

Case of Recurrent Exophthalmos: Angioedema Versus Idiopathic Orbital Pseudotumor

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A 67-year-old white woman with a medical history of aspirin-exacerbated respiratory disease, hypertension, and hyperlipidemia presented to the clinic with painless, isolated left orbital swelling and exophthalmos. The orbital swelling began a week before presentation and progressed, as depicted in Figures 1-3. Swelling was unassociated with aspirin and other nonsteroidal anti-inflammatory drugs, other medications, trauma, urticaria, or angioedema; there were no visual defects. The swelling resolved with prednisone 20 mg three times daily for 7 days, followed by a taper over 1 month. However, orbital swelling recurred within a week, and she again was placed on prednisone, 20 mg three times daily, tapered to a dose of 5 mg daily with complete resolution over a year.

Ten years later, the swelling and exophthalmos reappeared in the same eye. Again, a similar history was obtained, and a complete history and physical examination were normal. Her nasal polyps were controlled with mometasone 50 µg two sprays/nostril twice daily, and her asthma was controlled with budesonide/formoterol 160/4.5 µg two puffs twice daily and albuterol metered-dose inhaler 90 µg two puffs every 6 hours as needed. She is also on irbesartan/hydrochlorothiazide 300 mg/12.5 mg one tablet by mouth daily, Diltiazem CD (Lexicomp) 240 mg one tablet by mouth daily, and rosuvastatin 10 mg one tablet by mouth daily. There was no family history of urticaria, angioedema, or orbital swelling. There was no history of tobacco, alcohol, or illicit drug use.

A complete blood cell count, comprehensive metabolic panel, thyroid function tests, and the C1 esterase deficiency evaluation were negative. Magnetic resonance imaging of the orbits revealed a left isolated proptosis, swelling of the medial and inferior rectus muscles with mild hypertrophy, and swelling of the left lacrimal gland. Prednisone 20 mg three times daily until resolution was followed by a slow taper and a maintenance of 5 mg daily, with complete resolution over a year.

Idiopathic orbital pseudotumor (IOP) is usually a non-granulomatous inflammatory orbital mass without an identified local or systemic etiology.¹ It represents 4.7% to 6.3% of orbital disorders, with an age range from 2.5 weeks to 92 years.^{2,3} The

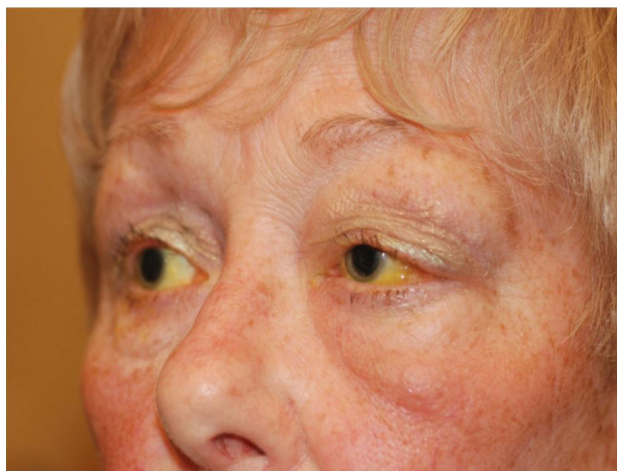


FIGURE 1. Left orbital swelling with exophthalmos, left lateral view.



FIGURE 2. Left orbital swelling with exophthalmos, right lateral view.

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mean age of onset is in the fifth decade, with no sex predilection.⁴ It is the third most common orbital disease after thyroid ophthalmopathy and lymphoproliferative disorders, and a diagnosis of exclusion based on the history, physical examination, laboratory tests, clinical course, and response to corticosteroid therapy. Biopsy is of limited use.⁴

Histopathology of IOP usually demonstrates a fibroinflammatory infiltrate. However, lesions with atypical histopathologic patterns, such as extensive sclerosis, vasculitis, granulomatous inflammation, and tissue eosinophilia also can be present.² Most commonly, it occurs in the orbit, with frequent involvement of the lacrimal gland.⁴ It is typically unilateral, rarely bilateral,⁴ and patients may present



FIGURE 3. Left orbital swelling with exophthalmos, front view.

with proptosis, diplopia, conjunctival chemosis, visual disability, and restriction of extraocular muscle function.⁴ Recurrent disease after a 10-year asymptomatic interval is reported.⁴ The computed tomography and magnetic resonance imaging findings were characterized by swelling of the affected intraorbital structures.

Differential diagnosis includes systemic inflammatory diseases, such as sarcoidosis, Sjögren syndrome, IgG4-related disease, Behçet disease, granulomatosis with polyangiitis, and Churg-Strauss syndrome.⁵⁻⁷ However, in this case, the absence of other systemic signs and symptoms, the benign localized progression, and the response to oral corticosteroids did not warrant further investigation. Physicians may confuse IOP with angioedema;

however, the absence of urticaria, prolonged duration of the swelling, and gradual response to corticosteroids rules out this diagnosis.

Approximately 80% of patients respond to oral corticosteroids.^{3,4} Radiotherapy may be useful in patients who do not respond to corticosteroid or those with a rapidly progressive course of disease.^{3,4} Biopsy is usually required with refractory and corticosteroid unresponsive IOP.^{2,4} Methotrexate; cyclosporin A; mycophenolate mofetil; cyclophosphamide; rituximab; and cytokine-specific biologic immunomodulatory agents, such as infliximab, adalimumab, etanercept, daclizumab, abatacept, tocilizumab, have been used with varying degrees of success.³ Prognosis is generally excellent, with complete resolution of disease.⁴

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