Short Communication

Isolated Noncompaction Left Ventricular Myocardium and Polymorphic Ventricular Tachycardia

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Summary: A 57-year-old woman with syncope was admitted. She had a family history of sudden death: two brothers had died suddenly at the age of 47. Transesophageal echocardiography showed numerous prominent trabeculations and deep intertrabecular recesses in the anterior and lateroapical zones. Isotopic left ventricular ejection fraction was 46%. Cardiac catheterization showed coronary arteries with no angiographic lesions. A prominent trabecular zone and deep intertrabecular recesses were seen in the anterior wall on left ventriculography. Right ventriculography was normal. The diagnosis of isolated noncompaction left ventricular myocardium was established. Continuous 24-h electrocardiographic registry showed episodes of polymorphic ventricular tachycardia. Programmed ventricular stimulation performed at the right ventricular apex with up to three extrastimuli failed to induce ventricular arrhythmias. Treatment with beta blockers was initiated, but short runs of polymorphic ventricular tachycardia persisted. A dual-chamber automatic implantable defibrillator was implanted. We discuss the pathophysiology of the arrhythmia. It appears that several factors could be responsible for the malignant arrhythmias in this entity.

Key words: noncompaction ventricular myocardium, polymorphic ventricular tachycardia

Introduction

Isolated noncompaction ventricular myocardium is a rare disorder of endomyocardial morphogenesis characterized by numerous, prominent trabeculations and conspicuous intertrabecular recesses that penetrate deep into the ventricular myocardium.¹ The disease typically involves the left ventricular (LV) myocardium, but the right ventricular (RV) myocardium may also be affected.^{2, 3} Histologic examination showed the intertrabecular recesses to be lined with endothelium.⁴ Clinical manifestations of the disorder included depressed LV systolic function, ventricular arrhythmias, and systemic embolization.^{3–5} Both sporadic and familial^{4, 6, 7} forms have been described.

The case of a patient with a family history of sudden death, in whom syncope secondary to polymorphic ventricular tachycardia was the first clinical manifestation, is reported.

Case Report

A 57-year-old woman with syncope was admitted. She had a family history of sudden death: two brothers died suddenly at the age of 47. Two months prior to admission she started with palpitations and dizziness of seconds' duration. On admission blood pressure was 130/85 mmHg; baseline electrocardiogram (ECG) revealed sinus rhythm with advanced left bundle-branch block and normal PR interval. Chest x-ray was normal. She was continuously monitored with telemetry recordings. Short repetitive episodes of polymorphic ventricular tachycardia were recorded some hours later. Continuous 24-h ECG registry also showed episodes of polymorphic ventricular tachycardia (Fig. 1). With these episodes, the patient suffered palpitations and dizziness similar to the symptoms she had had prior to the syncopal episode before admission. Two-dimensional echocardiography disclosed a nonhypertrophied, non-dilated left ventricle with global hypokinesia and ejection fraction 40–45%; echocardiographic abnormalities of the RV myocardium were not detected. Transesophageal echocardi-

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FIG. 1 Telemetry and 24-h continuous electrocardiographic recordings showing runs of polymorphic ventricular tachycardia.

graphy showed numerous prominent trabeculations and deep intertrabecular recesses in the anterior and lateroapical zones (Fig. 2). On Doppler color study, flow within the intertrabecular spaces was detected. Isotopic LV ejection fraction was 46%. Cardiac catheterization showed coronary arteries with no angiographic lesions. A prominent trabecular zone and deep intertrabecular recesses were seen in the anterior wall on left ventriculography (Fig. 3). Right ventriculography was normal. The diagnosis of isolated noncompaction LV myocardium was established. Intracardiac electrophysiologic

study showed normal AH interval (84 ms) and prolonged HV interval (62 ms). Programmed ventricular stimulation performed at the RV apex with up to three extrastimuli failed to induce ventricular arrhythmias. Treatment with beta blockers was initiated, but short runs of polymorphic ventricular tachycardia persisted. Given the family history of sudden death, syncope, and altered atrioventricular conduction parameters, a dual-chamber automatic implantable defibrillator (Ventak A-V II, Guidant Corporation, Indianapolis, Ind., USA) was implanted and anticoagulation with dicumarin was initiated. At

FIG. 2 Transesophageal echocardiography showing numerous prominent trabeculations and deep intertrabecular recesses in the anterior and lateroapical zones.

FIG. 3 Left ventriculography showing a prominent trabecular zone and deep intertrabecular recesses in the anterior wall.

1-year follow-up, the patient is doing well. The ECGs of one sister, three daughters, and one son were normal and long QT syndrome was ruled out.

Discussion

Ventricular arrhythmias ranging from premature ventricular beats to ventricular tachycardia and fibrillation have been documented in 18–63% of patients with isolated noncompaction myocardium.^{3,4,7} Ritter *et al.*³ also reported 18% sudden death in the follow-up of his series. It is unclear why this entity is arrhythmogenic. Chin *et al.*⁴ suggested that the intertrabecular recesses are reminiscent of the morphology of the right ventricle in arrhythmogenic RV dysplasia, but although both exhibit high arrhythmogenic potential, the morphologic characteristics of these conditions differ completely. Junga *et al.*⁸ demonstrated with positron emission tomography that myocardial perfusion and coronary flow reserve in areas of isolated ventricular noncompaction are impaired. The underlying mechanism might be failure of the coronary microcirculation to grow with the increase in ventricular mass. The same authors also described the presence of late potentials or prolonged QT dispersion in some cases which may explain the increased risk for subsequent occurrence of sudden cardiac death or sustained monomorphic ventricular tachycardia. The impairment of LV performance itself is one of the major causes of the increased incidence of arrhythmias. Another cause may be the morphologic features of isolated noncompaction myocardium with increased scarring and fibrosis in the zones of excessive trabecular hypertrophy.³ The value of the QT interval was difficult to ascertain in our patient since she had ad-

vanced left bundle-branch block; however, as the QT interval in family members still alive was normal, the diagnosis of long QT syndrome was highly unlikely. It would appear that several factors are responsible for the malignant arrhythmias in this entity.

This is the first case to show the association of isolated noncompaction LV myocardium with polymorphic ventricular tachycardia. Given the family history of sudden death and syncope, a cardioverter defibrillator with double-chamber cardiac pacing was implanted.

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