Orbital pseudotumour

Orbital pseudotumour (idiopathic orbital pseudotumour, idiopathic orbital inflammatory syndrome) is a benign inflammatory disease of the orbit without demonstrable local or systemic cause. It usually manifests clinically as a painful mass in the orbit, but more or less diffuse orbital involvement is also possible.

Epidemiology

It is the third most common orbital disease in adults, accounting for approximately 5 to 10% of orbital lesions. In terms of frequency of occurrence, it is nevertheless a relatively rare disease. The average age at diagnosis corresponds to the fifth decade, but it can occur at any age. The disease does not tend to show significant differences in incidence between the sexes.

Pathology

The actual mechanism of disease development is unknown, but is very likely mediated by the immune system. A number of infectious and environmental factors may be triggering factors for the disease. At least some cases are likely to be organ-specific manifestations of IgG4-associated disease.

Histopathological findings usually show a mixed cellular infiltration consisting of small lymphocytes, plasma cells, histiocytes, macrophages and neutrophils. Fibrosis is not uncommon; in some cases, fibrosis may be exaggerated and **sclerosing orbital pseudotumour** is then reported. Some authors consider sclerosing pseudotumour to be a separate clinical entity because it usually spreads to adjacent structures. In some cases, the pseudotumor may calcify focally, and then we can speak of a **calcifying orbital pseudotumor.**

According to the nature of the histopathological picture, three forms of pseudotumour can be distinguished:

- **lymphoid form,**
- **the granulomatous form,**
- **sclerosing form**.

Clinical picture

Orbital pseudotumor can manifest as acute, subacute, or chronic orbital involvement; the clinical picture depends on the specific localization and the intensity of the ongoing inflammation. Thus, the clinical picture may include a variety of manifestations such as proptosis (exophthalmos), diplopia, conjunctival chemosis, visual disturbances or limitation of ocular motility. Characteristic manifestations are unilateral periorbital pain, paresis of the relevant cranial nerves and dramatic improvement after corticosteroid administration.

Typical manifestations of lacrimal gland involvement are local pain, tenderness and swelling of the eyelid. Lymphoma, sarcoidosis, Wegener's granulomatosis and solid tumour are the main differential diagnosis.

Typical manifestations of scleral involvement are orbital pain and eyelid swelling and inflammation. In the differential diagnosis, infection and orbital trauma with a foreign body are mainly considered.

Typical manifestations of extrabulbar muscle involvement are orbital pain, proptosis, conjunctival edema, diplopia and limitation of ocular motility. In the differential diagnosis, endocrine orbitopathy and vasculitis come into particular consideration.

Typical manifestations of optic nerve sheath involvement are visual disturbance of varying degrees and color-vision disturbance with varying degrees of pain. In the differential diagnosis, optic neuritis and optic nerve meningioma are particularly important.

Typical manifestations of orbital apex involvement are orbital apex syndrome or painful ophthalmoplegia with minimal proptosis. In the differential diagnosis, Tolosa Hunt syndrome, lymphoma, glioma, and metastatic involvement are particularly relevant.

Diagnosis

Diagnosis is usually per exclusionem, supported by clinical picture and radiological findings. High-resolution computed tomography (HR-CT) can demonstrate soft tissue oedema, but is not as sensitive as magnetic resonance imaging, especially MRI with contrast agent. A relatively characteristic feature is the rapid response to corticosteroid therapy.

It is advisable to rule out causes of similar changes, especially autoimmune disorders (autoantibody testing, serum electrophoresis). Examination of cerebrospinal fluid is not diagnostically useful except when lymphoma is suspected. Biopsy makes sense only in very exceptional cases, e.g., in the case of rapid progression of neurological deficit, unresponsiveness to corticosteroids, or marked abnormalities on imaging findings.

Differential diagnosis

The more frequent endocrine orbitopathies in thyroid disorders differ from pseudotumor primarily in having milder pain, a more bilateral onset, a slow onset, and little response to corticosteroids.

Acute bacterial cellulitis may mimic the manifestations of orbital pseudotumour, especially with its rapid onset, severe pain and unilateral appearance. Patients with cellulitis are usually febrile, a more pronounced systemic inflammatory reaction is evident, and a different picture on magnetic resonance imaging is demonstrated.

Therapy and prognosis

Corticosteroids are an essential therapeutic modality. A characteristic feature of orbital tumour is that the response is truly dramatic, e.g. proptosis disappears within 24-48 hours of starting therapy. Prednisone is used at a dose of 60 to 100 mg per day followed by a slow to months-long withdrawal. Because of the small number of patients, clinical trials are problematic, but the optimal regimen appears to be two weeks of 60 mg prednisone in a single daily oral dose followed by several months of dose reduction.

After corticotherapy, a response to therapy can be expected in about four-fifths of patients, with complete cure in about one-third of patients and relapses in the remainder. If fibrosis develops, the response is worse. Surgery is often considered when there is an inadequate response to corticotherapy.

Other modalities such as radiotherapy and chemotherapy are also being tried, but data on their effectiveness are limited by the very small number of published studies.

Links

Literature

- CULVER, E.L. a A.C. BATEMAN. General principles of IgG4-related diseace. *Diag Histopatol.* 2013, vol. 19, no. 4, s. 111-118, ISSN 1756-2317.
- DING, Z.X., G. LIP a V. CHONG. Idiopathic orbital pseudotumour. *Clin Radiol.* 2011, vol. 66, no. 9, s. 886-92, ISSN 1365-229X.
- JACOBS, D. a S. GALETTA. Diagnosis and management of orbital pseudotumor. *Curr Opin Ophthalmol.* 2002, vol. 13, no. 6, s. 347-51, ISSN 1040-8738.
- SZABO, B., I. SZABO a D. CRIŞAN, et al. Idiopathic orbital inflammatory pseudotumor: case report and review of the literature. *Rom J Morphol Embryol* [online]. 2011, vol. 52, no. 3, s. 927-30, dostupné také z <http://www.rjme.ro/RJME/resources/files/520311927930.pdf>. ISSN 1220-0522.

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