Arrhythmogenic right ventricular dysplasia presenting as acute coronary syndrome: a case report

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Abstract Arrhythmogenic right ventricular dysplasia (ARVD) is underdiagnosed cardiomyopathy which commonly presents in young adults with ventricular tachycardia or sudden death. We report a case of ARVD presenting with features of acute coronary syndrome. The suspicion of ARVD came only when echocardiogram revealed abnormal shape and wall motion of right ventricle, which was later confirmed by right ventricular angiogram. The diagnosis of ARVD was discussed and the literature reviewed.

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KEYWORDS
Right ventricular cardiomyopathy; Dilated right ventricle; Ventricular tachycardia; Sudden death; Right ventriculography; Right ventricular dysplasia.

Arrhythmogenic right ventricular dysplasia (ARVD) is still poorly understood and is often underdiagnosed cardiomyopathy. It commonly presents as ventricular tachycardia or sudden death, but quite often palpitation may be the only presenting complaint. We report a case of ARVD presenting with acute chest pain and ECG showing ventricular tachycardia of left bundle branch block (LBBB) configuration. The initial management of the case was along the line of non-ST-segment elevation acute coronary syndrome.

Case report

A 49 years old male was admitted to hospital with retrosternal chest pain, palpitation and dizziness of one hour duration. There was history of similar episode about 10 years back and was treated with aspirin, diltiazem and beta-blocker for one year. There was no family history of heart disease or sudden death. On admission he was haemodynamically stable and was not in heart failure. ECG showed sustained ventricular tachycardia of LBBB pattern (Fig. 1a). Ventricular tachycardia failed to respond to intravenous lignocaine and amiodarone and suddenly became unstable and had to be cardioverted with synchronized DC shock. His post
conversion 12-lead ECG showed inverted T-waves in V1–V3 with Q-waves and inverted T-waves in inferior leads (Fig. 1b). CK, CKMB were modestly elevated and Troponin I was weakly positive. Other laboratory tests and chest X-ray were normal. Echo showed: dilated right ventricle with outpouching in the right ventricular cavity suggestive of right ventricular dysplasia (Fig. 2a, b). Left ventricle was normal. Based on chest pain of ischemic character, ECG findings of Q-waves with T-wave inversion in inferior leads and raised cardiac markers, patient was treated as a case of acute coronary syndrome with intravenous heparin, aspirin, atorvastatin and amiodarone. He was stabilized and was discharged on aspirin and amiodarone and with advice to do coronary angiography. Coronary and left ventricular angiogram did later show normal coronaries and normal left ventricle. Right ventriculogram showed dilated right ventricle with excessive wall thickening and dysplastic changes suggestive of right ventricular dysplasia. Patient was advised to continue amiodarone and has been asymptomatic ever since.

Discussion
ARVD commonly presents with arrhythmia or sudden death. Our patient presented misleadingly with acute chest pain, ECG changes suggestive of ischemia/infarction and raised levels of cardiac markers. The suspicion of ARVD came only when an echocardiogram revealed abnormal shape and wall motion of the right ventricle, which was later confirmed by right ventricular angiogram. It seems difficult to explain the occurrence of chest pain as the presenting symptom. It is possible that the episode was precipitated by coronary spasm leading to chest pain and eventually triggering ventricular tachycardia in a susceptible substrate. Bacior et al. reported a patient of ARVD who developed ST elevation followed by ventricular tachycardia during exercise stress test and who was found to have normal coronaries.1 Raised levels of cardiac markers are even more difficult to explain and led to initial mis-diagnosis of acute coronary syndrome. One explanation is that, as the first set of cardiac markers were normal but became high

Figure 1 (a) 12-Lead ECG showing ventricular tachycardia with LBBB configuration. (b) Post cardiovasive 12-Lead ECG.
Figure 1 (continued)
after cardioversion it is possible that markers were released from diseased apoptotic myocardium following DC shock.

Recently, a standardised diagnostic criterion for diagnosis of ARVD was proposed by McKenna et al.\(^2\) ARVD should be strongly suspected in a patient with ventricular tachycardia of LBBB morphology or in a young adult dying suddenly. Echocardiographic and angiographic studies showing dilated right ventricle with outpouching in the free wall are useful in making the diagnosis. Magnetic resonance imaging may be required for further confirmation showing typical fatty infiltration of right ventricular myocardium. At histological level disease is characterized by progressive fibro-fatty replacement of right ventricular myocardium, initially regional but later global with relative sparing of the septum. ARVD should be differentiated from idiopathic right ventricular outflow tract tachycardia, which has a better prognosis and occurs in the absence of any significant structural heart disease. ARVD is a common cause of sudden death in young adult. It can affect any age group but the typical patient is a male patient in the third decade of life.\(^3\) The inheritance of ARVD is primarily by autosomal dominant mode with variable expression and penetrance.\(^4\) However, an autosomal recessive pattern has been reported in Naxos disease and in Carvajal syndrome.\(^5,6\) Ten genes for ARVD have been identified on chromosomes 1, 2, 3, 6, 10, 14 and 17. Major candidate genes identified are involved in encoding for desmoplakin (ARVD8) and plakoglobin (Naxos disease), a protein for cell to cell adhesion and ryanodine receptor, RYR2 (ARVD2), involved in ion channels.\(^6\) Localization of genes has important implications in preclinical diagnosis of disease and diagnosis of carriers by gene mapping.\(^9\)

The treatment aims at the management of ventricular tachycardia and prevention of sudden death. Beta-blockers, sotalol and amiodarone remain the mainstay of treatment. For those at high risk of sudden death implantable cardioverter defibrillator is the treatment of choice.\(^10\)

Figure 2  (a) Parasternal long axis (PLAX) showing dilated right ventricle (RV). (b) Apical 4-chamber view (AP4) showing dilated RV with classical pouching of RV cavity.
References


