Catheter ablation of a right posterior accessory pathway in a patient with left ventricular noncompaction
A case report

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Abstract

Rationale: Left ventricular noncompaction (LVNC) is a genetic cardiomyopathy characterized by the presence of a thin compacted layer of myocardium and a spongy subendocardial layer with trabeculations and recesses. LVNC associated Wolf–Parkinson–White syndrome is very rare.

Patient concerns: A 32-year-old male presented with short episodes of palpitations and a syncope 6 months before his hospitalization.

Diagnosis: His ECG revealed the presence of a right posterior accessory pathway. Echocardiography identified trabeculations of the septal, apical, and lateral wall of the left ventricle, consistent with left ventricular noncompaction. Cardiac MRI confirmed the diagnosis, as the ratio between the noncompacted and compacted myocardial layer was 2.3.

Interventions: The electrophysiological study revealed a malignant right posterior accessory pathway. Catheter ablation was successfully performed at the level of posterior tricuspid annulus. Programmed ventricular stimulation could not induce any arrhythmia at the end of the procedure.

Outcomes: During 15 months of follow-up, the patient presented no more episodes of palpitations or syncope.

Lessons: Left ventricular noncompaction with right accessory pathway is a rare association with genetic basis and gives a higher risk of sudden cardiac death. Catheter ablation of the accessory pathway is a valuable way of treatment in this category of patients, lowering the risk of sudden cardiac death.

Abbreviations: HV = his-ventricular interval, ICD = implanted cardioverter defibrillator, LV = left ventricle, LVNC = left ventricular noncompaction, MRI = magnetic resonance imaging, RV = right ventricle, RVOT = right ventricular outflow tract, WPW = Wolff Parkinson–White syndrome

Keywords: accessory pathway, catheter ablation, left ventricular noncompaction

1. Introduction

Left ventricular noncompaction (LVNC) is a genetic cardiomyopathy characterized by a modified structure of the myocardium, with a thin epicardial and a noncompacted, thickened endocardial layer.[1] It results from incomplete embryogenesis[2] and can be associated with other cardiac abnormalities and neuromuscular disorders.[3] Patients with the disease can present with heart failure, arrhythmias or thromboembolic events.[4]

We describe the case of a patient with LVNC and a right accessory pathway that successfully underwent catheter ablation by radiofrequency. The patient provided informed consent for the publication of the case.

2. Case report

A 32-year-old male with dilated cardiomyopathy was hospitalized for recurring episodes of palpitations lasting <1 minute. Six months prior to the hospitalization, the patient had a syncope. He had no family history of sudden cardiac death, but has a 30-year-old brother with dilated cardiomyopathy. The physical examination revealed a grade II apical systolic murmur, and was otherwise normal, without any sign of left or right heart failure. The twelve-lead ECG showed sinus rhythm with a heart rate of 70 beats per minute, short PR interval (80 ms) and delta wave (Fig. 1) suggesting a right posterior accessory pathway. Echocardiography revealed a dilated left ventricle (LV) with a low ejection fraction of 30%. Multiple trabeculations separated...
Figure 1. Twelve-lead electrocardiogram at baseline shows a short PR interval of 80 ms and negative delta waves in lead II, III, aVF, and V1, consistent with a right posterior accessory pathway. PR interval = interval between the P wave and the QRS complex.

Figure 2. Two-dimensional echocardiography showing a dilated left ventricle with low ejection fraction and trabeculations separated by recesses of the anterior, septal, and lateral LV walls. LV = left ventricle.
by deep recesses were present on the septal, apical and lateral wall of the LV, consistent with LVNC (Fig. 2). Cardiac magnetic resonance imaging and computed tomography confirmed LVNC. The ratio between the noncompacted and compacted layers in diastole was above 2.3 (epicardial noncompacted layer—17 mm, endocardial compacted layer—7 mm) and there was no evidence of LV thrombus (Fig. 3). Contrast-enhanced computed tomography was also performed to exclude any coronary abnormalities, as the origin of the left main coronary artery was visible on echocardiography and appeared enlarged. However, the origin and course of the coronary arteries was normal. Neither the 24 hour Holter ECG nor the exercise stress test revealed any sustained supraventricular or ventricular arrhythmia.

Considering the history of syncope and the ECG pattern consistent with the presence of an accessory pathway, an electrophysiological study was performed. A negative HV interval of -20 ms was documented, suggesting an accessory pathway which had a short refractory period (<230 ms) (Fig. 4). Mapping of the tricuspid annulus revealed the localization of the pathway at the level of the posterior ring. Catherer ablation at this level abolished conduction through the accessory pathway (Fig. 5). The postablation ECG without delta wave is shown in Figure 6. Programmed ventricular stimulation was also performed, with aggressive stimulation to 180 ms using 3 extra stimuli from the RV apex and RVOT, before and after adrenaline infusion. We could not induce any arrhythmia.

Considering the underlying myocardial disease, the severely impaired systolic function and history of syncope, we decided to proceed with an implanted cardioverter-defibrillator (ICD). The patient was long-term anticoagulated to prevent peripheral and cerebral embolism. He received beta blockers and angiotensin converting enzyme inhibitors for severely impaired LV systolic function.

3. Discussion

LVNC is thought to be determined by the arrest of the normal process of compaction during intrauterine life, resulting in a spongy aspect of the endocardial ventricular myocardium, with large trabeculations separated by intraventricular recesses. In some cases, LVNC is accompanied by the abnormal closure of both annular fibrous tissues, facilitating the formation of accessory pathways. The presence of the WPW syndrome has been reported in 15% of the pediatric population with LVNC, and, although rarer, it has also been reported in adults.

In the currently presented case, the patient has been asymptomatic until the age of 32, when he experienced a syncope, prompting further investigation. The diagnosis of LVNC was made based on the echocardiogram and confirmed by cardiac magnetic resonance imaging, which was previously shown to be superior to echocardiography in diagnosing this condition. In our patient, the ratio of the diastolic thickness of the noncompacted to compacted area was 2.4; thus respecting the cardiac magnetic resonance imaging criteria for LVNC described by Petersen et al.

The ECG pattern consistent with the presence of an accessory pathway justified an electrophysiological study which confirmed...
the presence of a right posterior accessory pathway with a refractory period of 230 ms. Since the short refractory period (<250 ms) is a known risk factor for potentially life-threatening arrhythmias,[9] we proceeded with radiofrequency ablation, which was successfully performed. However, we could not safely assume that the syncope was triggered by arrhythmias associated with the presence of the accessory pathway. In patients with LVNC, ventricular arrhythmias are one of the most frequent causes of death. A previous study on 34 patients with LVNC reported ventricular

Figure 4. The electrophysiological study yields a negative –20 ms HV interval, consistent with the presence of an accessory pathway with short refractory period (<230 ms). HV=his-ventricular interval.

Figure 5. Catheter mapping at the level of the tricuspid annulus identified as pot where the atrial electrogram was fused with the ventricular electrogram. Catheter ablation at this level abolished the conduction through the accessory pathway.
arrhythmias in 47% of the participants and sudden cardiac death in 35%.\[10]\n
Although LVNC per se is not an indication for ICD,\[11]\nthe presence of LV dilatation and severely impaired systolic function\[11]\nprompted us to refer the patient for the implantation of an ICD.

We discharged the patient on beta-blockers. The decision to anticoagulate was a rather difficult one, considering the conflicting data on the subject. The study by Fazio et al\[12]\ndid not provide any evidence that LVNC by itself would be a risk factor for thromboembolic events, but Stöllberger et al\[13]\nnoticed a higher rate of strokes in LVNC patients with severe systolic dysfunction. Considering that our patient had severely impaired LV systolic function, we recommended long-term anticoagulation with coumarins. After a follow-up of 15 months the patient presented no more palpitations or syncope. He presented no episodes of peripheral or cerebral embolism.

LVNC is a challenging diagnosis and needs a high suspicion of the diagnosis is explained by a common genetic basis. Both LVNC and accessory pathways with short refractory periods have a risk of sudden cardiac death. The risk is even higher in patients that associate both diseases.

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