Congenital left ventricular diverticula: a rare cause of sudden cardiac death

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Summary: Congenital left ventricular diverticula are a rare cause of sudden cardiac death. We describe the first reported case of ventricular fibrillation in association with congenital diverticula of the heart. The diagnosis of left ventricular diverticula was made by cardiac catheterization and confirmed by magnetic resonance imaging. Treatment was initiated with anti-arrhythmic and anticoagulant drugs to prevent life-threatening arrhythmias and emboli.

Introduction

Coronary artery disease is the underlying cause of sudden cardiac death in at least 80% of cases in Western society.¹ Other causes include myocardial disease (such as hypertrophic cardiomyopathy and myocarditis), valvular disease (such as aortic stenosis) and electrophysiological or conduction system abnormalities.¹ Cardiac diverticula are a rare but important additional cause of sudden death.

Case report

A 43 year old nurse collapsed at home with a cardiac arrest. Her husband started cardio-pulmonary resuscitation and an ambulance crew defibrillated her from ventricular fibrillation into sinus rhythm. This was documented on an electrocardiogram recording. She was admitted to the local district general hospital where assisted ventilation was instituted because of cerebral oedema and resultant poor respiratory effort. After 6 days she had made a complete recovery and was neurologically normal. She was a non-smoker with no medical history of note. Physical examination and routine blood tests, including cardiac enzymes, were normal as were resting electrocardiogram and exercise stress testing. A 24 h tape showed occasional ventricular couplets, frequent triplets and bigeminy. Cross-sectional echocardiography suggested septal hypokinesia and basal paradoxical motion. The patient was discharged 17 days after

admission on atenolol and aspirin with a diagnosis of presumed myocardial infarction complicated by ventricular fibrillation.

One month later the patient was readmitted with central chest pain. A resting electrocardiogram showed sinus bradycardia but no changes suggestive of ischaemia. Atenolol was stopped and she was transferred to this hospital for cardiac catheterization and coronary angiography. The chest radiograph revealed an abnormal cardiac apical contour (Figure 1). Coronary angiography was normal but left ventricular cineangiography showed a diverticulum of the inferior wall and one of the anterior wall of the left ventricle (Figure 2). The right ventricular cineangiogram was normal. These findings were confirmed on magnetic resonance imaging (Figure 3). The following additional investigations were normal: viral titres, syphilis serology and auto-antibodies. Treatment was started with amiodarone as prophylaxis for life-threatening ventricular arrhythmias and with warfarin because of the risk of emboli. A conventional ventricular arrhythmia provocation protocol carried out after 3 weeks of treatment with amiodarone, failed to elicit any significant arrhythmia except for one salvo of non-sustained ventricular tachycardia. The patient remains well and free of symptoms 4 months after discharge.

Discussion

Congenital diverticula of the heart are rare, the reported incidence being 0.4% in a post-mortem study of cardiac deaths² and 0.26% in a group of patients undergoing routine cardiac catheterization.³ Isolated left ventricular diverticula can be single or multiple and have been associated with

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Figure 1 A posteroanterior view chest radiograph illustrating the abnormal cardiac apical contour.

Figure 3 Spin echo magnetic resonance image gated to the R wave in coronal view showing diverticula of the anterior and inferior walls of the left ventricle (arrows).

Figure 2 Left ventricular cineangiogram in the right anterior oblique projection in mid-systole showing diverticula of the anterior and inferior walls of the left ventricle.

cardiac rupture, emboli, cardiac failure, endocarditis and arrhythmias, though ventricular fibrillation is previously unreported.^{3,4} Ventricular tachycardia in association with ventricular diverticula has been reported in one other case but that patient died of congestive cardiac failure immediately after attempted surgical correction.⁵ Congenital diverticula have also been reported in association with other congenital abnormalities of the heart and of the abdominal wall, lower sternum, ventral dia-

phragm and pericardium. However, congenital diverticula are often asymptomatic and may be an incidental finding.^{3,6} In the case described, the cross-sectional echocardiogram was abnormal but the diagnosis was made on ventricular cineangiography. The patient underwent magnetic resonance imaging to confirm the angiographic results. Cineangiography is currently the diagnostic method of choice in view of its easy availability and its superiority over echocardiography, which can produce misleading results in these cases.⁴

This case illustrates the need to exclude the important finding of ventricular morphological abnormality in patients presenting with a life-threatening arrhythmia. Patients with congenital diverticula should be treated medically unless some other associated cardiac abnormality needs surgical correction, since the most recent review of the literature, in 1982, showed a high risk of post-operative cardiac failure and death following diverticulectomy.⁷ The only reported case of diverticulectomy for a congenital diverticulum since that review showed the subject alive 4 years after surgery⁸ and one may speculate that a review of the recent surgery would show better results.

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