CASE REPORT

Rheumatic involvement of all four cardiac valves

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Heart 2005;91:e50 (<http://www.heartjnl.com/cgi/content/full/91/6/e50>). doi: 10.1136/hrt.2005.060509

Rheumatic involvement of all four heart valves is rare. A 35 year old woman presented with gradually progressive exertional dyspnoea for the preceding 10 years. On evaluation she was in atrial fibrillation with congestive heart failure. Clinical examination found evidence of stenosis of the mitral aortic and tricuspid valves with a history of rheumatic fever in childhood. Transthoracic echocardiography showed the involvement of all four cardiac valves. Few reports are available in the literature describing rheumatic quadrivalvar damage. Operator awareness of possible rheumatic involvement of all four valves is essential for appropriate diagnosis.

A 35 year old woman presented with gradually progressive exertional dyspnoea of 10 years' duration. Initially she had New York Heart Association class II symptoms, which had gradually deteriorated to class IV at the time of presentation. She had a history of rheumatic fever 20 years previously. She was in atrial fibrillation and mean jugular venous pressure was raised with a prominent V wave and Y descent. The first heart sound was loud with a normally split second sound and an accentuated pulmonary component. There was an opening snap at the apex and lower left sternal border. Mid diastolic murmurs at the apex and lower left sternal border, a grade 3 ejection systolic murmur in the aortic area conducted to the carotids, and a grade 3 pansystolic murmur at the lower left sternal border increasing on inspiration were audible. The liver was palpable 4 cm below the right costal margin with ascites.

ECG showed right axis deviation, atrial fibrillation, and ST-T changes suggestive of a digitalis effect. A chest radiograph in the posteroanterior view showed cardiomegaly with a cardiothoracic ratio of 0.70 and right atrial, right ventricular, and left atrial enlargement with pulmonary venous hypertension.

Transthoracic echocardiography showed a speck of calcium on the tips of the pulmonary valve with doming (fig 1) and severe aortic, mitral, and tricuspid valve stenoses (fig 2). The mitral and tricuspid valve areas were 0.6 cm² and 0.9 cm², respectively. The mean gradient across both the mitral and tricuspid valves was 15 mm Hg. The peak systolic gradient was 128 mm Hg across the aortic valve and 16 mm Hg across the pulmonary valve. There was mild aortic and pulmonary regurgitation and severe tricuspid regurgitation. Biventricular function was normal with severe pulmonary artery hypertension, with a calculated right ventricular systolic pressure of 70 mm Hg.

DISCUSSION

Involvement of all the four cardiac valves due to a rheumatic process is rare¹ with stenosis in all valves being still more rare; few cases have been reported. Rheumatic quadrivalvar damage² has been found on necropsy (one in 586 patients with valve deformities)³ and by cardiac catheterisation. There is a high incidence of multivalvar damage when Aschoff

Figure 1 Transthoracic echocardiogram showing thickened and doming pulmonary valve (A) and thickened tricuspid valve (B). LA, left atrium; RA, right atrium; RVOT, right ventricular outflow tract.

bodies are identified at necropsy.⁴ Earlier, pulmonary valve involvement was diagnosed only at surgery.⁵ Organic tricuspid valve involvement is reported to occur in more than one third of patients with rheumatic heart disease studied at necropsy on the Indian subcontinent.⁶

There are a few reports of echocardiographic diagnosis of rheumatic cardiopathy affecting all four cardiac valves.^{7,8} Preoperative echocardiographic diagnosis of rheumatic involvement of all four cardiac valves and successful surgical treatment has also been reported.⁹

Figure 2 Transthoracic echocardiogram showing thickened and doming aortic valve (A) and thickened mitral valve (B). AO, aorta; LV, left ventricle; PE, pericardial effusion; RV, right ventricle.

It is important to realise that by the time echocardiography became prevalent and was commonly used to diagnose valve diseases, the severity of rheumatic heart disease had largely been declining. Also, operator awareness regarding possible quadrivalvar damage is essential for appropriate diagnosis. A resurgence of crippling rheumatic heart disease explains the extensive involvement of all four valves in this patient.

ACKNOWLEDGEMENTS

We thank our secretarial staff and echocardiographer, Mrs Meena Rani, for their help with this publication.

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Accepted 25 January 2005

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