Mediastinal fibrosis

Mediastinal fibrosis (idiopathic mediastinal fibrosis) is of unknown etiology, based on sclerotization of mediastinal connective tissue. It is a rare disease, usually affecting middle-aged and older adults, but can occur at any time. Disabilities of both sexes are about the same.

Origin

The very existence of idiopathic mediastinal fibrosis has been controversial in the past. It was thought to be a secondary condition of, e.g. tuberculosis mediastinal lymphadenitis, syphylis changes, after irradiation, or similar conditions. Another assumption was that it was a rare complication of histiocytosis. It is now thought to be an organ-specific manifestation IgG4-associated disease, at least in some cases. The disease is often associated with other chronic idiopathic sclerosing diseases.

Clinical manifestations

A typical clinical manifestation is the sudden onset of symptoms in full health. Manifestations are initially mild and may be intermittent. Swelling of the upper limbs and face usually occurs first, especially in the morning. Later appears manifestation of upper vena cava syndrome. A conspicuous manifestation is an ashen face, neck and arms of the patient, conjunctival dysfunction, dizziness, tinnitus, headache, epistaxis , hemoptysis, feelings of pressure in the head and intolerance to crowded areas. Manifestations are exacerbated by coughing, bending, exertion or exercise. After 6-9 months, there is usually a significant improvement, but there is no complete remission of symptoms. The development of the collateral vascular bed is probably at least partly responsible for this improvement. The disease is not painful in itself.

Other possible lesions of the mediastinal organs have also been described, which of course differ in the spectrum of manifestations: aortic compression, pulmonary artery obstruction with cor pulmonale development, pulmonary vein stenosis, pericarditis, tracheal obstruction with asthma-like symptoms, esophageal stenosis and obstruction of the coronary arteries.

Diagnosis

Biochemical and serological examination is usually without obvious abnormalities. Bronchoscopic and esophagoscopic examinations usually do not reveal pathology. No pathology may be seen on the chest X-ray, but dilation of the mediastinum may be evident. The actual diagnosis is usually based on the presence of upper vena cava syndrome and the absence of a clinically and radiologically demonstrable cause.

Macroscopically, the sclerotic lesion looks like a solid, white to off-white deposit. The size and shape vary from an ovoid formation with a diameter of only a few centimeters, sometimes even clearly demarcated, to a relatively large "pancake" structure. It can affect the surrounding organs, but it does not grow through.

Histologically, acute and chronic inflammatory changes with distinct fibroproduction can be detected. Acute changes represent an abundant number of inflammatory cells scattered between fibrous fibers. Chronic changes represent fibrous trees relatively low in vascular and low in cellular. So far, only in individual case reports is the presence of a larger number of IgG4 positive plasma cells in the lesion described.

Therapy

Therapy consists primarily of eliminating potential malignancies or infections, discontinuing drugs that may have mediastinal fibrosis as a potential side effect, and corticotherapy. In the case of corticoid resistance, more emphatic immunosuppression is appropriate . Surgical treatment of complications comes into play in patients with an inadequate response to immunosuppression, but i.e. Barrett (1958) notes that surgical therapy brings nothing but perioperative mortality.

Long-term observation of patients is necessary, relapse was observed in the interval from 3 months to 10 years. The disease itself does not pose a significant risk of shortening life. A life-threatening complication can be thrombosis of the superior vena cava.

Sources

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