophilic infiltration was made. Cardiac failure was treated with diuretics and beta blocker; the eosinophil count settled with pulsed intravenous methylprednisolone followed by cyclophosphamide.

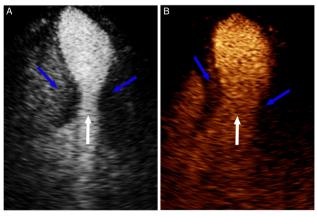


Figure 1 (A) Contrast echocardiography showing hypertrophy of the basal segments of the left ventricle (blue arrows) with mid-cavity obstruction (white arrow). (B) Contrast echocardiography 11 months later showing resolution of infiltration (blue arrows) and mid-cavity obstruction (white arrow).

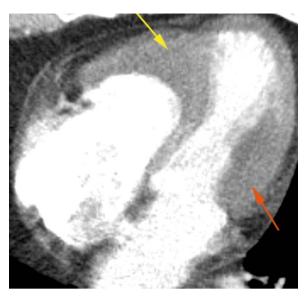


Figure 2 Contrast CT showing eosinophilic infiltration and hypertrophy of right ventricular apex (yellow arrow) and lateral wall of left ventricle (red arrow).

Cardiac involvement in EGPA has a poor prognosis—accounting for half of the deaths attributed to EGPA, and is seen in those with very high eosinophil counts. Imaging modalities include cardiac MRI with late gadolinium. In this case, contrast echocardiography and contrast CT provided early complimentary functional and anatomical information. Slow resolution of the left ventricular outflow tract obstruction (reduced to 6 mm Hg) and myocardial infiltration (figure 1B) were followed with contrast echocardiography alone.

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Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

► Additional material is available. To view please visit the journal online (http://dx.doi.org/10.1136/heartasia-2015-010714).



To cite Abraham D, Freeman LJ. *Heart Asia* 2016;8:21. doi:10.1136/heartasia-2015-010714

Heart Asia 2016;8:21. doi:10.1136/heartasia-2015-010714

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