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Clinical Communications: Adult

MAN WITH A SWOLLEN EYE: NONSPECIFIC ORBITAL INFLAMMATION IN AN ADULT IN THE EMERGENCY DEPARTMENT

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☐ **Abstract—Background: Nonspecific orbital inflammation (NSOI) is a rare idiopathic ocular pathology characterized by unilateral, painful orbital swelling without identifiable infectious or systemic disorders, which can be complicated by optic nerve compromise. Case Report: A 50-year-old man presented to the Emergency Department with recurring, progressive painless left eye swelling, decreased visual acuity, and binocular diplopia in the absence of trauma, infection, or known malignancy. His physical examination was notable for left-sided decreased visual acuity, an afferent pupillary defect, severe left eye proptosis and chemosis, and restricted extraocular movements; his dilatated funduscopic examination was notable for ipsilateral retinal folds within the macula, concerning for a disruption between the sclera and the retina. Ocular examination of the right eye was unremarkable. Laboratory data were unrevealing. Gadolinium-enhanced magnetic resonance imaging showed marked thickening of the left extraocular muscles associated with proptosis, dense inflammatory infiltration of the orbital fat, and characteristics consistent with perineuritis. The patient was diagnosed with NSOI with optic neuritis and admitted for systemic steroid therapy; he was discharged on hospital day 2 after receiving high-dose intravenous (i.v.) methylprednisolone with significant improvement. Why Should an Emergency Physician Be Aware of This?: NSOI is a rare and idiopathic ocular emergency, with clinical mimicry resembling a broad spectrum of systemic diseases such as malignancy, autoimmune diseases, endocrine disorders, and infection. Initial work-up for new-onset ocular proptosis should include comprehensive laboratory testing and gadolinium-enhanced magnetic resonance imaging. Timely evaluation by an ophthalmologist is crucial to assess for optic nerve involvement. Signs of optic nerve compromise include decreased visual acuity, afferent pupillary defect, or decreased color saturation. Patients with optic nerve compromise require admission for aggressive anti-inflammatory therapy with i.v. steroids in an attempt to reduce risk of long-term visual sequelae. Our case demonstrates a severe presentation of this disorder and exhibits remarkable visual recovery after 48 h of systemic i.v. steroid treatment. Published by Elsevier Inc.**

☐ **Keywords—nonspecific orbital inflammation; orbital pseudotumor; exophthalmos; emergency department; ophthalmology**

INTRODUCTION

Nonspecific orbital inflammation (NSOI), first described in early 20th century medical literature as "idiopathic orbital inflammatory syndrome" (or "orbital

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pseudotumor"), is a unique ocular disorder that is the third most common orbital disease and accounts for up to 11% of orbital tumors (1,2). It is characterized by unilateral polymorphous lymphoid infiltration of the orbit without identifiable infectious or systemic disorders (3). Despite its remarkable clinical features, evaluation and management of patients with NSOI can be challenging in the emergency department (ED).

CASE REPORT

A 50-year-old man from Guatemala with a vague history of prior similar eye swelling and vision changes, presented to the ED with 2 months of painless left eye swelling, binocular diplopia, and decreased vision. Review of systems was negative for trauma, fever, chills, night sweats, cough, hemoptysis, and weight loss. His past medical history included latent tuberculosis status post isoniazid therapy, complicated by self-resolving, asymptomatic transaminitis. His past surgical history included left orbital biopsies, performed in the United States, for a similar issue, which was reported to him as "normal." Medications included artificial tears as needed. The patient recalled a similar presentation 2 years prior that involved extensive but reassuring laboratory testing and imaging studies. His symptoms resolved after medication. Review of his chart confirmed the results of a prior orbital biopsy, which showed evidence of chronic inflammation with a single, non-caseating granuloma that was negative for acid-fast bacteria, fungi, and malignancy. Additional laboratory results obtained previously included normal laboratory work-up including complete blood count, thyroid function studies (thyroid-stimulating hormone [TSH]), erythrocyte sedimentation rate (ESR), antinuclear antibodies (ANA), antineutrophil cytoplasmic antibodies (ANCA), angiotensin-converting enzyme (ACE) level, rheumatoid factor, rapid plasma reagin test (RPR), human immunodeficiency virus (HIV), Lyme reflex, and serum protein electrophoresis. The orbital inflammation had been treated and responded well to oral prednisone.

On arrival to the ED, the patient's vital signs were blood pressure 121/70 mm Hg, pulse rate 74 beats/min, respiratory rate 18 breaths/min, temperature 36.8°C (98.3°F), and SaO_2 99% on room air. Ocular examination was notable for normal visual acuity in his right eye (20/20) and diminished visual acuity in his left eye (20/125). There was left-sided afferent pupillary defect present. Intraocular pressures were 10 and 15 (reference range 8–21 mm Hg), respectively, during initial ED presentation and throughout his hospitalization. Anterior examination was unremarkable in the right eye; the left eye exhibited proptosis, chemosis, and restriction of extraocular movements in all gazes (Figures 1 and 2). Dilatated

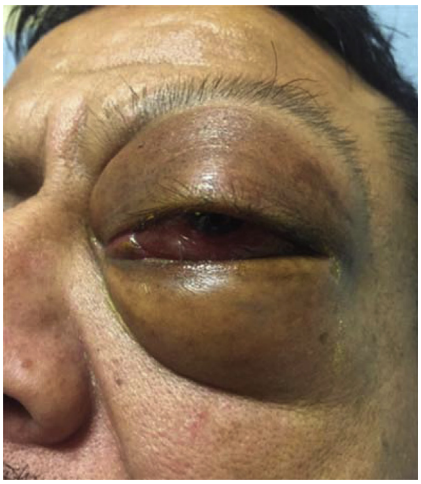

Figure 1. Patient image showing left eye proptosis and conjunctival chemosis.

fundus examination of the right eye was unremarkable, including normal optic disc appearance, normal retinal vasculature, and normal foveal light reflex. Fundoscopy of the left eye revealed normal optic disc, normal retinal vasculature, and retinal folds within the macula, suggesting disruption of the normal relationship between the sclera and the retina secondary to increased intraorbital pressure. His head and neck examination was notable for a supple neck without adenopathy and moist mucous membranes. His heart, lung, and abdominal examinations were normal. His extremities were warm, well perfused, without rashes, erythema, or pitting edema. On neurologic examination, the patient was anxious but he was alert and oriented to person, place, and time. With the exception of the previously mentioned left-eye ophthalmoplegia, this neurologic examination was otherwise unremarkable.

Laboratory results were significant for TSH level of 1.64 IU/mL (reference range 0.35–5.5 uIU/mL), an ESR of 20 mm/h (reference range 0–15 mm/h), and C-reactive protein of 1.24 mg/L (reference range 0–10 mg/L).

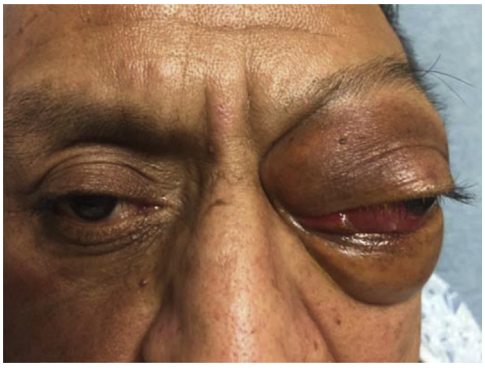

Figure 2. Patient image with disconjugate gaze secondary to left eye swelling.