Left ventricular noncompaction cardiomyopathy: diagnostic and therapeutic dilemmas

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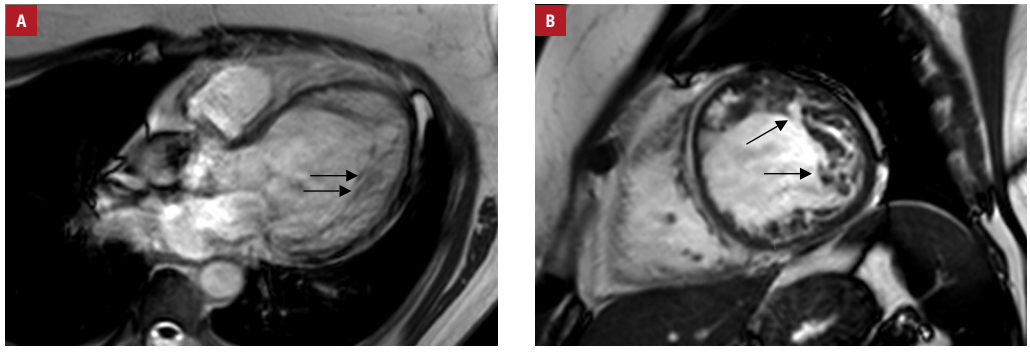

FIGURE 1 Long- (**A**) and short-axis (**B**) still frames from routine balanced steady-state free precession cine sequence (Philips Achieva TX 3.0T, Eindhoven, The Netherlands), showing marked thinning of the compacted layer, along with the thick, noncompacted layer and elongated trabeculae in the apex. Note the altered structure of the papillary muscles, which are largely composed of trabeculations and lack their regular solid base (arrows).

We present a case of a 62-year-old woman with left ventricular noncompaction cardiomyopathy (LVNC) and its 3 complications: heart failure (HF), ventricular arrhythmia, and stroke.

At the time of diagnosis, the patient's echocardiogram showed left ventricular ejection fraction (LVEF) of 28% and a 2-layered myocardium with a ratio of noncompacted to compacted layers of 3:1 (Supplementary material, *Figure S1*). In addition, cardiac magnetic resonance imaging demonstrated pronounced thinning of the compacted layer and abnormal (fragmented) bases of the papillary muscles (FIGURE 1). Neither late gadolinium enhancement nor thrombi were found. Despite optimal medical treatment, LVEF did not improve and an implantable cardioverter-defibrillator (ICD) was inserted for primary prevention of sudden cardiac death. Two years later, ventricular fibrillation was terminated by the ICD (Supplementary material, *Figure S2*).

Three years later, the patient presented with symptoms of stroke. Posterior cerebral artery territory acute ischemia was diagnosed on computed tomography and reperfusion therapy with alteplase was administered. Based on a comprehensive diagnostic workup, cardiogenic embolism was considered the most likely diagnosis and apixaban was prescribed for stroke prevention. Left ventricular noncompaction cardiomyopathy was also diagnosed in the patient's 2 daughters. Genetic testing of the proband is underway.

Left ventricular noncompaction cardiomyopathy is a rare congenital disease characterized by a spongy appearance of the myocardium and abnormal trabeculations occurring typically in the apical, mid-lateral, and inferior LV wall.[1] The disease can lead to progressive HF, ventricular arrhythmias, and systemic embolism

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(SE). Until now, no specific therapy for LVNC has been established. Patients with LVNC and asymptomatic LV systolic dysfunction or HF should be treated according to standard guidelines, and ICD placement for primary sudden cardiac death prevention is indicated in those with LVEF ≤35%.[1]

Patients with LVNC and a prior cardioembolic event should receive anticoagulation. However, there is much controversy around risk assessment and the optimal strategy for the prevention of thromboembolism. It has been postulated that patients with LVNC and atrial fibrillation or depressed LVEF may represent a group at high risk of developing SE, similar to hypertrophic cardiomyopathy. Thus, according to the recent Heart Rhythm Society guidelines, anticoagulation could be initiated in LVNC with LVEF <40%, even in the absence of any other specific indication,[1] which is at variance with the general recommendation in HF. The value of anticoagulation in LVNC patients with preserved LVEF is less clear, because no SE was reported in this group of patients.[1,2] Furthermore, antiplatelet therapy in patients with LVNC is not considered beneficial, except for pediatric use.[1] Given scant data on the efficacy of the therapy, the choice of an anticoagulant also remains uncertain. Results of the recent multicenter cohort study on ischemic and nonischemic HF showed that direct oral anticoagulant treatment was associated with a higher risk of stroke and/or SE compared with warfarin use, challenging the assumption of the equivalence of direct oral anticoagulants with warfarin in the treatment of LV thrombi.[3]

There is no gold standard for the diagnosis of LVNC. Therefore, preventing both under- and overdiagnosis is a priority. Other morphological findings, eg, maximal systolic compacted layer thickness <8 mm and absence of well-defined papillary muscles (FIGURE 1) can help avoid LVNC mislabeling.[4,5] The familial presentation of LVNC also improves the specificity of diagnosis.[1]

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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