Spongy heart: left ventricular non-compaction

A hypertensive 55-year-old gentleman was referred for lightheadedness, diaphoresis, dyspnoea, palpitations and chest pain. He was admitted in the past for symptomatic ventricular tachycardia and recurrent syncope, and was started on flecainide. He had no family history of arrhythmias or sudden cardiac death. Echocardiography revealed generalised left ventricular systolic dysfunction, without wall motion abnormalities and ejection fraction of 40%. Cardiac catherisation showed no coronary artery disease. Cardiac MRI revealed an increase in subendocardial trabeculations (figure 1), hypertrophic and non-compacted myocardium of the left ventricle (figure 2) suggesting left ventricular non-compaction (LVNC). The patient got an implantable cardioverter-defibrillator (ICD) and was started on sotalol and since then has not reported any arrhythmias.

Prominent trabeculae, deep endocardial recesses and a sponge-like appearance of the myocardium characterise LVNC. ¹ Clinical presentation varies from patients being asymptomatic to having conduction defects, thromboembolism, ventricular arrhythmias, severe heart failure or sudden cardiac death. ¹ Echocardiography, the most frequently used imaging modality, is measurement dependent, ratio dependent and reader dependent and has a high potential for false positivity. The cardiac MRI is another diagnostic tool used to validate the diagnosis of

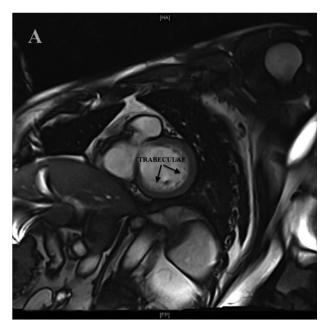


Figure 1 Cardiac MRI of the patient with short axis view of the heart: The left ventricular myocardium is significantly trabeculated with deep recesses extending along the entire cavity of the left ventricle including the apex.

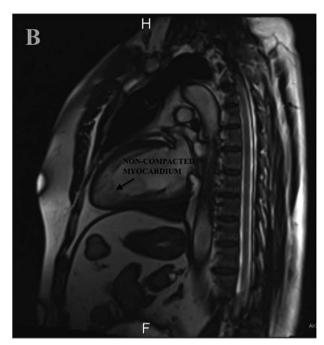


Figure 2 Cardiac MRI showing left ventricular myocardium with significant trabeculations which are extending along the entire cavity of the left ventricle including the apex. The ratio of the non-compacted to compacted myocardium is >20% suggesting left ventricular non-compaction.

LVNC. A non-compacted to compacted ratio of more than 20% in end-diastole distinguishes pathological non-compaction. ¹

Treatment options vary from medical management in mild cases to ICD placement to heart transplantation in those with refractory symptoms. Monitoring is encouraged in asymptomatic patients. Patients need to be on regular follow-up involving routine clinical assessment, echocardiography, Holter monitoring and screening of first-degree relatives.

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