Medical Expertise

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

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CASE SUMMARY

The authors presented a case of a 26-old-year male, professional football player, with pulmonary sarcoidosis and suspicion of cardiac sarcoidosis. In November 2013 the patient underwent a brief episode of fever, fatigue and vomiting. In January 2014 echocardiography revealed pericardial effusions. He did not report any symptoms. In previous cardiac echo studies performed in 2011, 2012 and 2013 any abnormalities were observed. On lung CT-scan in 02.2014 concominant mild pleural effusion and mediastinal lymphadenopathy were observed. He was than hospitalized in a Clinic in Switzerland where, based on positive results of IgG-reactivity for Adenovirus and Coxsackie B, viral pericarditis was diagnosed. CT scan in 04.2014 showed: mediastinal and cavity lymphadenopathy, no fluid in the pleural cavities, numerous micronodular changes in the lung parenchyma. Based on BAL(bronchoalveolar lavage) examination from 14.04.2014 suspicion of sarcoidosis was taken into consideration. Neoplastic process and tuberculosis were excluded.
When admitted to the Centre for Rare Cardiovascular Diseases he was asymptomatic, laboratory tests did not reveal any significant abnormalities. ECG at rest showed sinus rhythm 60 bpm, regular, normal heart axis. 24-hours ECG monitoring showed numerous episodes of bradycardia. The transthoracic echocardiogram (TTE) showed normal size of the heart chambers, preserved global and regional contractility of the left ventricular, (EF 65 %), increased enhancement of intraventricular septum, no valvular pathology, pericardial effusion max 17 mm in the area of anterior left ventricular wall, without signs of tamponade. Cardiac magnetic resonance with contrast (gadolinum) application showed slightly reduced mass and ejection fraction of the left ventricle (EF - 49 %), pericardial effusion, max 10 mm in the area of left ventricle inferior wall and right ventricule free wall. Furthermore, single, non-characteristic area of delayed hyperenhancement on the border of inferior wall and intraventricular septum; 6 mm diameter, was observed. Moreover, mediastinal and cavity limphadenopathy and pleural effusion, more on the right side, max 18 mm were detected. He was consulted by at the Department of Alergology and Immunology and the diagnosis of

pulmonary sarcoidosis was sustained. Because of the regression of lung lesions no treatment was recommended.

DISCUSSION

Sarcoidosis is a multisystem disorder of unknown ethiology [1]. The disease develops most frequently in hilar lymph nodes and lung parenchyma (90-97 % of cases). But it can affect almost every organ. It is most often diagnosed in people aged 20 to 60.

Cardiac involvement in the course of sarcoidosis is relatively rare. Symptomatic cardiac sarcoidosis is diagnosed in 5% of patients suffering from pulmonary sarcoidosis [2]. Sarcoid leasions most frequently occur in free wall of left and right ventricle, basal part of intraventricular septum, papillary muscles, left and right atrium, endocardium or pericardium [3, 4]. Main manifestations include complete atrio-ventricular block; ventricular and supraventricular tachyarrhythmias; systolic and diastolic heart failure; mitral or tricuspid valves insuficeinces; pericarditis or pulmonary hypertension.

Diagnosis of pulmonary sarcoidosis requires detailed history, physical examination, chest x-ray, pulmonary function tests, laboratory tests including tuberculin skin test, ecg, ophthalmological evaluation. It is important to eliminate alternative diagnoses: tuberculosis, HIV, histiocitosis, neoplasms etc. Final diagnosis is based on histopathological evaluation of biopsy. Cardiac sarcoidosis should be suspected in young people with diagnosed sarcoidosis of any organ, who have concomitant conduction disorders, tachyarrhythmias, heart failure symptoms or pericarditis.

The primary method of treatment for cardiac sarcoidosis is the systemic steroids. There is necessary the long-term use of high doses of medicines [4].

EXPERT'S OPINION

Histopathologic analysis of biopsy taken from enlarged mediastinal lymph nodes, by USG guided transbronchial biopsy is required in order to confirm the diagnosis of carcoidosis. This will guide further treatment strategy including systemic steroidotherapy. The patient should remain under close cardiological follow-up. Physical activity should be contraindicated until obtaining final diagnosis.

REFERENCES

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