Correspondence

Isolated Right Ventricular Cardiomyopathy of Unknown Etiology

Sir,

Right ventricular cardiomyopathy (RVCMP) is a rare and not a well defined entity. It presents with signs of right ventricular failure and ventricular arrhythmias. Most often patients have fibro-fatty infiltration on histopathology and the diagnosis is arrhythmogenic right ventricular dysplasia (ARVD).

We report a case of isolated, idiopathic RVCMP without features of ARVD or any other known etiology. Our patient was a 45 year old lady who presented to us with a 2 year history of class II dyspnea and irregular, infrequent and short lasting episodes of palpitations without syncope.

Physical evaluation was unremarkable except for the presence of a grade 2 pansystolic murmur at the left lower sternal border. There was T wave inversion in the inferior and precordial leads and there were no epsilon waves. Frontal chest X-ray was suggestive of right atrial (RA) enlargement.

The 2 D echocardiography with colour Doppler showed RA and right ventricle (RV) dilatation; and RV wall showed patchy areas of hyperechoic infiltration with hypokinesia. The overall RV systolic function was mildly depressed, while the left ventricle (LV) was normal in size and contractility. The tricuspid valve (TV) was mildly thickened and there was severe tricuspid regurgitation (TR), but no tricuspid stenosis. There was no pulmonary hypertension. The pulmonary arterial systolic pressure calculated by TR jet was 28 mm Hg. The other cardiac valves were normal and there was no intracardiac shunt. There was no pericardial effusion and the inferior vena cava was mildly congested. A transesophageal echocardiography (TEE) confirmed these 2D echocardiographic findings.

A signal averaged electrocardiogram was negative for late potential activity and a 24 hour ambulatory Holter study showed occasional atrial ectopics with a few couplets, but no atrial tachycardia. There was no ventricular ectopy noted during the entire study.

A cardiac magnetic resonance imaging (MRI) as well as computed tomography (CT) scan showed RA and RV dilatation with elevated end-diastolic volume, reduced RV ejection fraction and presence of TR. There was no fat infiltration nor was there any segmental wall abnormality. The LV was normal.

Coronary angiography revealed normal coronary arteries. RV endomyocardial biopsy (Figs. 1a & 1b) revealed histological features compatible with CMP i.e. endothelial thickening, increased connective tissue, decreased myocardial fibers with hypertrophy and atrophy of myofibres. But there was no evidence of myocarditis or fatty infiltration as is seen in ARVD.

Our patient had evidence of RVCMP, without any left ventricular involvement and also did not satisfy the conventional diagnostic criteria of ARVD1 or any other infiltrative disorder. In an extensive literature search, we were unable to find a case report of such clinical entity i.e. isolated, idiopathic RVCMP. Interestingly, we did find a single report of isolated right ventricular hypertrophic obstructive cardiomyopathy without left ventricular involvement.2

Our case seems to be a rare and as yet not well defined entity of isolated idiopathic right ventricular cardiomyopathy.

Fig. 1a : Biopsy showing bundles of myofibres. No obvious fatty tissue noted. (H & E 100).

Fig. 1b : Myofibres showing hypertrophic & atrophic fibres. (H & E x 400).
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