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Case Report

Cardiac sarcoidosis as a cause of sudden death

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ABSTRACT

Sarcoidosis, also called Besnier-Boeck disease, is a systemic granulomatous disease of unknown etiology which was classified as a separate unit in 1958. Histopathology of the disease is based on epithelioid granulomas which infiltrate and damage different organs and tissues. The disease is most commonly located in the lungs; however, first complaints may be related to its manifestation in other organs. Symptomatic involvement of the heart is found in approx. 5% of patients and is one of the most dangerous forms of the disease.

1. Introduction

Incidence of sarcoidosis in the Czech Republic reaches approx. 3 in 100,000 inhabitants, its prevalence amounted to as many as 60 in 100,000 inhabitants in the time of mass tuberculosis screening using abreugraphy (fluorography). Women are more commonly affected, usually between 30 and 50 years of age. The mortality of 3–5% has been reported, of which 80% of cardiac causes [1].

Sarcoidosis may be acute or chronic. Acute sarcoidosis is most commonly manifested by symptoms similar to a viral disease, a so-called Löfgren's syndrome (subfebrile temperature, polyarthritis, erythema nodosum, hilus lymphadenopathy, respiratory symptoms), and has a benign prognosis. Chronic sarcoidosis often leads to multi-organ involvement necessitating long-term therapy and has a less favorable prognosis.

Symptomatic involvement of the heart has been reported in 5% of patients with sarcoidosis. However, the studies show that asymptomatic cardiac involvement of different scope is present in as many as 20% of patients. In literature, there have been reports of an isolated involvement of the heart; however, no information is available about its prevalence [2]. Cardiac sarcoidosis may manifest itself by severe arrhythmia, heart failure, valve involvement, pericardial effusion or tamponade. Failure of the AV transmission or sudden cardiac death may also be the first manifestation of the disease [2].

We present a case report of a 49-year-old female patient in whom cardiac sarcoidosis was the cause of her death.

2. Case report

A 49-year-old female patient with a eufunctional struma and a history of cholecystectomy for lithiasis and a surgery of pleomorphic adenoma of the salivary gland has been

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followed in a cardiac out-patient department since 2004 for congenital aortic regurgitation. Valve replacement with metallic prosthesis was performed in 2005 due to the progression of the disease and clinical symptoms of heart failure. There were no perioperative abnormalities; postoperatively, there were no complications and follow-up echocardiography showed good functioning of the prosthesis.

The patient presented herself in January for severe weakness, fatigue, and slightly deteriorated breathing. The patient reported virus-like disease symptoms a month before comprising sore throat, joint and muscle pain, and subfebrile rise in temperature. Physical examination corresponded to the age of the patient, except for significant bradycardia. Her ECG revealed third-degree AV-block (Fig. 1) leading to patient's hospitalization. The patient had no history of bradycardia-causing therapy. Laboratory results revealed only slightly increased sedimentation, with no increase in inflammatory parameters. Thyroid hormones were normal, cardiac enzymes were negative. A chest X-ray showed no pulmonary pathology. Echocardiography verified good functioning of the prosthesis, with no signs of endocarditis or other pathology, good LV function, regular kinetics, with only a slightly asynchronous beat with artificial rhythm. Since there was no reversible cause of the AV-block, the patient was provided with a double-chamber pacemaker. The procedure had no complications and the patient was discharged for follow-up care in the out-patient unit. A history of myocarditis (however, with no conclusive serology; CMR or EMB was not indicated) or idiopathic degeneration of the transmission system were considered as possible causes of the AV-block.

The patient was regularly followed-up in our out-patient unit. She was fully dependent on pacemaking during pacemaker follow-up visits. The last follow-up visit was performed in November 2010. It was preceded by an echocardiography which revealed newly occurred mild left ventricular hypertrophy (13 mm, normal echogenicity); further findings were stable. The patient was clinically without any symptoms. The next follow-up visit was planned in 6 months, including updated echocardiography.

Further details of the case were received from health records and family members. In December 2010, the patient was examined by her general practitioner for an enlarged palpable right inguinal node (approx. 2×3 cm) which was hard and painless; with no other lymphadenopathy. Further examination, possibly including node extirpation, was recommended. However, the patient refused any further examinations. Since February 2011, the patient experienced progressive breathing difficulties and reduced exercise tolerance. She wanted to consult her physician with these complaints during her scheduled cardiac follow-up visit in April 2011.

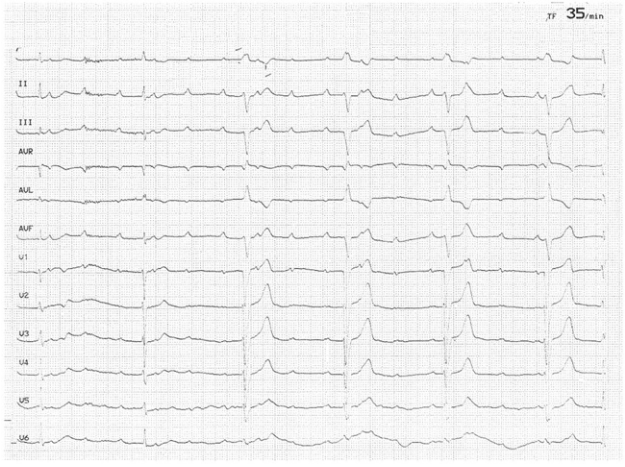

Fig. 1 – Third-degree AV block.

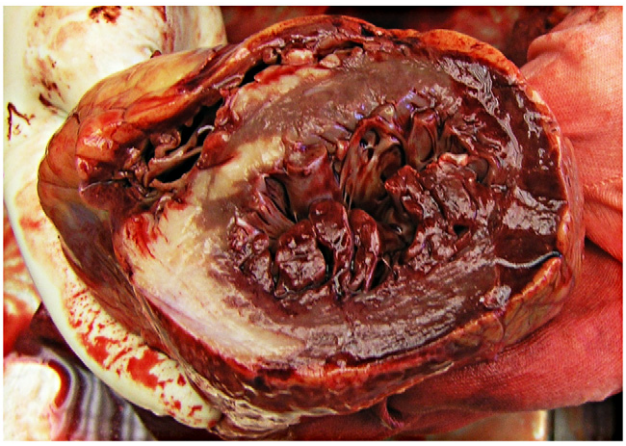

Fig. 2 – Sarcoidosis lesion in the wall of the left ventricle, anteroseptal position.

She suddenly lost consciousness a week before the scheduled follow-up visit. Her family called an ambulance and started cardiopulmonary resuscitation as instructed. Ventricular fibrillation was detected by paramedics. Resuscitation was unsuccessful and was finished after 30 min despite repeated defibrillation attempts and maximum pharmacologic support of circulation. Autopsy was indicated to clarify the cause of death.

Autopsy revealed the most robust findings in the heart with dilated left ventricle and LV hypertrophy which was most pronounced in the interventricular septum (anterior wall 12 mm, lateral wall 15 mm, posterior wall 12 mm, septum 22 mm). A yellow-whitish lesion ($50 \times 20 \times 48$ mm^3) was found in the area which affected the anterior wall of the right and left ventricles (Fig. 2). Other cardiac chambers had normal size, coronary arteries showed no significant anomalies. Additional similar macroscopic lesions were detected in the liver (Fig. 3) and the spleen, some smaller ones in both lung bases. Enlarged lymph nodes were found in the mediastinal, axillary, and both inguinal regions. Numerous epithelioid granulomas with a number of Langerhans-type giant cells and numerous fibrous areas, with no signs of caseous necrosis or microscopically detected infectious agents (Fig. 4) were found during histological examination. The character and location of these histopathological findings, together with the clinical manifestation of the disease, were suggestive of sarcoidosis. The cause of death was concluded as sudden death due to myocardial sarcoidosis.

3. Discussion

Isolated or predominantly cardiac sarcoidosis is rare and can be very difficult to diagnose. Given the symptoms of the acute phase of sarcoidosis, the complaints may often be considered to be of infectious origin. For this patient, the acute phase of