A rare unclassified cardiomyopathy: isolated right ventricle noncompaction

Sınıflandırılmamış nadir kardiyomiyopati: İzole sağ ventrikül süngerimsi miyokart

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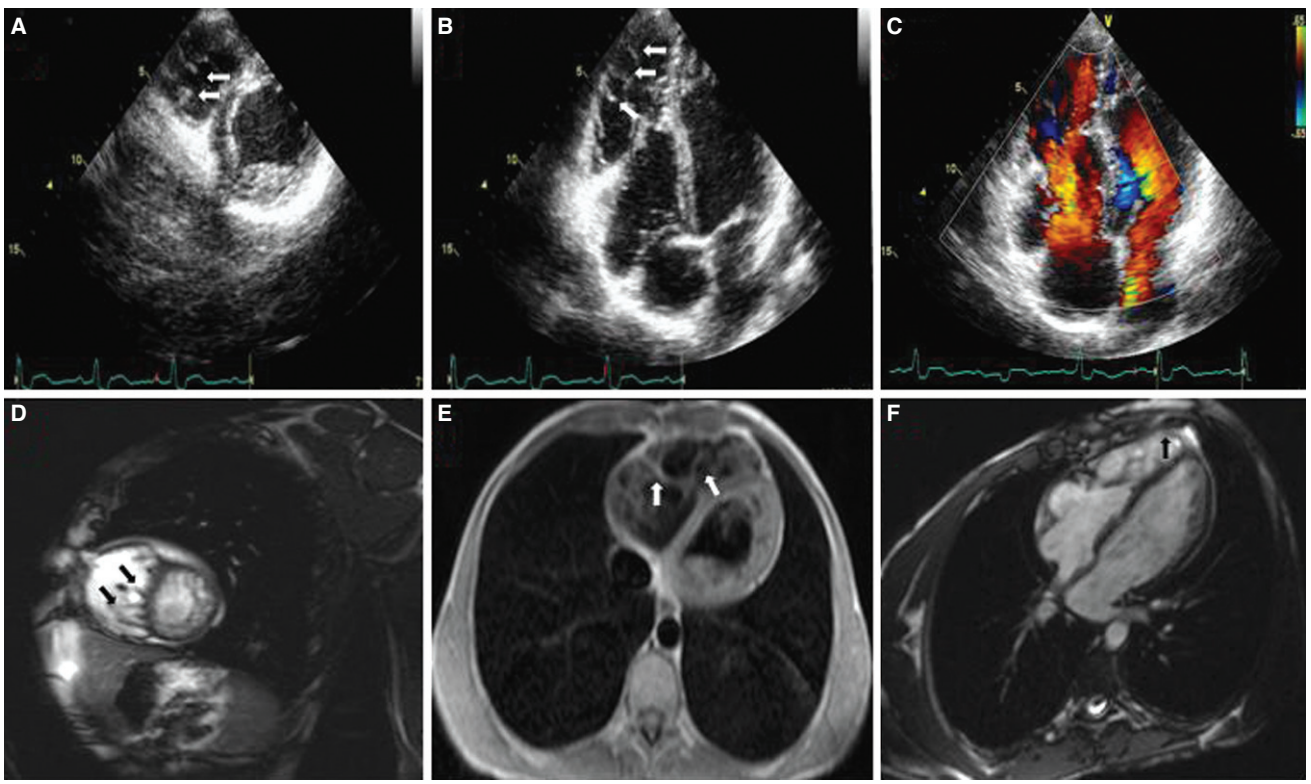
A 23-year-old man was referred to our hospital with palpitation and increasing exertional dyspnea. Two-dimensional echocardiography and color Doppler demonstrated marked right ventricular (RV) trabeculation and intratrabecular recesses (Fig. A-C, Supplementary Video 1-3*). Cardiac magnetic resonance imaging (cMRI) confirmed RV dilatation and depressed RV function. There was dyskinetic movement in the right ventricle apex and lateral segments. cMRI also showed a double layered appearance with non-compacted and compacted endomyocardial segment in the mid ventricle, apex and lateral free wall. Ratio of the maximum thickness of the non-compacted to compacted layer at the end systole was >2 consistent with the diagnosis of RV non-compaction cardiomyopathy. In addition, marked trabeculation and deep intratrabecular recesses were noted mid ventricle, apex and lateral free wall (Fig. D-F). All segments of the left ventricle were compacted in both echocardiography and cMRI. Based on echocardiography and cMRI, left ventricle and biventricular non-compaction are more frequently seen than isolated RV non-compaction. In our case, we could show isolated RV non-compaction that is rarely seen disease by using transthoracic echocardiography and cMRI.



Figures– Transthoracic echocardiography from the short **(A)** and apical 4-chamber axis **(B)** views showing marked trabeculation and deep recesses in the right ventricular apex, mid and lateral free wall with compacted left ventricle (white arrows). **(C)** The same figure as Figure B showing, intertrabecular spaces are filled by direct blood flow from the right ventricular cavity. **(D-F)** ECG-gated, cardiac MRI showing a double-layered appearance on the short axis view (black arrows) and marked trabeculations and deep intratrabecular recesses in the mid ventricle, apex and lateral free wall of the right ventricle. **(F)** Dyskinetic movement is observed in the right ventricular apical and lateral segments. **Supplementary video files associated with this presentation can be found in the online version of the journal.*