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ADVANCED

CASE REPORT: CLINICAL CASE

Thebesian Veins Draining to the Left Ventricle, Mimicking Left Ventricular Noncompaction

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ABSTRACT

Left ventricular noncompaction cardiomyopathy (LVNC) was diagnosed in a 59-year-old woman, based on echocardiography. Later, diagnostic criteria were also found positive by cardiac magnetic resonance (CMR). However, coronary angiography revealed thebesian veins were causing the noncompacted appearance. The complementary role of CMR and echocardiography criteria, including flow assessment in the recesses, is discussed. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:2085–9) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

PRESENTATION

A 59-year-old asymptomatic woman presented with lateral T-wave inversions in a routine electrocardiogram (ECG) (Figure 1A). Clinical examination results were normal.

LEARNING OBJECTIVES

- To integrate both CMR and echocardiography information for the diagnosis of LVNC, as the 2 modalities complement each other.
- To apply the full set of criteria for the correct diagnosis of LVNC, including flow assessment in the recesses.
- To consider differential diagnoses of LVNC (thebesian veins, hypertrabeculation, clefts/crypts) to ensure optimal therapy and risk assessment.

MEDICAL HISTORY

Medical history was unremarkable, and family history was negative for cardiomyopathies or sudden cardiac death.

DIFFERENTIAL DIAGNOSIS

Asymptomatic T-wave inversion can be associated with different kinds of cardiomyopathies, coronary artery disease, cardiomyopathy, or neurogenic causes or may reflect a normal variant. The initial investigative step is to confirm or rule out any structural heart disease.

INVESTIGATIONS

Echocardiography was performed, and isolated left ventricular noncompaction (LVNC) was suggested due to deep recesses and a systolic noncompacted-to-compacted ratio in the short axis of >2. Left ventricular ejection fraction (LVEF) and basal wall thickness

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ABBREVIATION AND ACRONYMS

CMR = cardiac magnetic resonance

CT = computed tomography

ECG = electrocardiography

HCM = hypertrophic cardiomyopathy

LV = left ventricle

LVEF = left ventricular ejection fraction

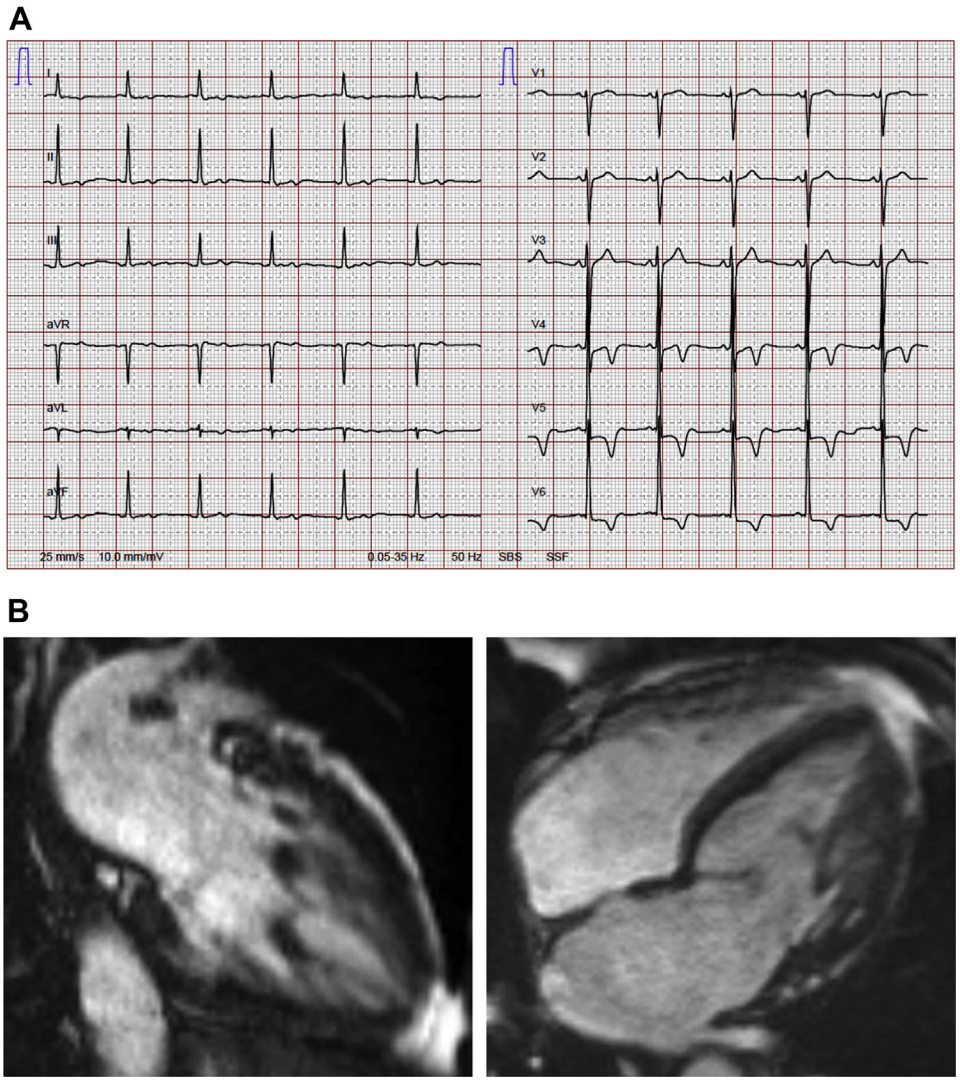
LVNC = left ventricular noncompaction

were normal. To confirm the diagnosis cardiac magnetic resonance (CMR) imaging was performed. Image quality was not optimal due to premature contractions and insufficient breath hold, but positive diagnostic criteria for LVNC (1) were reported (**Figure 1B**). LVEF was normal, and papillary muscles were displaced toward the apex.

Later, the patient was referred to the authors' institution for catheter ablation of a suspected paroxysmal supraventricular tachycardia, which was not confirmed. Because the patient at that time also complained about recurrent chest pain, a coronary angiogram was performed in the same session. A chronic total occlusion of the right coronary artery was detected. However, extensive shunting from the left coronary arteries to the LV through thebesian veins (sinusoids) was also observed (**Figure 2A**, Videos 1 and 2).

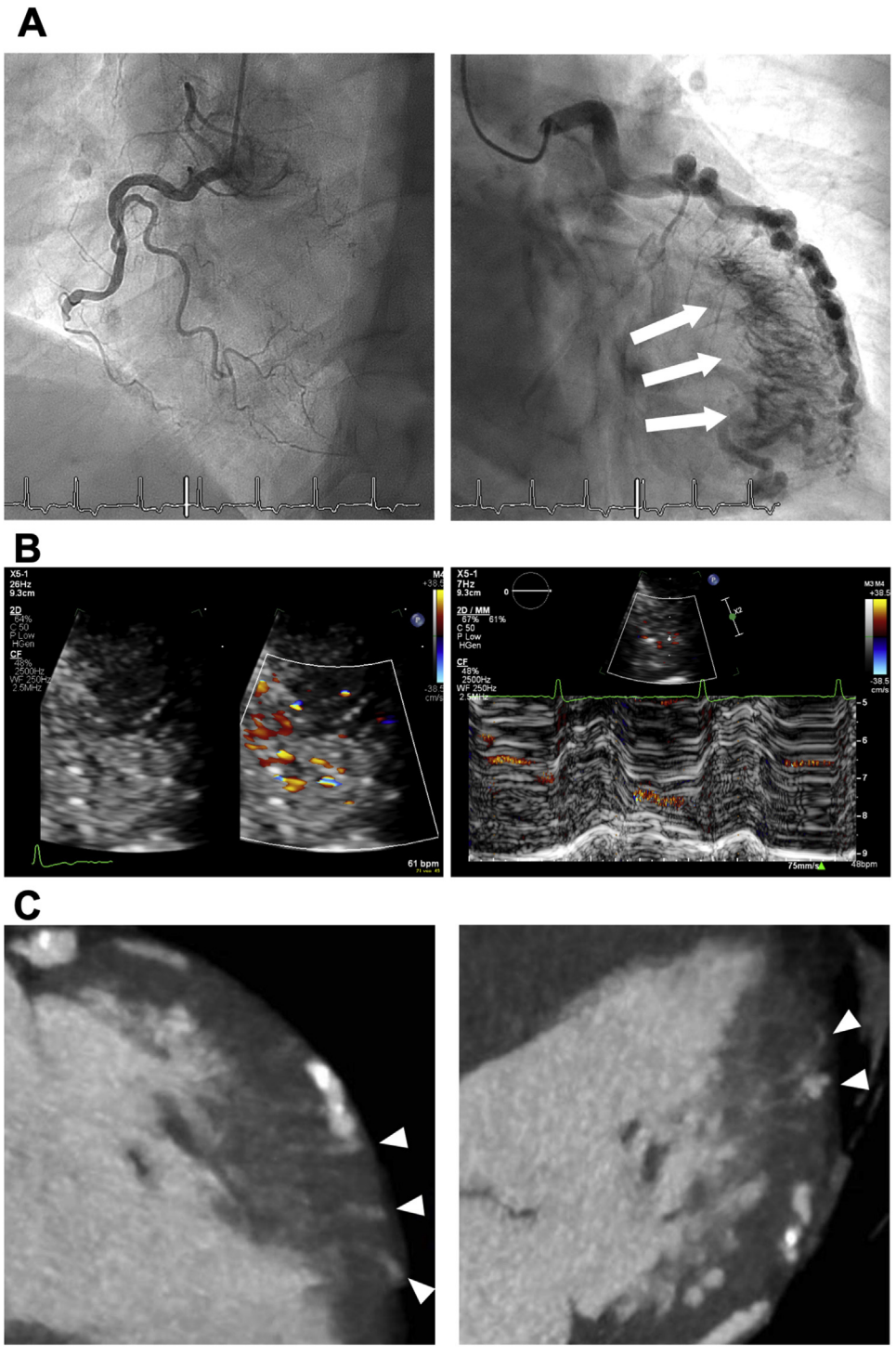
In light of this new finding, echocardiography was repeated, and color Doppler flow revealed diastolic filling of the recesses not from the LV cavity but from the coronary system, confirming that the irregular anatomy of the LV was due to the coronary anomaly

FIGURE 1 12-Lead Resting Electrocardiogram



(A) A 12-lead resting electrocardiogram with lateral T-wave inversions. **(B)** Cardiac magnetic resonance with positive diagnostic criteria for left ventricular noncompaction: 2-chamber view **(left)** and 4-chamber view **(right)**.

FIGURE 2 **Coronary Angiography of the Right Coronary Artery and the Left Coronary System**



(A) Chronic total occlusion of the right coronary artery **(left)**. Multiple shunts from the coronary system to the left ventricle in diastole, representing thebesian veins **(right)**. **(B)** Color Doppler echocardiography of the left ventricular apex (modified short-axis view). A still frame in diastole **(left)**, color M-mode in the same area **(right)**. Purely diastolic flow in the recesses in the left ventricular wall. **(C)** Cardiac computed tomography of the left ventricle. Recesses exhibit direct connections to the coronary arteries **(arrowheads, left and right)**.

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(**Figure 2B**, Videos 3, 4, and 5). To exclude concurrent LVNC, cardiac computed tomography (CT) was performed, confirming that the recesses which initially led to the diagnosis of LVNC were fed by small coronary fistulas exiting at their base (**Figure 2C**).

MANAGEMENT

At a later timepoint, adenosine stress CMR was performed and revealed no myocardial ischemia or scarring. However, as typical angina was still present under optimal medical treatment, revascularization of the chronically occluded right coronary artery was attempted but was unsuccessful.

DISCUSSION

Recesses within the LV myocardium can have different causes. Myocardial clefts or crypts derive from a congenital disarray of myocardial fibers and are seen in hypertrophic cardiomyopathy (HCM) (often multiple) or as a coincidental finding in healthy subjects (as singular lesions). Clefts or crypts are usually V-shaped, found mostly in the inferior wall or the interventricular septum and contract in systole (2). Hypertrabeculation of the LV apex corresponds to the presence of 3 or more apical trabeculations seen in 1 echocardiographic view. Unlike LVNC, the compacted layer of the myocardium is normal and not thinned (3). Thebesian veins are congenital coronary anomalies: venous endothelium-lined channels connect the coronary arteries with the LV cavity, thus leading to a shunt from the coronary system to the LV with strictly diastolic flow (4). Due to shunting of coronary blood into the LV cavity, ischemic chest pain may be present due to a "steal" phenomenon. Several case reports suggest that thebesian veins can occur in (apical forms) of HCM (5). In the present patient, solid LV wall thickness was normal. However, papillary muscles were displaced toward the apex, suggesting that some form (fruste) of apical HCM might have been present.

LVNC is a congenital anomaly which occurs when the compaction process of the inner trabecular layer of the left ventricular wall is disrupted. As the compaction process progresses from the base to the apex and from the septum to the free wall, the ventricular apex is always involved, as is, furthermore, primarily the apical and midventricular lateral wall (6). The echocardiography criteria for the diagnosis of LVNC as defined by Jenni et al. (7,8) take these features into account. Features include not only deep recesses in the expected areas and a noncompacted-to-compacted ratio of >2.0 (in systole) but also visualization of a thin, compacted layer and color Doppler assessment of the recesses to ensure that there is a to-and-fro flow from the LV and not a diastolic flow pattern as in the present case.

CMR is not limited by either acoustic windows or foreshortening and usually allows adequate differentiation between noncompacted and compacted LV layers. This makes CMR helpful in both confirmation and exclusion of LVNC. Echocardiography is broadly available and provides excellent temporal and spatial resolution. In contrast to CMR, echocardiography allows assessment of flow direction in the myocardial recesses and is thereby helpful to differentiate between sinusoids and noncompaction.

FOLLOW-UP

With time, LVEF decreased slightly to approximately 45%, and the patient still experienced chest pain on exertion under optimal medical therapy. In addition, atrial fibrillation developed, and the patient was treated with anticoagulation.

CONCLUSIONS

Different entities associated with recesses in the LV myocardium are associated with distinctive outcomes and symptoms. Therefore, it is crucial to establish the correct diagnosis to treat the patient adequately and improve risk assessment. This case exemplifies the fact that, for the diagnosis of isolated LVNC, all echocardiographic criteria as defined by Jenni et al. (6) need to be fulfilled, including color Doppler assessment of flow in the recesses. Consequently, it illustrates the fact that hypertrabecularization alone is not sufficient for the diagnosis of LVNC but both echocardiographic and CMR criteria need to be applied.

AUTHOR DISCLOSURES

Dr. Stämpfli has received consulting and speaker fees from Alnylam, AstraZeneca, and Pfizer. Dr. Cuculi has received consulting and speaker fees from SIS Medical. Dr. Kobza has received institutional grants from Abbott, Boston-Scientific, Biosense-Webster, Biotronik, Medtronic, and SIS Medical. Dr. De Boeck has reported that he has no relationships relevant to the contents of this paper to disclose.

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KEY WORDS cardiomyopathy, cleft, crypt, Doppler, LVNC, sinusoids

APPENDIX For supplemental videos, please see the online version of this paper.