CASE REPORT

Verapamil-sensitive Fascicular Ventricular Tachycardia in a Patient with Isolated Left Ventricular Noncompaction

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Abstract

Isolated left ventricular noncompaction (IVNC) is a rare congenital form of cardiomyopathy. Verapamil-sensitive fascicular ventricular tachycardia is a rare arrhythmogenic condition characterized by a right bundle-branch block pattern and left-axis deviation with a relatively narrow QRS complex. We herein present the case of a patient with IVNC who presented with verapamil-sensitive fascicular ventricular tachycardia.

Key words: isolated left ventricular noncompaction (IVNC), verapamil-sensitive, ventricular tachycardia (VT)

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Introduction

Isolated left ventricular noncompaction (IVNC) is a rare congenital cardiomyopathy resulting from the arrest of normal endomyocardial embryogenesis characterized by an excessively prominent trabecular meshwork that usually involves the left ventricle (1, 2). The clinical presentation includes systolic and diastolic dysfunction, systemic embolism and severe arrhythmias. Ventricular arrhythmia is a manifestation of IVNC. We herein present a case of IVNC that presented with verapamil-sensitive fascicular ventricular tachycardia.

Case Report

A 45-year-old man was admitted to our institution due to palpitations. He was a nonsmoker and had no significant history of alcohol use. There was no family history of cardiomyopathy. On a physical examination, the patient was mildly dyspneic without cyanosis or pallor. His pulse was 190 beats per minute (bpm), his blood pressure was 105/65 mmHg, his temperature was 37°C and his respiratory rate was 20 bpm. A cardiac examination revealed a grade 2/6 systolic murmur in the apical region. Laboratory blood tests showed normal white blood cell and platelet counts, with a hemoglobin level of 13.3 g. Normal electrolytes and negative cardiac enzymes were also observed.

A chest X-ray showed mild cardiomegaly. A 12-lead electrocardiogram (ECG) disclosed regular broad complex tachycardia and a right bundle-branch block pattern with left-axis deviation (Fig. 1). A transthoracic echocardiogram revealed enlargement of the left heart cavities. The dimension of the left ventricle was 57.6 mm in diastole and 44.9 mm in systole, with an ejection fraction of 43.4% and a shortening fraction of 22%. Marked trabeculation and intertrabecular recesses were present in the left ventricular apex (Fig. 2). Deep intertrabecular spaces communicating with main ventricular cavity were evident on color flow imaging (Fig. 3). Based on the echocardiographic findings, a diagnosis of ventricular noncompaction was established.

Due to the sustained ventricular tachycardia (VT), both intravenous amiodarone and lidocaine were administered; however, these drugs had no effect on the rhythm, while external cardioversion with 100J restored a sinus rhythm. The patient continued to experience frequent episodes of VT despite the administration of several combinations of antiarrhythmic drugs, including amiodarone, metoprolol and lidocaine. Adenosine was administered during one episode, although it did not affect the tachycardia. Despite the use of maximal doses of amiodarone and metoprolol, sustained tachycardia recurred, and intravenous verapamil, which had
not been previously administered, was given, followed by a decrease in the rate of tachycardia until resolution. Treatment with oral verapamil was continued, and no further episodes of tachycardia recurrence were observed.

The patient was discharged on oral verapamil, diuretics and an angiotensin-converting enzyme (ACE) inhibitor. Aspirin was added to the medication regimen in order to prevent embolism. The treatment with verapamil was continued, and the patient did not experience any recurrent episodes for one year.

**Discussion**

IVNC is a rare and unclassified congenital cardiomyopathy that is thought to be related to the arrest of myocardial development, resulting in the persistence of multiple areas of prominent ventricular trabeculation and deep intertrabecular recesses (3). The disease usually involves the left ventricle (2). The diagnosis of IVNC is often missed or delayed due to a lack of knowledge regarding this uncommon disease. It is thought that this type of cardiomyopathy has a higher prevalence than previously reported, possibly due to increased detection rates following improvements in cardiac imaging procedures and increasing awareness of the disease (4). Transthoracic echocardiography is the most useful noninvasive diagnostic test for diagnosing IVNC (5).

The clinical manifestations of IVNC are highly variable, ranging from no symptoms to disabling congestive heart failure, systemic thromboemboli, arrhythmias and sudden death (6, 7). VT is a manifestation of IVNC. The prevalence of complications has been reported to range from 12% to 41% (6). Subendocardial ischemia leading to localized necrosis, endocardial fibroelastosis and the subsequent formation of scar tissue in the atria and ventricles may account for heart failure and arrhythmias. Arrhythmias, particularly recurrent ventricular tachyarrhythmia, may be causes of sudden cardiac death. In such patients, cardiac transplantation or implantation of an implantable cardioverter defibrillator is an effective form of therapy for recurrent ventricular tachyarrhythmia (7).

Fascicular ventricular tachycardia belongs to a subclass of idiopathic VTs, which are thought to arise from a reentrant mechanism in the posterior fascicle of the left bundle branch (8). Classic ECG features include a right bundle-branch block pattern and left-axis deviation with a relatively narrow QRS complex (9). Belhassen et al. were the first to report the characteristic termination of this VT with intravenous verapamil, hence accounting for the terms Belhassen VT and verapamil-responsive VT to describe the condition (10). The use of vagal maneuvers and adenosine therapy is ineffective in converting this arrhythmia. The disease is also unresponsive to standard treatments (e.g. lidocaine) used for VTs caused by ischemic heart disease (9). The drug of choice for pharmacologic conversion of fascicular VTs is...
intravenous verapamil (11). Recurrent arrhythmia may be prevented with long-term oral verapamil therapy. The Purkinje fibers involved and damaged by the disease process of IVNC or trabeculation prominent in the area of the posterior fascicle of the left bundle branch may account for the mechanism underlying the development of fascicular VT in patients with IVNC. Radiofrequency ablation is effective and curative in most patients with this type of VT (12). Recurrent ventricular tachyarrhythmia is the most important cause of sudden cardiac death. Implantation of an implantable cardioverter defibrillator was refused by our patient.

In conclusion, IVNC is rarely associated with verapamil-sensitive fascicular ventricular tachycardia. The drug of choice for pharmacologic conversion is intravenous verapamil. The administration of long-term oral verapamil therapy is thought to be effective in preventing recurrent arrhythmia.

The authors state that they have no Conflict of Interest (COI).

References