Cardiac sarcoidosis

Cardiac sarcoidosis is a granulomatous inflammatory disease of the myocardium. It can occur together with other extracardiac lesions or alone. [1] [2] [3][4] Sarcoidosis itself is a multisystem disease of unknown cause. It usually affects individuals between the ages of 25 and 60.[1]Most often, granulomatous inflammation is located in the **lungs.** The eyes, liver, GIT, skin or nervous tissue can also be affected.[1][2][5]

Cardiac sarcoidosis is manifested by **AV conduction disorders and supraventricular and ventricular arrhythmias.** These are the main dangers of the disease. [1] [3] We also sometimes observe the development of Heart Failure. The presence of cardiac sarcoidosis and the degree of cardiac dysfunction are important predictors of patients' prognosis.[1][3]

Etiopathogenesis

The causes of sarcoidosis are unknown, probably an **autoimmune disorder.** The triggers considered include infectious and non-infectious agents. Genetic predisposition also plays a role.[1]

Pathologically, in the myocardium, we observe well-defined **granulomatous inflammatory deposits.** These are accompanied by **cardiac muscle fibrosis.** It leads to the development of cardiac dysfunction and arrhythmias.[1][2] As a result, the disease can progress to restrictive or dilated cardiomyopathy.[2][4] Histopathological changes are similar to those found in large cell myocarditis.[4] [6]

Epidemiology

Heart disease is detected in **5% of patients with sarcoidosis**. However, autopsy samples show that **clinically silent cardiac manifestations** are present in 20-25% of patients.[1] [2] The prevalence of sarcoidosis varies between 4.7-64 / 100 thousand inhabitants with a higher incidence in women. [4] In newly developed AV block of a higher degree of unclear etiology, cardiac sarcoidosis is confirmed in 19-34%.[1]

Clinical picture

The symptoms of the disease are very variable. It can manifest as **syncope**, **arrhythmia, congestive heart failure** to **sudden death**. Higher degree AV blockages and ventricular arrhythmias are typical of arrhythmias. [2] Due to pulmonary involvement or heart failure, pulmonary hypertension may develop. [2]

Diagnostics

ECG, Holter monitoring (intermittent arrhythmias) and echocardiography are used in the first line. These tests complement 18-FDG PET / CT (18-fluorodeoxyglucose positron emission tomography) and MRI.

Patients with the pre-diagnosed extracardiac disease are screened at annual intervals for possible myocardial involvement. Screening includes medical history, ECG, Holter monitoring, and echocardiography. If any of these tests are abnormal, a PET / CT or MRI scan is added.[1][2][3]

Cardiac sarcoidosis should be considered in patients with **advanced AV block, persistent ventricular tachycardia, and heart failure of unclear etiology.**[2] As in the previous group, there are patients examined by MRI or PET / CT. Endomyocardial biopsy is to be considered in selected patients. However, due to the focal nature of the inflammatory disorder, it has low sensitivity (25%).[2][4]

🔎 *For more information see cardiac sarcoidosis diagnosis details.*

Treatment

The treatment is aimed at **suppressing inflammatory and fibrous activity, treating cardiac dysfunction and preventing malignant arrhythmias.**

Immunosuppressive therapy

The basis of treatment is the administration of immunosuppressive therapy. In patients, it usually **suppresses inflammatory activity and improves systolic heart function.** Other effects are improved AV transmission and **reduced 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 predominantly given prednisone at an initial dose of 30-40 mg / day. In case of undesirable side effects or small treatment effects, treatment with methotraxate, azathioprine, cyclosporine or other preparations is chosen. [1] [4]

Treatment of heart failure and arrhythmias

Caution should be exercised when using beta-blockers due to the occurrence of AV blockages [2] and amiodarone. Class I antiarrhythmics (proarrhythmogenic effect) are completely contraindicated.

Serious arrhythmias (44-75% recurrence of ventricular arrhythmias) are a high risk of the disease. [3] ICD (implantable cardioverter-defibrillator) or Pacemaker. Such provision is necessary especially in patients with **persistent ventricular tachycardias, complete AV block** and **refractory** treatment. [1] [2] [3] [4]

Forecast

Cardiac involvement is an important 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 cardiac magnetic resonance imaging also applies in the prediction. [1] [3]

Links

Related articles

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

Literature

- KAUTZNER, Josef. *Srdeční selhání : news for clinical practice.* - edition. Mladá fronta, 2015. ISBN 9788020435736.
- MANN, Douglas L, et al. *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, vol. 60, no. 2, 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, vol. -, no. -, 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, vol. 68, no. 4, 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, vol. 138, no. 12, p. 1253-1264, ISSN 0009-7322. DOI: 10.1161/circulationaha.118.034687 (http://dx.doi.org/10.1161%2Fcirculationaha.118.034687).
- ANTON, Jan. Materials for the lecture "Sarcoidosis".

References

1. BIRNIE, David H. – KANDOLIN, Riina – NERY, Pablo B.. Cardiac manifestations of sarcoidosis: diagnosis and management. *European Heart Journal.* 2016, vol. -, no. -, p. ehw328, ISSN 0195-668X. DOI: 10.1093/eurheartj/ehw328 (http://dx.doi.org/10.1093%2Feurheartj%2Fehw328).
2. KOPŘIVA, Petr – GŘIVA, Martin – TÜDÖS, Zbyněk. Management of cardiac sarcoidosis - A practical guide. *Cor et Vasa.* 2018, vol. 60, no. 2, 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).
3. BIRNIE, David H. – NERY, Pablo B. – HA, Andrew C.. Cardiac Sarcoidosis. *Journal of the American College of Cardiology.* 2016, vol. 68, no. 4, 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).
4. MANN, Douglas L, et al. *Braunwald´s Heart Disease : A Textbook of Cardiovascular Medicine.* 10th Edition edition. 2015. ISBN 978-0-323-29429-4.
5. HUNNINGHAKE, G W – COSTABEL, U. , et al. ATS/ERS/WASOG statement on sarcoidosis. American Thoracic Society/European Respiratory Society/World Association of Sarcoidosis and other Granulomatous Disorders. *Sarcoidosis Vasc Diffuse Lung Dis* [online]. 1999, vol. 16, no. 2, p. 149-73, Available from <https://www.ncbi.nlm.nih.gov/pubmed/10560120>. ISSN 1124-0490.
6. KUBÁNEK, M. , et al. Giant cell myocarditis and cardiac sarcoidosis – update 2015. *Kardiologická revue.* 2015, vol. 4, no. 17, p. 295-299,