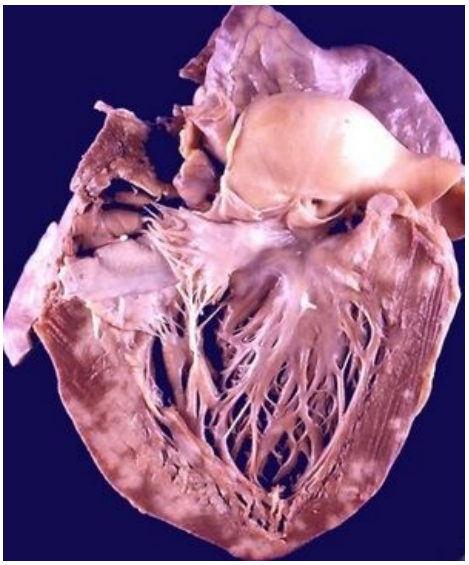
Sarcoidosis of the heart

Cardiac sarcoidosis is a granulomatous inflammatory disease of the myocardium. It can appear together with other extracardiac lesions or alone. Sarcoidosis itself is a multisystem disease of unknown cause. It usually affects individuals between the ages of 25 and 60. Most often, granulomatous inflammation is localized in the lungs. The eyes, liver, GIT, skin or nervous tissue can also be affected.

Cardiac sarcoidosis is manifested by **AV conduction disorders** and supraventricular and ventricular arrhythmias. These are the main danger of the disease. Sometimes we also observe the development of heart failure. The presence of cardiac sarcoidosis and the degree of cardiac dysfunction are important predictors of patient prognosis.



A heart affected by sarcoidosis with visible inflammatory-fibrotic changes in the myocardium. Sectional finding.

Etiopathogenesis

The causes of sarcoidosis are unknown, it is probably an a**utoimmune disorder.** Considered triggers include both infectious and non-infectious agents. Genetic predisposition also plays a role.

Pathologically, we observe well-defined granulomatous **inflammatory foci in the myocardium.** These are accompanied by **fibrotization of the heart muscle.** The mentioned changes lead to the development of cardiac dysfunction and heart rhythm disorders. As a result, the disease can progress to restrictive or dilated cardiomyopathy. Histopathological changes are similar to findings in large cell myocarditis.

Epidemiology

Cardiac involvement is found in **5% of patients with sarcoidosis**. However, autopsy specimens show that a **clinically silent cardiac manifestation** is present in 20-25% of patients. The prevalence of sarcoidosis fluctuates between 4.7-64/100 thousand inhabitants with a higher incidence in women. Cardiac sarcoidosis is confirmed in 19–34% of new-onset higher-degree AV blocks of unclear etiology.

Clinical picture

The symptoms of the disease are very variable. It can manifest itself in **syncope, arrhythmias, congestive heart failure** and even sudden death. Of the arrhythmias, AV blocks of a higher degree and ventricular arrhythmias are typical findings. Pulmonary hypertension may develop as a result of lung involvement or heart failure.

Diagnostics

ECG, Holter monitoring (intermittent occurrence of arrhythmias) and echocardiography are used in the first line. These examinations are complemented by 18-FDG PET/CT (positron emission tomography using 18-fluorodeoxyglucose) and MRI.

In patients with already diagnosed extracardiac involvement, a screening examination for possible involvement of the myocardium is performed at **annual intervals.** Screening includes history taking, ECG, Holter monitoring and echocardiography. If any of these examinations is abnormal, a PET/CT or MRI examination is added.

Cardiac sarcoidosis should be considered in patients with higher-degree **AV blocks, persistent ventricular tachycardias, and heart failure of unclear etiology.** As in the previous group, patients here are also examined using MRI or PET/CT. Endomyocardial biopsy may be considered in selected patients. However, it has a low sensitivity (25%) due to the focal nature of the inflammatory disease.

Therapy

Treatment is aimed at suppressing **inflammatory and fibrotic activity, treating cardiac dysfunction and preventing malignant arrhythmias.**

Immunosuppressive treatment

The basis of treatment is the administration of immunosuppressive therapy. In patients, it usually leads to suppression of **inflammatory activity and improvement of the systolic function of the heart.** Other effects are an improvement in AV conduction and a reduction in the risk of ventricular arrhythmias. Due to the considerable mortality of the disease and the risk of relapse after discontinuation of therapy, long-term (mostly

lifelong) **immunosuppression** is recommended. Patients are mainly given prednisone at an initial dose of 30–40 mg/day. In the case of unwanted side effects or a small effect of the treatment, treatment with methotrexate, azathioprine, cyclosporine or other preparations is chosen.

Treatment of heart failure and arrhythmias

Caution is needed in case of use of beta-blockers due to the occurrence of AV blocks and with amiodarone. Class I antiarrhythmics (proarrhythmogenic effect) are completely contraindicated. Heart transplantation is indicated for severe cardiac dysfunction refractory to pharmacotherapy.

A major danger of the disease is severe rhythm disturbances (recurrence of ventricular arrhythmias 44-75%). In certain situations, the administration of antiarrhythmic drugs and the implantation of an ICD (implantable cardioverter-defibrillator) or pacemaker are therefore recommended. Such provision is necessary mainly in patients with **persistent ventricular tachycardias, complete AV block and cardiac dysfunction** refractory to treatment.

Prognosis

Cardiac involvement is a significant predictor of prognosis in patients with sarcoidosis. Most deaths are due to ventricular arrhythmias. In addition to the severity of systolic dysfunction, the presence of fibrosis on magnetic resonance imaging of the heart is also used in the prediction. In patients with severe systolic dysfunction, the prognosis is very unfavorable.

Links

Related articles

- Sarcoidosis (pathology)
- Sarcoidosis (internal)
- Restrictive cardiomyopathy

Recommended literature

- KAUTZNER, Josef. *Heart failure : current affairs for clinical practice.* - edition. Mladá fronta, 2015. ISBN 9788020435736.
- MANN, Douglas L. *Braunwald´s Heart Disease : A Textbook of Cardiovascular Medicine.* 10th Edition edition. 2015. ISBN 978-0-323-29429-4.
- KOPŘIVA, Petr – GŘIVA, Martin – TÜDÖS, Zbyněk. Management of cardiac sarcoidosis - A practical guide. *Cor et Vasa.* 2018, y. 2, vol. 60, p. e155-e164, ISSN 0010-8650. DOI: 10.1016/j.crvasa.2017.05.012 (http://dx.doi.org/10.1016%2Fj.crvasa.2017.05.012).
- BIRNIE, David H. – KANDOLIN, Riina – NERY, Pablo B.. Cardiac manifestations of sarcoidosis: diagnosis and management. *European Heart Journal.* 2016, y. -, vol. -, p. ehw328, ISSN 0195-668X. DOI: 10.1093/eurheartj/ehw328 (http://dx.doi.org/10.1093%2Feurheartj%2Fehw328).
- BIRNIE, David H. – NERY, Pablo B. – HA, Andrew C.. Cardiac Sarcoidosis. *Journal of the American College of Cardiology.* 2016, y. 4, vol. 68, p. 411-421, ISSN 0735-1097. DOI: 10.1016/j.jacc.2016.03.605 (http://dx.doi.org/10.1016%2Fj.jacc.2016.03.605).
- OKADA, David R. – SMITH, John – DERAKHSHAN, Arsalan. Ventricular Arrhythmias in Cardiac Sarcoidosis. *Circulation.* 2018, y. 12, vol. 138, p. 1253-1264, ISSN 0009-7322. DOI: 10.1161/circulationaha.118.034687 (http://dx.doi.org/10.1161%2Fcirculationaha.118.034687).
- ANTON, Jan. Materiály k přednášce "Sarkoidóza".

Reference