IMAGES IN CARDIOLOGY

Left ventricular non-compaction in a patient with Becker's muscular dystrophy

This is a cardiac magnetic resonance image (MRI) of a 33 year old man with Becker's muscular dystrophy (BMD). From the age of three he had had wasting of the limbs that had progressed slowly. BMD was diagnosed when he was 29 when a muscle biopsy showed myopathic changes and an altered staining pattern for dystrophin. Dystrophin analysis revealed the expression of a truncated molecule of about 380 kDA caused by an inframe deletion of exon 45 to 48 within the dystrophin gene.

The patient underwent cardiological examination because of recent onset of resting dyspnoea, jaundice, and peripheral oedema. The electrocardiogram showed sinus rhythm with a rate of 120 beats/min, an incomplete right bundle branch block, and signs of left ventricular hypertrophy. Echocardiography showed biventricular and biatrial dilatation and a globally reduced ventricular systolic function. In the region of the apex and the adjacent lateral wall we detected abnormal structures that had the same echogenicity as the surrounding myocardium. They moved synchronously with the ventricular wall and were not attached to the mitral valve or false tendons.

The accompanying MRI confirmed the existence of these abnormal structures (arrowed), which were mainly located in the left ventricular apex and the adjacent lateral wall. In view of the morphology and the motion pattern of the structures we considered the diagnosis of a thrombus unlikely.

We interpret these structures as abnormal trabeculation also termed "left ventricular non-compaction". Non-compaction is believed to present an arrest in endomyocardial morphogenesis. Little is known about its association with skeletal muscle disorders. At present, we can not decide whether the combination of left ventricular non-compaction and BMD is a pure coincidence of two rare disorders or is due to a common underlying aetiology. We conclude that cardiologists and radiologists should consider non-compaction when they examine patients with skeletal muscle disorders—just as clinicians should look for skeletal muscle disorders in patients with non-compaction.

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