Letter to the Editor — Editöre Mektup

Do not only ablate but also look for myocardial fibrosis and myopathy in noncompaction

Dear Editor,

With interest we read the article by Alper et al. about a 20-year-old male with right ventricular and left ventricular hypertrabeculation/noncompaction (LVHT), frequent ventricular ectopic beats, and nonsustained ventricular tachycardia, who benefited from radiofrequency ablation of the midlateral portion of the tricuspid annulus.[1] We have the following comments and concerns.

We do not agree with the statement that LVHT generally results from abnormal embryogenesis of the endocardium and myocardium.[1] Though LVHT is frequently congenital, there are a number of cases in which LVHT develops postnatally or even in adulthood (acquired LVHT).[2] Acquired LVHT has been reported particularly in patients with neuromuscular disorders (NMDs), pregnant females, and training athletes.

We also do not agree with the statement that the main clinical manifestations of LVHT are arrhythmias, heart failure, or embolism.[1] LVHT is usually asymptomatic and frequently detected randomly but can occasionally be complicated by arrhythmias, heart failure, or embolism. The frequencies of these complications are quite variable in the literature and also vary between children and adults.

LVHT is frequently associated with subendocardial or myocardial fibrosis, which could be the cause of conduction defects or arrhythmias in the presented case.[3] Did the authors look for late gadolinium enhancement on cardiac magnetic resonance imaging, which is regarded as the morphological equivalent of myocardial or subendocardial fibrosis? Were there any areas of wall motion abnormalities attributable to myocardial fibrosis?

In up to 80% of LVHT patients, an underlying NMD can be detected if patients are systematically investigated by a mycologist.[4] Did the patient ever experience easy fatigability, exercise intolerance, myalgias, muscle cramps, muscle stiffness, or muscle weakness? Did he ever experience an episode of malignant hyperthermia during general anesthesia? Did the presented patient undergo a neurological exam or blood drawings for muscle enzymes? Did he ever undergo nerve conduction studies or needle electromyography?

Myocardial fibrosis can be a hallmark of cardiac involvement in NMDs and has been reported particularly in patients with dystrophinopathies,[4] mitochondrial disorders,[5] and myotonic dystrophy.[6] Myocardial fibrosis may not only be the cause of progressive dilatation of atria and ventricles in these patients but also the cause of conduction defects and arrhythmias.

In 15–30% of cases, LVHT can be detected in first-degree relatives of an index case (familial occurrence of LVHT).[7] Were first-degree relatives of the presented patient investigated for LVHT? Was the family history positive for palpitations, syncope, fainting, heart failure, or cardiac embolism? Was the family history positive for an NMD?

Overall, this interesting case could profit from more extensive investigations of the presence or absence of subendocardial or myocardial fibrosis, NMD in the index case and his relatives, and LVHT in other family members. LVHT patients require comprehensive, multiprofessional work-up to unravel the still enigmatic pathogenesis of this interesting morphological abnormality.

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Conflict-of-interest issues regarding the authorship or article: None declared

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Authors' reply

To the Editor,

We would like to thank the authors for their interest and valuable contribution to the case.[1] We agree with the points highlighted regarding the etiology and clinical presentation of left ventricular hypertrabeculation (LVHT). Unfortunately, an extensive workup for an underlying disease was not performed in the index case. However, the patient did not complain of symptoms related to neuromuscular disorders, and there was no family history of cardiomyopathy.

As reported, no wall motion abnormalities were observed in echocardiographic examination, and myocardial fibrosis was not observed in gadolinium-enhanced magnetic resonance imaging. Thus, we do not believe that myocardial fibrosis was the primary pathology that caused the tachycardia.

Arrhythmia is a common presentation and a leading cause of death in patients with LVHT.[2] However, benign ventricular arrhythmias can also be seen in LVHT. We had previously reported 2 cases of ventricular tachycardia (VT) originating in the right ventricle in patients with LVHT.[3,4] An intracardiac cardioverter defibrillator was not implanted in either patient, and radiofrequency (RF) ablation and medical therapy were adequate for relief of symptoms. In one case, left ventricular function normalized after RF ablation of ectopic beats originating in the right ventricle.[3] From a clinical perspective, the significance of the index case was the occurrence of VT originating from the tricuspid annulus, in which case catheter ablation is the recommended therapy. Electrocardiography was typical, and RF ablation was successful. While our results could not explain a pathophysiological basis for the occurrence of arrhythmia originating from the tricuspid, ours was the first reported case of this arrhythmia in a patient with LVHT.

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Conflict-of-interest issues regarding the authorship or article: None declared

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