CASE REPORT

Ulcerated conjunctival mass as an initial presentation of idiopathic orbital inflammatory disease: a case report

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Abstract

A 62-year-old Chinese woman, with a history of immunoglobulin A nephropathy, presented with an ulcerative conjunctival lesion and anterior orbital inflammation. No infective foci were found, and her symptoms were not alleviated by antibiotic treatment or drainage of the lesion. She was diagnosed with idiopathic orbital inflammatory disease, also known as orbital pseudotumor, which completely resolved with oral corticosteroid treatment. To the best of our knowledge, this is the first documented case of idiopathic orbital inflammatory disease that initially presented with an ulcerative conjunctival lesion. We illustrate our stepwise diagnostic approach and management of the disease.

Key words: Administration and dosage; Conjunctival disease; Diagnostic imaging; Drug therapy; Glucocorticoids; Orbital pseudotumor

Introduction

Idiopathic orbital inflammatory disease (IOID), also known as nonspecific orbital inflammation or orbital pseudotumor, is a benign, non-infectious, inflammatory condition of the orbit.[1] Its clinical presentation varies but generally includes periocular erythema, proptosis, chemosis, pain, extraocular motility limitation, and visual disturbance.[2] IOID can be categorized based on the predominantly inflamed anatomical area into anterior, posterior, diffuse, lacrimal, and myositic.[3] The anterior and diffuse subtypes are characterized by pain, eyelid swelling, ptosis, and occasionally distinct inflammation of the globe, presenting as uveitis, papillitis, optic neuropathy, and exudative retinal detachment. To the best of our knowledge, there has been no report of IOID that initially presented as ulcerative conjunctival lesions. Here, we present this atypical IOID case and illustrate our stepwise diagnostic approach and management of the disease.

Case presentation

In February 2022, a 62-year-old Chinese woman presented with a 5-day history of painful swelling of the left upper and lower lids and blurred vision. She denied any history of ocular or facial trauma, dental procedures, or sinusitis symptoms. Visual acuity of her left eye was finger-counting only. Intraocular pressure (IOP) was raised to 25 mmHg. There were diffuse swelling of the left upper and lower lids and copious discharge. There were a 4-mm proptosis and limitation of extraocular motility in all directions. A firm ulcerative conjunctival mass in the inferior bulbar and forniceal conjunctiva was noted (**Figure 1**). Her right eye

Figure 1. A firm ulcerative lesion located in the inferior bulbar and forniceal conjunctiva of the left eye.

was unaffected, with visual acuity of 1.0, normal IOP, and no orbital inflammation.

Salient past medical history included immunoglobulin A (IgA) nephropathy, which had been confirmed by a renal biopsy and immunofluorescence 9 years before the current presentation when she developed hematuria and acute renal failure. She had been treated with systemic corticosteroids and immunosuppression for 3 years until she refused further medication because she had general malaise and fatigue that she perceived to be related to immunosuppression. No relapse occurred after she stopped treatment. The patient had recurrent skin ulcers on her neck, trunk, hands, limbs, and axillary region, requiring multiple excisions over the past decade. Histopathology of the lesions was ulceration or granulation tissue. She had no history of oral or genital ulcers, inflammatory joint pain, abdominal pain, abnormal bowel output, or skin rash/purpura.

She was admitted for computed tomography and was administered empirical intravenous ceftriaxone and metronidazole. She remained afebrile although there was mild leukocytosis and elevated anti-nuclear antibody at a titer of 1:160. Test results for anti-neutrophil cytoplasmic antibodies and immunoglobulin G4 were negative. Computed tomography of the orbits, brain, and paranasal sinuses revealed increased fat stranding in the left anterior orbit and a gaseous rim-enhancing collection, suggestive of left orbital cellulitis with anterior orbital abscess formation without retrobulbar extension (**Figure 2**).

The lesion was incised and drained through the left lower lid. However, the patient developed orbital compartment syndrome, which was not alleviated by topical or systemic pressure-lowering medications. A lateral canthotomy and cantholysis was performed to reduce the orbital pressure. The patient's ears, nose, and throat were examined, and no signs of sinusitis or underlying infective foci were found.

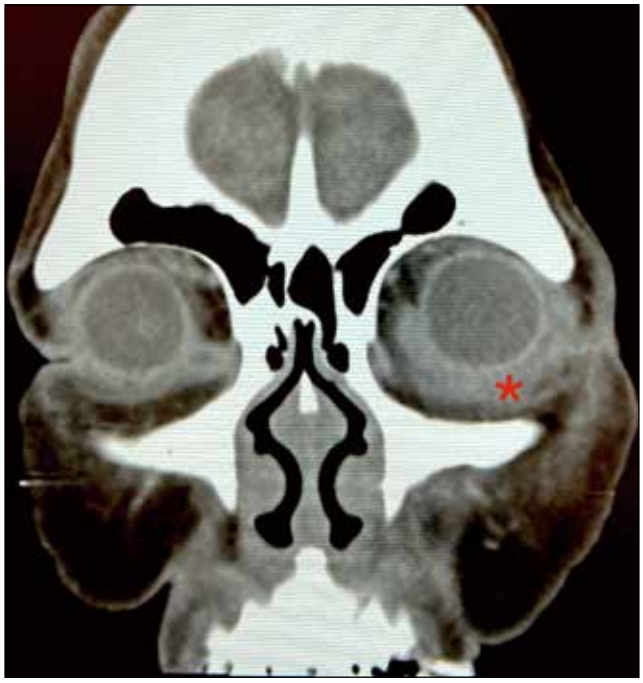

Figure 2. Computed tomography showing increased fat stranding of heterogenous density in the left inferior orbit (asterisk), soft-tissue thickening in the left periorbital region, and clear paranasal sinuses.

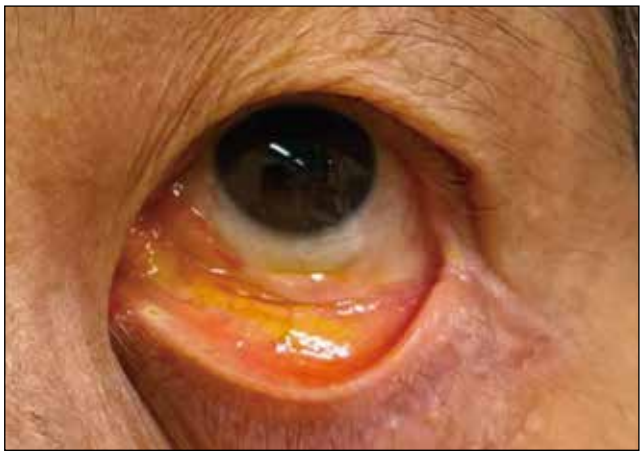

Figure 3. At 2 months after oral steroid treatment, the left eye showing a marked reduction in orbital inflammation and complete resolution of the ulcerative conjunctival lesion.

The conjunctival mass was biopsied; histological analysis showed chronically inflamed tissue but no evidence of underlying infection. Given the patient's limited improvement despite empirical antibiotic treatment, a negative eye swab, the abscess having been drained, and the absence of other active infective foci, she was administered 0.5 mg/kg of oral prednisone. Her pain, swelling, vision, and ocular motility rapidly improved and the conjunctival lesion gradually resolved (**Figure 3**). The oral prednisolone

regimen was tapered off within a month. Her visual acuity returned to 0.5 in 1 month and to 0.7 in 3 months. Residual corneal punctuate erosion and dry eye disease were treated with lubricating eye drops.

Discussion

IOID covers a broad spectrum of inflammatory disease and affects various locations of the orbit. Its variable nature poses a diagnostic and therapeutic challenge. IOID can be classified by the timing of presentation (acute, subacute, chronic), histopathology (classic, granulomatous, sclerotic, non-specific), or the main location (anterior orbit, posterior orbit, diffuse, lacrimal, and myositic).[3,4] However, none of the classifications has described a conjunctival ulcerative lesion.

IOID mainly affects middle-aged women.[5] The main symptom of anterior IOID is external inflammation. Patients often present with palpebral erythema accompanied by painful swelling, conjunctival hyperemia and chemosis, and limited ocular motility. Scleral involvement is possible.[6] Although our patient's presenting symptoms and signs were consistent with the classic description of anterior IOID, the conjunctival ulcerative lesion was peculiar. Differential diagnoses of an ulcerated conjunctival mass include neoplastic, autoimmune, and infective pathologies. Ocular surface tumors may present as either melanotic or non-melanotic conjunctival mass, which can undergo ulcerative changes.[7] Conjunctival ulcers are seen in autoimmune disorders (such as Behcet's disease[8] and Crohn's disease[9]) and in infections (such as tuberculosis and herpes simplex).[10] Although there are no prior reports of ulcerative conjunctival lesions in IOID or IgA nephropathy, autoimmune diseases (such as Crohn's disease, systemic lupus erythematosus, sarcoidosis, rheumatoid arthritis, and ankylosing spondylitis) are associated with IOID.[1] In fact, 10% to 84% of patients with IOID have a history of autoimmune or atopic disorders.[1] However, presentation of ulcerative conjunctival lesions as a sign of IOID in a patient with IgA nephropathy has never been reported.

IgA nephropathy is a common cause of glomerulonephritis.[11] It is characterized by clinically microscopic or macroscopic hematuria and, less commonly, acute renal failure and IgA antibody deposits in the glomeruli. Treatments with systemic corticosteroids or immunosuppressive agents have mixed results.[11] IgA vasculitis with nephritis involves IgA deposits in the glomeruli. It predominantly affects children and adolescents but can present at any age. It is characterized by purpuric rash over the lower extremities without thrombocytopenia or coagulopathy, with some lesions potentially becoming ulcerative.[12] Other clinical features include polyarthralgia, abdominal pain, and in 30% to 50% of patients, nephritis,[11] but presentation of conjunctival ulcerative lesions has never been described in either IgA nephropathy or IgA vasculitis with nephritis.

Systemic corticosteroids are first-line treatment for IOID, with a 75% response rate.[1] IOID can be diagnosed by a response to corticosteroids, which negates the need for a surgical biopsy of inflamed orbital tissues.[13] However, corticosteroids can alter cellularity and compromise diagnostic accuracy.[1] Furthermore, high-dose corticosteroids can deceptively improve both infective and inflammatory pathologies.[14] In our patient, we effectively managed and ruled out infective etiologies before commencing a course of oral corticosteroids; the patient responded favorably with complete resolution of the orbital inflammation and conjunctival lesion.

Conclusion

We successfully treated with oral corticosteroids an atypical IOID that initially presented as an ulcerative conjunctival lesion and anterior orbital inflammation. We illustrate our stepwise diagnostic approach and management of the disease. Further histopathological studies may elucidate the pathological association between IOID and ulcerative conjunctival lesions.

Contributors

All authors designed the study, acquired the data, analyzed the data, drafted the manuscript, and critically revised the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

As an editor of the journal, HKLY was not involved in the peer review process. Other authors have disclosed no conflicts of interest.

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Data availability

All data generated or analyzed during the present study are available from the corresponding author on reasonable request.

Ethics approval

The patient was treated in accordance with the tenets of the Declaration of Helsinki. The patient provided written informed consent for all treatments and procedures and for publication.

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