

Sjögren's syndrome

Sjögren's syndrome is an autoimmune disease that is chronic and slowly progressing. It manifests itself as a diffuse, chronic inflammation with the destruction of exocrine glands (salivary, lacrimal, or exocrine glands of the pancreas). We divide it into **primary** - separate disease and **secondary** - in another disease of an autoimmune nature (RA, SLE, MCTD - mixed connective tissue disease, systemic vasculitis).

Etiology

The etiology is idiopathic, there is a possible relation of viruses (EBV, CMV), association in the HLA system.

Clinical characteristics

Salivary glands

Swelling of the salivary glands, xerostomia (dry mouth) and dysphagia.

Eye affection

Due to insufficient tear production, a picture of dry keratoconjunctivitis (burning, foreign body sensation in the eye, photophobia), **xerophthalmia is formed**.

Other exocrine glands affection

- Decreased sweat production, dry skin, pruritus, loss of adnexa, **xerodermia**;
- achlorhydria gastritis, pancreatitis, hepatopathy;
- vaginal and vulvar epitosclerosis, tendency to candida infections;
- chronic bronchitis, irritating cough, interstitial pneumonia;
- vasculitis (petechiae), Raynaud's phenomenon;
- non-erosive, non-deforming arthritis .

Diagnosis

Elevated levels of FW, CRP, γ -globulins, ENA autoantibodies type SS / A and SS / B, rheumatoid factor (RF), antinuclear antibodies. Diagnosis is determined if 4 criteria are present (if 3, dg. Is also possible):

1. dry keratoconjunctivitis (demonstrated by Schirmer's test);
2. xerostomia;
3. biopsy of lymphocyte infiltrates in salivary glands; It is necessary to monitor - possible transition to lymphoproliferative disease (lymphoma). Monitoring of beta₂-microglobulin levels serologically.
4. detection of serum autoantibodies (rheumatoid factor, antinuclear antibodies, ENA antibodies).

Therapy

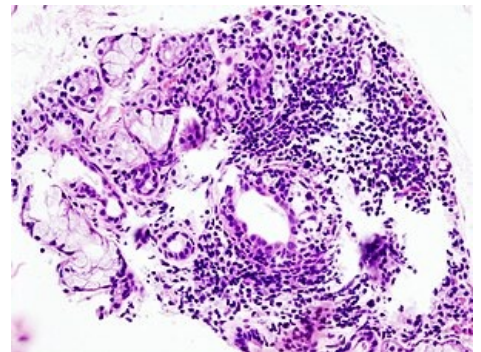
Mostly symptomatic:

- Substitution therapy: eye drops, lubricating gels, moisturizing lotions, chewing gum. The secondary requires proper treatment of the underlying disease:
- Antimalarials (hydroxychloroquine)
- Glucocorticoids + combinations with other immunosuppressive drugs (methotrexate, prednisone + cyclophosphamide).
- Careful dental care is important.

Prognosis

Patients with Sjögren's syndrome are limited in several respects. In the secondary form, the risk of malignancies increases. The most common malignancies are lymphoid malignancies - in about 5% ^[1] the most common are MALTomas and B-cell lymphomas of the salivary gland. ^[2] Despite this, the average age of patients with sicca syndrome is the same as in the normal population.

References



Biopsy - salivary gland (lymphocyte infiltration in the gland)

Related Articles

- Autoimmune diseases
- Sialoadenitis
- Pulmonary manifestations in systemic connective tissue diseases

Sources

- KLENER, Pavel. *Vnitřní lékařství*. 3. edition. Praha : Galén : Karolinum, 2006. pp. 1158. ISBN 80-7262-430-X.
- FUČÍKOVÁ, Terezie. *Klinická imunologie v praxi*. 2. edition. Praha : Galén, 1997. pp. 339. ISBN 8085824574.

Reference

1. TZIOUFAS, Athanasios G – VOULGARELIS, Michael. Update on Sjögren's syndrome autoimmune epithelitis: from classification to increased neoplasias. *Best Practice and Research Clinical Rheumatology* [online]. 2007, vol. 21, no. 6, p. 989-1010, Available from <<https://www.ncbi.nlm.nih.gov/pubmed/18068857>>. ISSN 1521-6942. PMID: 18068857 (<http://www.ncbi.nlm.nih.gov/pubmed/18068857>).DOI: 10.1016/j.berh.2007.09.001 (<http://dx.doi.org/10.1016%2Fj.berh.2007.09.001>).
2. VOULGARELIS, Michael – SKOPOULI, Fotini N. Clinical, immunologic, and molecular factors predicting lymphoma development in Sjogren's syndrome patients. *Clinical Reviews in Allergy and Immunology* [online]. 2007, vol. 32, no. 3, p. 265-74, Available from <www.ncbi.nlm.nih.gov/pubmed/17992593>. ISSN 1080-0549. PMID: 17992593 (<http://www.ncbi.nlm.nih.gov/pubmed/17992593>).DOI: 10.1007/s12016-007-8001-x (<http://dx.doi.org/10.1007%2Fs12016-007-8001-x>).

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