Aortic valve insufficiency in antinuclear antibody positive large vessel vasculitis

Introduction

The large vessel vasculitides are rare causes of aortic valve insufficiency through dilatation of the aortic root. This article presents a case of large vessel antinuclear antibody positive vasculitis through fibrous adhesions demonstrating a clinical improvement with immunosuppressant therapy.

Discussion

Large vessel vasculitis can occasionally cause aortic regurgitation through dilatation of the aortic root. This case demonstrates vasculitis causing aortic regurgitation through fibrous adhesions with a clinical improvement with immunosuppressant therapy.

Differential diagnosis include Lambl's excrescences (Roldan et al, 1997), although these are usually mobile filiform structures attached to the valve at the site of cusp closure but not the aortic wall, and Libman–Sacks endocarditis which is associated with systemic lupus erythematosus (Roldan et al, 1992). However, ds-DNA antiphospholipid antibodies were negative and there was normal coagulation profile and renal function. BJHM

Dr DP Ripley and Dr OE Gosling acknowledge the support of the Gawthorn Cardiac Trust.

Roldan CA, Shively BK, Lau CC, Gurule FT, Smith EA, Crawford MH (1992) Systemic lupus erythematosus valve disease by transesophageal echocardiography and the role of antiphospholipid antibodies. *J Am Coll Cardiol* **20**(5): 1127–34

Roldan CA, Sgively BK, Crawford MH (1997) Valve excrescence: prevalence, evolution and risk of cardioembolism. *J Am Coll Cardiol* **30:** 1308–14

Figure 1. ^{18}Fluorodeoxyglucose positron emission tomography illustrating widespread enhanced glucose uptake in the walls of the upper limb arteries (arrows).

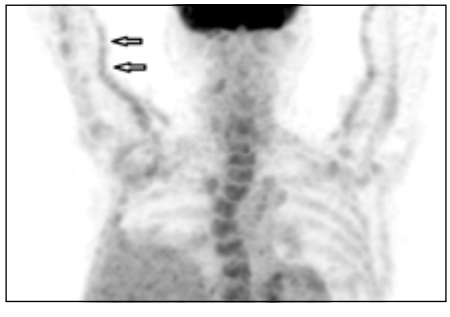

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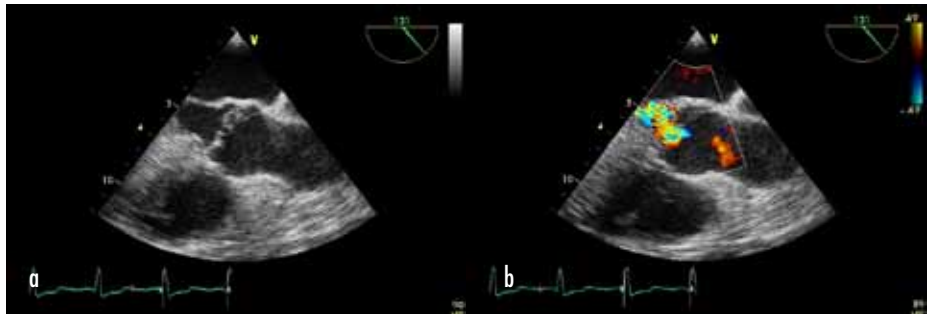
Figure 2. a. Transoesophageal echocardiography illustrating fibrous adhesion from the posterior aortic root attached to the tip of the non-coronary cusp resulting in failure of coaptation. b. Moderate jet of central aortic regurgitation.



LEARNING POINTS

- Large antinuclear antibody-positive vessel vasculitis is a rare cause of aortic valve disease.
- Treatment with immunosuppressant therapy can reverse aortic insufficiency in this context.

Case Report

A 76-year-old man was admitted with dyspnoea, pleuritic chest pain and a vasculitic rash. C-reactive protein was markedly elevated at 246 mg/litre and plain chest radiography was normal. Computed tomography pulmonary angiogram excluded pulmonary embolism. Clinical examination revealed a diastolic murmur with transthoracic echocardiography showing moderate aortic regurgitation. Transthoracic echocardiography 1 year previously was normal.

There was no bacterial growth in multiple blood cultures and antibodies for *Mycoplasma pneumoniae, Chlamydia psittaci* and *Coxiella burnetii* were also negative. Serum Na+ 129 mmol/litre, osmolality 258 mosmol/kg and urinary osmolality 543 mosmol/kg indicated syndrome of inappropriate antidiuretic hormone. There was a mild anaemia (9.9 g/dl) and abnormal liver function tests. ^{18}Fluorodeoxyglucose positron emission tomography excluded the possibility of malignancy but revealed widespread enhanced glucose uptake in the walls of the subclavian, carotid, upper limb arteries and the aortic arch as well as a 5 cm dilated abdominal aortic aneurysm, indicating large vessel vasculitis (*Figure 1*). Antinuclear antibodies staining pattern was positive at a titre < 1:64 (anti-neutrophil cytoplasmic antibody, ds-DNA, antiphospholipid and ENA negative).

Transoesophageal echocardiography demonstrated a fibrous adhesion from the posterior aortic root attached to the tip of the non-coronary cusp (*Figure 2a*), resulting in failure of coaptation and a moderate jet of central aortic regurgitation (*Figure 2b*) (Video 1 and 2 available from www.bjhm.co.uk). There was no evidence of endocarditis and left ventricular function was normal. He was commenced on oral steroids resulting in significant clinical improvement. Transthoracic echocardiography 6 months later showed the aortic regurgitation to be improved, graded as mild.