Left ventricle involvement in Arrhythmogenic Right Ventricular Dysplasia

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Abstract:
Arrhythmogenic right ventricle dysplasia ARVD is an unusual progressive cardiomyopathy characterized by genetically determined fibrofatty replacement of myocardium, predominantly right ventricle with LBBB pattern ventricular arrhythmias. We have encountered a case of arrhythmogenic right ventricular dysplasia with RBBB type of ventricular tachycardia. We report this case because of rare association of RBBB pattern ventricular tachycardia in ARVD.

Keyword: arrhythmogenic right ventricular dysplasia, epsilon wave, RBBB

INTRODUCTION Arrhythmogenic right ventricular dysplasia [ARVD] is an increasingly recognized cause of ventricular tachycardia and sudden cause of death in young people especially in adults. It is primarily a disorder of myocardium predominantly right ventricle leading to segmental and global myocardial dysfunction. We report one such case with rare association of RBBB [right bundle branch block] type ventricular tachycardia. CASE REPORT 55 year old man admitted in intensive care unit for palpitations and giddiness of 40 min duration. He was in state of hypotension with profuse sweating and a systolic blood pressure of 40 mm hg with feeble pulse. His past history did not reveal chest pain or breathlessness or syncop. Family history suggested sudden cardiac death of his elder brother at the age of 33 years of unknown cause. Examination suggested normal jugular venous pressure and no pedal edema. His ECG [fig-1] revealed monomorphic ventricular tachycardia of RBBB pattern, suggesting origin from left ventricle. Immediately ventricular tachycardia was reverted back to sinus rhythm by cardioversion, and hemodynamically stabilized.

![ECG Graph]
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Fig 1. ECG showing monomorphic ventricular tachycardia of RBBB pattern
The second ECG [fig-2] after recovery of sinus rhythm reveals a distinct terminal deflection immediately after QRS suggesting epsilon wave,[fig-3] a specific feature of ARVD.

Fig 2. ECG after cardioversion showing sinus rhythm with epsilon wave

Fig 3. ECG showing epsilon wave (indicated by arrow)
Echocardiogram revealed significant right ventricle enlargement [fig-4] with moderate right ventricle dysfunction [fig-5]. Hyper reflective moderator band also noted. RVOT diameter was 35 mm and apical dyskinesia noted. Left ventricle function was normal.

Fig 4. Echocardiography showing RV enlargement with mid and basal segmental dilatation

Subsequently patient underwent cardiac MRI, which suggested moderate RV enlargement with thinning of RV free wall [fig-6] and fatty infiltration of right ventricle myocardium, and severe segmental dilatation of basal right ventricle [fig-7].

Fig 5. Echocardiography showing RV dysfunction as evidenced by TAPSE of 12.7 mm
DISCUSSION:
According to the World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of Cardiomyopathies,[1] ARVC, also known as arrhythmogenic right ventricular dysplasia, is characterized by three features, progressive fibrofatty replacement of right ventricular myocardium, strong familial transmission and symptomatic ventricular arrhythmias. It is characterized by 30-50% familial occurrence with autosomal dominant inheritance. Fatty infiltration alone is not sufficient to diagnose ARVD/ARVC, but fibrosis should be accompanied which is more arrhythmogenic than fibrosis. Fibrofatty changes can be present anywhere in the “triangle of dysplasia”, formed by inflow, apex and outflow parts of the right ventricle.[2] Pathological changes may also migrate to free wall of left ventricle also.[3,4] ECG changes seen in 90% of patients with ARVD with inverted T wave in v1-v3 occurring in 50-70%. Epsilon wave occurs in 30-50% of patients. ARVD/C is a disease that may have a temporal progression, and the disease may present differently according to the time of presentation. [5].Echocardiogram forms an integral part of diagnostic evaluation. Echocardiographic evidence of diffuse right ventricular enlargement strongly supports ARVD in suspected cases.[6]. MRI is considered as the definite non invasive test for ARVD, which detects fatty infiltrations as high intensity signal in the right ventricle free wall.[7]. According to Task Force Criteria 2010, ARVD can be diagnosed if 2 major or one major with 2 minor or 4 minor are satisfied. In our case 3 major criteria are satisfied ie right ventricular enlargement in ECHO, epsilon wave in ECG and fibrofatty infiltrations with right ventricle enlargement in MRI and 1 minor criteria ie family history of sudden death [ 35 years]. The rare presentation in our case is ventricular tachycardia of RBBB type signalling the involvement of left ventricle. The prevalence of LV involvement in ARVD is from 16%[8] to 76%[9]. Clinical expression of left ventricle involvement occurs by extension of T wave inversion to lateral leads [V5, V6, L1, AVL], left ventricular arrhythmias, or LV dilatation or dysfunction.[10]. Family history of sudden death and involvement of left ventricle necessitates ICD placement in this patient. CONCLUSION: Even though ARVD can present with ventricular arrhythmias ranging from ventricular premature contractions to ventricular tachycardia of LBBB type, the possibility of occurrence of RBBB pattern ventricular tachycardia as a manifestation of left ventricular involvement should also be considered.
Usually occurrence of left ventricular involvement portends a poor prognosis because of advanced state of disease and warrants ICD implantation.

REFERENCES:


