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Review article

Management of cardiac sarcoidosis – A practical guide

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ABSTRACT

Sarcoidosis is a multi-system granulomatous disorder of unclear etiology which can affect any organ of the body including the heart. The heart is involved in up to 25% of sarcoidosis patients. In rare cases, the heart can be the only organ involved.

Involvement of the heart, called cardiac sarcoidosis, especially if symptomatic, significantly deteriorates the prognosis for sarcoidosis patients, which is why cardiac sarcoidosis should be not only considered, but also searched for actively. Despite recent advances in this field, diagnosis, risk-stratification, and treatment of cardiac sarcoidosis remains a challenging issue. Fortunately, several recommendations have been recently formulated which provide relatively clear guidance on the management of patients with cardiac sarcoidosis. The cornerstone of management of these patients is a multidisciplinary approach involving collaboration of cardiologists, pulmonologists, radiologists, rheumatologists, and other specialists.

Currently, diagnosis of cardiac sarcoidosis is based on an assessment of a patients' symptoms, physical examination and results of standard ECG, Holter monitoring and echocardiography. This series of examinations can identify individuals with possible cardiac sarcoidosis, who should undergo, as the next step, cardiac magnetic resonance and positron emission tomography, which are the techniques of choice for the diagnosis of cardiac sarcoidosis. Histological verification, critical for establishing a definitive diagnosis, is based – in cases with a typical picture documented by imaging techniques – on an extracardiac biopsy. In some cases, when an extracardiac biopsy is not feasible, an endomyocardial biopsy is needed.

The cornerstone of treatment remains corticosteroids, in some cases in combination with other immunosuppressives, although data on their efficacy and safety from randomized trials are lacking. As the most frequent causes of death from cardiac sarcoidosis are heart rhythm disorders, be it atrioventricular blocks or ventricular arrhythmias, an irreplaceable role in the management of these patients is played by implantation of pacemakers and implantable cardioverter/defibrillators (ICD). One of the most critical issues is risk

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stratification of patients who, while not meeting classic criteria for ICD implantation, continue to be at high risk of sudden cardiac death and therefore should still be considered for ICD implantation. The last option for patients with advanced sarcoidosis is heart transplantation.

The present paper is an overview of presentation, diagnosis, and treatment of cardiac sarcoidosis, with special emphasis on the use of algorithms applicable in routine clinical practice.

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Introduction

Sarcoidosis

Sarcoidosis is a multi-system granulomatous disorder of unclear etiology which can affect any organ within the body. The condition is characterized by the presence of noncaseating granulomas. The most frequently involved organs include the lungs, lymphatic nodes, skin, and eye. Half of patients with the most frequent form of the condition – referred to as lung sarcoidosis – remain asymptomatic, with only radiologically documented involvement of the hilar nodes. In the other half of patients, lung parenchyma is also involved and patients present with cough, exertional dyspnea and low-grade fever [1]. Active granulomatous inflammation may progress to tissue fibrosis [2]. Besides the lungs, sarcoidosis may affect any other organ system within the body. The prevalence of sarcoidosis is estimated at 10–20/100,000 persons [3]. However, the prevalence of sarcoidosis in some populations such as African Americans, Scandinavians or Japanese is markedly higher [4]. Women are affected more often than men. The disease most commonly develops between 25 and 45 years of age, with another peak occurring between ages 50 and 60 [4,5].

Despite intensive research in this field, the etiology of the condition remains unclear. It is most likely an exaggerated immunological response to an unknown antigenic stimulus. To date, both infectious and environmental factors (pesticides, aluminum, talc, etc.) have been implicated. An important role is believed to be played by genetic predisposition [6–8].

Cardiac sarcoidosis

Symptomatic involvement of the heart is relatively rare and reported to only occur in 2–5% of patients with pulmonary/systemic sarcoidosis [9,10]. However some pathological studies have suggested some involvement of the heart is present in up to 25% of patients with sarcoidosis [11]. Very rarely, the heart can be the only organ affected. Involvement of the heart in sarcoidosis patients suggests a less favorable prognosis [12]. The strongest prognostic marker is left ventricular (LV) dysfunction [13].

While sarcoidosis can affect any part of the heart and remain absolutely asymptomatic, it can also manifest itself as severe heart failure or arrhythmias resulting in sudden cardiac death; critical factors invariably include the extent and localization of cardiac tissue involvement and disease activity [2,14].

Clinical manifestations

Complete heart block is the most common finding in patients with cardiac sarcoidosis (CS) and can be present in up to 30% of these patients [15]. However, lower degree atrioventricular (AV) blocks as well as intraventricular conduction defects may also develop. From the pathophysiological point of view, the conduction system becomes infiltrated by sarcoid granulomas.

Ventricular arrhythmias are the second most common manifestation of CS. Triggered activity and abnormal automaticity secondary to inflammation can be present, though a macroreentrant mechanism around areas of a granulomatous scar is more common [16]. Supraventricular arrhythmias are also frequent and are typically caused by atrial dilation secondary to LV dysfunction or mitral regurgitation, although infiltration of atria by granulomatous process can play a role in some patients [15].

Extensive myocardial infiltration by granulomas may deteriorate both systolic and diastolic LV function with the patient subsequently developing heart failure [17].

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