Left ventricular non-compaction its Benign until It's not

Abstract

LVNC is a morphology that may be benign or may be a cardiomyopathy with potential for heart failure, ventricular arrhythmias and thromboembolic events. Additionally the various phenotypic expressions are heterogeneous and have the potential to transition from one morphological cardiomyopathy to another. This commentary gives a unique example of a transition from HCM to LVNC cardiomyopathy.

Commentary

The Imaging Vignette by Aung, et al. "LV Noncompaction Cardiomyopathy or Just a lot of Trabeculations" attempts to make a critical distinction between Left Ventricular Hypertrabeculation (LVHT) – a presumed "benign" clinical state and LVHT/LVNC cardiomyopathy, a disease state.

The distinction is important because we do not want to label healthy people as diseased, and when Left Ventricular Hypertrabeculation (LVHT) is associated with a cardiomyopathy it has the potential for significant clinical sequalae: including potentially heart failure, thromboembolism, and malignant/fatal arrhythmias. This latter condition requires important medical therapy, intense medical surveillance, potentially device therapy and recommendations regarding activity levels.

Left Ventricular Non-compaction (LVNC) is a heterogeneous clinical condition, which intersects/overlaps with other established cardiomyopathies. The heterogeneity is exemplified by a case of hypertrophy cardiomyopathy diagnosed in 2007 that "transitioned" to classical morphologic features of LVNC cardiomyopathy with heart failure in 2013. This transition had major implications for therapy and prognosis.

The widespread use of echocardiography and the increased awareness of LVNC has resulted in more patients meeting the "diagnostic criteria" of LVNC, characterized by a bi-layered "spongy looking" myocardium with a ratio of noncompacted to compacted myocardium >2. However, many of these individuals have normal LV systolic function, normal diastolic function parameters, tissue Doppler, and speckle tracking parameter including strain, strain rate and LV twist and rotation [1-3].

The query is whether morphological features without diastolic/mechanical features represent an early precursor form of LVNC/cardiomyopathy or simply a "benign" phenotype of LVNC best referred to as LV hypertrabeculation (LVHT) with an uncertain destiny? The query confronts the medical community and the patients presenting with the morphological features of LVNC. Arbustini et. al. proposed a nosology, MOGE(S), which improves our recognition and understanding of cardiomyopathies, including LVNC [4,5].

The MOGE(S) nosology comprehensively classifies five features for each cardiomyopathy: morphofunction phenotype (M), organ involvement (O), genetic or familial inheritance pattern (G), etiologic annotation (E), and functional classification (S). This systematic approach allows a distinction between pure LVNC (MLVNC/LVHT) - a "benign" condition from LVNC with dilatation and LV dysfunction (MLVNC + D) and hypertrophy (MLVNCC +H). The critical feature is

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distinguishing the "benign form" from a "disease state" to prevent over diagnosis, and prevention of healthy people being labeled with a disease and being mistakenly restricted in their activities of daily living [4,5].

The judicious use of echocardiography and Magnetic Resonance Imaging (MRI) by thoughtful physicians should allow accurate diagnosis, appropriate follow up, and meaningful prognostication. The MOGE(S) nosology may be used to aid in separating "benign" morphological condition from a cardiomyopathy.

An initial echocardiogram and MRI should allow us to identify LVHT from a cardiomyopathy with potential for adverse cardiac events. This "benign" subset will have normal LV systolic function and normal diastolic function parameters including tissue Doppler as well as normal speckle tracking parameter: including strain, strain rate, and LV twist and rotation. These benign patients should have no medical restrictions based upon LVHT.

How do we follow these patients with LVHT in clinical practice? Lifetime surveillance echoes performed yearly, or more frequently if the patient develops cardiac symptoms. This surveillance will allow us to understand the natural history of LVHT and understand what echocardiographic and MRI parameters change when there is a transition from "benign" to a cardiomyopathy.

The case we present keeps the clinician humble and reinforces the importance of clinical surveillance and cardiac imaging. In 2007, the patient had classic morphological features of hypertrophy cardiomyopathy. In 2013, the patient had transitioned to classic features of MLVNC +D. Shortly thereafter the patient was transitioned to the heart failure team. This reinforces the heterogeneous nature of the cardiomyopathies and the potential to transition from one phenotype to another in a short period of time.

The importance of comprehensive clinical and imaging surveillance can not be overemphasized because the evolution from "benign" to disease results in changing medical therapy, potential device therapy, and activity restriction that hopefully reduce the likelihood of morbidity and mortality. The future management of these complex, heterogeneous cardiomyopathy cases will require a deep insight into the genetics of these complex cardiomyopathies that have the potential to alter phenotypic expression over time [6,7].

A look to the future to develop a deeper understanding of these cardiomyopathies will require a historical review1 and a collaboration across medical researchers looking at the clinical features, phenotypiTc expressions and evolving genomics of these complex cardiomyopathies.

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