

CASE REPORTS

SCLEROSING INFLAMMATORY PSEUDOTUMOR OF THE EYELID: A CLINICAL CASE REPORT

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ABSTRACT

INTRODUCTION: Sclerosing inflammatory pseudotumor is a rare benign condition that mimics a malignant lesion or an inflammatory process. Its occurrence on the eyelids is an exception. It is of utmost importance to recognize its possible clinical manifestations, therapeutic approaches, and expected prognosis.

AIM: The aim of this case report is to present the pathomorphological characteristics, clinical findings, imaging studies, differential diagnosis, and therapeutic management of a sclerosing inflammatory pseudotumor mimicking a hordeolum (stye) localized in the upper eyelid.

METHODS: The study involved a 30-year-old patient with an eyelid abnormality. The initial contact with the participant occurred in an outpatient setting, followed by hospitalization, during which clinical examination, biometric and imaging studies were performed. These included visual acuity testing, intraocular pressure measurement using a pneumotonometer, biomicroscopy, ophthalmoscopy, perimetry, and optical coherence tomography (OCT). Additional laboratory tests were also conducted, including the Wassermann test and T-SPOT.TB. After computed tomography (CT) of the orbits with intravenous contrast and magnetic resonance imaging (MRI) with contrast enhancement, a biopsy was performed and histological material was examined. A diagnosis of sclerosing inflammatory pseudotumor was established, and further tests were carried out—c-ANCA, p-ANCA, and antiphospholipid antibodies. Treatment was continued in a hospital setting.

RESULTS: Following clinical examination, a granuloma, cyst, or tumor process was suspected. Imaging studies suggested the possibility of a lacrimal lymphoma, inflammatory pseudotumor, or pleomorphic adenoma. Laboratory investigations ruled out tuberculosis, syphilis, and vasculitic etiology. The histological result confirmed a sclerosing inflammatory pseudotumor.

DIAGNOSIS: Sclerosing inflammatory pseudotumor.

TREATMENT: The patient was hospitalized and underwent conservative treatment.

CONCLUSION: After extensive investigations, surgical intervention and removal of the lesion were avoided. The patient was successfully treated conservatively.

Keywords: orbit, hordeolum, chameleon, histology, sclerosing inflammatory pseudotumor, eyelid

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INTRODUCTION

Sclerosing inflammatory pseudotumor (SIP) is a term describing a benign and rare condition that can occur in any organ of the human body. Its most common locations are the lungs and the orbit. In 1954, Umiker and Iverson introduced the term *inflammatory pseudotumor*, because its clinical and



imaging features mimic those of malignant tumors (1).

Informed consent was obtained for the use of the presented materials in this article.

1. Medical History

A 30-year-old man was admitted for the first time to the University Specialized Hospital for Active Treatment of Eye Diseases in Varna with complaints of gradually increasing swelling of the upper right eyelid, leading to drooping of the eyelid, pain, and reduced vision in the affected eye, particularly when the forehead and face were relaxed. He was referred for surgical treatment of a hordeolum by another medical specialist. The patient reported no trauma, recent illnesses (fever, chills, weight loss), or injuries. The symptoms began 3–4 weeks prior as mild discomfort in the area of the upper right eyelid and progressively worsened over time. No signs of respiratory tract infection, joint pain, morning stiffness, or neurological deficits were found. He denied smoking, regular alcohol consumption, or any comorbid conditions. The patient had a history of glaucoma in the left eye during childhood, for which he underwent anti-glaucoma surgery—trabeculectomy. At the time of admission, he was not undergoing any treatment.

2. Clinical Examination Results

The clinical examination revealed visual acuity of 20/20 in both eyes, intraocular pressure (IOP) of 16.7 mmHg in the right eye and 15.6 mmHg in the left eye (normal range: 10–21 mmHg), with corneal pachymetry showing 523 μ m in the right eye and 541 μ m in the left eye (normal range: 522–574 μ m). Ptosis of the right upper eyelid was observed (MRD1 = -1 mm, normal range: 3–4 mm), causing visual field obstruction in the right eye when the face is at rest. The upper eyelid was swollen and hyperemic, while the conjunctiva showed no signs of chemosis or injection. The anterior ocular surface was smooth and shiny; the cornea was clear, without epithelial defects or infiltrates. Both direct and consensual pupillary reactions were normal. Ocular motility was normal. No significant abnormalities were observed in the anterior segment, although a mild anterior chamber reaction was noted.

Fundus examination showed a vital optic disc, macula with a clear reflex, normal vessel course and

caliber, and an unremarkable peripheral retina. No functional or structural abnormalities were found on specialized tests (computerized perimetry and optical coherence tomography) of peripheral vision, the optic nerve, or the retinal ganglion cell layer.

Due to the elevated position of the lesion, differential diagnosis included: granuloma, cyst, or tumor. Therefore, imaging studies were performed—CT scan of the orbits with intravenous contrast and MRI with contrast enhancement.

3. Imaging Studies

The CT scan revealed a parenchymal mass in the area of the right lacrimal gland, adjacent to approximately one-third of the lateral contour of the right eyeball. The average density was around 40 Hounsfield Units (HU), increasing to 60 HU in the arterial phase and approximately 75 HU in the late phase. The bony structures and paranasal sinuses appeared normal, as did the ocular globes and extraocular muscles. Part of the lesion extended slightly into the upper right retrobulbar space. The initial conclusion suggested a probable lacrimal lymphoma, with a recommendation for MRI and PET/CT for confirmation and further staging.

Subsequent MRI with contrast enhancement revealed that the lesion in the right lacrimal gland was isointense on T1 and T2, hypointense on FLAIR, with a slightly heterogeneous structure. The transverse dimensions of the right lacrimal gland measured 10 mm compared to 4 mm on the left side, and the longitudinal size was 28 mm. After contrast administration, moderate hyperintensity and increased heterogeneity of the structure were noted. The retrobulbar spaces appeared normal. The lateral rectus muscle was slightly displaced inward near the lesion. The ocular globes, optic nerves, and other extraocular muscles were unremarkable. The conclusion pointed toward a likely inflammatory pseudotumor or pleomorphic adenoma, with lymphoma considered significantly less probable based on imaging data.

4. Laboratory Tests

Additional tests were performed to clarify the etiology of the lesion:

- ◆ Wassermann test (syphilis): negative
- ◆ T-SPOT.TB (tuberculosis): negative

- ◆ Complete blood count with differential was within normal limits.
- ◆ ESR = 2 mm/h
- ◆ Bleeding time and clotting time: normal
- ◆ Urea: normal

5. Histological Examination

An interdisciplinary decision was made to refer the patient for a biopsy and histopathological examination to determine the etiology of the lesion in the right upper eyelid.

The biopsy revealed, macroscopically, a rounded lesion about the size of a penny, dense and firm in consistency, attached to the underlying tarsus. Tissue was taken for histopathological analysis, including a small sample from the lacrimal gland.

The histological result confirmed a sclerosing inflammatory pseudotumor.

Further testing was recommended for c-ANCA, p-ANCA, and antiphospholipid antibodies to rule out a vasculitic etiology—all results were negative.

DIAGNOSIS

Based on the clinical presentation, imaging studies (Fig. 1), laboratory results, and histopathological findings (Fig. 2), the patient was diagnosed with an orbital pseudotumor. Supporting this diagnosis were signs of orbital inflammation, normal intraocular pressure, absence of optic nerve involvement, and lack of elevated inflammatory markers.



Fig. 1. A 30-year-old man with swelling of the right upper eyelid and pain for 1 month.

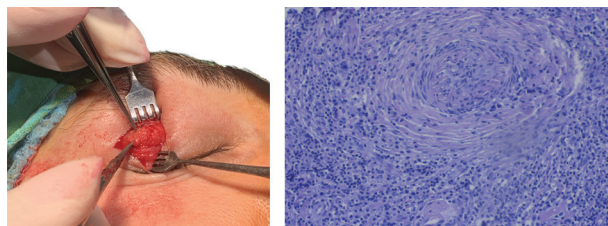


Fig. 2. Biopsy material taken from the right supraorbital area of the patient (A), illustrating a picture of granulomatous inflammatory process (B).

Informed consent was obtained for the presentation of the current materials in this article.

TREATMENT

The patient was hospitalized and underwent conservative treatment with corticosteroids: methylprednisolone intravenously according to the following regimen:

- ◆ 500 mg for 3 days i.v.
- ◆ 1.5 mg/kg for 3 days i.v.
- ◆ 1 mg/kg for 3 days i.v.
- ◆ 60 mg for 3 days orally (p.o.)
- ◆ 40 mg for 3 days orally (p.o.)
- ◆ 20 mg for 15 days orally (p.o.)

During continuous monitoring and multiple IOP measurements—considering the patient's history of childhood glaucoma and ongoing corticosteroid therapy—an improvement in the condition was observed without changes in IOP.

One month after starting treatment, the patient reported that the pain had disappeared. There was no eyelid swelling or visual disturbance. Resolution of eyelid edema and hyperemia was noted; the anterior chamber was clear with normal depth, and visual acuity was 20/20 without the need for forehead elevation. Laboratory blood tests were within normal limits. A follow-up MRI was recommended to confirm regression of the condition. Surgical intervention was avoided.

DISCUSSION

Sclerosing inflammatory pseudotumor can present as a solitary mass or as multiple masses with a polymorphic inflammatory cellular infiltrate, exhibiting varying degrees of fibrosis, necrosis, granulomatous reaction, and myofibroblastic spindle cells.

Although imaging studies of SIP may be non-specific, they are extremely important for preoperative differential diagnosis, as they can help avoid unnecessary surgical interventions (2).

Pathogenesis and Clinical Manifestation of SIP

Histologically, SIP is composed of acute and chronic inflammatory cells (lymphocytes, plasma cells, neutrophils), myofibroblastic spindle cells, and collagen (fibrotic reaction). The etiology of SIP remains unclear. Possible causes include trauma, iat-

rogenically induced inflammation, autoimmune response, or an inflammatory myofibroblastic tumor (3).

Some SIPs are associated with IgG4-related sclerosing disease, a systemic condition characterized by the presence of IgG4-positive plasma cells and T-cell infiltration in various tissues. This condition may manifest as autoimmune pancreatitis, sclerosing cholangitis, cholecystitis, sialoadenitis, retroperitoneal fibrosis, tubulointerstitial nephritis, interstitial pneumonia, prostatitis, and lymphadenopathy.

The condition can be differentiated from lymphoma, as SIP contains both T and B cells, while lymphoma typically involves clonal proliferation of only one cell type. Some pseudotumors are considered to be a secondary reaction following infection. Interleukin-1, a cytokine produced mainly by monocytes and macrophages, contributes to the local and systemic response to this type of pseudotumor.

Clinically, patients may present with fever, weight loss, thrombocytosis, iron deficiency anemia, hypergammaglobulinemia, symptoms related to the mass effect, or a combination of these signs (1).

Patients are usually in their second or third decade of life and generally in good overall health. Exophthalmos has a relatively acute onset and, in half of the cases, presents with moderate to severe pain or with eyelid and conjunctival edema. Pain typically responds well to corticosteroid therapy, although some sclerotic cases are resistant to steroids.

Diplopia is a common symptom, resulting from restricted ocular motility in one or more directions of gaze, although visual acuity is preserved. Elevated intraocular pressure may also be present (4).

The lesion is often palpable through the eyelid, making it easily accessible for biopsy by the surgeon. If the lesion is located more deeply and the clinical and imaging findings point toward pseudotumor, biopsy may be avoided. In such cases, corticosteroid treatment is initiated directly, which usually results in dramatic relief of symptoms and signs. CT imaging is used to help localize the lesion.

Differential Diagnosis

In differential diagnostic terms, SIP is a diagnosis of exclusion. Conditions that must first be ruled out include: lymphoma, vasculitis, infection, systemic lupus erythematosus, sarcoidosis, Wegener's gran-

ulomatosis, Rosai-Dorfman disease, Erdheim-Chester disease, rhino-orbito-cerebral mucormycosis (an ocular infection often leading to orbital exenteration), and Waldenström macroglobulinemia (5).

CONCLUSION

Sclerosing inflammatory pseudotumor presents with a wide range of clinical features that can mimic other disease entities, including neoplastic processes. Imaging studies are nonspecific, and the lesion can occur in virtually any tissue of the human body. It should be considered in the differential diagnosis of any similar lesion. Diagnosis is usually confirmed histologically, and in some cases, surgical intervention is unnecessary.

The localization of SIP on the eyelid is rare. The literature primarily describes findings in the liver, lungs, and kidneys, but not on the eyelid. In this case, the patient responded successfully to conservative treatment—a systemic corticosteroid regimen.

It is important to recognize the specific diagnostic features of SIP to avoid unnecessary surgical procedures.

REFERENCES

1. Ding ZX, Lip G, Chong V. Idiopathic orbital pseudotumour. *Clin Radiol*. 2011;66:886–92. doi:10.1016/j.crad.2011.03.018.
2. Espinoza GM. Orbital inflammatory pseudotumors: etiology, differential diagnosis, and management. *Curr Rheumatol Rep*. 2010;12:443–7. doi:10.1007/s11926-010-0128-8.
3. Chaudhry IA, Shamsi FA, Arat YO, Riley FC. Orbital pseudotumor: distinct diagnostic features and management. *Middle East Afr J Ophthalmol*. 2008;15:17–27. doi:10.4103/0974-9233.53370.
4. Jacobs D, Galetta S. Diagnosis and management of orbital pseudotumor. *Curr Opin Ophthalmol*. 2002;13:347–51. doi:10.1097/00055735-200212000-00001
5. Yan J, Wu Z, Li Y. The differentiation of idiopathic inflammatory pseudotumor from lymphoid tumors of orbit: analysis of 319 cases. *Orbit*. 2004;23:245–54. doi:10.1080/01676830490487980.