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Brief Report

Formation of pannus on prosthetic valves in a child with pseudoxanthoma elasticum

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Abstract A child with pseudoxanthoma elasticum had mitral valvar prolapse with severe regurgitation necessitating replacement with a prosthetic valve. Extensive formation of pannus caused obstruction of two mechanical valves, one after twenty months and the second after three years. Histology of the pannus was similar to the endocardial lesions that are considered unique to pseudoxanthoma elasticum.

Keywords: Pseudoxanthoma elasticum; pannus formation; mitral valve replacement; echocardiography

PSEUDOXANTHOMA ELASTICUM IS A RARE autosomal recessive condition involving the elastic tissues and affecting the skin, eye, and cardiovascular system. It is infrequently diagnosed in childhood.[1] We report a 6-year-old child with this condition who had involvement of the mitral valve and endocardium. Two prostheses inserted to replace the mitral valve became obstructed with extensive formation of pannus.

Case report

The female child was first assessed at the age of seven months, when she was found to have cardiac failure associated with severe mitral regurgitation. Her echocardiogram showed prolapse of both leaflets of the mitral valve with severe regurgitation, good left ventricular contractility with a fractional shortening of 47%, and increased echodensity of the endocardium and papillary muscles (Fig. 1). She initially responded to medical management with frusemide and captopril. By the age of seventeen months, however, her cardiac failure was poorly controlled, and she was failing to thrive. She was therefore referred for surgery.

At operation, the echocardiographic findings were confirmed. Proliferative tissue in the leaflets, and cordal elongation, were also noted. Repair was not possible, so the valve was replaced with a 23 mm St Jude prosthesis. Her post-operative course was uncomplicated, and she was stabilised on warfarin anticoagulant therapy.

At the age of 2 years, she was rehospitalised with dyspnoea and signs of cardiac failure. Echocardiography showed left ventricular dilation and poor contractility. The fractional shortening was only 12%, and the left ventricular end diastolic dimension was 49 mm. The valvar prosthesis was functioning normally. Medical therapy was recommenced.

Cardiac catheterisation demonstrated proximal obstruction of the left anterior interventricular coronary artery, with some retrograde filling of the distal vessel from the right coronary artery. It was assumed that the occlusion resulted from an embolus related to thrombus on the valvar prosthesis at a time of suboptimal anticoagulation. Medical therapy was continued in the hope that collateral arteries would continue to develop. The site of femoral arterial puncture for cardiac catheterisation developed a false aneurysm, necessitating surgical closure.

The child began to experience episodes of typical angina despite maximal medical therapy. At the age of thirty months, she underwent angioplasty to the left anterior interventricular coronary artery. This was successful, and was the subject of another report.[2] There was some improvement in echocardiographic measures of contractility, with fractional shortening increasing to 18–20%.

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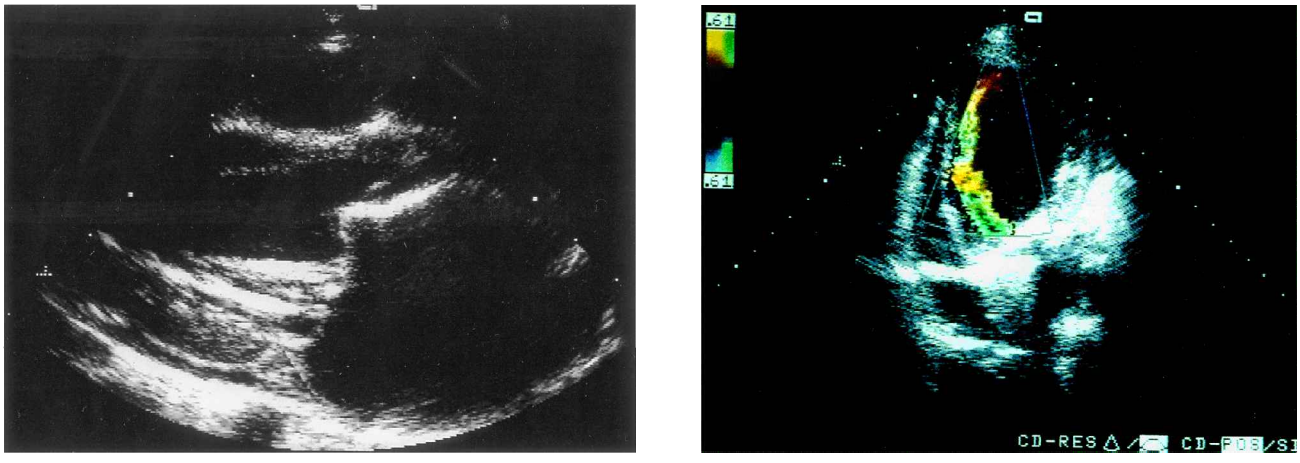

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Figure 1.
The echocardiogram at the age of seven months (a) shows prolapse of both leaflets of the mitral valve, which caused severe mitral regurgitation with left atrial and left ventricular enlargement. Note the increased echodensity of the endocardium of the papillary muscles. The echocardiogram in the four chamber view at the age of 3 years (b) shows restriction of movement of the discs of the mitral valvar prosthesis, causing an eccentric inflow jet. The mean gradient across the prosthesis was 18 mmHg pressure.

At the age of 3 years, she was re-admitted with symptoms. Echocardiography then suggested thrombus on the discs of the prosthetic valve, restricting their motion. The mean pressure gradient across the prosthesis was 18 mmHg, with an eccentric inflow jet (Fig. 1). At her second operation, the prosthesis was seen to be obstructed, with thrombus fixing the posterior leaflet in the closed position. Formation of pannus on the outflow portion of the valve produced marked narrowing. The valve was replaced with a 23 mm ATS aortic valvar prosthesis inserted in the reverse position because of unavailability of a small mitral prosthesis. A left ventricular assist device was made ready, but was not needed as the patient was satisfactorily weaned from bypass. She was weaned from ventilation on the fourth post-operative day. On discharge, her fractional shortening was 15% and she was prescribed warfarin, aspirin, frusemide, enalapril and amiodarone.

At the age of 3 years and 6 months, she had a transient cerebrovascular event despite good anticoagulative control. An infusion of heparin was used, but echocardiography did not show further valvar or intracardiac thrombus. Computerized scanning of the head was normal, and all neurological signs resolved. Over the next 4 months, she developed progressive left ventricular failure and her fractional shortening reverted to 12%. She was assessed for cardiac transplantation, and carvedilol added to her therapeutic regime. She stabilised and medical management was continued.

At the age of 5 years, she developed flexural skin lesions, and a biopsy indicated the diagnosis of pseudoxanthoma elasticum. No ocular lesions were found. She remained stable and was asymptomatic. At a routine outpatient review at the age of 6 years, the echocardiogram demonstrated restenosis of the valvar prosthesis, with movement of only one disc. There was a gradient of 16 mmHg across the valve with associated pulmonary hypertension. The fractional shortening was 27%.

At a third valvar operation, there was fixation of the anterior disc of the prosthesis due to formation of thrombus and pannus. The posterior leaflet was mobile, but restricted by pannus, particularly on the left ventricular aspect (Fig. 2). Histology of the valve showed that the pannus consisted of partly calcified fibrosis with overlying organising thrombus. An elastin stain demonstrated focal fragmentation of elastic laminas. This valve was replaced with a 25 mm ATS mitral prosthesis. She came off bypass with the aid of inotropes, and made a rapid post-operative recovery. She is currently well with a fractional shortening of 25%.

Discussion

Pseudoxanthoma elasticum is an inherited disorder of elastic tissue in which there is progressive fragmentation and calcification of elastic fibres in the skin, eye, and blood vessels.[1] Diagnosis is rarely made in childhood, apart from sometimes at autopsy.[3,4] The condition is characterised by flexural skin lesions of "plucked chicken" appearance, angioid streaks in the retina and cardiovascular problems including mitral valvar prolapse,[5] accelerated atherosclerosis, calcification of peripheral arteries, renovascular hypertension,[6] and fibrous thickening of the endocardium of

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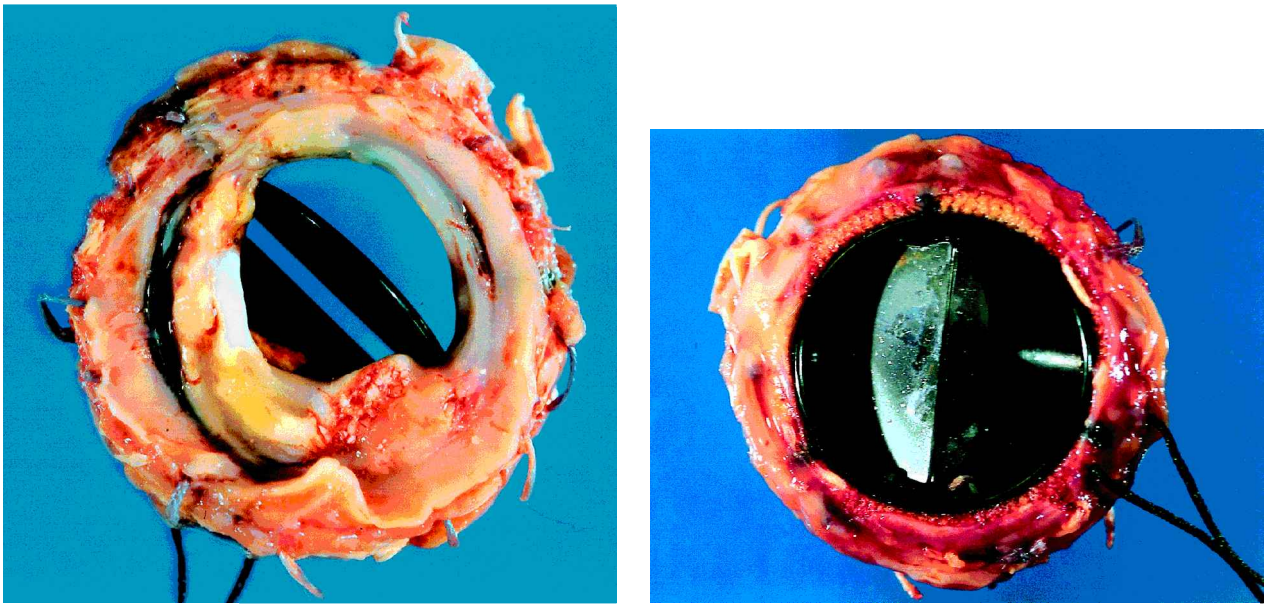

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Figure 2.
The explanted ATS mitral valvar prosthesis shows obstruction of one disc by extensive formation of pannus on the ventricular side (a). Only one disc is able to open. Figure 2b shows the left atrial side of the explanted valve.

the ventricles, atriums and atrioventricular valves.[7] Endocardial lesions of intimal fibroelastic thickening, with disorganisation and calcification of elastic fibres, are considered specific to the diagnosis.[8]

Our patient had cutaneous lesions, mitral valvar prolapse, and endocardial lesions. Formation of pannus was extensive, and caused obstruction of two valvar prostheses. A previous report of formation of pannus in a 14 year old boy with endocardial fibroelastosis who underwent replacement of the mitral valve suggested that the complication was rare.[9] In a series of 100 patients with obstructed valvar prostheses, nonetheless, almost half had pannus. Of these, 4 were under 10 years of age, and 20 were below the age of 20 years.[10]

We believe that the formation of pannus on the prosthetic valves inserted in our patient was directly related to her pseudoxanthoma elasticum. The histology of extensive fibrotic thickening, calcification, and fragmentation of the elastic laminas is typical of the condition.[8]

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