Consider the neuromuscular and non-congenital implications of left ventricular noncompaction

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Keywords
non-compaction, unclassified cardiomyopathy, myopathy, neuropathy, muscular dystrophy, cardiac involvement

Letter to the Editor

In a recent article, Alihanoglu et al. reported about a 25yo female with left ventricular hypertrabeculation / noncompaction (LVHT), heart failure and atrial fibrillation who benefitted from heart failure therapy, oral anticoagulation, and implantation of an implantable cardioverter defibrillator (ICD). We have the following comments and concerns.

The authors do not refer to the frequent association of LVHT with neuromuscular disease. In up to 80% of the cases with LVHT a NMD can be detected if LVHT patients are systematically screened by a myologist. Since presence of NMD in LVHT patients strongly determines their outcome, it is important to refer these patients systematically to the myologist. Did the patient, her mother, or other relatives present with clinical manifestations of a NMD, such as muscle weakness, wasting, reduced tendon reflexes, muscle cramping, muscle stiffness, myalgia, sore muscles, easy fatigability, fasciculations, myokymia, muscle rupture, elevated creatine-kinase, or myoglobinuria? Did she ever undergo nerve conduction studies or needle electromyography?

The reported patient presented with tachycardious atrial fibrillation (AF) why she received oral anticoagulation. AF has been identified as a predictor of mortality in LVHT patients. Which type of agent did she receive? Was AF also recorded in the patient’s mother before decease? Was the mother also put on oral anticoagulation? Was “sudden death” actually due to embolic stroke? Did she undergo autopsy? Was ever a MRI of the cerebrum carried to look for subclinical cerebrovascular events? Did the mother or the presented patient ever develop cardio-embolic events outside the cerebrum?

We do not agree with the frequently cited phrase that LVHT is a congenital disorder in all cases. Though LVHT is presumably congenital in the majority of the cases, it may also develop during adult life (acquired LVHT) as has been shown particularly in patients with NMDs, pregnant females and training athletes. It is also noteworthy that LVHT may be a temporary condition disappearing after some time or after delivery. Acquired LVHT may result from increased preload conditions during pregnancy (adaptation to increased stroke volumes), reduced cellular adhesion of cardiomyocytes, an unsuccessful attempt to overcome a metabolic defect leading to systolic dysfunction, enlargement of the endocardial surface to improve oxygenation from the ventricular side, or from micro-infarctions.

There is also disagreement concerning the statement that LVHT has been first described in 1984. Though Engberding et al. were the first to detect and describe isolated LVHT on echocardiography, biventricular hypertrabeculation has been already demonstrated by Westwood et al. in 1975 and by Feldt et al. in 1969. Recently, Udeoji et al. reported that LVHT was even described in 1932 at autopsy of a newborn with aortic atresia and a coronary-ventricular fistula. Overall, this interesting case could profit from more comprehensive examinations of the patient and his relatives to eventually contribute to the many unsolved issues concerning etiology, pathogenesis, diagnosis, prognosis, and management of LVHT. Also the neuromuscular and non-congenital implications of LVHT should be addressed.

References

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