Sarcoidosis mimicking arrhythmogenic right ventricular cardiomyopathy

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Introduction: Cardiac sarcoidosis predominantly affects the left ventricle (LV). Here we describe an atypical case of sarcoidosis with predominant right ventricular (RV) involvement, mimicking arrhythmogenic right ventricular cardiomyopathy (ARVC) clinically as well as on investigations.

Methods: A 36-year-old male presented with recurrent episodic palpitations associated with dizziness for the last three months. There were no syncopal attacks. Baseline electrocardiogram (ECG) showed presence of right bundle branch block (RBBB) and inverted T waves in anterior chest leads (Figure 1a) with documented ventricular tachycardia (VT) during the episodic palpitations. ECG during VT showed left bundle branch block pattern with inferior axis (Figure 1b). Echocardiogram showed normal LV function and mild RV dysfunction with thinning of RV myocardium. On the basis of baseline ECG, morphology of VT and echocardiographic findings, a possibility of ARVC was considered. Cardiac Magnetic Resonance Imaging (MRI), however, suggested the diagnosis of acute sarcoidosis with mediastinal lymphadenopathy. Subsequently PET/CT and bronchoscopic biopsy of lymph node were conducted for confirmation.

Result: Cardiac MRI revealed global RV hypokinesia with diffuse T2 hyperintensity and nodular late gadolinium enhancement (LGE) involving the interventricular septum and RV free wall (Figure 1c) along with multiple lymph nodes in the mediastinum largest measuring 3 cms. Cardiac 68Ga-DOTANOC PET/CT showed mediastinal lymphadenopathy (Figure 1d) with intense uptake in the lymph nodes and RV myocardium. Bronchoscopic ultrasound-guided lymph node core biopsy revealed the presence of non-necrotizing epithelioid cell granulomas consistent with sarcoidosis. AFB staining, Gene Xpert for TB and tubercular bacterial culture were negative. The patient was started on oral steroids (Wysolone 1 mg/kg per day) and was advised intracardiac cardioverter-defibrillator. The episodic palpitations and VTs completely subsided following treatment initiation and follow-up cardiac MRI showed no evidence of acute sarcoidosis with reduction in the size of lymphadenopathy and intensity of LGE.

Conclusion: Cardiac sarcoidosis can very rarely involve predominantly the RV, causing RV dysfunction and typical ECG features of ARVC with right-sided ventricular arrhythmias. The distinction between sarcoidosis and ARVC here is very important because only sarcoidosis improves with corticosteroid treatment, unlike ARVC.