Content available at: https://www.ipinnovative.com/open-access-journals

IP International Journal of Ocular Oncology and Oculoplasty

Journal homepage: https://ijooo.org/

Case Report

Case report of lymphovascular malformation

Thummathati Sivapriya[1]*

[1]*Dept. of Ophthalmology, Minto ophthalmic institutions, Bengaluru, Karnataka, India*

ARTICLE INFO

Article history:
Received 01-11-2024
Accepted 02-12-2024
Available online 14-01-2025

Keywords:
Lymphovascualr malformation
Hemangioma
Orbitotomy

ABSTRACT

To report a rare presentation of lympho-vascular malformation, a 42-year-old female presented with a one year history of progressive left eye protrusion, restricted ocular movements, and diminished vision. Clinical examination revealed facial asymmetry and axial proptosis of the left eye. Contrast-enhanced computed tomography demonstrated a well-defined, lobulated, soft-tissue attenuating lesion in the intraconal compartment of the left retrobulbar region, suggestive of a lymphovascular malformation. An anterior orbitotomy was performed via a lateral approach, and the excised lesion was sent for histopathological examination (HPE), which confirmed the diagnosis of cavernous hemangioma. Our case highlights importance of keeping differential diagnosis of cavernous hemangioma in cases of lympho vascular malformation. It is very much important to have differential diagnosis of cavernous hemangioma in cases of lympho vascular malformation. Misdiagnosing cavernous hemangioma as another orbital lymphovascular malformation can lead to inappropriate management, such as unnecessary interventions or delayed treatment, increasing the risk of vision loss or complications. Accurate differentiation via imaging and clinical evaluation is crucial for optimal outcomes.

1. Introduction

Vascular malformations are classified into four types: arteriovenous malformations, hemangiomas, venous angiomas, and capillary telangiectasias. Hemangiomas are the second most common among these. They are benign, slow-growing lesions that are typically well-circumscribed.[1]

Approximately 80% of orbital hemangiomas are located in the intraconal compartment, situated between the Tenon's capsule and the extraocular muscles, often in the lateral region. They are most commonly observed in middle-aged women.[2] They arise from the development of new blood vessels, the growth of tissue elements within the vessel wall, and the hyperplasia of cells typically involved in the formation of vascular tissue.[3] Case reports on cavernous hemangiomas are critical for enhancing clinical knowledge, improving diagnosis, and refining treatment approaches for this common yet potentially impactful orbital lesion.

2. Case Presentation

A 43-year-old female with no significant medical or surgical history presented with a one-year history of gradual, insidious protrusion of the left eye. She also reported diminished vision in the left eye for the past four months, accompanied by restricted extraocular movements. There were no complaints of pain or double vision.

On examination, axial proptosis of the left eye was observed, accompanied by lagophthalmos and scleral show, with the upper eyelid positioned higher than that of the right eye. (Figure 1) Extraocular movements were restricted in the left eye. Intra ocular pressure and fundus examination in both eyes normal. The remainder of the ophthalmologic

* *Corresponding author*.
E-mail address: t.sivapriya27@gmail.com (T. Sivapriya).

https://doi.org/10.18231/j.ijooo.2024.039

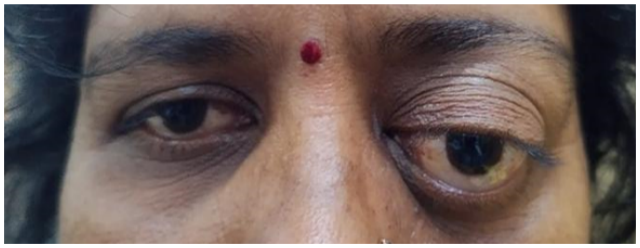

Figure 1: Clinical photograph of patient with left eye showing axial proptosis.

evaluation and systemic evaluation was unremarkable, with no evidence of inflammatory signs.

On contrast enhanced CT well defined lobulated soft tissue attenuating lesion of 2.8 * 3.0 * 2.6 cm in intra conal compartment of left retrobulbar region -likely of lymphovascular malformation was noted. On post constrast study the lesion showed few patchy areas of enhancement with average HU -84 with few non enhancing areas within. (Figure 2a&b)

CT brain was normal with no significant neuroparenchymal abnormality. (Figure 3)

The tumor excision was done under general anaesthesia. A lateral canthotomy and cantholysis performed to access the lateral orbital rim. Lateral rectus and superior oblique are dissected and intraconal space approached. Tumor identified and carefully dissected from surrounding structures using blunt dissection and lateral rectus, superior oblique were resutured. (Figure 4)

Sample was sent for histopathological examination. Patient withstood the procedure well and Surgery was uneventful. (Figure 5) Histopathological examination was suggestive of cavernous hemangioma.

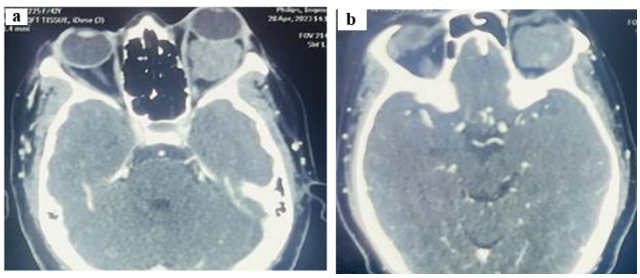

Figure 2: a & b: CECT orbit showing well defined lobulated soft tissue attenuating lesion in intra conal compartment of left retrobulbar region at different cross sections with no evidence of calcification, bony erosion or remodelling.

No evidence of extension in to optic canal or orbital apex.

Close monitoring was done to detect signs of infection, swelling, or bleeding. Eye movements and vision were assessed regularly, and anti-inflammatory medications may be prescribed to reduce swelling.

Figure 3: CT Brain showing no significant neuroparanchymal abnormality. No obvious evidence of mass effect / hemorrhage /any parenchymal lesion noted.

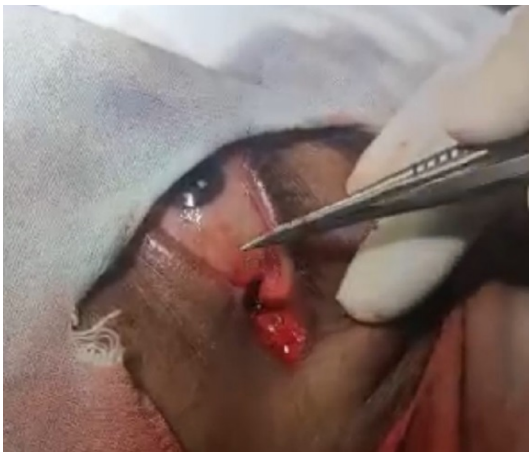

Figure 4: Surgical excision through lateral approach anterior orbitotomy

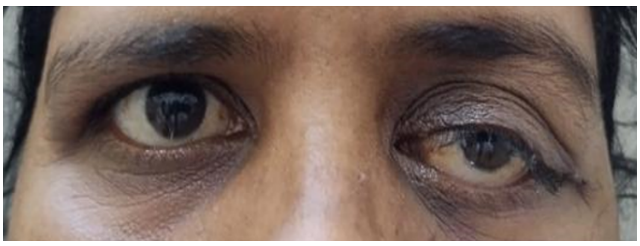

Figure 5: Clinical photograph of patient postoperatively

Potential Complications which include recurrence, diplopia (double vision), vision loss, and ptosis (drooping eyelid) due to surgical manipulation of adjacent orbital structures are assessed in the follow up period.

Importance of Periodic imaging to monitor for residual or recurrent hemangioma and ensure no impact on the optic nerve or surrounding tissues.

3. Discussion

Hemangiomas are categorized into capillary and cavernous types. They are non-capsulated poorly circumscribed lesions. Capillary hemangiomas typically appear within the first year of life.

In contrast, cavernous hemangiomas usually develop between the second and fourth decades of life and can be asymptomatic or symptomatic. Due to their asymptomatic nature in early stages, these tumors may go undiagnosed for years before clinical signs become apparent . It can present with features like proptosis, diplopia, and visual disturbances by optic nerve compression or as a result of high degree of relative hyperopia.[4]

Studies state that this lesion is more common in women, probably due to circulating estrogen/progesterone levels affecting the progression of orbicular cavernous hemangioma Hormones like estrogen may promote endothelial proliferation and angiogenesis, contributing to tumor growth. Studies have observed tumor enlargement during periods of hormonal surges, such as pregnancy. However, data are limited, and further research is needed.[5]

The middle third of the orbit is the most common site often occurring within the intraconal space. This typically results in progressive axial proptosis, which is the primary sign and symptom.[6]

Diagnosis is majorly by imaging modalities like computed tomography with contrast, magnetic resonance and angiography.

Computed tomography (CT) reveals the lesion as a well-defined, encapsulated, hyperdense structure with mild enhancement following contrast injection, though less than the surrounding muscles. Contrast-enhanced imaging can help differentiate cavernous hemangiomas from other orbital masses, such as optic nerve gliomas or orbital lymphoma, which may present with similar clinical features. Magnetic resonance imaging (MRI) is essential for evaluating potential compressive effects, particularly on the optic nerve.[7]

Depending on the case treatment can either be conservative or surgical. For asymptomatic cases, management is typically conservative. However, when the lesion causes symptoms, especially those affecting vision or resulting in noticeable cosmetic changes like proptosis, surgical removal is recommended. With advancements in surgical approaches, the procedure now carries a lower risk of complications, and the outlook for vision recovery post-surgery is generally better. Post operative impairment of vision was mainly attributed to incomplete or difficulty in removal, adhesion to periosteum, optic nerve or muscle.[8]

Histological examination often reveals a well-encapsulated lesion, which helps in distinguishing orbital cavernous hemangiomas from other orbital tumors. There is minimal inflammatory infiltrate, and the lesion typically lacks the aggressive features seen in malignant vascular tumors.[9]

3.1. Differential diagnosis

Pseudotumor which is an inflammatory condition with acute onset, causing pain, swelling, and possible restriction of eye movements, MRI shows diffuse orbital involvement without a well-defined mass, often with muscle involvement.

In Optic Nerve Sheath Meningioma gradual onset of vision loss and proptosis, with MRI showing a fusiform mass surrounding the optic nerve, often with a "tram-track" appearance and enhancing margins are noted.

4. Conclusion

This case underscores the importance of considering cavernous hemangioma as a differential diagnosis in suspected lymphovascular malformations.

Orbital cavernous hemangioma is a common diagnosis in adults presenting with progressive proptosis and an orbital mass. Diagnosis is primarily based on clinical evaluation and imaging techniques, with MRI being the gold standard for identifying the tumor's location and characteristics. Histopathological examination confirms the diagnosis, revealing characteristic blood-filled vascular spaces surrounded by fibrous tissue.

Surgical management should be tailored to the specific characteristics of each case for symptomatic lesions, especially those affecting vision or causing cosmetic concerns. Incomplete Lymphovascular excision of the lesion increases the risk of recurrence, necessitating close monitoring of the patient's clinical course.

With advances in surgical techniques, the prognosis for visual recovery is generally favorable, and complications are rare. Timely diagnosis and appropriate management are key to ensuring positive outcomes for patients with orbital cavernous hemangiomas.

5. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

6. Source of Funding

None.

7. Conflicts of Interest

None.

References

1. Steiner JE, Drolet BA. Classification of vascular anomalies: an update. *Semin Intervent Radiol*. 2017;34(3):225–32.
2. Anand R, Deria K, Sharma P, Narula M, Garg R. Extraconal cavernous hemangioma of orbit: a case report. *Indian J Radiol Imaging*. 2008;18(4):310–2.
3. Rosca TI, Pop MI, Curca M, Vladescu TG, Tihoan CS, Serban AT, et al. Vascular tumors of the orbit-cavernous and capillary hemangiomas. *Ann Diagn Pathol*. 2006;10(1):13–9.
4. Harris GJ, Jakobiec FA. Cavernous hemangioma of the orbit: an analysis of 66 cases. *J Neurosurg*. 1979;51(2):219–28.
5. Rootman J, Hay E, Graeb D, Miller R, Robertson W. Orbital-adnexal cavernous hemangiomas: a spectrum of apparently related lesions. *Ophthalmology*. 1986;93(3):1558–70.
6. Mcnab AA, Selva D, Hardy TG, O'Donnell B. The anatomical location and laterality of orbital cavernous haemangiomas. *Orbit*. 2014;33(5):359–62.
7. Dallaudiere B, Benayoun Y, Boncoeur-Martel MP, Robert PY, Adenis JP, Maubon A, et al. Imaging features of cavernous hemangiomas of the orbit. *J Radiol*. 2009;90(9):1039–45.
8. Missori P, Tarantino R, Delfini R, Lunardi P, Cantore G. Surgical management of orbital cavernous angiomas: prognosis for visual function after removal. *Neurosurgery*. 1994;35(1):34–8.
9. Bilaniuk LT. Orbital vascular lesions: role of imaging. *Radiol Clin North Am*. 1999;37(1):169–83.

Author's biography

Thummathati Sivapriya, Junior Resident Ophthalmology https://orcid.org/0009-0009-1879-405X

Cite this article: Sivapriya T. Case report of lymphovascular malformation. *IP Int J Ocul Oncol Oculoplasty* 2024;10(4):220-223.