

Cavernous Hemangioma, Presumed To Be Orbital Schwannoma: Diagnostic Challenge And Successful Surgical Management

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Abstract

Objectives:

Orbital tumors represent a heterogeneous group of lesions that can mimic one another clinically and radiologically. Cavernous venous malformation, schwannoma, and optic nerve sheath meningioma are among the most common benign orbital masses in adults. These tumors may lead to compressive optic neuropathy, resulting in progressive visual loss. We present a case of a longstanding orbital mass with compressive optic neuropathy in which surgical excision restored visual function.

Methods:

A 52-year-old woman presented with a four-year history of progressive left eyelid protrusion and gradual decline in visual acuity. Clinical evaluation demonstrated visual acuity of 6/19 in the right eye and 1/60 in the left eye, with a relative afferent pupillary defect and optic disc pallor. Magnetic resonance imaging of the orbit revealed a solid, well-circumscribed intraconal mass in the left orbit measuring 2.79 × 2.35 × 2.16 cm. The differential diagnoses included optic nerve sheath meningioma, cavernous venous malformation, and schwannoma.

Results:

The patient underwent surgical excision of the orbital tumor using a transconjunctival inferomedial orbitotomy with cryoprobe assistance. Intraoperatively, a well-encapsulated purplish mass was identified, extending to the orbital apex and compressing the optic nerve. The mass was successfully excised in toto with an intact capsule. Postoperative recovery was uneventful, and visual improvement was observed.

Conclusion:

This case underscores the diagnostic challenges of orbital tumors and emphasizes that surgical intervention remains critical in the setting of compressive optic neuropathy. Even in longstanding lesions, complete excision can restore visual function and prevent further irreversible damage..

Keywords: Orbital tumors, compressive optic neuropathy, visual improvement, surgical excision, cavernous venous malformation, optic nerve sheath meningioma, schwannoma, orbitotomy, cryoprobe.

INTRODUCTION:

Orbital tumors, although relatively uncommon, represent an important cause of morbidity due to their proximity to the globe and optic nerve. They can lead to disfiguring proptosis, diplopia, pain, and in many cases progressive visual loss, thereby exerting a profound impact on patients' functional status and quality of life (Nassiri et al., 2015). The burden is particularly significant when tumors cause compressive optic neuropathy, which may result in irreversible blindness if not recognized and managed in time.

Among these causes of these lesions, orbital lymphatic malformations (lymphangiomas) are rare, benign, non-encapsulated vascular anomalies that account for approximately 1–4% of orbital tumors (Mansour et al., 1988; Nassiri et al., 2015). Lymphatic malformations typically present in childhood but may also manifest in adults, producing slowly progressive or acute proptosis, restricted motility, and visual decline. Their infiltrative growth, association with recurrent intralesional hemorrhage, and lack of a defined capsule make diagnosis and treatment particularly challenging. Multiple therapeutic strategies—including observation, sclerotherapy, systemic medical therapy, and surgical debulking—have been reported, yet no universally accepted standard of care exists (Cheng et al., 2019)..

Other benign orbital tumors such as cavernous venous malformations, schwannomas, and optic nerve sheath meningiomas occur more frequently in adults and often mimic lymphatic malformations in both clinical and imaging presentation. CVMs are low-flow vascular malformations typically located in the intraconal space. On MRI, CVM's generally appear as well-circumscribed, homogeneous T2-hyperintense lesions with nodular early enhancement and delayed filling (Park et al., 2024).

ONSMs classically present with painless progressive visual loss and the characteristic “tram-track” and “doughnut” MRI signs (Park et al., 2024; Horowitz et al., 2023; Cameron et al., 2022). Because of these overlaps, preoperative differentiation can be difficult.

Schwannomas arise from Schwann cells of peripheral nerves and may occur in both intraconal and extraconal compartments. Imaging features overlap with CVMs, but comparative analyses suggest that schwannomas more frequently show heterogeneous T2 signal, diffuse early enhancement, and “tail” or “target” signs, whereas CVMs remain more homogeneous (Park et al., 2024).

This case report describes a middle-aged patient with a longstanding orbital mass complicated by compressive optic neuropathy. Imaging initially suggested CVM, schwannoma, or ONSM. However, histopathology examination confirmed a lymphatic malformation (capillary hemangioma subtype). Significant visual improvement following surgical excision highlights both the diagnostic complexity and therapeutic potential in selected cases of orbital lymphatic malformation..

CASE ILLUSTRATION

A 52-year-old woman presented with a chief complaint of progressive protrusion of the left upper eyelid for approximately four years. The swelling had enlarged gradually over time and was accompanied by a decline in visual acuity of the left eye. The patient denied any associated pain, redness, or history of trauma.

Her past medical history revealed a previously diagnosed intraconal orbital tumor of the left eye, with differential considerations including optic nerve sheath meningioma (ONSM), cavernous venous malformation, or schwannoma. She also had a history of type 2 diabetes mellitus but no hypertension. At the time of presentation, her medication regimen included oral glimepiride 4 mg once daily and oral metformin 500 mg three times daily. On clinical examination, the patient's general condition and vital signs were within normal limits. Visual acuity was 6/19 in the right eye and 1/60 in the left eye. Hertel exophthalmometry demonstrated asymmetry, with values of 15–105–19 mm, indicating left-sided proptosis. Intraocular pressure was 12 mmHg in the right eye by palpation, while non-contact tonometry readings were not measurable in the left eye.

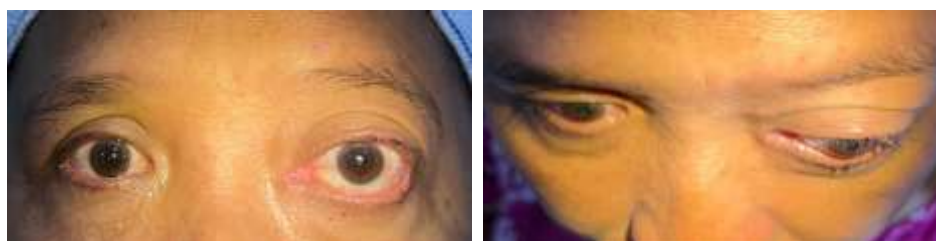


Figure 1. Clinical presentation before surgery

Anterior segment evaluation showed no eyelid edema or spasm, no conjunctival injection, and clear corneas bilaterally. The anterior chamber was deep and quiet in both eyes, and the irides demonstrated a normal radial pattern. Both pupils were round and measured 3 mm in diameter, with positive light reflexes. A relative afferent pupillary defect was absent in the right eye but present in the left eye. Both crystalline lenses were clear.

Fundusoscopic examination of the right eye revealed a normal optic disc with sharp margins and healthy coloration. In contrast, the left optic disc appeared pale, consistent with optic nerve involvement, while the foveal reflex was preserved and the retina appeared otherwise unremarkable.

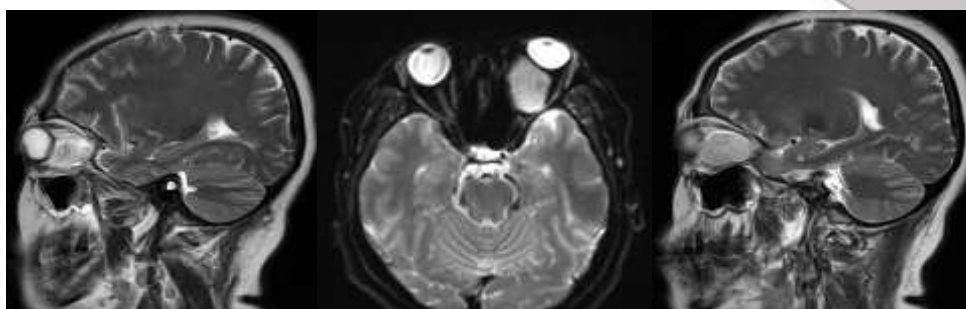


Figure 2. MRI Findings

Magnetic resonance imaging (MRI) of the orbit identified a solid intraconal mass in the left orbit measuring approximately $2.79 \times 2.35 \times 2.16$ cm. The lesion was well defined and showed contrast enhancement. The radiological impression suggested the possibility of orbital lymphoma, orbital meningioma, or orbital schwannoma. Additional findings included left maxillary sinusitis. Thoracic imaging demonstrated no evidence of metastasis to the lungs or visualized skeletal structures, although cardiomegaly was noted.

Pre-surgery laboratory investigations showed a hemoglobin level of 14.8 g/dL, leukocyte count of 6,860/ μ L, and platelet count of 342,000/ μ L. Coagulation studies revealed a prothrombin time of 9.8 seconds and an activated partial thromboplastin time of 23.2 seconds, both within normal ranges. Renal function tests showed a blood urea nitrogen of 11 mg/dL and creatinine of 0.85 mg/dL. Liver function tests were unremarkable, with SGOT at 13 U/L and SGPT at 22 U/L. Electrolyte values were within normal limits. However, blood glucose control was poor, with a random blood glucose of 373 mg/dL and an HbA1C of 11.4%. Hepatitis B surface antigen was non-reactive.

Based on the clinical, radiological, and laboratory findings, the patient was diagnosed with a left orbital tumor involving both extraconal and intraconal compartments. The differential diagnoses included cavernous venous malformation, schwannoma, and optic nerve sheath meningioma. The markedly reduced visual acuity, presence of a relative afferent pupillary defect, and optic disc pallor confirmed the presence of compressive optic neuropathy in the left eye.

The patient was planned for surgical excision of the orbital tumor through a medial orbitotomy with cryoprobe assistance under general anesthesia.

Table 1. Lab Results at Pre-Operation

Parameter	Result	Reference Range
Hemoglobin	14.8 g/dL	12–16 g/dL
Leukocytes	6,860/ μ L	4,000–11,000/ μ L
Platelets	342,000/ μ L	150–450 $\times 10^3$ / μ L
PT	9.8 sec	9–12 sec
aPTT	23.2 sec	23–35 sec
SGOT	13 U/L	<35 U/L
SGPT	22 U/L	<35 U/L
Random Blood Glucose	373 mg/dL	70–140 mg/dL
HbA1C	11.4%	<6.5%
BUN	11 mg/dL	7–20 mg/dL

Creatinine	0.85 mg/dL	0.6–1.2 mg/dL
Electrolytes (Na/K/Cl)	Na 140 / K 4.4 / Cl 101	Within normal limits
HBsAg	Non-reactive	Negative

The patient underwent surgical excision of the orbital mass. A transconjunctival inferomedial approach was selected. A marking was made between the fornix and the tarsus along a 5-mm segment, followed by infiltration of local anesthetic using pehacaine along the incision line. A conjunctival incision was performed along the marking, and blunt dissection was carefully advanced until the anterior surface of the tumor was identified. The posterior boundary of the lesion was delineated in relation to the periosteum, and adhesions of the tumor capsule to surrounding structures were meticulously freed. A combination of periosteal dissection and gentle spreading allowed separation of adherent orbital fat from the tumor surface.

The mass was then mobilized and delivered using a cryoprobe, which facilitated en bloc removal. The lesion was excised in toto with its capsule intact. Intraoperative exploration revealed a well-encapsulated mass with distinct borders, displaying a purplish hue. The lesion extended medially to the orbital apex, closely abutting and compressing the optic nerve.



Figure 3. Surgical mass excision

Hemostasis was secured, and the orbital cavity was irrigated with povidone iodine solution. A surgical pack was placed within the orbit. The Tenon's capsule was reapproximated with two interrupted 6-0 Vicryl sutures, and the conjunctiva was closed with five sutures along the inferior and medial margins. Gentamycin ophthalmic ointment was applied to the ocular surface, and a force suture was placed. The procedure was completed with a pressure dressing.

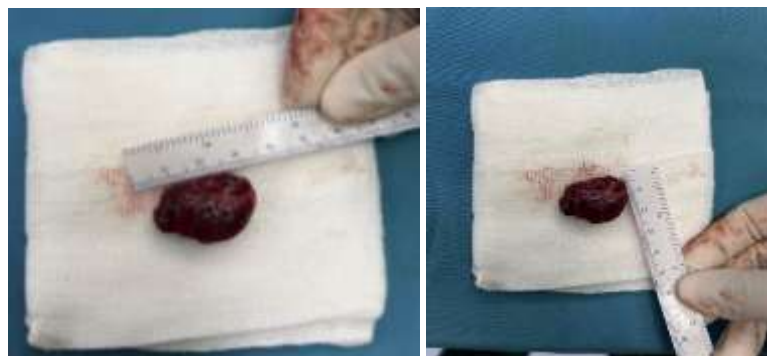


Figure 4. Extracted mass

On the first postoperative day, the patient was stable with no evidence of orbital bleeding and intact ocular surface. At one week, the left eye showed significant improvement in visual acuity to 5/40 with a pinhole correction to 5/30, while the right eye maintained 5/5. Confrontation testing confirmed full visual fields

bilaterally, and there was no recurrence of pain or marked inflammation, although minimal conjunctival hyperemia and chemosis were noted. At two weeks, the patient reported subjective visual improvement, describing her vision as brighter and clearer compared with preoperative status. Examination confirmed stable ocular alignment with mild motility restrictions and residual diplopia in extreme gaze, but without significant pain.

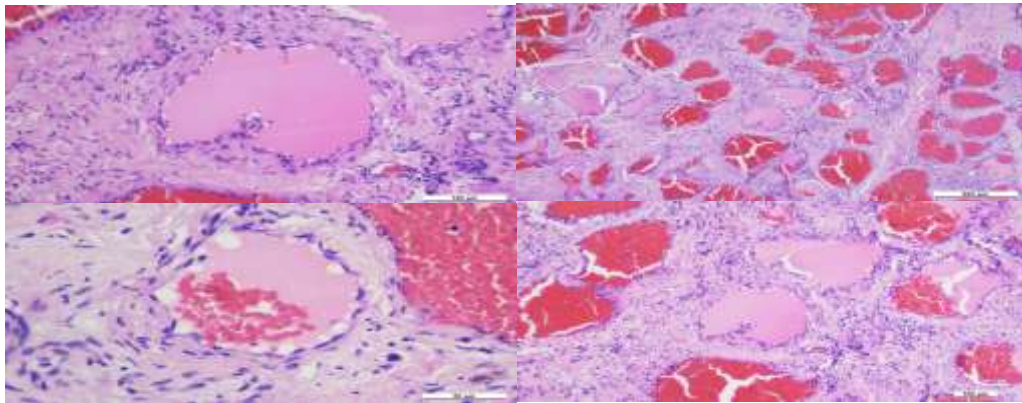


Figure 5. Histopathological result showing signs for lymphatic malformation

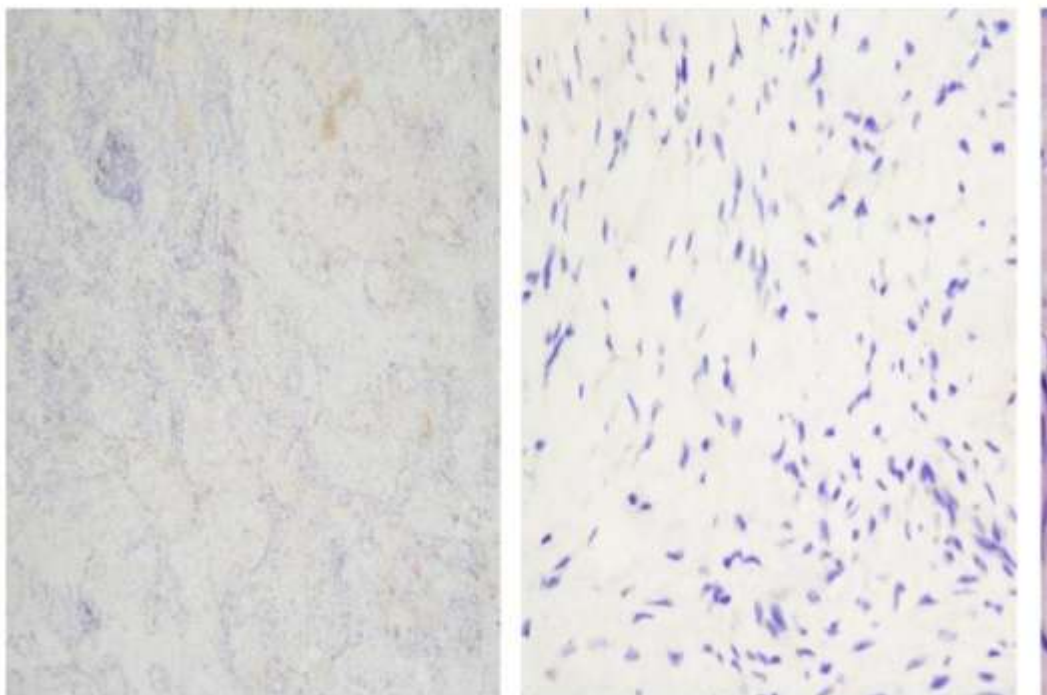


Figure 6. Histopathological with S-100. Immunohistochemical staining shows negative S-100 expression in the tumor cells, excluding schwannoma and supporting the diagnosis of orbital lymphatic malformation

By two months post-operation, best-corrected visual acuity in the left eye had stabilized at 5/40 with pinhole to 5/20, demonstrating sustained improvement compared to the preoperative level of 1/60. Ishihara testing remained limited in the left eye, consistent with chronic optic nerve involvement. Histopathological examination confirmed the lesion as a capillary lymphangioma.

ONH and RNFL OU Analysis: Optic Disc Cube 200x200

OD

OS

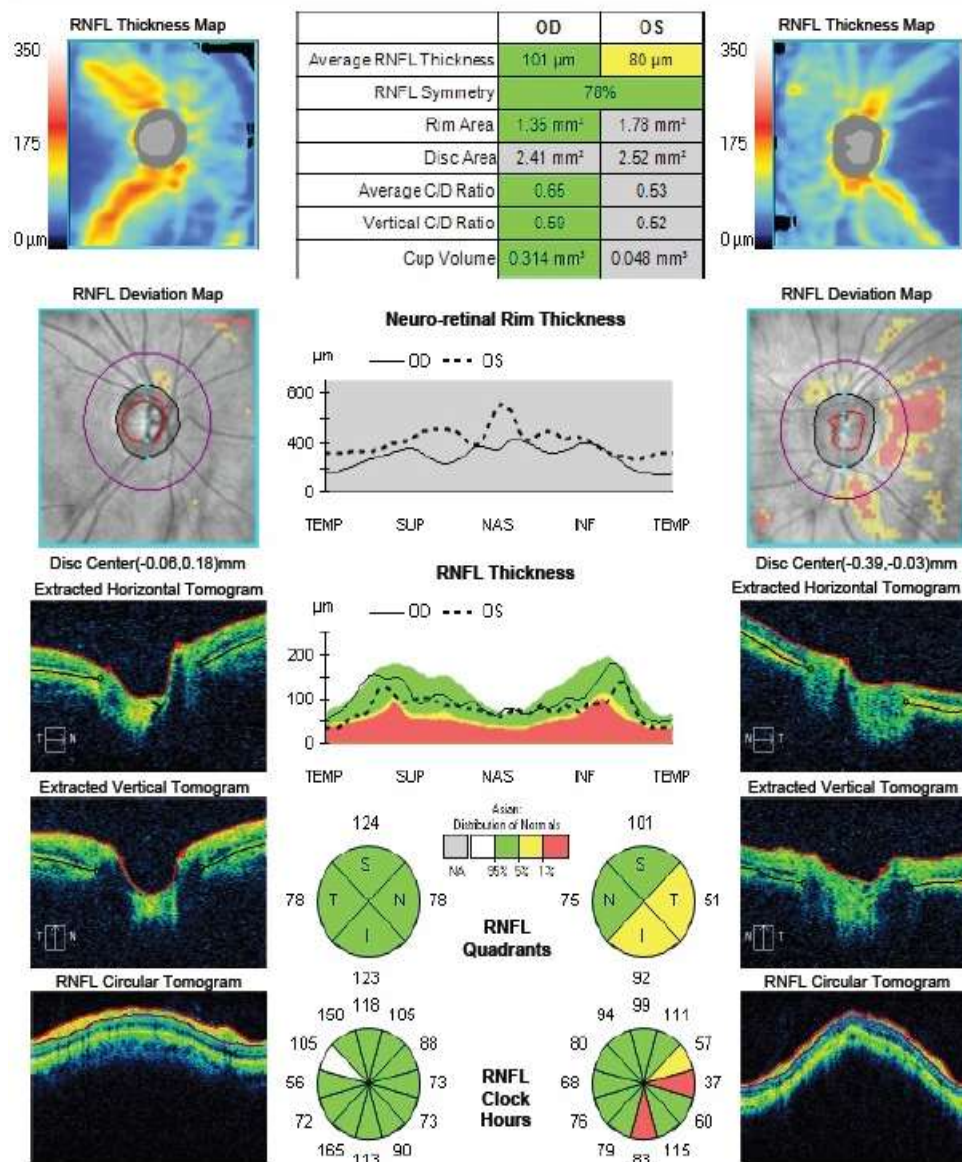


Figure 7. Post Operative RNFL examination (3 months)

Optical coherence tomography (OCT) performed after surgery revealed persistent structural changes in the left eye (OS) consistent with longstanding compressive optic neuropathy. Retinal nerve fiber layer (RNFL) analysis demonstrated significant thinning, particularly in the inferior and temporal quadrants, while ganglion cell complex analysis showed diffuse thinning across all quadrants. Macular cube assessment further indicated reduced macular thickness in OS compared to the fellow eye. These findings reflect chronic axonal and ganglion cell loss secondary to prolonged optic nerve compression.

Macula Thickness OU: Macular Cube 512x128 **OD** **OS**

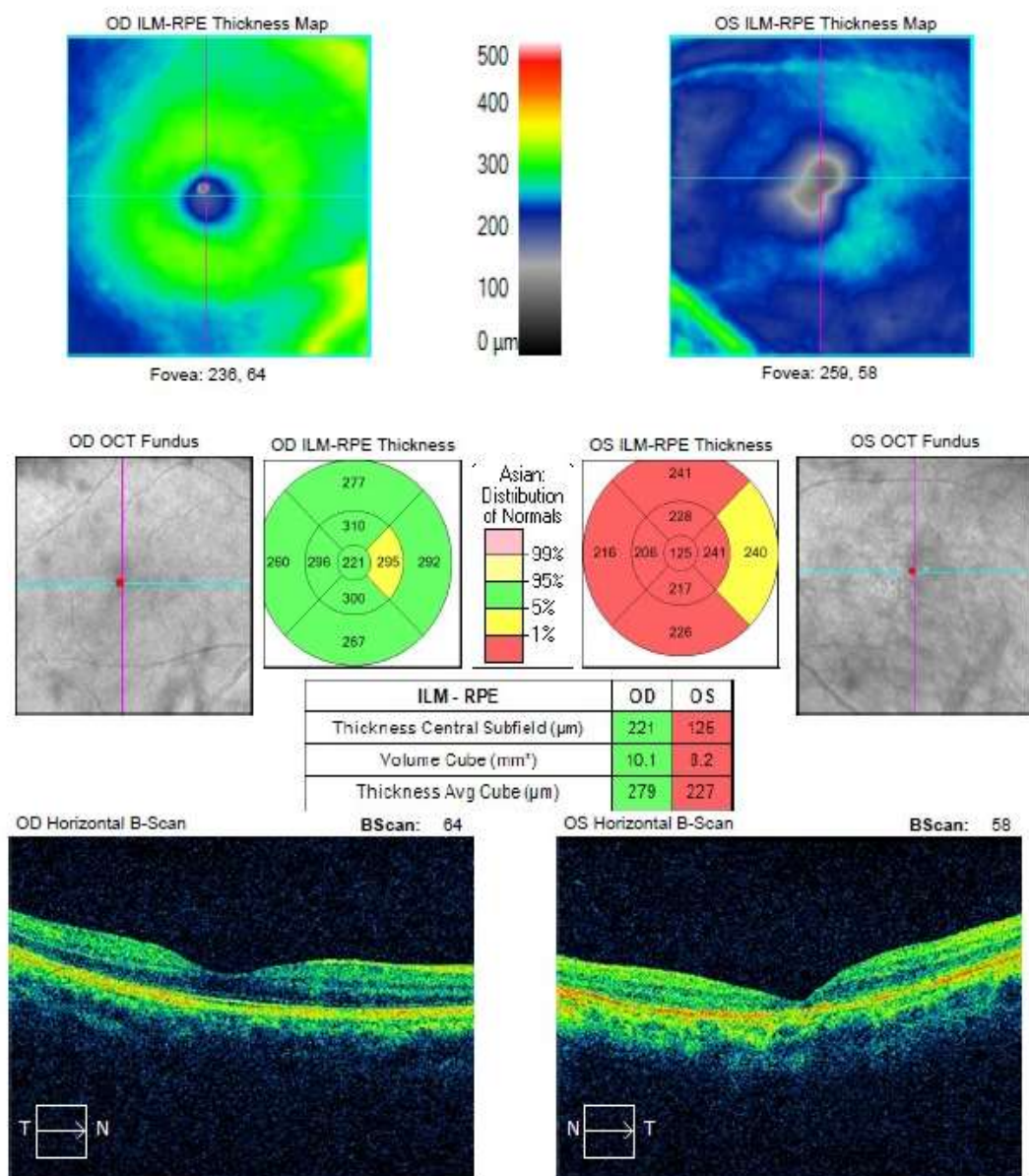


Figure 8. Post Operative OCT (3 months)

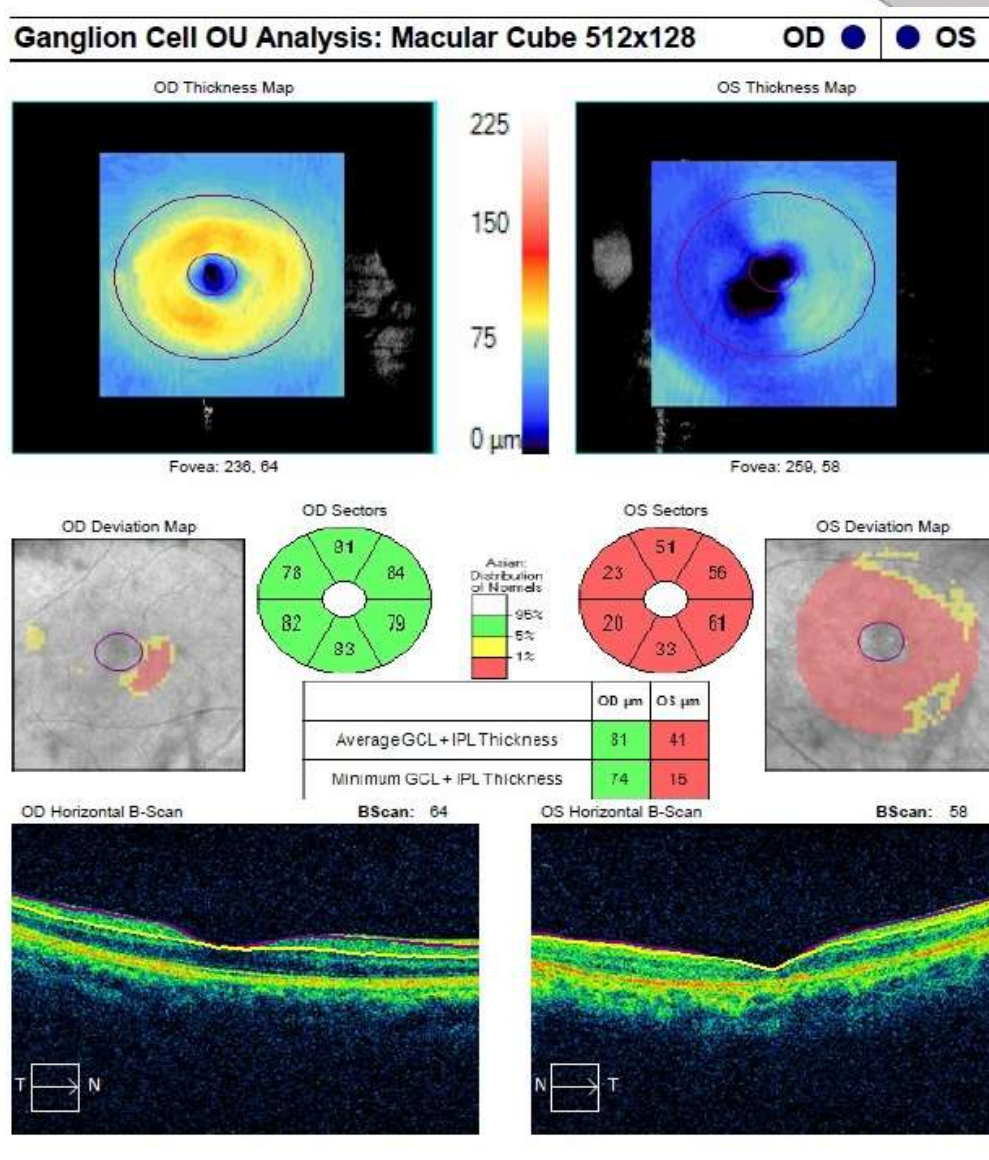


Figure 9. Post Operative macular cube examination (3 Months)

Humphrey Field Analyzer (HFA) 30-2 threshold testing was performed with reliable fixation and reading indices. The right eye (OD) demonstrated visual fields within normal limits, with a Visual Field Index (VFI) of 99%. In contrast, the left eye (OS) showed significant field defects involving the superomedial and inferomedial quadrants, as well as additional deficits in the superotemporal and inferotemporal areas, corresponding to the structural damage seen on OCT. The VFI in OS was 73%, with marked reductions in mean deviation (MD: -11.56 dB) and pattern standard deviation (PSD: 6.50 dB), consistent with functional impairment due to compressive optic neuropathy. These findings highlight the discrepancy between persistent structural and functional deficits and the patient's subjective visual improvement, underlining the value of multimodal assessment in postoperative follow-up.

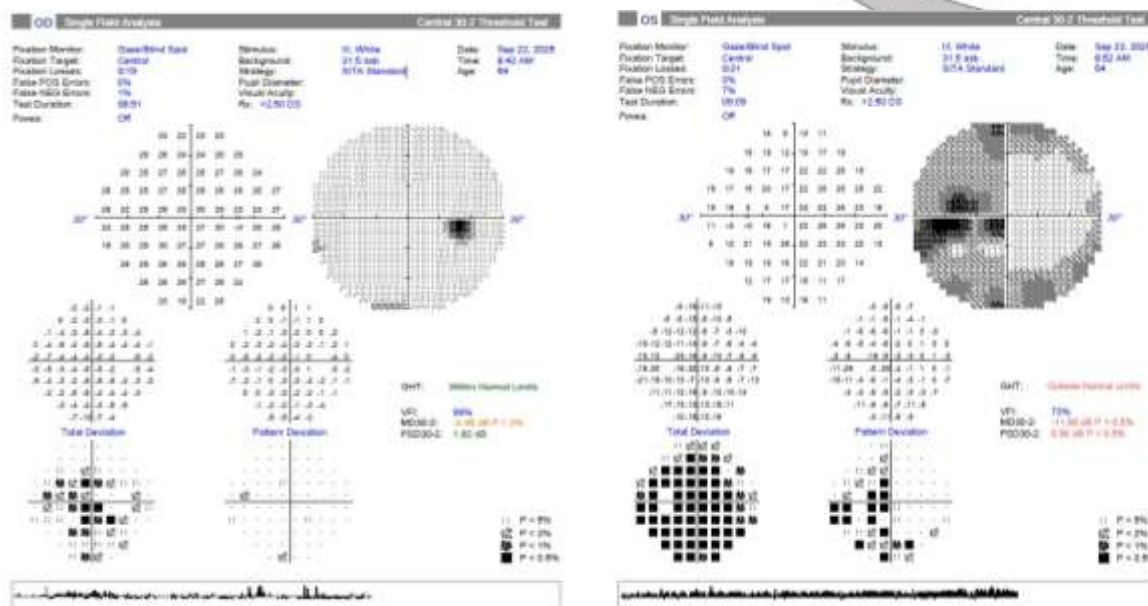
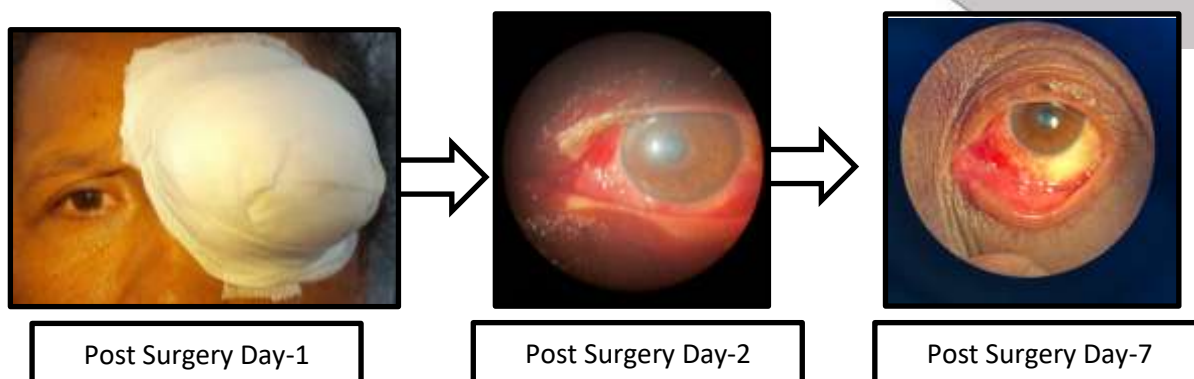


Figure 10. Post Operative HFA analysis (3 months)

This case illustrates the potential for visual recovery even after prolonged compressive optic neuropathy. The patient initially presented with severe visual loss (1/60) due to a large orbital mass compressing the optic nerve for nearly three years. Following meticulous excision of the encapsulated tumor via a transconjunctival orbitotomy, visual acuity improved to 5/40 with pinhole 5/20. Although some functional deficits persisted, including optic disc pallor and limited color discrimination, the restoration of measurable vision underscores the reversibility of optic nerve dysfunction when decompression is achieved. This highlights the importance of timely surgical intervention in orbital tumors with compressive optic neuropathy, even in longstanding cases.

Table 2. Patient's timeline post-operation

Time Point	Visual Acuity OD	Visual Acuity OS	Key Clinical Findings
Day 1	6/19	Not tested (eye under dressing)	Stable vitals, no orbital bleeding, clear cornea, RAPD negative OD, positive OS.
2 months	5/8 (pinhole 5/5)	5/40 (pinhole 5/20)	OS: mild motility limitation (–1 superolateral/lateral), no pain. Persistent optic disc pallor, Ishihara OS 1/14.
3 months	6/12 cc S+1.25 → 6/7.5	6/40 PH 6/19 cc S+1.75 → 6/15 PHNI	Binocular VA 6/7.5. Subjective improvement reported. OS: further functional recovery despite persistent structural changes.



DISCUSSION

In this case, a middle-aged patient presented with a large left orbital tumor involving both extraconal and intraconal compartments, with longstanding proptosis and severe visual loss (preoperative VA 1/60 OS), features consistent with compressive optic neuropathy. After surgical excision via an inferomedial transconjunctival orbitotomy with cryoprobe assistance, vision improved to 5/40 (pinhole 5/20) over two months. The tumor was well encapsulated, allowing complete en bloc removal with minimal disruption of surrounding tissues.

Initially, the differential diagnosis included cavernous venous malformation (CVM), schwannoma, and optic nerve sheath meningioma (ONSM). Orbital schwannomas, in particular, share overlapping imaging features and clinical presentations. Yong et al. (2020) reported that 28% of patients with orbital schwannoma presented with optic neuropathy, with several cases demonstrating postoperative visual improvement. This parallels our case, supporting the concept that decompression—even in longstanding cases—can yield functional recovery. Similarly, Vogele et al. (2022) emphasized that earlier surgical intervention in benign orbital tumors is associated with better visual outcomes.

Histopathological examination in this case confirmed a lymphatic malformation (capillary hemangioma subtype), a rare finding in adults. Orbital lymphangiomas are uncommon overall, representing only 1–4% of orbital tumors, and are typically diagnosed in childhood rather than later in life (Nassiri et al., 2015). Their rarity in adults contributes to diagnostic difficulty, as clinicians often prioritize more common entities such as CVM or schwannoma in the differential. Moreover, lymphatic malformations usually demonstrate an infiltrative, non-encapsulated pattern with poorly defined margins, features that complicate surgical excision and increase recurrence risk (Mansour et al., 1988; Nassiri et al., 2015).

The present case was unusual in that the lesion appeared well encapsulated, permitting safe en bloc removal and deviating from the classic description. Clinically, the patient's history of slow, progressive proptosis with compressive optic neuropathy more closely resembled CVM or schwannoma, rather than the typical presentation of lymphatic malformations with acute swelling episodes or recurrent hemorrhage. This underscores that lymphatic malformations should remain in the differential diagnosis even when clinical and imaging findings do not align perfectly with their usual profile, as pathology remains the gold standard for diagnosis.

Management of orbital lymphatic malformations continues to be debated. The Cochrane review by Cheng et al. (2019) highlighted the absence of randomized controlled trials and reliance on case series. Options include observation, sclerotherapy for macrocystic lesions, systemic agents such as sirolimus for diffuse disease, and surgical debulking in vision-threatening cases. Complete excision is rarely feasible due to their infiltrative nature; however, as demonstrated here, certain subtypes with circumscribed features may be amenable to safe removal.

This case reinforces several key lessons. First, even after three to four years of mass effect and severe preoperative vision loss, meaningful visual recovery can occur if compressive neuropathy is relieved before irreversible axonal loss. Second, detailed preoperative imaging remains essential to define tumor extent and its relationship to the optic nerve, while intraoperative findings such as capsule integrity ultimately dictate surgical feasibility. The inferomedial transconjunctival approach in this case provided safe and direct access, and when combined with capsule-preserving dissection, enabled complete excision. Finally, adjunctive perioperative measures—including high-dose corticosteroids and optimization of systemic comorbidities such as diabetes—

likely supported postoperative recovery.

This case also illustrates the limitations of single-patient reports. Visual outcomes are strongly influenced by the duration and severity of optic nerve atrophy, and follow-up in this case remains relatively short. Longer observation is needed to evaluate long-term stability, recurrence risk, and late complications such as motility restriction or diplopia. Nevertheless, the histopathological confirmation of a lymphatic malformation in an adult expands the clinical spectrum of orbital lymphangiomas and emphasizes the need for clinicians to consider this rare entity, even in atypical presentations.

CONCLUSION

This case highlights the diagnostic challenges associated with orbital tumors and underscores the importance of excisional biopsy in the evaluation of indeterminate orbital lesions. In addition to establishing a definitive histopathological diagnosis, surgical excision can prevent further irreversible tumor-related damage and restore visual function. In the present case, despite years of progressive visual loss due to compressive optic neuropathy from a longstanding orbital lymphatic malformation, the patient experienced meaningful visual recovery following complete surgical excision.

Although orbital lymphatic malformations are typically infiltrative and often difficult to manage surgically, this case demonstrates that complete en bloc excision can be safely achieved when the lesion is well encapsulated and surgically accessible. Detailed preoperative imaging was essential for narrowing the differential diagnosis, anticipating surgical complexity, and guiding the optimal surgical approach. Capsule-preserving excision, together with meticulous perioperative management, contributed to a favorable functional outcome. This report also emphasizes that delayed presentation should not preclude surgical intervention. While early recognition remains important to minimize irreversible optic nerve damage, selected patients may still achieve significant visual improvement after surgery. Overall, this case adds to the growing evidence that atypical adult orbital lymphatic malformations can be successfully managed with complete surgical excision when supported by careful preoperative evaluation and multidisciplinary planning.

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