DOI: 10.7860/JCDR/2016/16290.8070

Letter to Editor

Internal Medicine Section

Consider Differentials of MRI Myocarditis in Noncompaction

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Sir,

We were appreciated to read the report by Karaca et al., about a 21-year-old male with presumed myocarditis and left ventricular hypertrabeculation/ noncompaction (LVHT) [1]. The paper raises a number of questions and concerns. We do not share the view that LVHT is a rare condition [1]. Meanwhile, LVHT is regarded as the third most frequent of the cardiomyopathies. However, most frequently LVHT remains undetected because it remains asymptomatic during a long period of time.

We also do not share the view that LVHT is exclusively congenital [1]. In a number of patients LVHT has been shown to occur after birth or during adult life (acquired LVHT). Though the pathogenesis of acquired LVHT remains elusive, there are indications that it may reflect hormonal changes or may represent an adaptive mechanism to volume or pressure overload, since it occurs in professionally training athletes or pregnant females [2]. There are also rare cases in which LVHT disappears over time. The diagnosis of myocarditis was only established upon the clinical presentation, blood chemical values and the cardiac MRI findings. Since no biopsy of the right ventricular myocardium had been carried out, myocarditis can be suspected but is not definitively proven. The strongest argument for myocarditis is the diffuse oedema on cardiac MRI [1], which has not been reported in association with LVHT but in association with other conditions. Late gadolinium enhancement (LGE) in patients with LVHT is a frequent finding and can be also due to scarring or subendocardial fibrosis, frequently associated with LVHT [3]. LGE most frequently shows a midventricular or transmural distribution but only rarely a subepicardial or subendocardial distribution [3].

We do not share the approach to implant an implantable cardioverter defibrillator (ICD) in a patient with LVHT just because he has LVHT. LVHT may be asymptomatic in most of the cases but can be complicated by malignant rhythm abnormalities. This is why patients with LVHT need to be frequently monitored for ventricular arrhythmias. In case the family history is positive for sudden cardiac death, recurrent syncopes occur, or long-term ECG recordings disclose a predisposition for ventricular arrhythmias, implantation of an ICD can be proposed. Since LVHT is frequently associated with neuromuscular disorders (NMDs) or chromosomal defects [4], the patient should undergo neurological investigations. Was there any indication for muscle disease, such as elevated creatine-kinase, muscle cramps, muscle weakness, muscle wasting, or easy fatigability? Were dysmorphic features obvious on clinical examination?

In a number of cases LVHT occurs familiarly [5]. Were other family members screened for LVHT? Was LVHT found in any of the relatives of the patient? Was there consanguinity between the parents?

Overall, this interesting case merits further diagnostic work-up with regard to the genetic background, neurological comorbidity, and familiarity of LVHT. Certainly, not each LVHT patient requires implantation of an ICD which should meet established criteria for indicating such an approach. LGE and diffuse T2A hyperintensity should be evaluated for potential differential diagnoses, such as vasculitis, rheumatoid arthritis, amyloidosis, or NMD. Coronary-angiography findings should be reported.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 16, 2015**
Date of Peer Review: **Oct 12, 2015**
Date of Acceptance: **Oct 30, 2015**
Date of Publishing: **Jul 01, 2016**

REPLY FROM THE AUTHOR

We read the letter with great interest that led to criticisms regarding our previous report. Although we agree with some parts of the comments derived by the authors, we believe that several important issues relating to the topic should be clarified. First, non-compaction cardiomyopathy (NC-CMP) as described by the American Heart Association (AHA) [1] is a rare form of primary genetic cardiomyopathy believed to result from the failure of myocardial development process during embryogenesis. It must be differentiated from acquired forms of left ventricular hyper-trabeculations. NC-CMP is a congenital disease associated with systolic/diastolic dysfunction of the left ventricle. The related mutations were previously well-defined [2]. The ongoing debate regarding the incidence is derived from the diagnostic dilemma between true NC-CMP and other forms of LV hyper-trabeculations. Second, myocarditis is a clinical diagnosis most of the time that's achieved by combination of history, electrocardiography, increased cardiac troponins and associated systolic dysfunction with imaging modalities. It does not always require an endomyocardial biopsy,

especially in a patient with haemodynamically stable clinical status [3].

Third, due to the limited word count of the journal for a case image, we could not have given information regarding the treatment process of the given patient. The decision for implanting an ICD was given based on the documentation of non-sustained ventricular tachycardia on holter monitoring and subsequently the induction of haemodynamically unstable ventricular tachycardia on electrophysiological study. Again, our patient was a sporadic case without any associations of a neuromuscular disorder. First degree relatives were scanned with echocardiography but no signs of LV hyper-trabeculation or systolic dysfunction were reported.

Thanks,
Dr. Oguz Karaca

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