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Correspondence

QJM

Isolated left ventricular non-compaction, not hypertrophic cardiomyopathy

Sir,

Hypertrophic cardiomyopathy (HCM) is a relatively common cardiac disease (1:500 in the general population), which both general physicians and cardiologists encounter frequently in clinical practice. However, there are conditions that may mimic HCM (Table 1). We describe a patient with an initial diagnosis of hypertrophic cardiomyopathy. Further investigations several years later revealed an alternative diagnosis of isolated left ventricular non-compaction (IVNC).

In 1991, an asymptomatic 24-year-old woman was diagnosed with HCM, based on an echocardiogram that showed moderate left ventricular hypertrophy particularly affecting the septum, part of familial screening following the diagnosis of HCM in her mother.

Over the following 11 years, she developed mild dyspnoea. Ambulatory ECG monitoring showed atrial flutter and a short burst of ventricular tachycardia. Treatment with a beta blocker was initiated, but had to be withdrawn following side-effects. Her symptoms worsened appreciably over the next 12–18 months, and she began to experience breathlessness at rest and chest pain.

Transthoracic echocardiography showed global hypokinesia, especially involving the inferior wall. There was moderate LV hypertrophy affecting the posterior wall. Trabeculations were seen at the left ventricular apex as well as in the right ventricle. These features were now more in keeping with left ventricular non-compaction rather than hypertrophic cardiomyopathy. Contrast cardiac magnetic resonance imaging confirmed the diagnosis, demonstrating several areas of myocardial hyper-enhancement mostly sparing the subendocardium. These areas of hyper-enhancement were associated with a wall motion abnormality.

Treatment with an ACE inhibitor was commenced, and a nitrate preparation was added to relieve symptoms of chest pain. In view of the previously documented atrial tachy-arrhythmia and left atrial enlargement, she was anticoagulated with warfarin. Amiodarone was given to suppress ventricular tachycardia. Heart failure medication was optimized with the addition of spironolactone.

It is likely that the incidence of IVNC is being under-estimated because of its phenotypic similarity to HCM. It is important to discriminate between these cardiomyopathies as they differ in terms of natural history, prognosis and treatment. IVNC is a progressive disorder with a poor prognosis, resulting in systolic and diastolic failure in the long term. The largest study of patients with IVNC to date reported a 58% event-free survival rate at 5 years post diagnosis (combined end-point of heart transplantation and death).[1] In comparison, HCM confers a 1% annual mortality rate, and most patients have little or no disability and a normal life expectancy. Subsets may be identified with higher morbidity and mortality, and are linked to the complications of

Table 1 Differential diagnoses to consider when echocardiographic features of HCM are present

Left ventricular non-compaction
Athlete's heart
Fabry disease (particularly in male patients with late-onset HCM)
Cardiac sarcoidosis
Primary amyloidosis
Cardiac tumours
Friedrich's ataxia
Hyperparathyroidism
Hypereosinophilic syndrome
Noonan's syndrome
Kleinfelter's syndrome
Pompe disease
Glycogen storage disorders
Phaeochromocytoma

sudden death, progressive heart failure and atrial fibrillation.[2]

Treatment options for IVNC mostly comprise heart failure therapy, including the use of cardioselective beta blockers, which have beneficial effects on LV function, mass, and scintigraphic findings, leading to reversal of ventricular dilatation and hypertrophy.[3] All patients should be anticoagulated in order to prevent the increased incidence of stroke, pulmonary embolism, and other systemic emboli. Early implantation of automated defibrillator/cardioverters in patients who are symptomatic of NYHA classes III or IV, those who have atrial fibrillation and/or severely dilated left ventricles (features associated with poor prognosis and increased incidence of death) reduces morbidity and mortality, as does early heart transplantation.[1]

When considering the diagnosis of HCM or re-evaluating patients who have been labelled with this disorder, it is important to be aware of other conditions such as IVNC that may manifest as HCM. Increased awareness of the differential diagnosis of HCM will ensure that patients receive the most appropriate management.

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