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CASE REPORT

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Palpitations as a presenting feature of multisystem sarcoidosis

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ABSTRACT

Introduction: Sarcoidosis is described as a systemic condition characterized by non-caseating granulomas in multiple organs. In this report, we present an unusual manifestation of cardiac sarcoidosis and review management strategies.

Case presentation: A 29-year-old African-American man presented with weight loss, fatigue, dyspnea, palpitations, night sweats, painless left eye redness and bilateral leg pain over the course of three months. His physical exam revealed left conjunctival congestion and bilateral crackles on auscultation. Computerized tomography of the chest showed severe parenchymal disease with bilateral fibrotic bands. Bronchoscopy and transbronchial biopsy revealed non-caseating granulomas and multinucleated giant cells, confirming sarcoidosis. Non-sustained ventricular tachycardia developed. Cardiac MRI showed myocardial delayed gadolinium enhancement. He responded to methotrexate and steroid therapy. An implantable cardioverter-defibrillator was placed.

Discussion: Although cardiac sarcoidosis manifests in only 5% of sarcoidosis, autopsy reports indicate subclinical cardiac involvement in up to 30%. There are no established criteria for diagnosis of cardiac sarcoidosis.

Conclusion: Early recognition and diagnosis of cardiac sarcoidosis is challenging but vital due to unpredictability and high risk for malignant cardiac involvement. Newer diagnostic imaging modalities have further aided in earlier identification and prevention of sudden cardiac death.

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1. Introduction

Sarcoidosis is a systemic condition characterized by non-caseating granulomas in multiple organs. The heart is involved in about 20–30 percent of cases [1]. In this report, we present an unusual presenting manifestation of cardiac sarcoidosis and review management strategies.

2. Case presentation

A 29 year old African-American man presented with a three month history of palpitations, weight loss, fatigue, shortness of breath, progressive exercise intolerance, night sweats, painless left eye redness and bilateral leg pain. He did not use tobacco or illicit drugs, and drank alcohol only on social occasions. Physical examination revealed left conjunctival congestion (Figure 1) and bilateral crackles on lung auscultation.

Laboratory investigations showed an elevated creatine kinase at 1095 units/L and a creatine kinase-MB level of 113.7 mcg/L. The erythrocyte sedimentation rate was 5 mm/hour and serum creatinine was 1.09 mg/dL without a previously available baseline. Investigations for tuberculosis, viral hepatitis, and the human immunodeficiency virus proved negative.

His electrocardiogram (EKG) showed nonsustained ventricular tachycardia (VT) (Figure 2). A transthoracic echocardiogram revealed a dilated left ventricle with an ejection fraction of 30% and no evidence of valvular heart disease. Irregular bilateral fibrotic bands in the pulmonary parenchyma were seen on computerized tomography (Figure 3). ACE (angiotensin converting enzyme) levels were elevated at 198 U/L (reference range: 9–67 U/L). Bronchoscopy and transbronchial biopsy showed non-caseating granulomas with multinucleated giant cells, confirming the diagnosis of sarcoidosis. The patient continued to have several episodes of nonsustained VT over the course of his hospital stay. Cardiac magnetic resonance imaging (CMR) showed delayed gadolinium enhancement in the myocardium, highly suggestive of sarcoid involvement. Due to his decreased left ventricular ejection fraction, electrophysiologic testing was pursued. It revealed inducible sustained polymorphic ventricular tachycardia and subsequently an implantable cardioverter-defibrillator was placed. The patient had been started on intravenous methylprednisolone at a dose of 40 mg every eight hours. He was treated with prednisolone acetate eye drops for his left eye uveitis. Steroids were tapered down gradually and he was

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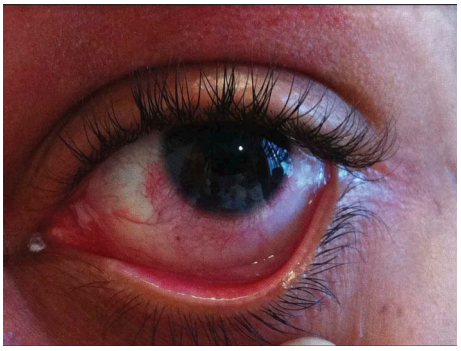

Figure 1. The patient's left eye showing congestion.

discharged on prednisone 40 mg twice daily at the end of his eight-day hospitalization.

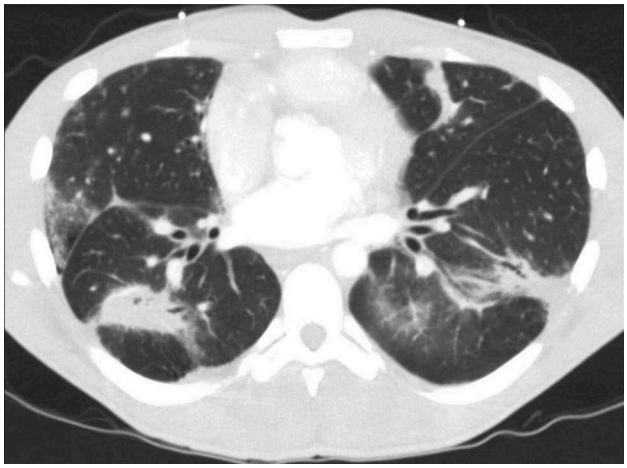

Figure 3. CT chest showing irregular bilateral fibrotic bands in the pulmonary parenchyma.

3. Discussion

Cardiac involvement may present in only 5% of cases of sarcoidosis. However, autopsy reports indicate subclinical cardiac involvement in up to 20–30% of cases. In Japanese studies, there are reports of rates up to 60%. Cardiac involvement is not related to pulmonary involvement and may be seen asynchronously. Hence, unexplained cardiac symptoms or irregularities on electrocardiography in any patient aged below 55 years without any significant medical history should prompt an evaluation for possible cardiac sarcoidosis.

A further clue may be provided by the patient's ethnicity as a higher incidence of sarcoidosis has been found in certain ethnicities. The Scandinavian countries, Ireland, and Japan have reported the highest prevalence. In the United States, the highest rates have been seen in black women, with an incidence of 39.1 per 100 000. Black men are second highest at 29.8 per 100 000. White women and men have rates of 12.1 and 9.6 cases per 100 000, revealing a clear racial predilection [2].

Conduction abnormalities are the most common manifestation of cardiac sarcoidosis. The most commonly documented arrhythmias include complete atrioventricular block, ventricular tachycardia, and ventricular fibrillation [3]. However, atrial arrhythmias, including atrial fibrillation, are emerging as more common that previously considered [4]. The pathophysiology is thought to be that the non-caseating sarcoid granulomas act as niduses for re-entrant arrhythmias. These arrhyhthmias are seen in up to 22% of patients with sarcoidosis [1]. The exact locations of the granulomas are variable, but the myocardium is most frequently involved. Other observed sites include coronary arteries and the valve leaflets.

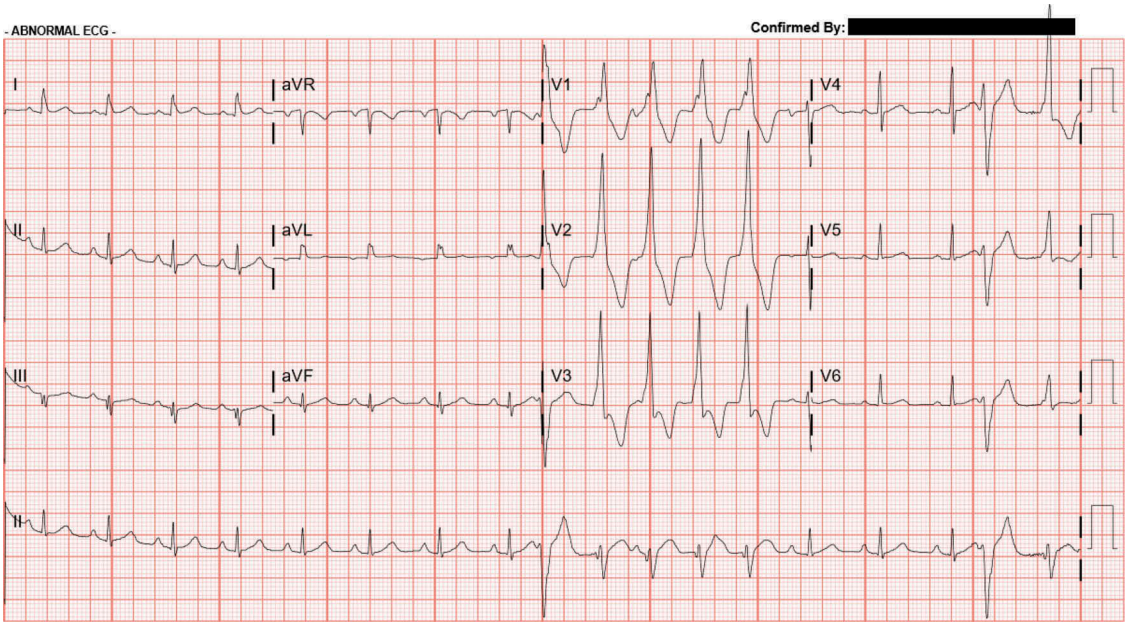

Figure 2. The patient's EKG showing non-sustained ventricular tachycardia.

When granulomas infiltrate the sinus node, sinus arrest may occur.

There are no established criteria for a specific diagnosis of cardiac sarcoidosis. The Japanese Ministry of Health and Welfare have proposed guidelines which have been commonly used. One diagnostic criterion is histological confirmation with endomyocardial biopsy, although sensitivity for this criterion is low due to the patchy involvement. Histological confirmation of extra-cardiac sarcoidosis plus presence of conduction abnormalities or ventricular arrhythmias is also considered diagnostic. Our patient met the second set of diagnostic criteria.

As the heart is involved in a significant percentage of patients with sarcoidosis, an EKG should be performed to detect conduction and repolarization abnormalities in all patients diagnosed with sarcoidosis. Up to 70% of these EKGs may be abnormal and may require further investigations.

Isolated cardiac sarcoidosis is an emerging entity, and may comprise up to 25% of all cases of cardiac sarcoidosis [5]. In this condition, sarcoidosis involves only the heart, prompting the need for a low clinical threshold when faced with an abnormal EKG in younger patients. Early detection is crucial as management will be affected – over 70% of patients with cardiac sarcoidosis will require a pacemaker or a defibrillator.

Cardiac magnetic resonance imaging facilitated the diagnosis in our case. The presence of late gadolinium enhancement can highlight even small areas of damage in patients with suspected cardiac sarcoidosis. Guidelines proposed by the Heart Rhythm Society suggest that CMR or PET (positron emission tomography) should follow if history, EKG or echocardiography is suspicious for cardiac involvement [6]. A recent study shows that the presence of LGE on CMR has a sensitivity of 97% and a specificity of 100% in the detection of cardiac sarcoidosis [7]. Although CMR is promising, inter-observer variation will need to be improved if it is to take on a greater role in screening purposes [8].

Initial management of cardiac sarcoidosis comprises systemic corticosteroids at moderate to high doses. Positive effects include improved symptoms, decreased risk of ventricular arrhythmia, and curtailed negative remodeling of the ventricles. Arrhythmia may persist despite immunosuppressive therapy. In these patients, antiarrhythmic drugs such as amiodarone or sotalol should be started. These medications may suppress arrhythmia in up to 50% of patients [9]. However, amiodarone may negatively affect patients with active pulmonary sarcoidosis and its use has been discouraged in these patients.

Further therapy in refractory cases can include implantable cardioverter defibrillators (ICDs), radiofrequency ablation, and sympathectomy. Patients with a pacemaker or defibrillator demonstrate significantly improved survival compared to patients without either device [10]. ICDs are considered definitive therapy for spontaneous sustained ventricular arrhythmias or reduced left ventricular ejection fraction equal to or below 35% [11]. Our patient did have a low ejection fraction of 30% which did not improve despite immunosuppressive therapy, and hence met criteria for ICD implantation as per Heart Rhythm Society guidelines [6].

4. Conclusion

Early recognition and diagnosis of cardiac sarcoidosis is challenging but vital due to its inherent high risk and lack of predictability for sudden cardiac death. All patients with sarcoidosis should be evaluated for the presence of conduction abnormalities and arrhythmias, even in the absence of any symptoms. The use of newer diagnostic imaging modalities can aid in identifying sarcoidosis patients with cardiac involvement, facilitating the initiation of treatment modalities in the prevention of sudden cardiac death.

Disclosure statement

No potential conflict of interest was reported by the authors.

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