# Idiopathic Orbital Inflammation Leading to Unilateral Blindness Over a 2-Day Presentation in a Child

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***Abstract:*** **An 8-year-old boy developed vision loss to no light perception on the left side over a 2-day period. He initially presented with unilateral eyelid swelling, which progressed to bilateral edema and an eventual left-sided orbital apex syndrome. Orbital imaging revealed enlarged extraocular muscles, and biopsy confirmed idiopathic orbital inflammation. Despite subsequent orbital decompression, high-dose steroids, and additional steroid-sparing therapy, he did not regain vision after 9 months of follow up.**

Idiopathic orbital inflammatory syndrome (IOIS) accounts for roughly 8% of orbital lesions.[1] It is primarily a disease of middle-aged individuals, but between 6–16% of cases occur in individuals under the age of 20. In the pediatric population, bilateral disease is more common, affecting up to 45% of pediatric IOIS cases. Permanent decreased vision as a consequence of IOIS has been described.[2] However, severe loss of vision at the initial presentation of the disease has not been reported. We report the case of a child who presented with bilateral IOIS and progressed to no light perception (NLP) vision in one eye over a 2-day period.

**Case Description.** A healthy 8-year-old boy with a history of 20/20 vision developed left eye pain and eyelid edema over a 24-hour period. The following day, he presented to an optometrist who measured a visual acuity of 20/20 in the right eye but 20/80 in the left. He was prescribed prednisolone eyedrops for a presumed allergy and referred to an ophthalmologist.

Upon presenting to the ophthalmologist—now 48 hours after initial symptoms—the patient had NLP vision in the left eye, severe proptosis, and an intraocular pressure of 47 mm Hg on the left side. He was given pressure-reducing eyedrops and underwent a lateral canthotomy with inferior cantholysis. The intraocular pressure decreased to 32 mm Hg. A CT scan revealed muscle thickening and bilateral proptosis, worse on the left (Fig. 1).

The patient was admitted to an outside institution that same day, where laboratory studies showed an elevated white blood cell count at 16.2 K/$\mu$l with a left shift and a C-reactive protein of 33 mg/dl. He was initially given intravenous antibiotics and intravenous steroids (dexamethasone 0.26 mg/kg/day) with the presumptive diagnosis of orbital cellulitis or IOIS.

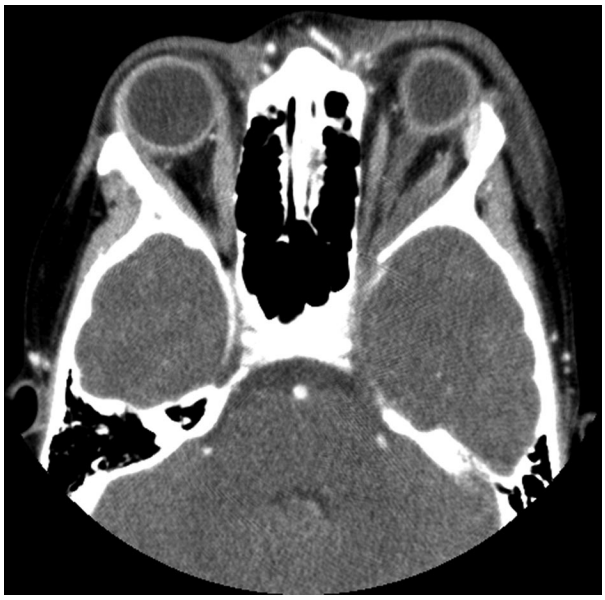

**FIG. 1.** Preoperative axial CT on day 2 of presentation revealed 4 mm of relative proptosis and significant tenting of the posterior sclera with a posterior globe angle of 71°.

After overnight observation, the patient had not improved and was transferred to the Children's Hospital of Philadelphia. On admission, his vision was 20/80 on the right and still NLP on the left side. In addition, the left eye exhibited 8 mm of relative proptosis, complete ptosis with no levator function, an enlarged nonreactive pupil, and total ophthalmoplegia. The patient was taken to the operating room to obtain biopsies to rule out a neoplastic process. Intraoperative frozen section biopsies of the enlarged left medial rectus muscle and orbital fat were consistent with IOIS, and the patient underwent ethmoidal decompression via an anterior approach. A postoperative CT of the orbits displayed an adequate posterior decompression with reduction in left-sided proptosis and decreased tenting of the posterior globe (Fig. 2).

Pathology showed a non-neoplastic, necrotizing inflammatory myositis (Fig. 3) that was consistent with IOIS. The patient was treated with high-dose intravenous steroids (methylprednisolone) for 4 days with no improvement in vision. After consultation with rheumatology, the patient was treated with infliximab and methotrexate due to concern for recurrence and to preserve vision in the right eye. After 9 months of follow up, he had no improvement in vision on the left, although all other symptoms subsided.

## DISCUSSION

Vision loss to NLP is not typically seen in IOIS. A 1978 case series of pediatric IOIS reported 53% of patients had decreased visual acuity at the initial onset of an attack. However, all of these patients maintained a visual acuity of at least 20/60. There were no patients with NLP vision at initial presentation.[2] There is a single report of NLP vision loss in a pediatric patient with extension of IOIS in the cavernous sinus. His clinical course progressed to NLP vision in one eye over a 4-month period.[3]

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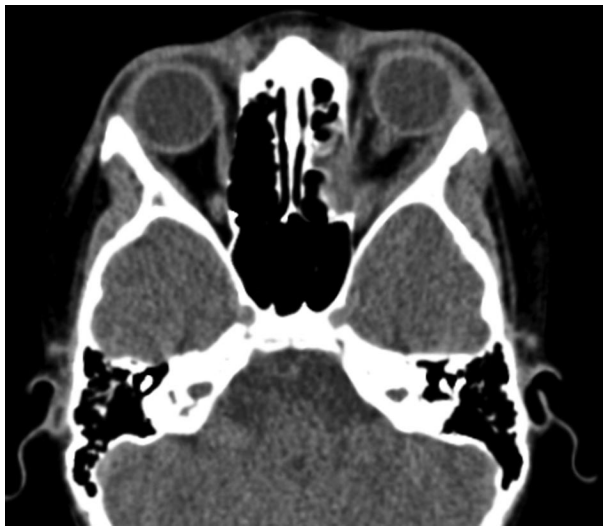

**FIG. 2.** Postoperative axial CT on day 4 of presentation showed ethmoidal decompression, decreased proptosis, and improved globe morphology.

In our patient, the cause of vision loss could be attributed to a left-sided orbital apex syndrome with optic nerve compression. An alternative etiology for vision loss could have been optic nerve stretching secondary to severe proptosis.[4] This patient's initial CT scan showed a posterior globe angle of less than 90° (Fig. 1), suggesting a poor prognosis for vision. The tenting of the posterior globe reflected the stretching force placed on the optic nerve and also the acuteness of his presentation.[5] Finally, an orbital compartment syndrome has been described with features that include proptosis, visual loss, and posterior globe tenting.[6] Indeed, despite adequate medical and surgical interventions, he did not regain vision on the left. This case highlights the potential for rapid, progressive vision loss in IOIS.

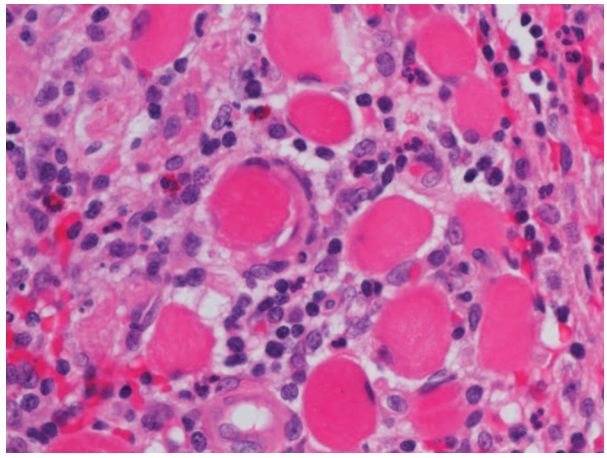

**FIG. 3.** Hematoxylin–eosin stain at 60× magnification of biopsied medial rectus showed a non-neoplastic, necrotizing mixed inflammatory myositis, consistent with idiopathic orbital inflammation.

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