


CLINICAL VIGNETTE

Non-compaction cardiomyopathy with low-gradient aortic valve stenosis

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A 73-year-old man presented to the emergency department with dyspnea of 2 days duration. Auscultation of the precordium was notable for a grade 2/6 systolic murmur. Echocardiography revealed a highly impaired left ventricular function and a calcified aortic valve stenosis with a mean pressure gradient of 25 mmHg and a valve area of 0.8 cm². Eye-catching was a thickened mid-ventricular and apical myocardium with a spongy appearance. On magnetic resonance imaging (MRI) multiple, prominent muscular trabeculations of the left ventricular myocardium (white arrows) with deep intertrabecular recesses were evident (Panel A, LV: left ventricle, LA: left atrium). These findings were consistent with non-compaction cardiomyopathy with accompanying low-gradient aortic valve stenosis. The patient underwent mechanical aortic valve replacement, during which the spongy left ventricular myocardium was visualized by video endoscopy (Panel B). The histological workup of an endomyocardial biopsy specimen showed irregular heart muscle fibres with vacuolar changes and large chromatin-dense nuclei as well as moderate interstitial fibrosis (Panel C, haematoxylin and eosin staining, magnification: original ×20). The patient made a full recovery and was discharged on heart failure medication and oral anticoagulation. Screening of first-degree relatives by MRI revealed left ventricular non-compaction with a preserved ejection fraction in the patient’s asymptomatic son (Panel D, LV: left ventricle, LA: left atrium).

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