

Cardiac Sarcoidosis: An Underestimated Cause of Congestive Heart Failure

Gülker JE*, Bansemir L, Klues H and Bufe A

Department of Cardiology, Germany University, Germany

Corresponding author: Gülker JE, Helios Clinics Krefeld, Department of Cardiology, Lutherplatz 40, 47805 Krefeld, University of Witten-Herdecke, Alfred-Herrhausen-Strasse 50, 58448 Witten, Germany, **E-mail:** jan-erik.guelker@helios-kliniken.de

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Introduction

Sarcoidosis is a multisystemic disorder and a systemic granulomatous disease. It can affect many organs; the predominant manifestation is the chest with a bilateral hilar lymphadenopathy. In most cases the lymphoreticular system, the eyes and the skin are involved as well. The etiology of sarcoidosis still remains unclear [1]. The incidence of heart involvement is with 2% a rare entity [2]. However, heart involvement is the main cause of a poor outcome. Sudden cardiac death is frequent even in previously asymptomatic patients [3]. Congestive heart failure is the other severe consequence.

Keywords: Cardiac sarcoidosis; Congestive heart failure

Case Report

We report the case of a previously healthy 47 years old male who was referred with exertional dyspnae (NYHA III) which progressively worsened within the last eight weeks. Medical history, electrocardiogram (ECG) and chest X-ray were inconspicuous. No history of coronary artery disease or pulmonary disease. The physical and pulmonary evaluation failed to reveal any abnormalities. A transthoracic echocardiography proved a left ventricular systolic and diastolic dysfunction and the left ventricular ejection fraction was reduced to 40%. No relevant atherosclerosis in the coronary angiography which was performed additionally.

The cardiac magnetic resonance imaging showed extensive transmural late enhancement in the anterior, apical and basilar posterior and septal walls of the left ventricle corresponding to excessive regional fibrosis (Figures 1 and 2). As there were no subepicardial lesions this pattern was highly suggestive of cardiac sarcoidosis which was confirmed histologically

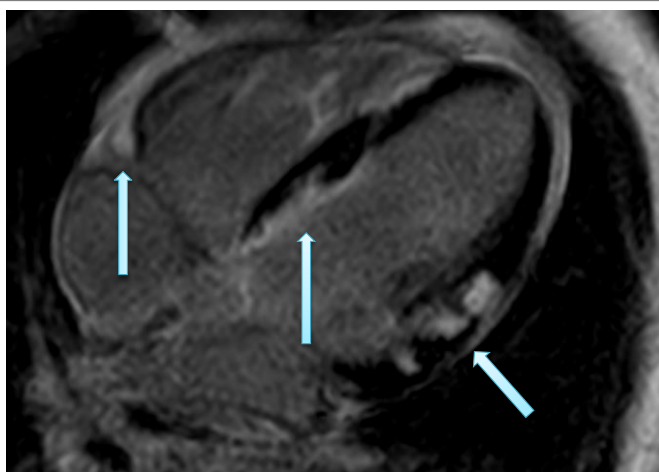


Figure 1: Cardiac magnetic resonance imaging showed extensive transmural enhancement

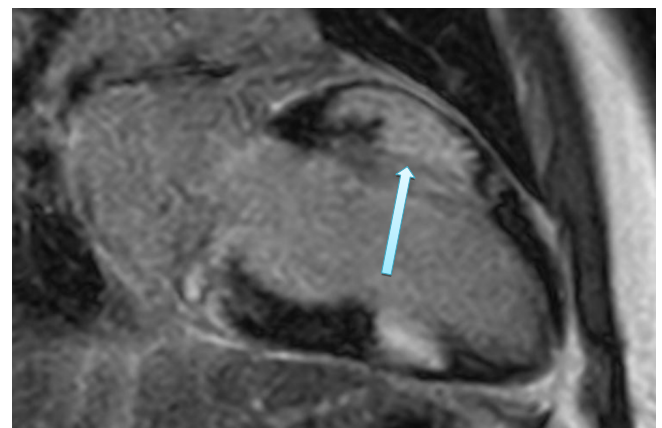


Figure 2: Cardiac magnetic resonance imaging showed extensive transmural enhancement

after endomyocardial biopsy (EMB) of the left ventricle. The EMB could reveal myocardial sarcoid granulomas and inflammatory mononuclear cell infiltrates. Immunohistochemical analysis showed numerous of CD3-positive T-lymphocytes, a few CD-20 positive B-lymphocytes and several CD-68 positive B-lymphocytes.

Treatment was started immediately with 60 mg corticosteroids daily. This dose was tapered gradually to a maintenance level of 15 mg per day over the next months. A reevaluation of clinical symptoms and left ventricular function should be mandatory during the further course of the disease.

Doppler echocardiography after 2 months showed an improved left ventricular function and in several ECG there were no signs of any severe arrhythmia.

Discussion

Primary cardiac sarcoidosis should be suspected in patients with dyspnae or arrhythmia and no signs of primary heart disease particularly

at younger age. The histological verification of cardiac sarcoidosis can be obtained with endomyocardial biopsy with a high specificity. Early diagnosis is a challenge as clinical manifestations are not specific and diagnostics have no sensitivity and specificity in spite of recent advances [4]. Therapy consists of corticosteroids and additionally the use of other immunosuppressive drugs in cardiac sarcoidosis as azathioprine, methotrexate, cyclophosphamide can be discussed; in selected cases even the implantation of an Implantable Cardioverter-Defibrillator (ICD) should be considered.

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