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Orbital pseudotumor after an upper respiratory infection: a comprehensive review

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Idiopathic orbital inflammation;
Orbit;
Periorbital edema;
Eye pain;
Extraocular restrictions;
Proptosis;
Upper respiratory infection;
Steroids

Abstract

BACKGROUND: Idiopathic orbital inflammation (IOI), also known as *orbital pseudotumor*, is a nonspecific orbital inflammation of unknown etiology. IOI can clinically mimic many other orbital pathologies, some of which can be life-threatening, as in the case of orbital cellulitis. Thus, it is imperative for the clinician to emergently arrive at the correct diagnosis. In many cases, however, IOI presents as a clinical and therapeutic challenge, and conclusive diagnosis is only confirmed after all other etiologies have been ruled out.

CASE REPORT: A 63-year-old man presented urgently with a red, proptotic, painful eye. After history, blood tests, radiologic testing, and ruling out other emergent etiologies, such as orbital cellulitis, the patient was placed on oral steroids. He responded immediately to the steroid treatment. However, the patient did have a recurrence. With the second IOI episode, the patient noted, as previously, a preceding upper respiratory infection. This case may possibly show an association between an infectious trigger leading to a nonspecific aberrant immune response in the orbit.

CONCLUSION: IOI is a difficult condition to diagnose and treat. After ruling out other orbital pathologies, it is appropriate to begin treatment with oral steroids. In this case report, the patient noted an upper respiratory infection before each episode of IOI. Although there is no proof of cause, there is a strong case for the consideration of a viral respiratory infection leading to IOI.

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In 1905, Birch-Hirschfeld described an "orbital mass clinically mistaken for a neoplasm that was histologically inflammatory" in nature and coined the term *orbital pseudotumor.*[1] This term continues to be accepted and used widely; however, it is a misrepresentation of the disease process. Recent literature prefers to call this disease entity *idiopathic nonspecific orbital inflammation*. Therefore, for the remainder of this report, idiopathic orbital inflammation (IOI) will be used in place of orbital pseudotumor.[1]

IOI is a nonmalignant orbital inflammation without a known local or systemic cause.[2] It is the third most common orbital disorder after Graves' disease and lymphoproliferative disease.[3] IOI is a diagnosis of exclusion; therefore, it is imperative to arrive at the correct diagnosis. Diagnosis is complicated by the diverse range of disease entities that can mimic IOI, some life-threatening, such as orbital cellulitis. A thorough history, specific laboratory tests tailored to diagnostic suspicion, radiologic evaluation, response to steroid treatment, and, in limited cases, biopsy, are essential in making an accurate diagnosis.[2,4]

The etiology remains unknown, with possible speculation that infection, immune mediation, or trauma, may trigger IOI or lead to an aberrant wound-healing response.[5] We present a case of IOI in a 63-year-old man who has

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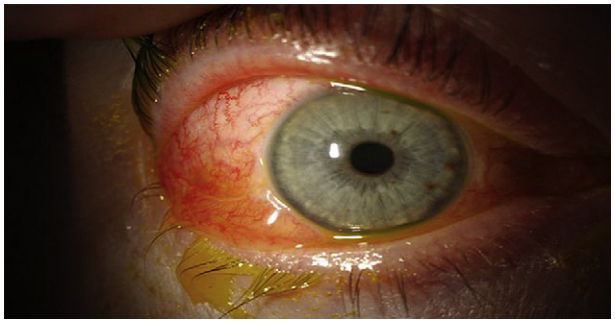

Figure 1 Red eye: unchanged.

recurrent IOI episodes, each time after an upper respiratory infection (URI). This case possibly shows an association between infection and an aberrant immune response leading to IOI. We also provide a thorough analysis of IOI differentials, preferred radiologic testing, and treatment strategies in the management of IOI.

Case report

A 63-year-old white man presented emergently to the Primary Optometry Eye Clinic at the Veterans Affairs Hospital in West Haven, Connecticut, on November 16, 2006, with an acute red painful right eye. The patient reported a rapid onset of pain and pressure within the eye, photophobia, and decreased vision that began the evening before (at 6 PM). The symptoms progressively worsened to wake him from sleep at 1 AM. The patient denied any recent trauma, eye surgery, or giant cell arteritis symptoms. Medical history was significant for prostate cancer but negative for all other systemic diseases. Entrance examination found visual acuities of 20/40 in the right eye (O.D.), a decrease from 20/20 at his last examination and 20/20 in the left eye (O.S.). No afferent pupillary defect was noted, but restriction was seen on extraocular motility (EOM) testing in all fields of gaze O.D. (worse in right gaze), and no restrictions O.S. Slit lamp examination found edematous right upper and lower lids with grade 3+ injection and chemosis; O.S. was unremarkable. No cells or flare were seen in both eyes (OU; *see* Figure 1). Goldmann tonometry (at 1:38 PM) was 29 mmHg O.D. and 14 mmHg O.S. Dilated examination found a healthy fundus OU without nerve pallor or edema and no vitritis. Asymmetry of IOPs was not of concern, as a dilated examination found small cup-to-disc ratios. Thus, close monitoring of the patient's IOPs was appropriate in the future. Auxillary testing found normal color vision O.D. and O.S., but exophthalmometry showed a 6-mm asymmetry between the 2 eyes (with the right greater), indicating significant proptosis of the right eye. The patient was afebrile and vital signs were all normal.

An ophthalmology consultation was obtained emergently, and immediate tests ordered were complete blood count (CBC) with differential, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and a computed tomography (CT) scan. Results are listed in Table 1.

Table 1 Blood test results after first visit

Test	Result
CBC with differential	Normal
ESR	62 (High)
CT scan	Intraconal and extraconal fat stranding/no muscle or lacrimal gland involvement/sinuses were clear

CBC = complete blood count; ESR = erythrocyte sedimentation rate; CT = computed tomography.

The ophthalmologist tentatively made a diagnosis of orbital cellulitis; however, IOI was considered as a potential differential. The patient was admitted immediately as an inpatient at the Veterans Affairs Hospital and was started on Unasyn® (Pfizer, New York, New York), 3 G intravenously (IV), every 6 hours, and monitored closely overnight. Follow-up the next morning did not show improvement but worsening of symptoms. The patient was continued on the IV drug and monitored closely overnight as an inpatient. The following morning he returned and presented with similar clinical findings showing no change in signs or symptoms. A magnetic resonance image (MRI) found an obvious hyperintense signal of the intraconal fat with a minimal hypertintense signal of the right lacrimal gland, signifying inflammation; no fluid collection or abscess was seen (*see* Figure 2), and no muscle or optic nerve involvement was noted. B-scan was also done and showed no scleral thickening, and a negative T-sign indicated no scleritis. The diagnosis was orbital cellulitis versus IOI, and the patient was continued on IV antibiotics. Prednisone was initiated at 80 mg orally every day. By the next day, the patient reported a marked improvement in symptoms, confirming the diagnosis of IOI. Vision returned to 20/20 O.D., and exophthalmometry found symmetrical findings OU. Goldmann tonometry

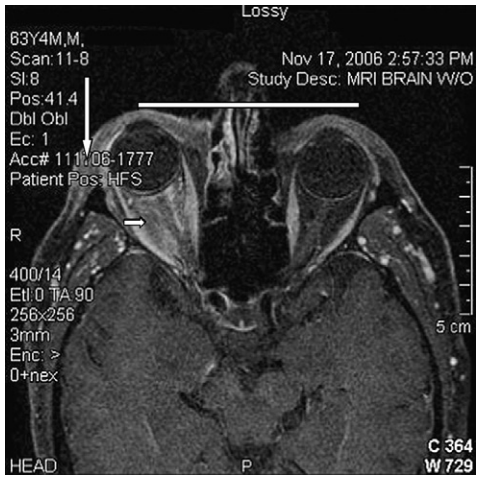

Figure 2 Axial T1-weighted MRI of the orbit with contrast and fat suppression. The *smaller arrow* points to a hyperintense, ill-defined mass within the muscle cone, representing inflammation of the intraconal fat. The longer arrow shows a mild increased signal of the lacrimal gland O.D., compared with O.S. Note proptosis of O.D. in comparison with O.S. (*line*).