Rapid and complete vision loss due to idiopathic orbital inflammation with partial recovery following orbital decompression

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ABSTRACT

We present a case of a 36-year-old man who developed rapid onset of proptosis and orbital compartment syndrome resulting in no light perception vision. He was treated with surgery and subsequent rituximab therapy with some recovery of vision and motility. Though the range of presentations of idiopathic orbital inflammation is broad, such rapid evolution of proptosis with complete vision loss is very uncommon.

KEYWORDS Chronic nongranulomatous inflammation; idiopathic orbital inflammation; orbital compartment syndrome; orbital decompression; proptosis; vision loss

Idiopathic orbital inflammation (IOI), a nonspecific inflammation of orbital tissues, is the third most common orbital disease, accounting for 8% of orbital lesions presenting in the middle-aged population.^{1–3} IOI has no known local or systemic cause and is commonly a diagnosis of exclusion.^{2,4} IOI's characteristic symptoms are pain, swelling, and proptosis. Only 10% to 20% of patients with IOI experience decreased vision, and complete vision loss is exceptionally rare.^{1,5} The mainstay of treatment involves systemic steroids, which typically results in remission. Although the efficacy of treatment can vary, absolute failure to improve after steroid administration is uncommon.^{1,2,6}

CASE DESCRIPTION

An obese 36-year-old Latin American man presented to an outside facility following an injury to the right eyelid with a stone. There was no evidence of laceration or globe malposition at the time. After treatment with tobramycin/dexamethasone ointment for 3 days, he noted worsening erythema, periorbital edema, and chemosis on the right side. He was started on a methylprednisolone dose pack, ofloxacin, and prednisolone eye drops. He continued to deteriorate on this therapy and treatment was escalated to 60 mg of oral prednisone, which failed to improve the clinical course. The patient was subsequently admitted to the hospital for intravenous antibiotic therapy for suspected dacryadenitis. Examination in our

emergency department 8 days after the patient initially sought care disclosed periorbital edema and erythema around the lacrimal gland on the right, with minimal tenderness to palpation (*Figure 1*). He had a mechanical ptosis on the right. Intraocular pressure (IOP) was within normal limits, visual acuity was 20/25, the pupillary exam was normal without relative afferent pupillary defect, and extraocular motility examination demonstrated mild abduction deficit on the right. The left side was unremarkable. Slit lamp exam and dilated funduscopic exam were also unremarkable bilaterally with the exception of conjunctival injection on the right. A computed tomography scan of the orbits with and without contrast revealed abnormal reticular enhancement involving intraconal and extraconal fat in the superior lateral right orbit, enlargement of right superior and lateral rectus muscles,^{7,8} and preseptal soft tissue thickening (*Figure 2*).

Intravenous antibiotics were continued for coverage of bacterial dacryadenitis. Neither intravenous steroids nor oral steroids were initiated in concordance with the patient's reported history of worsening following each steroid treatment, including treatment with intravenous steroids. Over the subsequent 7 hours while asleep, the patient developed rapidly progressive painless proptosis. When awoken in the morning, the patient had no light perception visual acuity on the right, with an IOP of 50. There was complete restriction of all motility of the globe. The pupil was 6 mm and nonreactive.

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Figure 1. Presentation 8 days after the patient initially sought care.

An emergent lateral canthotomy and inferior and superior cantholysis was performed at the bedside. The IOP decreased to 34 and he had improvement in visual acuity to hand motions at the face. Afterwards, a transcaruncular medial orbitotomy was performed. The medial wall was decompressed from the level of the posterior lacrimal crest to the sphenoid sinus. The sphenoid sinus was opened. A subtotal ethmoidectomy was carried out. The mid- and posterior orbital strut was removed for additional decompression. An incision in the

Figure 2. Computed tomography scan showing enlargement of the right superior and lateral rectus muscles, which are commonly inflamed in idiopathic orbital inflammation.^{7,8} Abnormal tissue posterior to the lacrimal gland fossa was biopsied along with the fat in the inferolateral orbit.

periorbital was made to access the abnormal tissue just posterior to the lacrimal gland fossa. Clinically, the tissue was gray, rubbery, and denser than a typical lacrimal gland. Orbital fat in the inferolateral orbit was also biopsied. Intraoperatively, the patient received 12 mg intravenous dexamethasone.

Upon waking from anesthesia, the patient's retina and optic nerve were perfused and the nerve demonstrated spontaneous venous pulsations. There were mild choroidal folds and striations in the posterior retina. Visual acuity had improved, allowing the patient to count his fingers at the face. Treatment with 6 mg dexamethasone every 6 hours was continued for 2 days following surgery.

Histopathologic analysis revealed a predominance of lymphocytes but no clonal lymphocyte population. Immunoglobulin G-positive cells were exceptionally rare. Cytometry revealed a small nonhemopoietic population, consistent with stromal and epithelial cells. The final pathology diagnosis was chronic nongranulomatous inflammation.

The steroids were tapered and the patient was discharged on 10 mg of oral prednisone daily for 8 days. A serologic workup failed to identify any alternative etiology aside from IOI. Rituximab was initiated at a dose of 375 mg every 2 weeks for four doses. The patient's visual acuity stabilized at 20/300, and his motility returned to normal.

DISCUSSION

Our patient experienced many atypical findings such as absence of pain, initial steroid ineffectiveness, rapid acceleration of symptoms, and rapid decline in vision to no light perception. The symptoms developed shortly after a head contusion; however, the development of IOI as a result of the inflammatory cascade resulting from trauma is still left to be elucidated.⁹ Furthermore, IOI resulting in complete loss of vision has been mentioned in only two previously published reports.^{10,11} These cases have inferred that nerve inflammation, apical crowding, and either optic nerve stretching or compression are possible reasons for vision loss in IOI.^{10,11} In our patient, sudden severe proptosis resulting in optic stretching may have contributed to vision loss. Painless progression resulting in severe vision loss to no light perception is exquisitely rare given that pain is considered the chief characteristic of inflammation in this entity.^{12,13}

A Delphi study of expert panelists agreed that in nonmyositic IOI, biopsy is key for diagnostic confirmation, and in myositic IOI corticosteroid response is preferred.¹⁴ However, relying on this method has been considered paradoxical, because corticosteroids can switch off inflammatory genes and induce apoptosis in lymphocytic lesions.¹⁵ Some reports propose that if there is steroid ineffectiveness and/or persistent imaging abnormalities, tissue sampling should be considered for a definitive diagnosis. To further complicate the issue, the timing of biopsy is also controversial. Recent studies have claimed that biopsy utilizing local anesthesia and noninvasive techniques should be a first-line strategy in confirming the diagnosis rather than a subsequent tool.¹⁵

Typically, biopsy of IOI lesions reveals a high prevalence of lymphocytes, which necessitates the exclusion of an alternative

diagnosis of lymphoma. Although our serological results revealed IOI as the leading diagnosis, serological screening is limited because inflammatory markers are not necessarily diagnostic and match inflammation regardless of cause. Serological tests are necessary to exclude systematic causes.

Other treatments utilized in steroid refractory cases include chemotherapy, external beam radiation, and/or antiproliferative drugs, such as cyclophosphamide or chlorambucil and prednisone.^{16–18} Radiotherapy has shown a modest efficacy rate of 70% in treatment of IOI and is described as adequate in cases of steroid refraction.¹⁹

Diagnosis by exclusion is necessary for identification of IOI, and exhaustive serologic and histopathological analysis is required. Here, orbital decompression was required to stabilize the patient's condition. Following an exhaustive workup, with rituximab therapy, the patient's ocular mobility recovered and visual acuity improved to 20/400.

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