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Biventricular non-compaction cardiomyopathy: Rare disease and far rarer case

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Isolated ventricular non-compaction (IVNC) is congenital cardiomyopathy defined by the appearance of prominent ventricular trabeculae and deep intertrabecular recesses on cardiac imaging [1]. Although biventricular non-compaction (BiVNC) has been reported rarely [2], its strict diagnostic criteria remain unclear. In the study by Lutokhina et al. [3], a combination of IVNC with arrhythmogenic right ventricular cardiomyopathy (ARVC) was found in 14.8% of the patient subset. The below-mentioned report presents a challenging case of the biventricular hypertrabeculation phenotype and its unforeseen consequences.

A 36-year-old Caucasian man without prior medical history presented with dyspnea at rest and, on admission, angiographically documented intermediate-high risk pulmonary embolism (PE) complicated with pulmonary and renal infarction as well as pneumonia. Reperfusion treatment was not considered due to the patient's hemodynamic stability. Laboratory examination revealed: C-reactive protein level 369 mg/l (normal range [NR] <6 mg/l); D-dimers 4.5 mg/l (NR <0.5 mg/l); creatinine 0.97 mg/dL (NR 0.6–1.6 mg/dl); N-terminal pro-B-type natriuretic peptide 8649 pg/ml (NR <125 ng/l); high-sensitivity cardiac troponin T 203 ng/l (NR <14 ng/l).

An electrocardiogram on admission revealed a right bundle branch block with epsilon wave (Supplementary material, *Figure S1* [arrows]).

Transthoracic echocardiogram at admission showed right ventricle (RV) systolic function at a lower range limit (Tricuspid Annular Plane Systolic Excursion 17 mm, S' 10 cm/s), lack of tricuspid regurgitation, severely reduced left ventricle (LV) ejection fraction (EF) as much as 15%, increased LV trabeculation and thrombus 22 × 17 mm in the LV apex (Figure 1A, Supplementary material, *Video S1*). Cardiac magnetic resonance confirmed LV non-compaction with non-compacted/compacted (NC/C) ratio 4.2 and revealed right ventricular non-compaction with NC/C ratio 4.2, left and right ventricular EF impairment (14% and 32%, respectively), and the presence of thrombus in the LV apex 20 × 12 mm (Figure 1B–C, Supplementary material, *Video S2*). Moreover, biventricular circumferential subendocardial distribution of late gadolinium enhancement, less common than mid-myocardial [4], was observed (Figure 1D). Late gadolinium enhancement pattern may result from connective tissue diseases but is also highly specific for coexisting dilated cardiomyopathy. A complete package of rheumatological tests has been performed, and connective tissue diseases have been excluded. The course of BiVNC was complicated with prehospital thromboembolic events diagnosed during hospitalization deriving both from RV (PE) and LV (pulmonary infarction, clinically silent renal infarction) (Figure 1E–F). Initial treatment included B-blocker, mineralocorticoid receptor antagonist, diuretics, ivabradine, broad-spectrum antibiotic therapy, anticoagulation primarily with low molecular weight heparin, and then Vitamin K Antagonist, resulting in the resolution of the LV apex thrombus. Sacubitril/valsartan and the sodium-glucose co-transporter 2 inhibitor were initiated early after acute decompensation of heart failure, resulting in LV EF improvement of up to 25% and concomitant symptoms reduction (New York Heart Association class IV to II) within the next 3 months. Chronic anticoagulation with dabigatran was subsequently introduced. The patient was applied as a potential heart transplant recipient and electively implanted with a cardiac resynchronization therapy defibrillator.

Reported complications in IVNC are heterogeneous and poorly understood [5]. The present case study shows that IVNC may include both ventricles and present as thromboembolic events deriving from both sides of a heart. This unusual case of BiVNC shows, the borderline features of ARVC [3] (regional RV akinesia, RV EF ≤40%, and epsilon wave), which should be considered while managing the patient.

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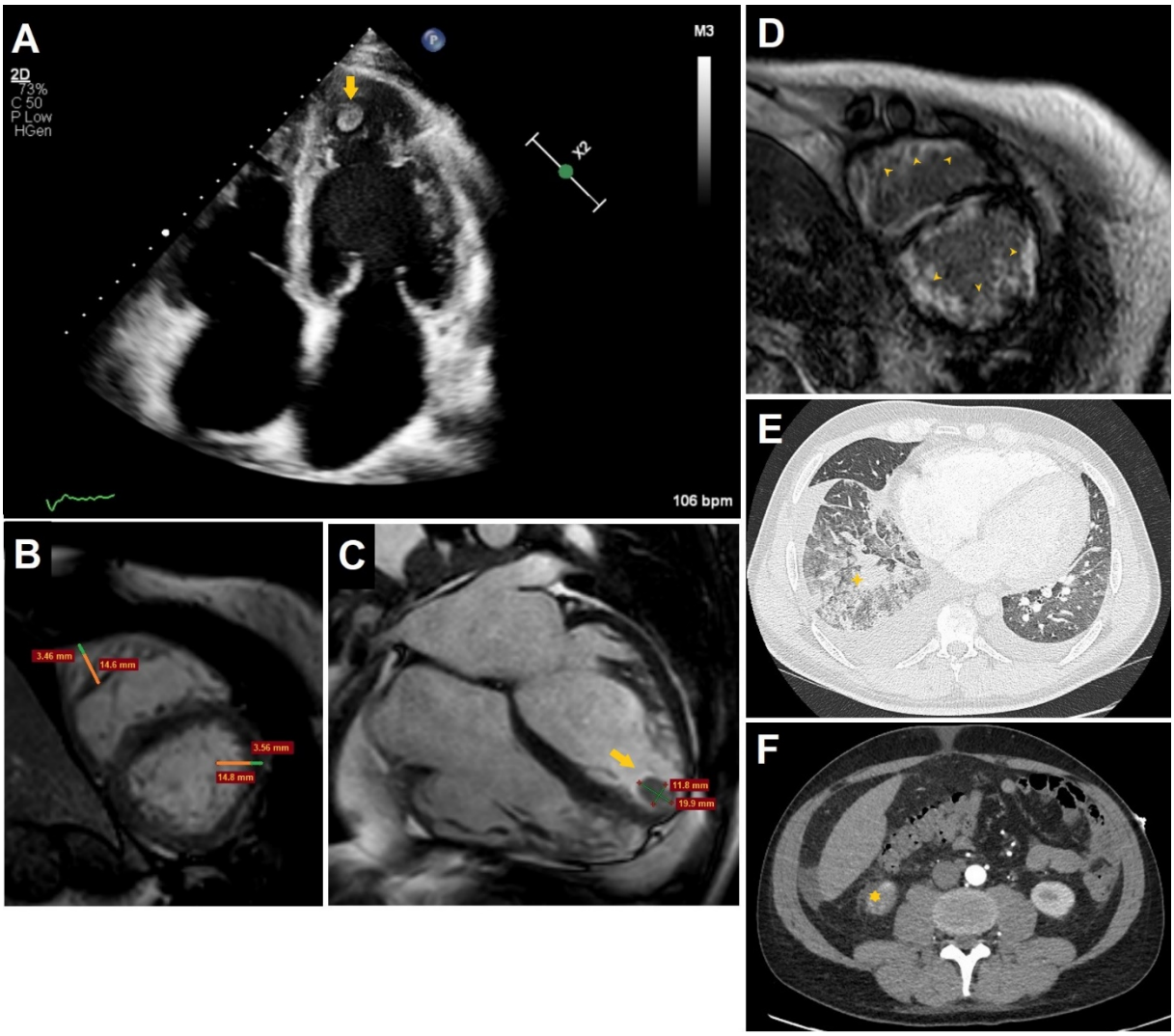

Figure 1. A. Transthoracic echocardiography in apical 4-chamber view showing LV hypertrabeculation and thrombus in the LV apex (arrow). **B, C.** CMR cinematographic sequences in short axis (**B**) and 4-chamber (**C**) views showing non-compacted to compacted myocardium ratio for both ventricles and thrombus in the LV apex (arrow). **D.** CMR short tau inversion recovery sequence showing biventricular endocardial circumferential late gadolinium enhancement (arrowheads). **E.** High resolution computed tomography viewed in lung window with mixed lesions in the right lower lobe typical for infarction and pneumonia coexistence (asterisk). **F.** Contrast-enhanced computed tomography reveals a hypodense lesion in the right kidney's lower pole related to the infarction area (star)

Abbreviations: CMR, cardiac magnetic resonance; LV, left ventricle