An unusual case of isolated non-compacted right ventricular myocardium

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Isolated ventricular non-compaction is a rare type of cardiomyopathy resulting from arrested myocardial development during embryogenesis. This rare entity can be easily diagnosed by characteristic appearance of prominent myocardial trabeculations and deep inter-trabecular spaces. The clinical manifestations include heart failure signs, ventricular arrhythmias, and cardio-embolic events. Although the usual site of involvement is the left ventricle, the right ventricle (RV) can rarely be affected. Here, we report a case of 23-year-old male patient with isolated RV non-compaction.

KEYWORDS
RHF; Cardiomyopathy; IVNC

A 23-year-old male came with the history of exertional dyspnoea for the past 1.5 years. He has class II dyspnoea at present. He had a history of facial puffiness and pedal oedema in the past and also has exertional palpitation. There was no family history of cardiac illness. On examination, he was found to have an irregular pulse rate of 92/min, with a blood pressure of 110/70 mmHg. He had elevated jugular venous pressure with the prominent ‘y’ descent. Cardiovascular examination was unremarkable except muffled heart sounds with tricuspid systolic murmur. Respiratory system examination revealed normal findings. Chest X-ray revealed enlarged right heart chambers with normal pulmonary vasculature. Surface electrocardiogram revealed atrial fibrillation with a controlled ventricular rate with polymorphic QRS in the anterior leads. Echocardiogram revealed enlarged right heart chambers with normal pulmonary vasculature. Surface electrocardiogram revealed atrial fibrillation with a controlled ventricular rate with polymorphic QRS in the anterior leads. Echocardiogram revealed enlarged right heart chambers with honeycomb appearance in apical RV and free wall of the RV. Colour Doppler demonstrated flow from RV cavity into the trabecular recesses. He also had severe low-pressure tricuspid regurgitation with severe RV dysfunction. He was diagnosed to have isolated non-compaction of RV and he was treated with decongestive measures with oral anticoagulation and rate-controlling medications for atrial fibrillation.

Discussion

Myocardial non-compaction is a rare disorder with uncertain aetiology.1 According to the report of the World Health Organization/International Society and Federation of Cardiology Task Force in 1995, isolated ventricular non-compaction (IVNC) is considered an unclassified cardiomyopathy.2 Normally between the foetal 5th week and 8th week, inter-trabecular spaces are obliterated and ventricular compaction occurs from the base towards the apex and from epicardium to endocardium, and an arrest in the progression of ventricular compaction results in non-compaction.3–5 The left ventricle (LV) is the usual site of involvement, but involvement of both ventricles, and rarely isolated RV non-compaction, can be seen.6 Both familial7,8 and sporadic1 forms of IVNC have been described. The familial form was observed in 18% of the adult population with IVNC.7 Because of the risk of familial occurrence, the first-degree relatives should be screened by echocardiography to identify asymptomatic patients. Our patient’s first-degree relatives were screened and found to be disease-free.

The following diagnostic criteria for IVNC have been defined: (i) the absence of coexisting cardiac anomalies; (ii) a two-layered structure of LV wall, with the end-systolic ratio of the non-compacted to compacted myocardial layer >2; (iii) finding this structure predominantly in the apical and midventricular areas; and (iv) blood flow directly from the ventricular cavity into deep inter-trabecular recesses as assessed by Doppler echocardiography.7 In our patient, all four echocardiographic criteria of IVNC were present. A two-layered structure in the right ventricle (RV) was found in the apical region; mid and superior regions of the right ventricle revealed trabeculations and deep inter-trabecular spaces.
Ventricular arrhythmias are the major, sometimes fatal, complications of IVNC. Ventricular tachycardia has been found in up to 41% of the patients with IVNC. Other arrhythmias such as atrial fibrillation and ventricular premature beats also have been found in patients with IVNC. Our patients had not shown any arrhythmias on surface ECG as well as Holter monitoring.

Cardio-embolic events are reported in many patients of IVNC. In a series by Oeschslin et al., cardio-embolic events have been reported in 24% of the patients. Prevalence of these events was independent of the LV dimensions and function. Endomyocardial morphology in IVNC is responsible for the development of mural thrombi within the inter-trabecular spaces. All adult patients are recommended oral anticoagulation, irrespective of ventricular size and function. In our case, we started oral anticoagulation with warfarin, when IVNC was diagnosed.

IVNC patients had variable prognosis, ranging from prolonged asymptomatic course to severe cardiac disability, leading to heart transplantation and death. Approximately 50% of the patients died suddenly. Prognosis is worse in patients with heart failure NYHA classes III–IV, the LV end-diastolic diameter >60 mm, the left bundle branch block, and chronic atrial fibrillation. No such criteria for prognosis have been proposed for isolated RV non-compaction.

Our patient was treated with thiazide diuretic, aldosterone antagonist, ACE-inhibitor, digoxin, and warfarin.

References