

Cardiac involvement as a presenting feature of eosinophilic granulomatosis with polyangiitis in childhood

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Eosinophilic granulomatosis with polyangiitis (EGPA) is considered primarily a disease of adulthood with a mean age of symptomatic onset of around 40–45 years in most case series (1). Cardiac involvement in EGPA affects between 17 and 92% of adult patients and may account for as many as half of the deaths, particularly in the early stages (2). Nonetheless, cases of paediatric EGPA have been reported more frequently and cardiac involvement seems to be an important cause of mortality (3, 4). We present a 14-year-old Caucasian girl who was found to have significant cardiac involvement on diagnosis with EGPA. She developed exercise-induced cough and wheeze that initially improved with corticosteroid and salbutamol inhalers however she subsequently required several hospital admissions with cough and pleuritic chest pain. She responded to oral glucocorticoids and antibiotics but deteriorated upon discontinuation of glucocorticoids.

She demonstrated eosinophilia (more than 50% of total white cell count) and elevated cardiac troponin levels but negative ANCA testing. A chest CT scan showed interstitial shadowing with multi-lobar consolidation, pleural and pericardial effusions (Fig. 1). Echocardiography confirmed severely impaired left ventricular function.

She received pulsed intravenous methylprednisolone but continued to deteriorate with signs of cardiogenic shock. Repeat echocardiography showed further deterioration of left ventricular systolic function (ejection fraction 8–11%). She was transferred to the intensive care unit for inotropic and ventilatory support along with high dose oral glucocorticoids, intravenous immunoglobulin and rituximab (two doses of 1g each). She commenced maintenance mycophenolate mofetil alongside oral glucocorticoids and made a gradual recovery. After 9 months her eosinophilia had normalised with significant improvement in cardiac function on echocardiography (left ventricular ejection fraction 61%).

Although paediatric EGPA is rare, around 50% of those reported in the medical literature demonstrate evidence of cardiac involvement and at least 40% of deaths are related to cardiac complications (3, 4). Reported manifestations include pericardial effusions, cardiomyopathy, myocarditis, valvular lesions and cardiac thrombosis (3, 4). Our understanding of paediatric EGPA is still developing. Whether patterns of disease reported amongst adults are generalis-



Fig. 1. Chest CT scan showing perihilar consolidation with pericardial and right-sided pleural effusion.

able to those seen amongst younger patients is still not entirely clear (5). There do appear to be some important differences in the reported literature. At least 75% of children with EGPA are ANCA negative which is a larger proportion than amongst adults (3, 4). Furthermore the rates of cardiac and pulmonary involvement may be higher amongst children with lower rates of neurological involvement than adults (4, 6).

More extensive cardiac investigations including serum cardiac biomarkers (for example troponin levels), electrocardiography, echocardiography and cardiac MRI are important amongst children in view of these different patterns of disease progression and organ involvement.

Contrast-enhanced cardiac MRI has been used in adults with EGPA to non-invasively distinguish between irreversible fibrosis (which typically displays patterns of late enhancement that remain after treatment) and active inflammation (which usually manifests as sub-endocardial oedema and early enhancement that resolves after treatment) (7). Cardiac MRI has been used to find sites for biopsy or even prevent the need for these to be done in some cases. Our patient was too unwell to undergo cardiac MRI at the time of diagnosis but this has provided a useful adjunct in the assessment of disease activity during follow-up. This case adds to an emerging number of paediatric patients with cardiac involvement in EGPA. As in adults, cardiac disease in EGPA is an important cause of mortality and we need to carefully screen for cardiac disease in all paediatric patients and to gain more experience using imaging techniques such as cardiac MRI in the assessment of disease activity non-invasively. Early recognition of active disease with prompt immunosuppressive treatment is vital in preventing disease progression.

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