Clinical vignette

Biventricular non-compaction and giant left atrial appendage

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A young man with unexplained symptoms of breathlessness presented for cardiovascular magnetic resonance imaging (MRI) following a non-contrast echocardiogram that had suggested mild impairment of function. As he was being placed on the MRI table, the technologist spontaneously commented upon his unusual facial features with the words 'he's a funny looking kid'. Cardiac MRI revealed dramatic evidence of non-compaction cardiomyopathy affecting both ventricles. The diagnostic appearance of excessive and long trabeculations with deep intratrabecular recesses is shown in axial (Panel A) and short axis (Panel B) steady state free precession (SSFP) images together with a 4 chamber ‘delayed enhancement’ image post-gadolinium (Panel C). A thin outer layer of normally compacted myocardium was also apparent. From the MRI examination, it was clear that the pathological process was affecting both ventricles. The other striking abnormality in the MRI was the unusual size and prominence of the left atrial appendage (Panel D).

Non-compaction affecting both ventricles is only rarely described. It is likely that there exists a direct genetic linkage between the patient’s dysmorphic appearance and his cardiac phenotype. Non-compaction certainly occurs in association with a range of congenital disorders. Giant left atrial appendage has not been previously described in association with this condition. It may be important given the prevalence of systemic embolism, which can be a presenting feature of left ventricular non-compaction. It may be important given the prevalence of systemic embolism, which can be a presenting feature of left ventricular non-compaction.

Panel A. Axial SSFP image.
Panel B. Short-axis SSFP image.
Panel C. Four-chamber delayed enhancement image without evidence of myocardial fibrosis.
Panel D. Giant left atrial appendage on axial SSFP image.

Supplementary video clips demonstrating left and right ventricular contraction in axial and short axis oblique imaging planes are available at European Heart Journal online.