

Case Report

Congenital Ventricular Diverticulum Associated with Ventricular Tachycardia

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Congenital ventricular diverticula are rare. Clinically, they may be asymptomatic or cause systemic embolization, heart failure, valvular regurgitation, ventricular rupture, ventricular arrhythmia, or sudden death. We report the case of a 56-year-old woman with sustained ventricular tachycardia, who, during investigation, was diagnosed with a diverticulum in the inferobasal portion of the left ventricle. The clinical characteristics and treatment of this rare disease are discussed.

Congenital ventricular diverticulum is an extremely rare cardiac abnormality¹. The initial reports resulted from autopsies, and many of those deaths occurred due to rupture of the diverticulum. Such a case was first described in 1838².

Clinically, congenital ventricular diverticulum may be asymptomatic or cause systemic embolization, heart failure, valvular regurgitation, ventricular rupture, ventricular arrhythmia, or sudden death^{1,2}. Ventricular tachycardia is a rare and severe complication, ours being the eighth case reported in the literature.

Case Report

The patient is 56-year-old woman who sought the emergency department due to clinical findings of sustained monomorphic ventricular tachycardia (fig. 1) with hemodynamic repercussions. Synchronous electrical cardioversion was performed, and, later, amiodarone was administered intravenously.

The patient had a history of previous treatment for systemic arterial hypertension with propranolol, enalapril, and hydrochlorothiazide, and reported being hospitalized at another institution 3 years earlier, due to a similar arrhythmic episode. On that occasion, an electrophysiologic study was carried out with ventricular extrastimulation at 400-millisecond cycles and reproduction of the arrhythmia after an extrastimulus. She was then prescribed amiodarone, but used it irregularly.

During the investigation, the patient's physical examination showed some alterations. After controlling the acute setting, the electrocardiogram, chest X-ray, and serum electrolytes were within

the normal range. The transthoracic echocardiography indicated the presence of left ventricular posterobasal bulging, which, on left ventriculography (fig. 2 and 3), was characterized as a protrusion in the posterobasal region in close connection with the ventricular cavity and preserved contractility, which eliminated all its contrast medium content after 3 beats. The imaging findings were compatible with the diagnosis of diverticulum. Myocardial magnetic resonance imaging (fig. 4) confirmed the findings of the left ventriculography. The reaction of hemagglutination for Chagas' disease was negative, and, on cine coronary angiography, the coronary arteries were normal. Ventricular mapping for the arrhythmogenic focus was not performed.

The patient underwent surgical resection of the left ventricular diverticulum with placement of a bovine pericardial flap. The anatomicopathological examination of the surgical specimen revealed the presence of epicardium, myocardium with marked dystrophic alterations (interstitial fibrosis, hypertrophy, and vacuolization of fibers), and endocardial fibrosis (fig. 5).

The patient had an uneventful postoperative period and was discharged from the hospital with antihypertensive medications and amiodarone. One year after the procedure, she is asymptomatic.

Discussion

The difference between aneurysm and congenital diverticulum of the heart is not clear. It is not only a question of semantics, but of definition and classification³.

Abbot et al⁴ reported congenital cardiac aneurysm as cardiac diverticulum, and those 2 terms have often been used interchangeably. Treisman et al³ used the term "diverticulum" when the defect in the ventricular wall was associated with other congenital alterations, both cardiac and noncardiac. In 1958, Cantrel described it with details as part of a pentalogy (associated with changes in the abdominal wall, sternum, diaphragm, pericardium, and heart)^{3,5,6} and reserved the term "aneurysm" when no association occurred¹⁻³. However, 30% of the cases of left ventricular diverticulum are not associated with congenital malformations, being called isolated ventricular diverticulum⁶, and the lack of association of congenital defects does not exclude its congenital origin³. The largest series of congenital ventricular diverticula has been reported by Okereke et al², with 10 cases between 1965 and 1984 at the Texas Heart Institute. On ventriculography, those cases had characteristics that differentiated them from aneurysms: close connection with the true ventricular cavity, contractility, rapid contrast medium filling, and complete emptying

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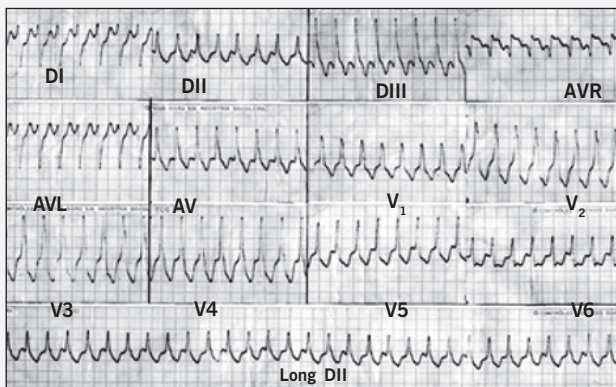


Fig. 1 - Electrocardiogram showing monomorphic ventricular tachycardia.

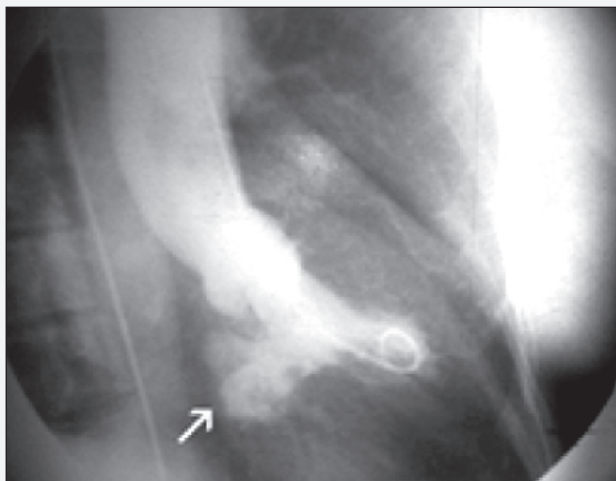


Fig. 2 - Left ventriculography during systole showing posterobasal diverticulum (arrow).

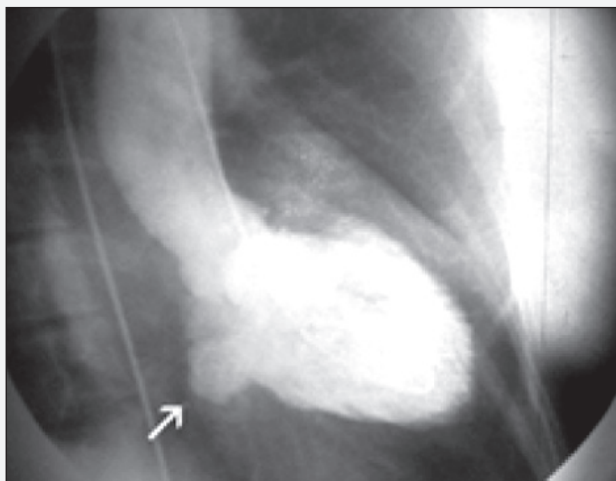


Fig. 3 - Left ventriculography during diastole showing posterobasal diverticulum (arrow).

after 2 to 4 contractions¹. Ventriculography of the case reported has a similar description.

Most ventricular diverticula are found in the apical region of the left ventricle, but they may occur in any topography, in the right ventricle or in both ventricles^{1,2}. From the pathological viewpoint, they may be muscular or fibrous. The muscular type originates in the ventricular apex, is surrounded by myocardium, endocardium, and, occasionally, pericardium, and the point of connection with the ventricle is usually narrow. This type is often

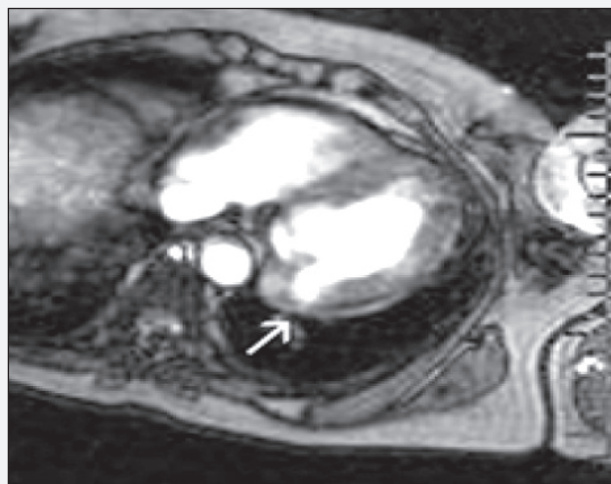


Fig. 4 - Myocardial magnetic resonance imaging showing posterobasal left ventricular diverticulum (arrow).

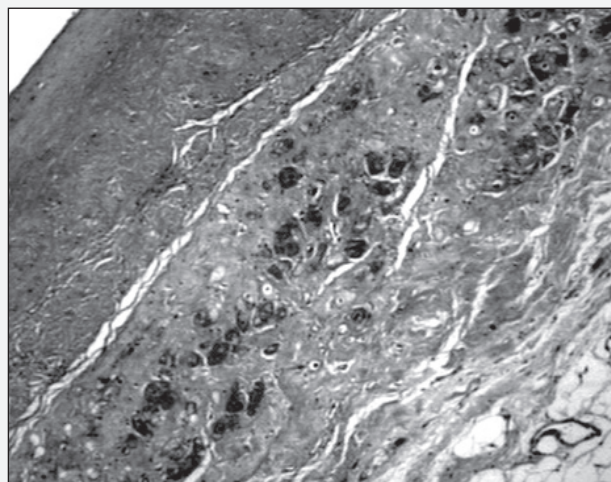


Fig. 5 - Histological section, magnification of 100x, showing the presence of epicardium, myocardium with dystrophic changes, and endocardial fibrosis.

accompanied by abnormalities in the median line (defects in the pericardium, diaphragm, or abdominal wall) and by congenital cyanotic heart disease, being diagnosed at birth. The fibrous type originates in the apical or subvalvular position, and occurs predominantly in black people and Africans. The subvalvular form, the most common, may be accompanied by aortic or mitral regurgitation and systemic embolism. The fibrous diverticula are associated with neither defects in the median line nor congenital cardiac malformations^{1-3,7}.

The histopathological differentiation between congenital or acquired diverticula and aneurysms is not possible, because specific alterations do not exist^{3,9}. According to Treistman et al³, the histological findings may be similar to those of the normal ventricular wall or connective tissue may predominate. Those authors reported 2 cases, one had the 3 layers of the ventricular wall, the myocardium being atrophic and with fibrous tissue, and, in the other, the connective tissue predominated³.

The etiology of the diverticula is uncertain. They may represent congenital epicardial cysts or result from a weakness in the ventricular muscle, with gradual bulging due to the high intraventricular pressure^{1-3,7}.

The diagnosis may be performed hours or even years after birth⁸,



in asymptomatic patients when assessing an abnormal electrocardiogram or chest X-ray during the investigation of congenital heart diseases or abnormalities in the median line ¹. In symptomatic patients, it presents as a cause of embolic events, heart failure, valvular regurgitation, or sudden death ^{1,2,9}. Sudden death may occur due to rupture of the diverticulum, which is more common in the fibrous type, or due to ventricular tachyarrhythmias ¹.

Few cases of congenital diverticula and aneurysms with ventricular tachycardia have been reported ^{1,8,10}. Chesler et al ⁸ reported the cases of 2 Bantu women with ventricular tachycardia and congenital subannular aneurysm. Maloy et al ¹¹ reported the case of a 26-year-old woman with apical aneurysm and sustained ventricular tachycardia, who underwent aneurysmectomy and remained asymptomatic for 6 months. Fellows et al ¹⁰ reported the cases of 3 patients with congenital ventricular aneurysm, 2 patients who experienced sudden death but were revived, and the third with recurring nonsustained ventricular tachycardia. Shen et al ¹ reported a patient with subvalvular fibrous diverticulum and sustained ventricular tachycardia refractory to medicamentous treatment, who underwent mapping and surgical ablation of the arrhythmogenic focus but recurred one and a half years after the procedure.

The natural history of patients with congenital diverticulum is uncertain ^{1,7}. Because of the likelihood of complications, such as the risk of spontaneous rupture, some authors have indicated surgical resection even in asymptomatic patients ². The surgical

technique depends on the type and extension of the diverticulum. The diverticula with well-defined connection and true ventricular cavity and no other associated defects may be resected without extracorporeal circulation ². Okereke et al ² reported the surgical treatment in 10 cases of ventricular diverticula. Three were resected without extracorporeal circulation, and one required ventricular repair with dacron. In the remaining cases, 2 required mitral valve repair, one required mitral valve replacement, and one required aortic valve replacement.

In patients with ventricular tachycardia, the electrophysiological study and mapping are part of the diagnosis and treatment ¹, because, so far, no uniform management has been established for those cases in the literature.

Our patient had 2 episodes of ventricular tachycardia with hemodynamic repercussion and previous electrophysiological study with reproduction of the arrhythmia. During the investigation, a left ventricular subvalvular mitral diverticulum was identified. No ventricular mapping of the arrhythmogenic focus was performed, due to local difficulties for the procedure. This fact did not change the therapeutic decision due to the risk of other complications (embolic events, valvular regurgitation, and rupture). Later, the patient underwent resection of the diverticulum and had a good postoperative evolution. Surgical resection was considered the most adequate treatment, due to the probability of the complications described.

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