

Non-compaction Cardiomyopathy Presenting with Classical Angina Pectoris

Dear Editor,

Non-compaction cardiomyopathy (NCC) is an unclassified cardiomyopathy, previously known as “spongy left ventricular myocardium”. It is characterised by prominent myocardial trabeculations and deep intertrabecular recesses which lie in continuity with the left ventricular cavity. This failure in the normal compaction of the ventricular endomyocardium results from an arrest in cardiac embryogenesis. NCC is a genetically heterogeneous condition with both familial and sporadic forms described.¹ It commonly presents as heart failure, ventricular arrhythmia or cardiac thromboembolism. Presentation with angina pectoris has been documented but not frequently recognised.

Case Report

A 57-year-old postmenopausal female presented with 3 episodes of exertional central chest pain which were compressive in nature, radiated to the jaw and associated with dyspnoea and diaphoresis. Each episode abated within half an hour of rest. She had no known medical problems or reversible cardiovascular risk factors. Physical examination was unremarkable.

The electrocardiogram (ECG) showed sinus rhythm with widespread downsloping ST segment depression in leads

I, II, aVL and V2 to V6 (Fig. 1A) which were minimally reversible after resolution of symptoms (Fig. 1B). Cardiac biomarkers were not elevated. As acute coronary syndrome was initially suspected, the patient had coronary angiography which demonstrated normal epicardial arteries. Left ventriculography showed a grossly thickened myocardium with a spongiform appearance (Fig. 1C). Echocardiography confirmed isolated left ventricular non-compaction with classical findings of a thin compacted layer, thicker highly trabeculated layer and deep intertrabecular recesses (Fig. 1D). The left ventricular ejection fraction was 50%.

Discussion

Although patients with symptomatic NCC often present with arrhythmia, heart failure or cardioembolism, chest pain is not uncommon. Up to 40% of patients present with anginal chest pain, most of whom have a normal coronary angiogram.² Repolarisation abnormalities on ECG are frequently reported. Reduced myocardial perfusion has been demonstrated in non-compacted myocardial regions using positron emission tomography.³ In the absence of epicardial coronary artery disease, postulated mechanisms of angina include decreased coronary flow reserve arising from underdeveloped microcirculation or compression of the intramural vasculature.³

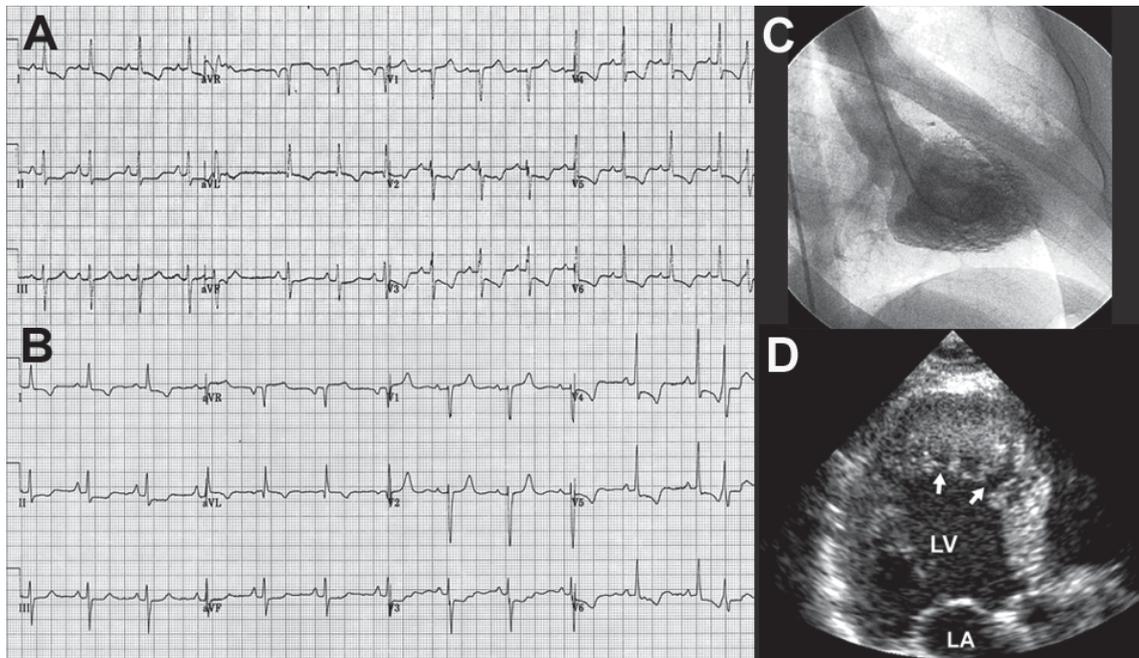


Fig. 1A. Presenting electrocardiogram (ECG) showing widespread downsloping ST segment depression in leads I, II, aVL, V2 to V6.

Fig. 1B. ECG showing partial resolution of ST segment depression after chest pain had subsided.

Fig. 1C. Left ventriculography documenting the characteristic spongiform appearance of the myocardium in non-compaction cardiomyopathy.

Fig. 1D. Echocardiography demonstrating a highly trabeculated left ventricle. Solid arrows indicate deep intertrabecular recesses. LA indicates the left atrium and LV indicates the left ventricle.

Conclusion

NCC is associated with a broad spectrum of clinical manifestations. Angina pectoris is an under-appreciated presentation of this uncommon condition, the diagnosis of which is frequently delayed. Early recognition of NCC is important to facilitate long-term management and screening of first degree relatives of affected individuals.

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