Case report – Congenital Ventricular tachycardia with congenital left ventricular aneurysm in an adult

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Abstract

A 60-year-old woman without any risk factors for coronary artery disease presented with symptomatic, recurrent, non-sustained ventricular tachycardia. She was found to have a postero-lateral left ventricular aneurysm. Coronary angiography revealed normal coronary arteries. The 12-lead electrocardiogram showed sinus rhythm with frequent premature ventricular contraction. Her non-sustained ventricular tachycardia was reproduced by programmed electrical stimulation and was unresponsive to procainamide, mexiletine, and disopyramide. Aneurysmal resection and cryoablative surgery was performed. After surgery, ventricular programmed stimulation was negative, and premature ventricular contraction was disappeared. The patient discharged on no antiarrhythmic therapy.

Keywords: Congenital left ventricular aneurysm; Ventricular tachycardia; Ablating surgery

1. Introduction

True congenital aneurysm of the left ventricle, which discovered in adult, is very rare. They may be asymptomatic or may present with systemic embolization, congestive heart failure, valvular regurgitation, and ventricular rupture [1,2]. Ventricular tachyarrhythmias are an unusual but significant complication, with only a few cases reported previously [3–5]. We present the case successfully treated with cryoablating surgery for recurrent non-sustained ventricular tachycardia (NSVT).

2. Case report

The patient was a previously healthy 60-year-old woman. In February 2001, she had a first time episode of lightheadedness, flushing and syncope. It resolved spontaneously. On physical examination, no abnormal findings were obtained. Laboratory examination results were within normal limits. The brain computed tomography was normal. The 12-lead electrocardiogram (ECG) revealed sinus rhythm with frequent premature ventricular contraction (PVC). Subsequently, her cardiac examination was started. Two-dimensional echocardiography revealed normal right and left ventricular size and function. The long axial images revealed an aneurysm extending from posterior left ventricular (LV) wall. This pouch showed no contraction, with relative thin wall. Thrombus was not recognized in the left atrium and ventricle including in aneurysm. Coronary angiography revealed normal coronary anatomy. A left ventriculogram showed a wide-mouthed, multilobulated 3–4 cm aneurysm of the posterior wall, which had asynchronous contraction with LV (Fig. 1). Over all LV contractility was preserved. The hemodynamic data obtained by Swan-Ganz catheter were as follows: right atrial pressure 3 mmHg, pulmonary arterial pressure 25/10 mmHg, pulmonary capillary wedge pressure 9 mmHg, and cardiac index 3.79 l/min per m². Electrophysiological testing was performed while the patient was off antiarrhythmic agents. SVT, identical in morphology to the clinical tachycardia, was induced with intrastimuli from the right ventricular apex (Fig. 2). The patient was started on therapy with mexiletine.

Three month later, the patient again had lightheadedness with syncope. She was found to have a wide complex tachycardia with left bundle-branch block configuration, and her rate 200 beats per minute. In addition, a 24 h Holter ECG elicited several two-beats to three beats runs of repetitive ventricular complexes.

Finally, ablating surgery was chosen. During surgery, it was identified that NSVT had been induced with extrastri-
muli from the right ventricular apex. The aneurysm was found to consist entirely of grayish-white fibrous tissue, arising postero-lateral wall of left ventricle bordered by the posterior descending and distal circumflex marginal vessels. Its motion was asynchronous with the rest of the left ventricle. Extensive aneurysmal resection was performed. Thrombus was not presented. Through the open ventricle, the fibrous endocardial scar determines the border zone between the totally fibrous tissue and the beginning of muscular tissue. Then a cryoprobe was used to freeze the edge of resection concentrating on the focus of ventricular tachyarrhythmias. Endoaneurysmorrhaphy was performed using Fontan suture and Dacron patch graft to reconstructing the left ventricle. A double layer of continuously running suture placed over the aneurysmorrhaphy area with reinforcing felt strips. The pathological examination of aneurysmal wall revealed focal defect of muscle fibers. Various degree of vacuolization of muscle fibers is present, and cross striations may be lost in some areas. After surgery, ventricular-programmed stimulation was negative, and PVC was disappeared on 24 h Holter ECG. The patient was discharged on no antiarrhythmic therapy.

3. Discussion

Aneurysm of the left ventricle is usually due to myocardial infarction. It has been recognized that this complication can occur in 24–35% of patients with coronary artery disease. Rarely ventricular aneurysms can occur as a result of congenital defects, trauma, myocardial abscess or ulcerating bacterial endocarditis [3]. However, it is not well recognized that ventricular aneurysms can also occur in patients with primary myocardial disease. Treisman et al. classified the defect as an aneurysm when its root of connection to the left ventricle was wide, and diverticulum if the connection was narrow [6]. The wall of true congenital diverticulum is formed by all three cardiac layers and contracts normally, whereas an aneurysm is generally a fibrous saccular lesion with paradoxical contraction [7]. The fibrous aneurysm arises either in apical or subvalvular positions. The etiology of these aneurysm is unclear. They have been postulated to represent congenital epicardial cysts, to derive from abnormal attachment of the heart tube to the yolk sac, or to arise from weakness in the ventricular muscle with gradual outpouching from high ventricular pressures [4].

Ventricular tachyarrhythmias associated with the congenital LV aneurysm are an unusual but significant complication, with only a few cases reported previously [3–5]. Maloy et al. reported the findings in 26-year-old woman with an apical aneurysm and refractory VT [1]. She had a blind aneurysmectomy and was asymptomatic on 5-month follow-up. Fellows et al. described three patients, two with aborted sudden deaths and one with NSVT and syncope [2]. The patient with syncope had inducible VT and unsuccessful epicardial cryoablation of the VT focus.
This patient was stable on therapy with imipramine. Shen et al. reported successfully treated with mapping-guided surgery [4]. Even though the patient had a recurrence 11/2 years later, the focus was probably sufficiently modified to allow a prolonged arrhythmia-free duration and eventual suppression by previously ineffective agents. Our patient had a posterolateral left ventricular aneurysm, which was symptomatic with recurrent NSVT and syncope refractory to antiarrhythmic therapy. We performed aneurysmal resection and cryoablative surgery without mapping. To avoid recurrence of ventricular arrhythmia, we ablated aneurysmal base, which was fibrous change, sufficiently. Although further long-term follow-up is required, the patient had been discharged on no antiarrhythmic therapy and has free of symptoms with follow-up over 6 months.

The natural history of LV aneurysm is unknown. Therefore, patients with LV aneurysm should be treated medically unless some other associated cardiac anomalies need surgical correction or medical treatment cannot control the symptoms [6]. However, some cases of sudden death probably result from ventricular tachyarrhythmias [4]. Moreover, sudden death may occur by actual rupture of the aneurysm because of its thickness wall [1–6]. In view of their high rate of complications and the risk of spontaneous rupture, some authors advocate surgical resection, even if asymptomatic [8]. In patient with associated VT, electrophysiological testing and ablative therapy should be an integral part of the diagnostic and therapeutic regimen.

References