Arrhythmogenic right ventricular cardiomyopathy (ARVC) with right ventricular clot: The best timing for ICD implant

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Introduction: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a relatively rare inherited cardiac disease characterized by non-ischemic ventricular arrhythmias originating from the right ventricle and abnormal ventricular pathology. Macroscopically, there is a scarred appearance with fibrous or fibro-fatty replacement of myocardium.

Methods: A 19 year old university student collapsed while playing football on 16th February 2019 morning. CPR was commenced and automatic external defibrillator was attached. He was direct-current cardioverted twice. In Emergency department, he developed ventricular tachycardia and was given another 2 direct-current cardioversion. However, he progressed to ventricular fibrillation. CPR was commenced for another 15 minutes. He was intubated for airway protection. The patient was cardioverted due to another episode of supraventricular tachycardia and ventricular tachycardia in CCU. He was given intravenous amiodarone infusion and was transferred to a cardiac referral center on day 3 for further management. This was his first episode of collapse. There was no previous history of medical illness, no family history of sudden death or cardiac disease. Echocardiogram showed good left ventricular function with ejection fraction of 66% and good right ventricular function with TAPSE of 24mm. There was no regional wall motion abnormality, all chamber size and valves were normal. No clots and no pericardial effusion. Electrocardiography (ECG) showed T wave inversion in V1 to V3 and widening of QRS complexes, 720ms. Cardiac magnetic resonance (CMR) showed right ventricular clots. The regional right ventricular dyskinesia and akinesia with dilated right ventricle (indexed end diastolic volume of 233) and mildly impaired systolic function fitted in as major CMR criteria for ARVC as per 2010 Task Force criteria.

Result: The patient was given warfarin for 6 weeks. Repeated CMR 6 weeks later showed resolved right ventricular clot. The patient was finally implanted an implantable cardioverter defibrillator (Ellipse VR St Jude) after 2 months. He was then discharged well the following day. We managed to screen all his family member although all of them were asymptomatic. Echocardiogram of his mother, brother and 2 sisters showed dilated right atrium and right ventricle.

Conclusion: ARVC with right ventricular thrombus is an uncommon phenomenon which the patient experienced both. Right ventricular thrombi usually represent clots traveling from the legs to the lungs. The diagnosis of ARVC is definitely challenging and it can be easily misdiagnosed. Thus, it is imperative to make a thorough investigation for all possible causes so as not to miss the diagnosis of this rare
condition and avoid delay in treatment.