Arrhythmogenic Right Ventricular Dysplasia With Epsilon Potential—Case Report With Review*

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Summary:

Arrhythmogenic right ventricular dysplasia is a form of cardiomyopathy which causes life threatening ventricular tachyarrhythmias in adults. We present a case who ultimately expired along with review of literature of this entity.

Arrhythmogenic right ventricular dysplasia (ARVD) is a type of right sided cardiomyopathy, possibly familial in some patients[2,3], with hypokinetic areas in the right ventricular musculature. It is considered an oddity and can be an important cause of life threatening ventricular tachyarrhythmias in children and young adults with an apparently normal heart[4, 5]. The surface ECG may have a ventricular post excitation wave[6]. We report a young patient of ARVD who presented with recurrent ventricular tachyarrhythmias and had epsilon potential on surface ECG.

Case Report:

A 36-year old man presented to us with history of palpitations off and on and dyspnoea on exertion NYHA Cl II for 3 months. He also had frequent dizziness and a syncopal attack once about 15 days prior to admission. The general examination was unremarkable. The first and second heart sounds were normal. A loud right ventricular fourth heart sound (RVS_4) was auscultable. There were no murmurs.

Electrocardiograms recorded during sinus rhythm showed an obvious ventricular post excitation wave termed as Epsilon potential (Fig. I). During an episode of palpitation, a ventricular tachycardia with a LBBB contour was recorded with a mean QRS axis of -60° (Fig. II). Echocardiography revealed a dilated right ventricle with a RV/LV ratio of 1.2:1 (Fig. III). The septal motion was flat and there was global hypokinesia of right ventricle.

The patient was put on Amiodarone therapy in view of his symtomatic ventricular tachycardia and after effective control was discharged after 2 weeks.

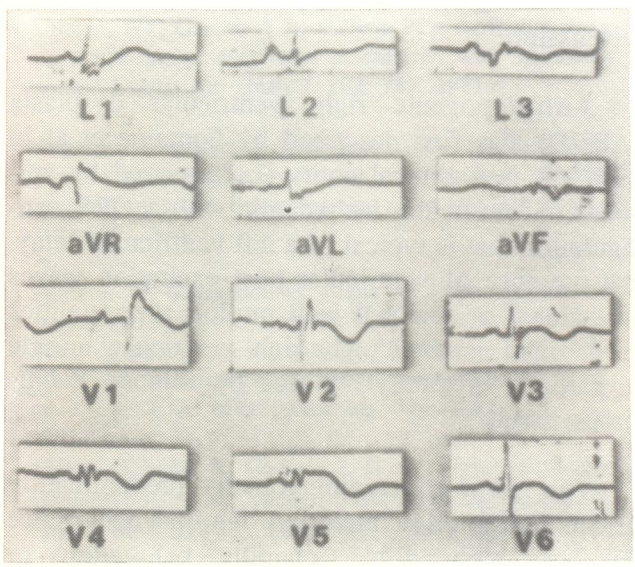

Fig. I: ECG recorded during sinus rhythm showing an obvious ventricular post excitation wave termed as "Epsilon potential".

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After a month, the patient again presented to us with ventricular tachycardia and haemodynamic compromise. It was found that the patient had stopped taking amiodarone of his own accord, due to gastrointestinal upset, for the last 2 weeks. Though his tachycardia was reverted by D C shock and he was given intravenous amiodarone, he continued to have sustained episodes of ventricular tachycardia with varying QRS axes over the next three days. He ultimately developed ventricular fibrillation and we were unable to salvage him.

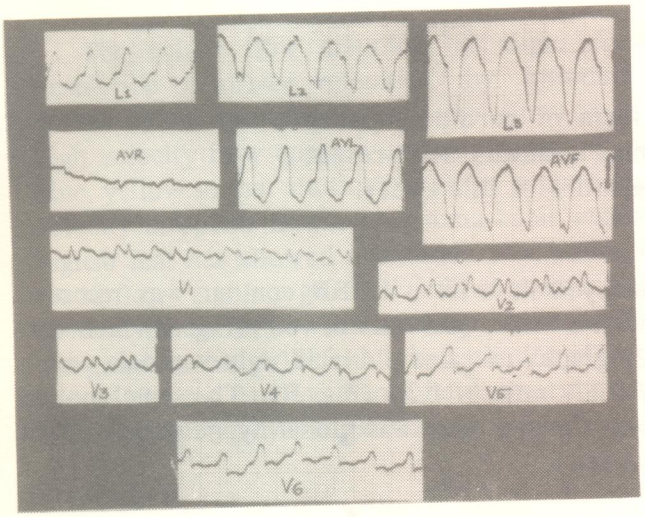

Fig. II: ECG during ventricular tachycardia with a LBBB contour and mean QRS axis of -60°.

Discussion:

Arrhythmogenic right ventricular dysplasia (ARVD) was first described by Fontaine et al in 1977[7]. It is a clinical entity characterized by episodes of ventricular tachycardia with a QRS configuration that is typical of a left ventricular delay, the presence of ventricular post excitation waves and global or regional wall motion abnormalities of the right ventricle[4]. The right ventricular muscle is partially or totally replaced by adipose and fibrous tissues[8].

This form of cardiomyopathy predominantly affects young adult males. A familial occurrence has been reported[2,3,5]. Generally, palpitations or syncope due to recurrent sustained ventricular tachyarrhythmia bring the patient to the hospital. Physical examination is usually normal, but a fourth heart sound may be audible. Patients may present in right heart failure or with asymtomatic right ventricular enlargement.

Our patient was a 36-year old man who presented with history of palpitations off and on and dyspnoea on exertion NYHA II. He also gave H/O dizziness and one episode of syncope. There were no signs of right ventricular failure; only a right sided fourth heart sound was audible.

During sinus rhythm, a form of right ventricular conduction delay appears in patients with ARVD. The delayed activation is inscribed in the form of a sharp deflection after termination of the QRS, during the ST segment or upstroke of the T wave, termed as the "Epsilon potential"[4, 6]. The visibility may be enhanced by doubling the gain of ECG while recording leads V_1 to V_3. These ventricular potentials can extend beyond the refractory period of the adjacent healthy myocardium and reactivate the adjacent myocardium leading to reentrant ventricular tachyarrhythmias[9]. Ventricular tachycardias generally have a left bundle branch block contour, often with right axis deviation, with T wave inverted over the right precordial leads[10]. A LBBB pattern with a mean QRS axis betwen +60° and +135° during tachycardia suggest an origin in the right ventricular infundibulum. Some patients may have different QRS configurations implying a widespread right ventricular electrical disturbance. Spontaneous and inducible ventricular fibrillation have been reported in ARVD[11,12]. Supraventricular arrhythmias and atrioventricular blocks may also occur[10]. Our patient had an epsilon potential on the ST segment on surface ECG. During ventricular tachycardias, a LBBB pattern with a QRS axis varying from -60° to +30° was observed.

Two dimensional echocardiography shows increased right ventricular diastolic diameter; the RV/LV ratio is greater than normal. Septal motion is abnormal. There may be regional or global wall motion abnormalities of the right ventricle. Echocardiography can effectively diagnose patients with ARVD in the presence of gross structural abnormalities of the right ventricle[13]. Our patient had a dilated RV with global hypokinesia. The RV/LV ratio was 1.2:1.

Haemodynamic study reveals a prominent right atrial 'a' wave; the other pressures are normal. The right ventricular chamber has global or segmental wall motion abnormalities. There may be aneurysms with paradoxical systolic motion, usually over the anterior surface of the pulmonary infundibulum, at the apex and inferior wall of right ventricle. Electrophysiology reveals normal atrial, nodal and ventricular conduction. Ventricular tachycardia can be induced by programmed stimulation and can be shown to have LBBB configuration. Endocardial post excitation waves may be recorded[4]. Haemodynamic studies could not be done in our patient due to uncontrolled ventricular tachyarrhythmias.

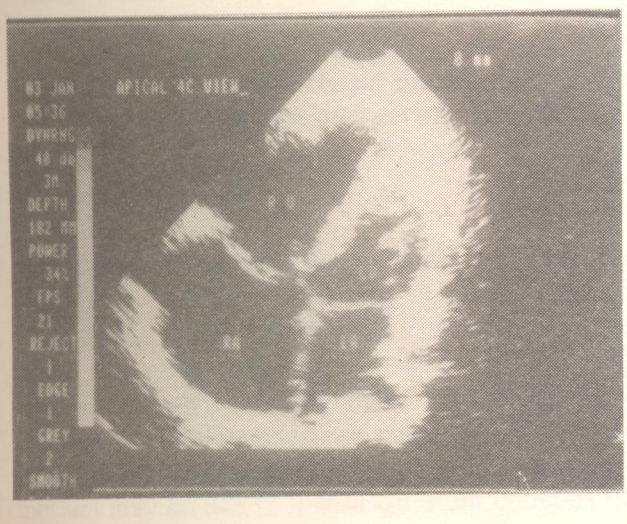

Fig. III: Apical four chamber view (2DE) showing a dilated right ventricle with RV/LV ratio of 1.2:1.

The exact aetiology of this entity is largely unknown. Right ventricular endocardial biopsy may be helpful in the diagnosis of ARVD[1]. The free wall of the right ventricle is not parchment like as in Uhl's anomaly. Histology reveals myocardial replacement by lipomatous and/or fibrous tissue along with foci of inflammation, degeneration and necrosis[8].

Antiarrhythmic drugs are effective in abolishing the ventricular tachyarrhythmias in majority of patients. The efficacy of various antiarrhythmic drugs has not been systematically evaluated. Amiodarone and class IC drugs are found to be effective. Surgical treatment in the form of right ventriculotomy is recommended when patients are disabled with recurrent ventricular tachycardia after failure of antiarrhythmic drug therapy[14]. Because of the variable course and instability of ventricular arrhythmias, if is difficult to evaluate medical or surgical therapy in these cases.

The few long term follow up studies suggest a favourable prognosis[14, 15], though the course is highly variable amongst individual patients. Our patient died within three months of presentation. So it is evident that such patients have the potential for sudden arrhythmic death and warrant further investigations to identify such high risk cases.

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