Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol. 14, Issue 25s (2025)
https://www.jneonatalsurg.com

OPEN ACCESS

Secondary Glaucoma Associated With Orbital Pseudotumor

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Cite this paper as: Camilla Amanda Prakoeswa, Evelyn Komaratih, Susy Fatmariyanti, (2025) Secondary Glaucoma Associated With Orbital Pseudotumor. *Journal of Neonatal Surgery*, 14 (25s), 251-256.

ABSTRACT

Orbital pseudotumor is a non-infectious and benign inflammatory condition of the orbit and the surrounding tissues. This case report aims to emphasize the importance of understanding the cause-effect relationship between orbital inflammation and raised intraocular pressure (IOP). An 18-year-old girl was previously diagnosed with primary angle-closure glaucoma and had a history of twice trabeculectomy surgeries done elsewhere two months before she came to our hospital. She came with left eye pain, eyelid edema, ptosis, conjunctival chemosis, ophthalmoplegia, and elevated IOP despite a positive seidel test for a leaking trabeculectomy bleb. Five months prior to her first surgery she realized an inward squint on her left eye. She also felt joint pain for a year before the ophthalmic symptoms. After comprehensive diagnostics, she was diagnosed with left eye orbital pseudotumor and secondary glaucoma with spondyloarthropathy. Daily 30 mg of oral prednisone was given to reduce the inflammation. After a week, her conditions improved with decreased IOP although there was no additional anti-glaucoma medication. The raised IOP in orbital pseudotumor is often caused by the swelling of orbital tissue and extraocular muscles, that result in choroidal circulation problems from the superior and inferior ophthalmic veins compression. The IOP was greatly reduced once the inflammation was treated Diagnosing orbital pseudotumor may be challenging since it can mimic various conditions. Secondary glaucoma should be acknowledged as a complication in patients with orbital pseudotumor. Once orbital pseudotumor is diagnosed, a systemic steroid should be given to manage the inflammatory conditions as well as resultant glaucoma.

Keywords: *Orbital pseudotumor, glaucoma, trabeculectomy*

1. INTRODUCTION

Orbital pseudotumor is an inflammatory condition of unknown origin, although an underlying immune process is suspected. Orbital pseudotumor may manifest with sudden onset of orbital pain, eyelid edema, extraocular muscles disturbances, conjunctival chemosis, and increased intraocular pressure.[1] Orbital pseudotumor may cause secondary glaucoma due to the extraocular muscle and orbital tissue swelling, which compresses the superior and inferior ophthalmic veins, thus elevating the episcleral venous pressure.[2] This study is to illustrate a challenging case of orbital pseudotumor with glaucoma and the importance of understanding the cause-effect relationship between the orbital inflammation and the secondary glaucoma.

Patient and observation

An 18-year-old girl was referred with severe left ocular pain with swollen and drooping eyelid after she underwent two trabeculectomy surgeries two and three months prior, elsewhere. Her left eyelid started to droop after the first trabeculectomy surgery. She could not move her left eyeball in any direction in the last two months. She had no history of fever, weight loss, or known malignancy. However, she stated that she often felt joint pains and muscle aches, and she had acne all over her face since a year ago. She was diagnosed with LE (left eye) glaucoma elsewhere and was given timolol 0.5% eyedrops LE and acetazolamide tablets that she took three times daily for two years. In April 2022, she admitted she had difficulty moving her left eyeball medially, and she started to have an outward squint in the left eye. In the next five months, she had severe left eye pain with LE IOP around 50 mmHg, thus a trabeculectomy was planned for her. After the surgery, she complained of decreased vision and droopy eyelid. Since there was no reduction of her IOP, the ophthalmologist decided to do another trabeculectomy surgery on the same eye a month later and resulted in a worse condition. The patient was then given ciprofloxacin eye drops six times daily LE, timolol 0.5% eyedrops twice daily LE, acetazolamide 250 mg tablets four times daily orally, potassium chloride tablets once daily orally, and cefixime 250 mg tablets twice daily. Since there was no improvement, she was eventually referred to our hospital with the diagnosis of LE primary angle-closure glaucoma and LE flat bleb and blebitis.

The patient's left eye visual acuity was four-meters counting fingers with an IOP of 38 mmHg. The left eye had total ophthalmoplegia, exotropia, and mild proptosis. It had ptotic, spastic, and edematous eyelid with lower eyelid entropion and hyperaemic conjunctiva with chemosis from eight to ten o'clock position and caruncle swelling. The trabeculectomy bleb was visible in the eleven o'clock positions with a seidel-positive leaking bleb. The cornea was hazy, and the fluorescein staining showed multiple punctates on every quadrant. Anterior chamber depth was shallow around grade I-II Van Herrick. The iris was radial with posterior synechiae around the twelve to seven o'clock position and two iridectomy holes at the eleven and one o'clock position. The pupil had decreased light reflex, and the lens had iris pigment scattered on its surface. Funduscopy were hard to evaluate.

Moreover, the anterior segment Optical Coherence Tomography (OCT) for the left eye showed a 2.09 mm anterior chamber depth. B-scan ultrasonography was normal for both eyes. Orbital CT scan resulted in diffuse left intraconal soft tissue thickening (41-54 HU) in the medial, lateral, superior, inferior rectus and superior and inferior oblique muscles with contrast enhancement and left eye proptosis. The laboratory results showed normal levels of leucocytes, C-reactive protein, antinuclear antibody, complement 4, and thyroid blood test. However, she had an abnormally high erythrocyte sedimentation rate (86 mm/hr) and high Complement 3 (132 mg/dL), which suggest an inflammatory process. Consultation to the internal medicine department resulted in the diagnosis of peripheral spondyloarthritis with extraarticular (ocular) manifestation. The findings were suggestive of orbital pseudotumor, so we initiated the systemic steroid therapy immediately with 1 mg/kg weight of oral prednisone daily.

One week after steroid therapy, the patient showed improvement of the left eye with 0.2 visual acuity and 16 mmHg IOP. Although she still had restricted ocular motility in the left eye, she then could move her eyeball nasally with resolved proptosis and lacrimal caruncle swelling. While the left eye was still ptotic, she could lift her upper eyelid better since it was not as edematous as before. The trabeculectomy bleb still leaked though the flow was minimum. In the fourth week of therapy, the patient was scheduled for a Magnetic Resonance Imaging (MRI) evaluation. The results were left orbital extrinsic muscle thickening (left lateral, medial, superior, and inferior rectus muscle) and orbital soft tissue swelling. The prednisone was tapered after six week and oral Azathioprine, an immunosuppresant was given 25 mg daily by the internal medicine department. We diagnose this patient with orbital pseudotumor with complications of secondary glaucoma, total ophthalmoplegia, and corneal erosion in the left eye, and peripheral spondyloarthritis with extraarticular (ocular) manifestation. The patient has given her consent to be included in this report.

2. DISCUSSION

Orbital pseudotumor, also known as non-specific orbital inflammation or idiopathic orbital inflammation, is a non-infectious and benign inflammatory condition occupying the orbit and the tissues surrounding it.[3] It has an unknown cause and is commonly regarded as a diagnostic of exclusion with testing done to rule out systemic diseases, infections, and neoplasms.[4,5] No conclusive pathogenesis of orbital pseudotumor has been established, although immune-mediated mechanisms are most likely to be the underlying ocular mechanism.[3] Orbital pseudotumor is the third most prevalent orbital disease following graves ophthalmopathy and lymphoproliferative disease.[4] Its incidence is estimated to be 6 to 16% of all orbital tumors.[6-8] Diffuse orbital pseudotumor, a sub-type of orbital pseudotumor with inflammation throughout the orbit, as seen in this case, is the least common type of orbital pseudotumor.[8, 9] Orbital pseudotumor is typically unilateral, as seen in the patient in this case report. However, 8%–20% of cases is bilateral.[7] Orbital pseudotumor may manifest with acute onset of eyelid pain, ptosis, ocular motility disturbances, eyelid edema, conjunctival injection, conjunctival chemosis, and raised IOP, which can be found in this patient.[1, 6, 10]

The occurrence of comparable prior episodes, trauma, illness, systemic or immunocompromised disorders, symptom duration, and a comprehensive eye examination need to be evaluated.[10] Orbital pseudotumor is a diagnosis of exclusion, therefore, a thorough laboratory evaluation for orbital pseudotumor suspect should consist of a complete blood count, metabolic panel, thyroid function tests, erythrocyte sedimentation rate, antinuclear antibodies, antineutrophil cytoplasmic antibodies, angiotensin-converting enzyme level, rapid plasma reagin test, and rheumatoid factor.[11]

Did glaucoma and the surgery later cause orbital pseudotumor, or was it orbital pseudotumor mimicking glaucoma with increased IOP all along? For the first possibility, complex inflammatory responses at several levels involving numerous pathways are a part of the pathophysiology of glaucoma. In the eye, glial cells in the retinal/neuronal tissue and TM cells in the trabecular meshwork play a significant role in controlling inflammatory reactions.[12] As a result of the persistent stress caused by mechanical, vascular, and oxidative stress in glaucoma, this feedback mechanism to recovery is likely impeded, and chronic inflammation may develop if the stress is sustained over time.[12] Furthermore, surgical intervention will carry the risk of postoperative inflammation although the extent of inflammation cannot be predicted. Several factors that make the inflammation more likely are the invasiveness of the surgery, predisposition to inflammation, and past surgical history.[13] The patient in this report was diagnosed with glaucoma for two years before she started showing signs and symptoms of orbital pseudotumor, triggered after trabeculectomy surgeries was done twice for her left eye.

For the second possibility, the raised IOP in patients with orbital pseudotumor can also be caused by extraocular muscle and orbital tissue swelling, which compresses the superior and inferior ophthalmic veins, thus causing a problem in the choroidal

circulation.[2] A quick IOP decline can be seen in this patient when the inflammation is managed.[14] Diffuse type of orbital pseudotumor, seen in this case, was more likely to have increased IOP due to elevated venous pressure than other subtypes of orbital pseudotumor.[15] A study found that their diffuse orbital pseudotumor cases all had an IOP of over 21 mmHg before treatment.[15] Raised IOP in orbital pseudotumor can be managed by steroid therapy.[15] Although a study did not report elevated IOP secondary to steroid therapy in orbital pseudotumor, IOP monitoring must always be considered, especially for younger patients since they are likely to develop steroid-induced glaucoma.[15]

This patient had worse symptoms after trabeculectomy surgery with a leaking bleb. In the beginning phase of inflammatory disease, an elevated episcleral venous pressure (EVP) causes increased IOP. The inflammatory condition might also cause fibrosis of the area surrounding the bleb, thus reducing the drainage, and further increasing the IOP.[16] It is postulated that the thin cystic wall of the bleb could not sustain the grossly increasing pressure, which results in the bleb leakage. The most detrimental effect of bleb leakage is an infection, estimated to be over 25 times more likely in patients with a leaking filtering bleb than not.16 Topical instillations of aqueous suppressants may aid in reducing the flow, thus allowing the holes to heal.[17] A conjunctival advancement and compression sutures may be offered to the patient to lessen the cystic bleb complication and establish adequate filtration.[16] For the patient, it is crucial to ensure no infection on the bleb before steroid therapy is started.

While only a small percentage of individuals had their symptoms resolved without treatment, orbital pseudotumor responds remarkably to steroids.[6] About 80% of orbital pseudotumor cases had a good response to first-line steroid therapy, with 50% of the cases showing relapse.[4] The initial dose for moderate to severe orbital pseudotumor in adults is 1 mg/kg of prednisone, with the total dose ranging from 60 mg to 100 mg a day for a week or two and slowly tapered in six to eight weeks soon after the clinical improvement is completed.[3, 4] In most cases, steroid therapy resulted in substantial improvement within 48-72 hours, primarily in pain and signs of inflammation.[18] Although the patient did not show total remission of orbital pseudotumor after steroid therapy, one week post steroid therapy, she showed an improved condition and decreased IOP (16 mmHg) despite the same regimen of anti-glaucoma therapy. A longer and slower steroid taper may be necessary for this particular type of orbital pseudotumor.[19] For the next step, we will evaluate her condition now that the internal department has given her 25 mg azathioprine, an immunosuppressant, twice daily, for the extraarticular manifestation of peripheral spondyloarthropathy.[20]

3. CONCLUSION

We illustrated a case of an eighteen-year-old girl with a challenging case of orbital pseudotumor with the complication of secondary glaucoma. The presented case of orbital pseudotumor and glaucoma may be considered an immunological process affecting the orbit after the surgery even though the cause-effect relationship remains unclear. Diagnosing orbital pseudotumor is challenging due to its ability to mimic a wide spectrum of diseases. It is crucial to involve an interprofessional team of ophthalmologists, internists, and radiologists to promptly manage patients suspected of orbital pseudotumor. In patients with orbital pseudotumor, secondary glaucoma should be considered as a differential diagnosis of secondary angle-closure glaucoma, especially if the presented signs and symptoms are similar. Once orbital pseudotumor is diagnosed, a systemic steroid is indicated for managing the inflammatory conditions as well as resultant glaucoma.

Competing interests

The authors declare no competing interest.

Figures

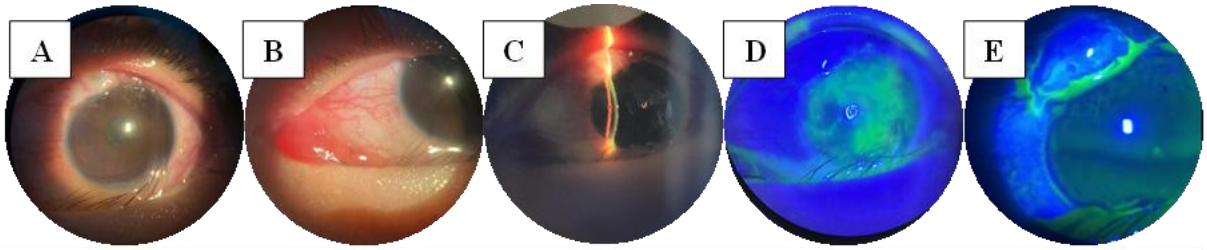
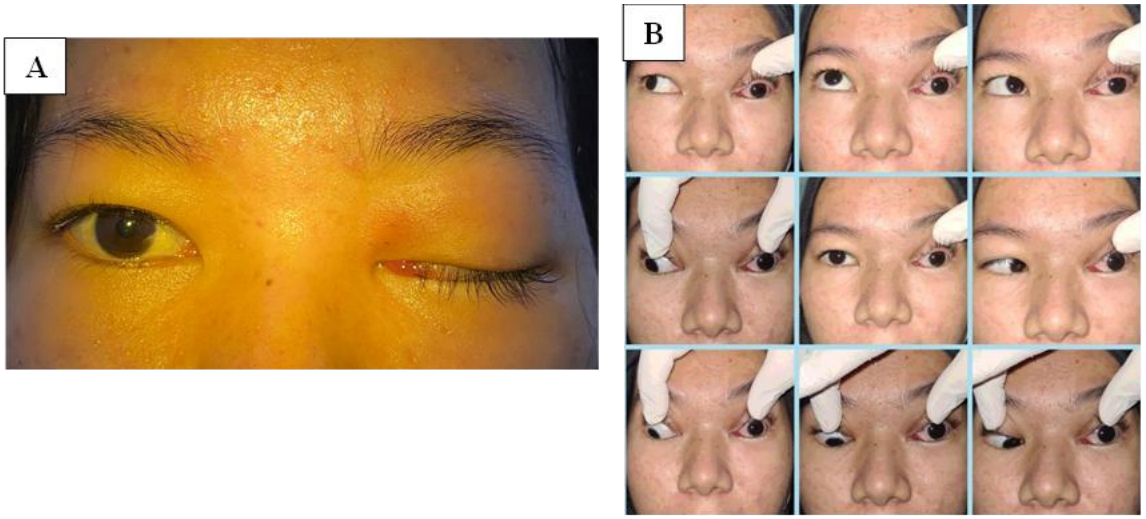


Figure 1: Slit lamp examination of the left eye (A) demonstrated caruncle swelling (B), trabeculectomy bleb (C), positive fluorescein staining (D), and Seidel-positive leakage of the bleb (E)



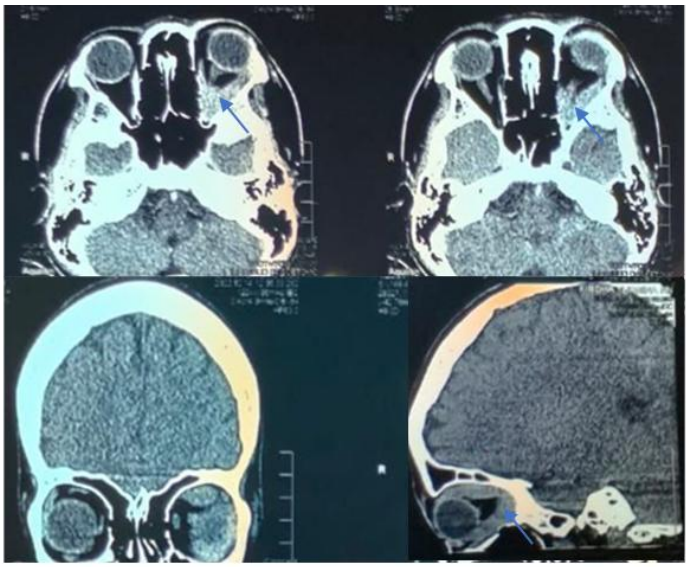


Figure 2: External clinical photos of the patient, initial examination demonstrating left eye ptosis (A) and total ophthalmoplegia (B)

Figure 3: Orbital CT scan indicated intraconal soft tissue thickening on the left eye (blue arrows)

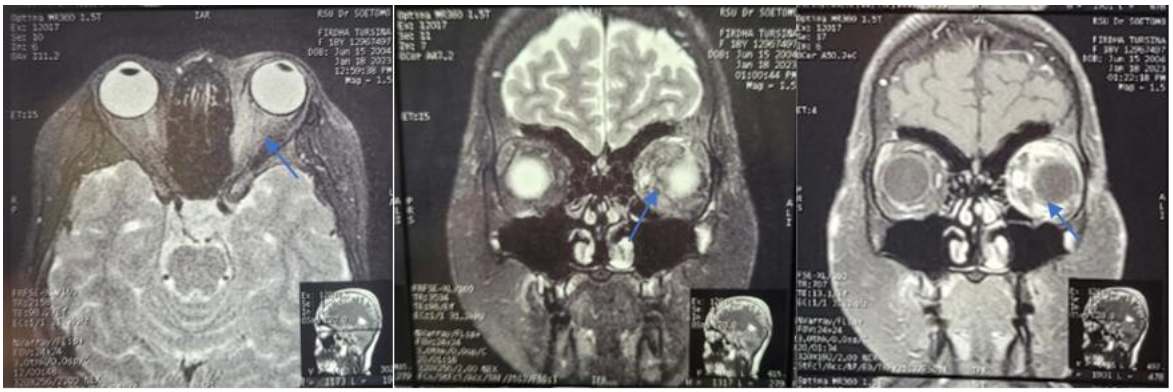


Figure 4: MRI evaluation one month after steroid therapy showed muscle thickening of all four rectus muscles on the left eye as well as swelling of the orbital tissue, consistent with orbital inflammation (blue arrows)

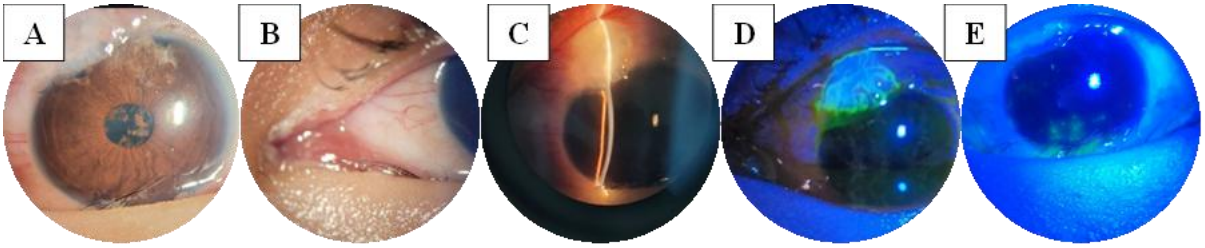


Figure 5: Sixth week of follow up anterior segment evaluation. Inflammatory signs were starting to worsen after the prednisone was tappered off for 5 mg a day (A, B). The bleb became more cystic with positive Seidel test (C, D). Fluorescein staining positive for corneal erosion (E)

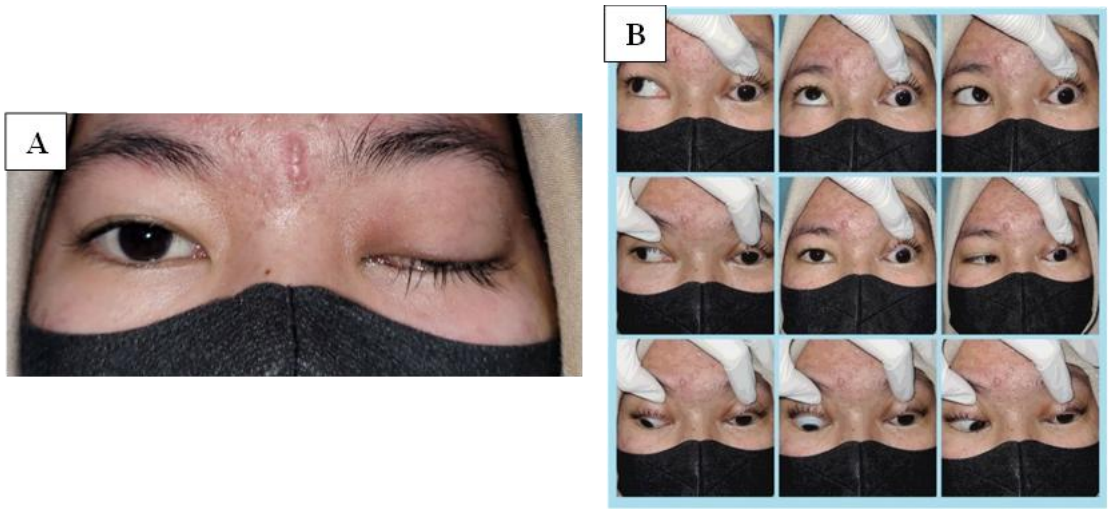


Figure 8 External examination on the sixth week of steroid therapy with recurring lacrimal caruncle swelling and more spastic and edematous left eyelid (A) and total ophthalmoplegia (B)

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