VENTRICULAR TACHYCARDIA HARBINGER OF AN INTERESTING CASE OF CARDIOMYOPATHY

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Abstract:
Ventricular tachycardia can be a harbin-ger of cardiomyopathy. Arrhythmogenic right ventricular dysplasia (ARVD) can present as ventricular tachycardia. ARVD is grouped under primary cardiomyopathies of genetic origin. ARVD is an uncommon form of inheritable heart muscle disease (estimated 1 in 5000). ARVD involves predominantly the right ventricle with progressive loss of myocytes and fatty or fibrofatty tissue replacement.

Keyword: cardiomyopathy, arrhythmogenic right ventricular dysplasia (ARVD), Ventricular tachycardia

INRODUCTION: Ventricular tachycardia (VT) is a manifestation of underlying heart disease-acute myocardial infarction, cardiomyopathies, heart failure etc. Here, arrhythmogenic right ventricular dysplasia (ARVD) presents as VT. ARVD is grouped under primary cardiomyopathies of genetic origin. ARVC/D is an uncommon form of inheritable heart muscle disease (estimated 1:5000). ARVC/D involves predominantly the right ventricle with progressive loss of myocytes and fatty or fibrofatty tissue replacement, resulting in regional (segmental) or global abnormalities. ARVC/D has a broad clinical spectrum, usually presenting clinically with ventricular tachyarrhythmias (eg. monomorphic ventricular tachycardia). In early stage of the disease, structural changes may be absent or subtle and confined to a localized region of the RV, typically the inflow tract, outflow tract, or apex of the RV, the ‘triangle of dysplasia’.

CASE REPORT:
42 year old man presented to the intensive care unit with h/o palpitations for past 30 minutes in a drowsy state with no history of chest pain, breathlessness or syncope. ECG(fig.1) revealed monomorphic ventricular tachycardia of LBBB morphology with right axis deviation suggestive of RVOT origin. Pulse-feeble, thready and BP not recordable. Immediately, he was cardioverted to sinus rhythm. ECG(fig.2) showed sinus rhythm, normal axis with T inversion from V1 to V5. He was stabilised and further evaluated.
His chest xray(fig.3) showed cardiomegaly.

ECHO(fig.4,5,6) revealed dilated right atrium and right ventricle with dilated RVOT. RV free wall was hypokinetic.
Cardiac MRI (fig.7,8) demonstrated dilated right atrium, right ventricle and RVOT. RV myocardium was thinned out with a few dyskinetic bulges. Holter monitoring for 24 hours showed about 250 monomorphic VPDs of RV origin. He was treated with Inj.Amiodarone infusion followed by oral amiodarone. DISCUSSION: ARVD is a cause of sudden cardiac death in young persons, as the most common presentation is ventricular tachycardia. One series accounted for 20% of sudden deaths in all individuals younger than 35 years and 22% of sudden deaths in young athletes. Our case presented with VT which was subsequently diagnosed as ARVD. By ECHO (fig.4,5), patient had dyskinetic RV free wall with dilated RVOT-proximal 37 mm and distal 37 mm. MRI (fig.7,8) also fulfilled features of ARVD. ECG (fig.2) showed terminal QRS duration from nadir of S wave to end of QRS duration of 70 milliseconds with T inversion from V1 to V5 leads. Arrhythmogenic right ventricular dysplasia (ARVD) is a myocardial disorder of primarily the right ventricle, with unknown cause with a frequent familial occurrence. The typical clinical manifestation consists of ventricular arrhythmias with a left bundle branch block (LBBB) pattern that occur predominantly in young adults. Other manifestations are electrocardiographic repolarization and depolarization changes, structural abnormalities that range from subtle wall abnormalities within the “triangle of dysplasia” to biventricular regional or global dysfunction, and localized or widespread fibrofatty infiltration of the right ventricular myocardium. The diagnosis of ARVD is based on the presence of major and minor criteria encompassing genetic, electrocardiographic, pathophysiologic, and histopathologic factors., i.e. 2 major, 1 major and 2 minor or 4 minor. Our patient had 3 major criteria fulfilling the criteria laid by Task Force 2010.

CONCLUSION:
As ARVD can cause sudden cardiac death, patients are advised to avoid vigorous athletic activity because of the strain on the right heart and to take anti-arrhythmic drugs continuously. Therefore, an early and accurate diagnosis followed by appropriate therapy for this condition is increasingly important for it may prevent lethal arrhythmias.

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