

Non-Compaction of the Right Atrium and Left Ventricle in Ebstein's Malformation

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Ebstein's malformation (EM) is characterized by dysplasia and displacement of the tricuspid inferior and septal leaflets from the true atrioventricular (AV) junction. Left ventricular hypertrabeculation/non-compaction (LVHT) including the 'atrialized' portion in EM has not been described. A 42-year-old man with a history of radiofrequency ablation of a Mahaim-like bundle suffered from chest pain. Coronary angiography was normal, but echocardiog-

raphy showed a septal tricuspid leaflet inserting 3.5-cm apically beyond the AV junction, deep recesses of the atrialized interventricular septum, and a heavily trabeculated left ventricle; these were confirmed by cardiac magnetic resonance imaging. Neurologically, hypoacusis, positive pyramidal signs, postural tremor and brisk tendon reflexes were identified.

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Ebstein's malformation (EM) is a rare cardiac lesion, characterized by dysplasia and displacement of the proximal attachments of the tricuspid inferior and septal leaflets from the true atrioventricular junction. EM has been reported to be associated with left heart lesions including left ventricular hypertrabeculation/non-compaction (LVHT) (1,2). LVHT affecting both sides of the interventricular septum including the 'atrialized' portion in EM has not been described to date.

Case report

A 42-year-old HIV-negative male patient was admitted because of sudden onset of chest pain. Since childhood, he had suffered from exercise-induced palpitations, but two years earlier EM had been diagnosed and a Mahaim-like bundle had been removed using radiofrequency ablation. Since then the palpitations had decreased. On examination, the patient's blood pressure was 140/80 mmHg, while the electrocardiogram showed sinus rhythm (heart rate 50 beats/min), Q waves in leads II and III, and AVF and ST-elevations in V₁-V₃. Coronary angiography was normal, but echocardiography demonstrated a normal-sized left ventricle with thickened myocardium

(interventricular septum 17 mm; left ventricular posterior wall 13 mm), an excellent systolic function, and more than four trabeculations of the left ventricular apex and interventricular septum (Fig. 1). The septal tricuspid leaflet inserted 3.5 cm apically beyond the atrioventricular junction, and deep recesses of the atrialized interventricular septum were visible within the right atrium. There was only slight tricuspid insufficiency. The diagnosis of LVHT was confirmed by cardiac magnetic resonance imaging (Fig. 2).

Since LVHT is often associated with neuromuscular disorders (3), the patient was also examined neurologically. These investigations revealed left hypoacusis, positive pyramidal signs on the upper limbs, postural tremor and brisk tendon reflexes.

Twenty months later, the patient was in a stable clinical condition, though further follow up investigations are scheduled to assess the long-term outcome.

References

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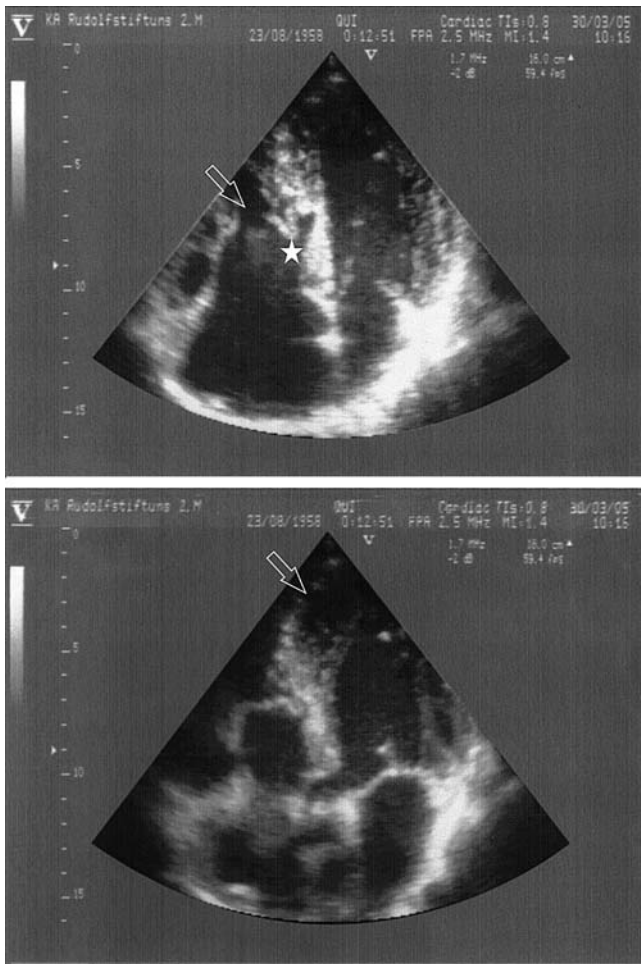


Figure 1: Upper panel: Echocardiographic apical four-chamber view showing apical displacement of the septal tricuspid leaflet (arrow) and a deep intramural recess of the atrialized portion of the interventricular septum within the right atrium (asterisk). Lower panel: Echocardiographic apical five-chamber view of the same patient, showing left ventricular hypertrabeculation/non-compaction of the left ventricular lateral wall, apex and interventricular septum (arrow).

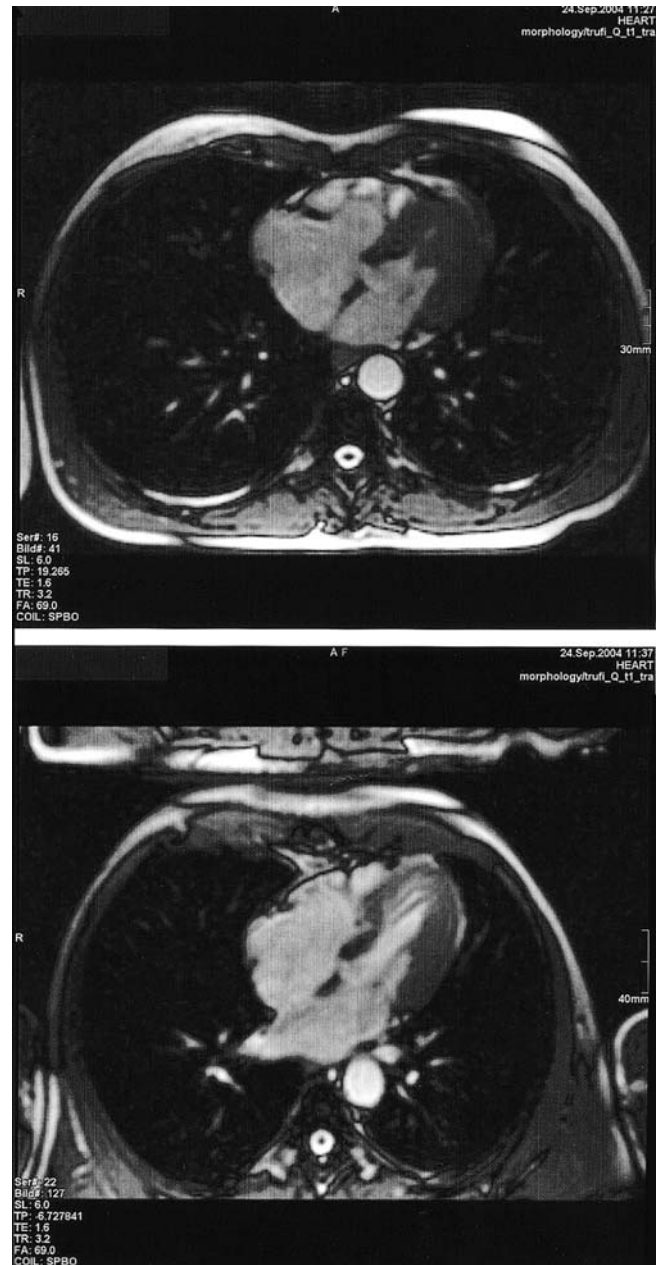


Figure 2: Upper panel: Cardiac magnetic resonance axial TrueFisp T₁ bright blood four-chamber views showing apical displacement of the septal tricuspid leaflet and prominent trabeculation of the right atrium and 'true' right ventricle. Lower panel: Thickening and hypertrabeculation of the left ventricular walls.