

Isolated cardiac sarcoidosis: a failed diagnosis?

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Introduction: Isolated cardiac sarcoidosis (iCS) is an infiltrative cardiomyopathy that is the result of granulomatous inflammation that manifests predominantly in the myocardium. The annual incidence of sarcoidosis varies between 1 and 15 per 100,000 depending on the region¹. The prevalence of iCS among patients with systemic sarcoidosis varies widely (23–54%) because of differences in the definitions used². Establishing a diagnosis of iCS is extremely difficult, since there is no unique echocardiographic, radiological or laboratory test to confirm the diagnosis. iCS may present with symptoms of heart failure, sudden cardiac death, ventricular arrhythmia, myocardial infarction or atrio-ventricular block³.

Case report: 60-year-old female patient was recently hospitalized in our institution who presented with symptoms of heart failure without a history of previous cardiac diseases. Echocardiography showed a dilated left ventricle with hypokinesia of the basal segments of the septum, posterior, inferior and lateral walls with a reduced ejection fraction (EF) of 30%. Obstructive coronary artery disease was ruled out with coronary angiography. Cardiac magnetic resonance imaging (CMRI) was performed, which suspected the diagnosis of iCS. Positron emission tomography (PET) computed tomography (CT) showed increased metabolism of glucose analogues in the greater part of the left ventricle, which is consistent with iCS. Corticosteroid therapy was also introduced into the therapy in addition to the optimal medical therapy of heart failure. As part of the primary prevention of sudden cardiac death, a two-chamber ICD device was implanted.

Conclusion: Patients with iCS have poor prognosis. If left untreated, iCS leads to progressive failure of the left ventricle with frequent ventricular arrhythmias and sudden cardiac death. iCS is a frequently misdiagnosed due its rarity and high index of suspicion needed to make the diagnosis. It should be noted that this is the only diagnosed case of iCS in the last 5 years in our institution, which, considering the incidence, leads us to the question of an adequate diagnosis confirmation. It is of utmost importance to increase the use of non-invasive diagnostic methods such as PET CT scan and CMRI to detect all patients with iCS.

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