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Brief Report

Left ventricular mass in Wegener's granulomatosis: a brief report

Sunita J. Ferns,[1] Nguyenvu V. Nguyen,[2] Hyde M. Russell,[3] Carl L. Backer[4]

[1]Hope Children's Hospital, University of Illinois at Chicago, Illinois; [2]Division of Pediatric Cardiology; [3]Division of Cardiovascular and Thoracic Surgery, Children's Memorial Hospital, 2300 Children's Plaza; [4]Division of Cardiovascular and Thoracic Surgery, Children's Memorial Hospital, Northwestern University's Feinberg School of Medicine, Chicago, Illinois, United States of America

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WEGENER'S GRANULOMATOSIS IS A SYSTEMIC necrotising small vessel vasculitis of unknown aetiology, characterised by granulomatous lesions.[1] The prevalence in the general population has been reported to be 20–50 per million.[2,3] Although the most frequently involved organs are the respiratory tract and kidneys, virtually any system in the body can be affected. With advances in imaging techniques, cardiac involvement is increasingly being recognised.[4] The coronary arteries, pericardium, myocardium, endocardium, valves, conduction system, and great vessels are all reported to be involved.[5] Presentation in the form of a cardiac mass is extremely uncommon.[6] We report a patient with Wegener's granulomatosis with a left ventricular mass.

Case report

A 14 year-old girl presented with a 4-week history of low-grade fevers, nasal congestion, intermittent epistaxis, weakness, and joint pains. On examination at admission she was pale with mild swelling of her ankle joints and purpuric lesions on her toes and distal metacarpophalengeal joints. Initial investigations were suggestive of renal failure with a blood urine nitrogen of 105 milligrams per decilitre, and a creatinine of 10 milligrams per decilitre. Within the next few hours she developed symptoms of fluid overload and respiratory distress. She was transferred to the intensive care unit for initiation of ventilation. Haemodialysis was initiated and respiratory support was successfully weaned with improvement in her fluid status while on dialysis. Given the symptoms of sinusitis, arthralgia, vasculitic rash, and acute renal failure, a preliminary diagnosis of Wegener's granulomatosis was made. This was later confirmed by a positive C-anti-neutrophil cytoplasmic antibody pattern and a renal biopsy and therapy with plasmapheresis, cyclophosphamide and methylprednisolone was initiated. An echocardiogram was obtained due to concerns of embolic skin manifestations at initial presentation and showed mildly depressed left ventricular systolic function, ejection fraction 40%, and a moderate size mobile pedunculated mass in the left ventricular apex (Fig 1a, b). A cardiac magnetic resonance imaging study was performed to better delineate boundaries of the mass and to evaluate for additional lesions (Fig 2a, b). Because of the concern for embolisation, the patient was taken to the operating room to resect the left ventricular mass. On cardiopulmonary bypass with cardioplegic arrest, the aorta was opened and the mass was approached through the aortic valve. A 5-millimetre, 30 degree thoracoscope was used to aid visualisation of the left ventricular apex. There was a pedunculated light tan to white soft tissue mass in the apex of the left ventricular cavity measuring 1×1.1 centimetre and adherent to the left ventricular trabeculations. The mass was excised with biopsy forceps, and the left ventricular cavity irrigated and examined for additional lesions. Transeosophageal

Correspondence to: S. J. Ferns, MD, MRCPCH, Division of Pediatric Cardiology, Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60613, USA. Tel: 001 312 545 4837; Fax: 001 773 880 8111; E-mail: sferns@childrensmemorial.org

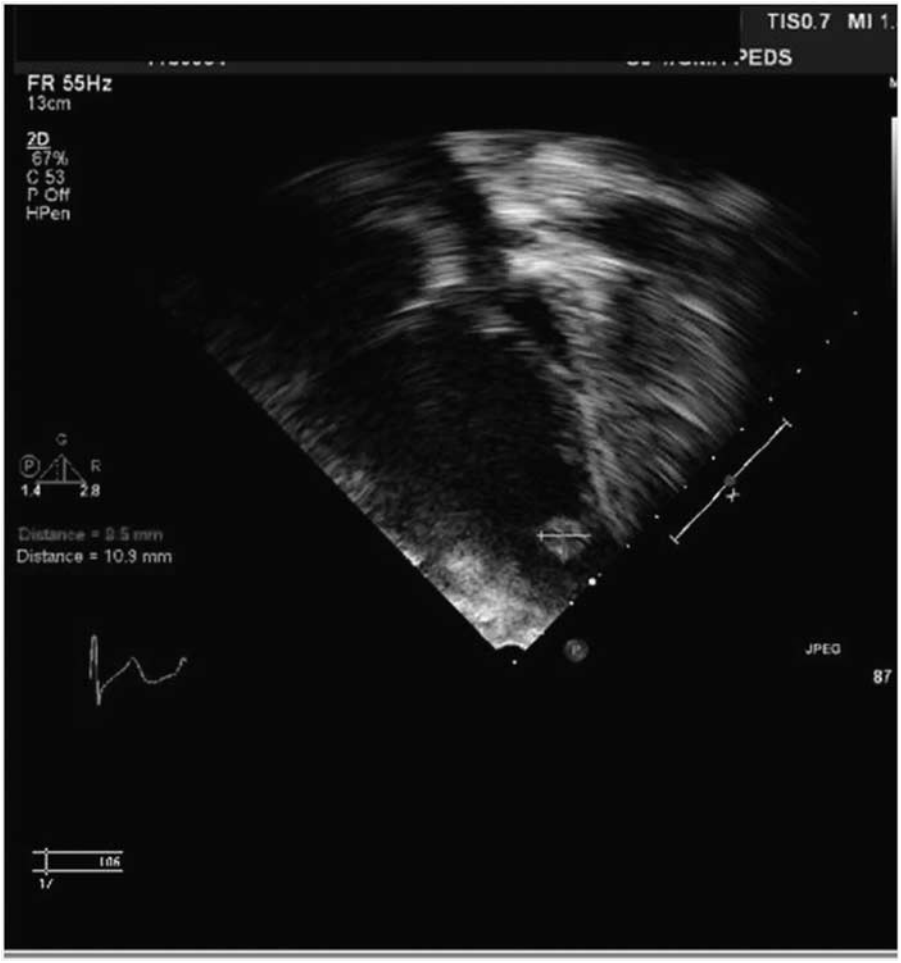

Figure 1.
*(**a**) Transthoracic two-dimensional echocardiogram. Apical four-chamber view showing a pedunculated mass near the apex of the left ventricle measuring 0.9 × 1.1 centimetre (arrow). (**b**) Transthoracic three-dimensional reconstruction. Apical four-chamber view showing a mass in the cavity of the left ventricle.*

echocardiography at the conclusion of the case revealed no residual left ventricular mass, no aortic insufficiency, and normal biventricular function. Histopathology of this mass showed necrotic tissue with marked inflammatory infiltrate consisting primarily of neutrophils and eosinophils. In addition, there was necrosis and fibrosis of adjacent myocardial tissue. She made an uneventful recovery, although she has continued with haemodialysis as an outpatient. She was discharged from the hospital on post-operative day 14 with outpatient immunology, renal, and cardiac follow-up.

Discussion

Wegener's granulomatosis is a systemic inflammatory disorder of unknown origin that was first described 70 years ago.[1] It affects both men and women across a wide age range and is characterised by granulomatous inflammation and vasculitis of small-to-medium-sized blood vessels and the presence of circulating antineutrophil cytoplasmic antibodies. Various organ systems are involved with the respiratory and the renal system being the most common.[2,3] Cardiac involvement is increasingly being recognised and has been reported in 4–44% of these patients.[4,7] Pericarditis, conduction disturbances, and supraventricular arrhythmias constitute the most clinically apparent entities, while coronary, myocardial, and valvular abnormalities, commonly reported on necropsy findings, are often not recognised clinically. Regional wall motion abnormalities may be reported, but these do not follow coronary artery distribution and generally improve after disease activity is suppressed.[5] The overall mortality for Wegener's granulomatosis is higher in patients with echocardiographic findings.[5] Anti-inflammatory therapy with immunosupressants like cyclophosphamide,

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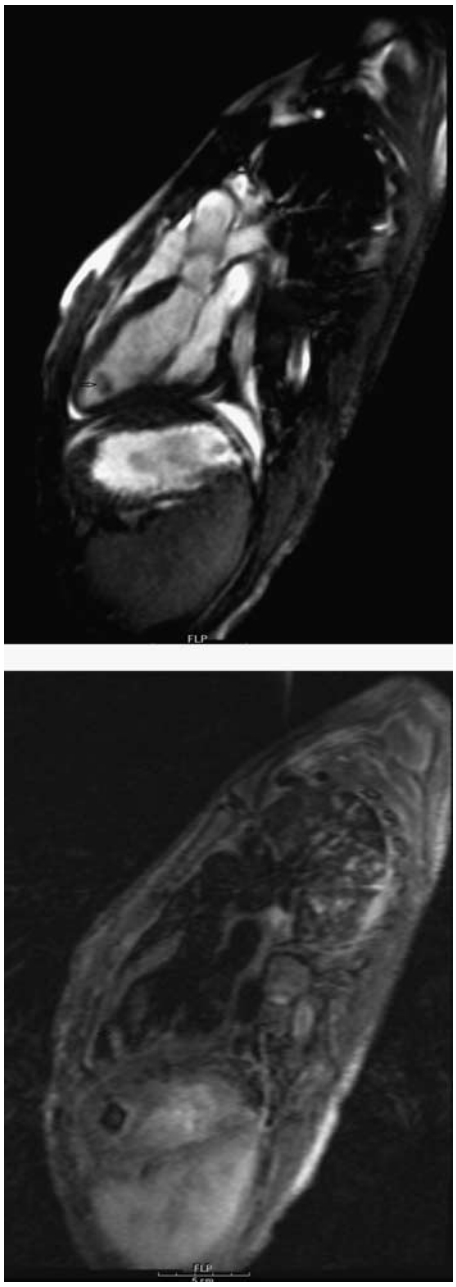

Figure 2.
*(**a**) Cardiac magnetic resonance imaging (MRI), bright blood imaging with Steady-state free precession (SSFP). A three-chamber view of the heart showing mass near the apex of the left ventricle (arrow). (**b**) Cardiac MRI, T1-weighted dark blood delayed enhancement sequence showing mass (arrow).*

methotrexate, and steroids usually lead to remission.[8] The diagnosis of Wegener's granulomatosis in our patient was based on a typical medical history, positive C-anti-neutrophil cytoplasmic antibody and findings on renal biopsy. Because of the thin stalk noted on an echocardiogram there was concern for a potential risk of embolization and thrombus formation and we proceeded with early surgical resection. Our patient was monitored closely for conduction disturbances; but, apart from a brief run of non-sustained ventricular tachycardia post-cardiac surgery, we did not note any arrhythmias. Given the rare presentation with intra-cardiac masses in Wegener's granulomatosis, the natural history of this lesion is unclear. The anticipated remission rate of Wegener's with therapy is 90%. We postulate that control of primary disease would prevent recurrence of the mass, and would monitor this patient with an echocardiogram at times of acute flare up of inflammatory disease. In addition, annual echocardiogram for surveillance after resection would seem appropriate.

In summary, Wegener's granulomatosis can be associated with a wide variety of intra-cardiac lesions. A high index of suspicion is necessary for prompt recognition and management. Imaging with two-dimensional, three-dimensional echo-cardiogram, or cardiac magnetic resonance plays an important role in assessing myocardial function and characterising cardiac lesions.

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