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Rare case of Isolated left ventricular apical hypoplasia accompanied by Left ventricular noncompaction

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Background: Isolated left ventricular apical hypoplasia (ILVAH) was first described as congenital heart disease with an unusual type of cardiomyopathy in 2004 (Cleveland Clinic, USA). The review the data of worldwide literature contains the description of 42 clinical cases in patients with ILVAH. This rare phenomenon frequently presents with different clinical forms from no symptoms in children to heart failure (HF), pulmonary edema, atrial fibrillation (AF) or malignant tachycardia in adults. The early TTE and CMR diagnosis is particularly important due to the difference in the prognosis and treatment of ILVAH from those of other diseases with symptoms similar to this rare entity. We present a first case of surgical treatment in an adult with ILVAH complicated by persistent AF and biventricular HF.

Case Presentation: A 32-year-old male patient with exertional dyspnea and poorly controlled AF was admitted to our hospital for the treatment of a persistent AF. He had no history of congenital HF, and there was no family history of premature coronary artery disease, cardiomyopathy or sudden cardiac death. A TTE revealed a mildly dilated LV with moderately to severely reduced function; End-diastolic dimensions: LA: 65 ml/m2; RA: 52 ml/m2; LV: 108 ml/m2; LV diameters: basal - 65 mm; mid - 67 mm; longitudinal – 43 mm; RV diameters: basal - 38mm, mid – 30 mm, longitudinal – 96 mm. LV ejection fraction (LVEF) was 32% by 3D quantification and CMR. The LV had a spherical appearance with a thin-walled, truncated, and akinetic LV apex. The RV appeared elongated and was noted to wrap around the distal LV, RV systolic function was abnormal (FAC -29%). There were significant tricuspid regurgitation and high pulmonary artery systolic pressure as an important marker for RV dysfunction and pulmonary hypertension. The CMR and TTE demonstrated all of the phenotypical features of ILVAH: 1) a spherical truncated LV with impaired function; 2) replacement of the LV apex with fatty material contiguous with epicardial fat; 3) anteroapical origin of the papillary muscle network; and 4) an elongated RV. Non-compacted endomyocardial layer and deep myocardial trabeculae, particularly in the mid-cavity of the LV was determined by both methods. The surgical treatment that was done consistently by implantation of ICD device in primary sudden cardiac death prevention and pulmonary vein isolation (PVI) for AF treatment. After one-year follow-up, there are sinus rhythm, improvement of EFLV and FAC. Importantly, LA and RA volumes are the same in comparison with first exam, and our patient still display moderate TR and pulmonary hypertension.

Conclusion: This is the first description of rare combination of ILVAH and LV noncompaction with a persistent AF and biventricular HF with successful results of surgical treatment. This case also demonstrates the degree to which such patients are highly dependent on atrial contractile function because of altered LV geometry.
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