Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

EXPERT: Krzysztof Bederski MD, thoracic surgeon

Affiliation: *Department of Thoracic Surgery, John Paul II Hospital, Krakow, Poland*

Medical Expertise

CASE SUMMARY

The authors presented a case of a 32-old-year patient with pulmonary sarcoidosis. Despite initiation of conventional treatment the patient experienced progression of physical intolerance and shortness of breath. Additional cardiovascular evaluation was performed and the patient was diagnosed with cardiac sarcoidosis based on magnetic resonance study. Steroid therapy was started, what resulted in reduction of dyspnea and improvement in exercise capacity. Follow-up MRI showed partial remissions of previously described changes in heart. Prednisone dose was gradually reduced to 20 mg / day.

On current admission the patient's general condition was good. On physical examination no abnormalities were observed. ECG at rest did not reveal any significant abnormalities. 24-hours ECG monitoring showed no rythms and conduction pathologies. No abnormalities in blood tests were detected.

6 minute walk test – 615 m, without decrease in oxygen saturation. In cardiopulmonary exercise test the peak oxygen consumption was 38 ml/kg/min. The transthoracic echocardiogram (TTE) showed normal size of the heart chambers, preserved left ventricular ejection fraction (EF 60 %), increased echogenicity of intraventricular septum, no valvular pathology, no fluid in pericardium. Cardiac magnetic resonance with contrast (gadolinum) application was performed and delayed hyperenhancement area of intraventricular septum was detected. Compared to previous MRI images partial remission of changes in intraventricular septum was observed.

The authors discuss the further management strategy.

DISCUSSION

Cardiac involvement in the course of sarcoidosis is relatively rare. Symptomatic cardiac sarcoidosis is diagnosed in 5% of pulmonary sarcoidosis patient [1]. However, the autopsy data suggest that the heart may be involved in up to 78% of patients, depending on the population [2-4]. Sarcoid lesions are most frequently seen in left ventricular free wall, interventricular septum, and rarely in right ventricular free wall, left and right atrium, endocardium, or pericardium [3-6]. Because of initially asymptomatic course most of cardiac sarcoidosis cases remain undiagnosed. The most common manifestations of cardiac sarcoidosis include arrhythmias and conduction abnormalities [1]. Others are: dilated cardiomyopathy, heart failure, pulmonary hypertension, pericarditis, valvular defects due to papillary muscles involvement [1, 7- 9]. The risk of death among patients with symptomatic cardiac involvement is high. The five-year survival in various research ranges from 60 to 90% [10]. Treatment of cardiac sarcoidosis is aimed at controlling the inflammation, preventing fibrosis, and preventing compromise of cardiac structure or function. Corticosteroids are the cornerstone of treatment for cardiac sarcoidosis despite the lack of confirmatory randomized clinical trials.

EXPERT'S OPINION

The benefit of steroid treatment on cardiac function and structure seen in this patient is promising. Therefore, this therapy should be kept for at least 1 or two years. Optimal dose of glucocorticoids is not known, and choosing a dose requires balancing the risk of side effects with the likelihood of response. Gradual dose reduction to a maintenance level of 10 to 15 mg/day over one year should be recommended. If reemission is seen steroid discontinuation may be suggested. Regular cardiac and pulmonary monitoring is mandatory. In case of recurrence of symptoms the dose adjustment should be performed. Currently, there are no indications for HF specific treatment nor for electrotherapy. He should be followed-up closely with clinical assessment, echocardiography, electrocardiography and exercise testing done every 6-12 months.

REFERENCES

1. Ayyala U., Nair A., Padilla M.: Cardiac sarcoidosis. Clin. Chest Med., 2008; 29: 493-508.
2. Iwai K., Scklguti M., Hosoda Y., DeRemee R.A., Tazelaar H.D., Sharma 0. I'., Mahcahwari. A., Noguchi T.I.; Racial difference in cardiac sarcoidosis incidence observed at autopsy. Sarcoidosis. 1994 Mar;11(1):26-31.

John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu

3. Schulte W., Kirsten D., Drent M., Costabel U.: Cardiac involvement in sarcoidosis. Eur. Respir. Mon. 2005; 32: 130–149.

4. Sharma O.P., Maheshwari A., Thaker K. Myocardial sarcoidosis. Chest 1993; 103: 253–258.

5. Gozo E.G.Jr, Cosnow I., Cohen H.C., Okun L.: The heart in sarcoidosis. Chest 1971; 60:379-388.

6 . Nelson J.E., Kirschner P.A., Teirstein A.S.: Sarcoidosis presenting as heart disease. Sarcoidosis Vasc. Diffuse Lung Dis. 1996; 13: 178–182.

7. Roberts W.C., McAllister H.A. Jr., Ferrans V.J.: Sarcoidosis of the heart: a clinicopathologic study of 35 necropsy patients (group 1) and review of 78 previously described necropsy patients (group 11). Am J Med 1977;63(1):86-108.

8. Yazaki Y., Isobe M., Hiroe M., Morimoto S., Hiramitsu S., Nakano T., Izumi T., Sekiguchi M.: Prognostic determinantsof long-term survival in Japanese patients with cardiac sarcoidosis treated with prednisone. Am J Cardiol 2001 ;88(9): 1006-10.

9. 40. Fahy G.J., Marwick T., McCreery C.J., Quigley P.J., Maurer B.J.: Doppler echocardiographic detection of left ventricular diastolic dysfunction in patients with pulmonary sarcoidosis. Chest 1996;109(l):62-6.

10. Szczeklik A., Tendera M. redaktorzy: Kardiologia podręcznik oparty na zasadach EBM. Tom I. Medycyna Praktyczna, Kraków. 2009; 653.

John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu