Isolated ventricular non-compaction or 'spongy cardiomyopathy'

Gamini Galappatthy¹, Ruvan Ekanayaka² and D M Dhanapala³

(Index words: Echocardiographic features, symptoms, dysrhythmia)

Isolated ventricular non-compaction (IVNC) is a recently described rare congenital cardiomyopathy resulting from an arrest of normal endomyocardial embryogenesis. Non-compaction of the myocardium is diagnosed by echocardiography which shows a typical 'spongy' appearance of the myocardium with trabeculations and recesses. Left ventricular (LV) myocardium is commonly affected, with or without right ventricular involvement (1,2).

Symptoms are frequently delayed until adulthood and manifest as a triad of clinical heart failure due to systolic and diastolic ventricular dysfunction, cardiac arrhythmias (including ventricular tachycardia (VT)), sudden cardiac death, and systemic embolism from emboli formed in the inter-trabecular recesses (1,2). Familial occurrence of the disease has been observed.

We describe a case of non-compaction of the ventricular myocardium in a 37-year old man with the typical echocardiographic features of this disease and a suggestive family history. This is the first such reported patient in Sri Lanka.

Case report

The patient presented with a 19-year history of easy fatigability, shortness of breath with exertion, atypical chest pain, palpitations lasting 1 to 2 min, faintishness, loss of appetite, and loss of weight. He was apparently well till he was 18 years of age and noticed unusual fatigue which progressed gradually thereafter. He was a smoker and

consumed alcohol moderately but had stopped both 5 years ago.

He gave a suggestive family history of arterial embolism and heart failure in his mother who had breathlessness on exertion for 3 years, monoparesis of the right upper limb and dense right side hemiplegia a year later, and death at the age of 42 years following an above knee amputation for gangrene. His father also died suddenly at the age of 42 years of unknown cause but 2 siblings are healthy.

The heart apex was not displaced or heaving, with no parasternal heave or thrills. The heart sounds were normal. ECG showed atrial fibrillation with a controlled ventricular rate. Holter monitoring showed a few runs of supraventricular tachycardia and non-sustained ventricular tachycardia. Coronary angiogram showed normal coronary arteries and good LV systolic function.

Discussion

Four morphological criteria diagnostic of IVNC by echocardiography have been proposed (1,2). They are absence of coexisting cardiac abnormalities (by definition), a compacted thin epicardial band and a thicker (>2:1) non-compacted endocardial layer of trabeculations and deep endomyocardial spaces, and predominant localisation of the pathology to LV mid-lateral, apical and mid-inferior areas and 4 colour. Doppler evidence of deep perfused intertrabecular recesses. Our patient had all these features with good LV systolic function and diastolic dysfunction (Figures 1 and 2).

¹Acting Resident Cardiologist, ²Cardiologist, ³Registrar, Institute of Cardiology, National Hospital of Sri Lanka, Colombo. (Correspondence: GG, Telephones + 94 1 691407, + 94 1 854682, fax + 94 1 691407, e-mail: gaminiks@sltnet.lk. Received 16 October 2002, accepted 6 February 2003).

Figure 1. Echocardiogram

Figure 2. Echocardiogram

LV = Left ventricle; SP = Spongy myocardium; E = Compact epicardial layer; LA = Left atrium

Mortality and morbidity of IVNC are high due to heart failure, thrombo-embolic events and ventricular arrhythmias (3). Management of IVNC includes treating heart failure when present, oral anticoagulants and amiodarone which were started in this patient. Definitive treatment at present to prevent arrhythmic death is artificial implantable cardio-verter defibrillator (AICD) implantation presently not available in Sri Lanka. Heart transplantation is also described in literature (1,2,3).

Knowledge regarding diagnosis, morbidity and prognosis of IVNC is still limited. Many cases could have been previously mistaken for LV hypertrophy, dilated cardiomyopathy or cardiac tumour, and awareness of this condition will improve diagnosis and our understanding of the

disease. It has been suggested that the WHO classification of cardiomyopathies be reconsidered to include IVNC as a distinct cardiomyopathy (1,2,3).

References

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