



Case Report

Thrombosed orbital varix of the inferior ophthalmic vein: A rare cause of acute unilateral proptosis

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ABSTRACT

Background: Orbital varices are rare, accounting for only 0–1.3% of orbital masses. They can be found incidentally or cause mild to serious sequelae, including hemorrhage and optic nerve compression.

Case Description: We report a case of a 74-year-old male with progressively painful unilateral proptosis. Imaging revealed the presence of an orbital mass compatible with a thrombosed orbital varix of the inferior ophthalmic vein in the left inferior intraconal space. The patient was medically managed. On a follow-up outpatient clinic visit, he demonstrated remarkable clinical recovery and denied experiencing any symptoms. Follow-up computed tomography scan showed a stable mass with decreased proptosis in the left orbit consistent with the previously diagnosed orbital varix. One-year follow-up orbital magnetic resonance imaging without contrast showed slight increase in the intraconal mass.

Conclusion: An orbital varix may present with mild to severe symptoms and management, depending on case severity, ranges from medical treatment to escalated surgical innervation. Our case is one of few progressive unilateral proptosis caused by a thrombosed varix of the inferior ophthalmic vein described in the literature. We encourage further investigation into the causes and epidemiology of orbital varices.

Keywords: Ophthalmic vein, Orbit, Proptosis, Thrombosis, Varix, Orbital, Thrombosed

INTRODUCTION

An orbital varix is a rare pathology that has abnormally distensible, thin-walled veins. It can be primary due to congenital venous malformation or secondary due to orbital venous outflow obstruction.^[4] It equally affects males and females and can occur in different age groups.^[5] Most cases are unilateral, affecting superior or inferior ophthalmic veins.^[5] It can be asymptomatic or present with intermittent painless proptosis.^[5]

Orbital varices may be found in the intraconal space, a conical area posterior to the eyeball which includes arteries, nerves, and central orbital fat. The anterior boundary includes Tenon's capsule of the posterior half of the eye; the posterior border is the orbital apex tendinous ring, and the

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peripheral boundary includes the four recti muscles with intermuscular septa and extraconal space. The intraconal space itself can be further separated into the central, medial, and lateral orbital spaces based on location in relation to the optic nerve. Due to the extremely innervated and vascular compact area of the intraconal space, it may be described as a rather limited and challenging surgical area.

We present a rare case of unilateral progressive proptosis caused by a secondary orbital varix due to thrombosis of the inferior ophthalmic vein.

CASE PRESENTATION

A 74-year-old male was admitted to the emergency department with a 3-day history of progressive painful proptosis of the left eye. His medical history included hypertension and hyperlipidemia with no history of trauma. Clinical examination revealed a left proptotic orbit with chemosis and scleral injection [Figure 1]. Visual acuity of the left eye was 20/150. The right eye was unremarkable; the patient had no fever nor other infectious signs.

Imaging

Computed tomography (CT) scan showed an ovoid mass in the lateral intraconal space with surrounding infiltration and increased vascularity. CT angiogram revealed a well-circumscribed heterogeneously enhancing mass within the inferior intraconal space of the left orbit (measuring 1.8×1.7 cm round) with periorbital soft-tissue swelling, suggesting inflammation [Figure 2]. Magnetic resonance imaging (MRI) angiogram confirmed the findings of past studies: orbital cellulitis was apparent, the left inferior ophthalmic vein was prominent as compared to the right, and there was a partly thrombosed venous varix of the inferior ophthalmic vein [Figure 3]. Imaging did not show any evidence of arteriovenous fistula.

Differential diagnosis

At this point, potential considerations included orbital cavernous malformation, metastatic tumor causing venous outflow stenosis, arteriovenous malformation, dural arteriovenous fistula, and thrombosed venous varix due to orbital cellulitis.

Management

A neurosurgical consult was requested by the ophthalmology team. To further delineate the nature of the lesion, a diagnostic cerebral angiogram was performed, which revealed an intraconal venous varix [Figure 4]. Consideration was made for transvenous embolization or direct orbital puncture and embolization along with

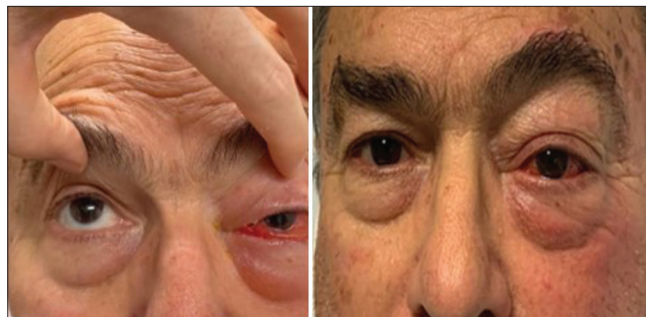


Figure 1: Photographs showing left eye proptosis with chemosis and scleral injection.



Figure 2: Axial computed tomography angiogram showing a well-circumscribed soft-tissue mass within the inferior intraconal space of the left orbit measuring 1.8 ± 1.7 cm.

surgical removal. A mutual decision of conservative treatment without intervention was reached, with aggressive interventions reserved if the patient declined further. Intravenous methylprednisolone for 5 days was initiated, as well as a maintained course of oral corticosteroids, doxycycline, and levofloxacin thereafter.

Patient status and follow-up

The patient was seen in the clinic 2 weeks after discharge. His symptoms had significantly improved, and he denied any eye redness, diplopia, or pain. On examination, there was no chemosis or scleral injection, but there was a significantly improved mild, painless proptosis [Figure 5]. A 2-week follow-up CT scan of the orbit with contrast demonstrated a stable soft-tissue intraconal mass in the left orbit with peripheral venous phase enhancement in addition to central enhancement, consistent with the previously diagnosed thrombosed orbital venous varix of the inferior ophthalmic vein [Figure 6]. At this time, the patient had completed his

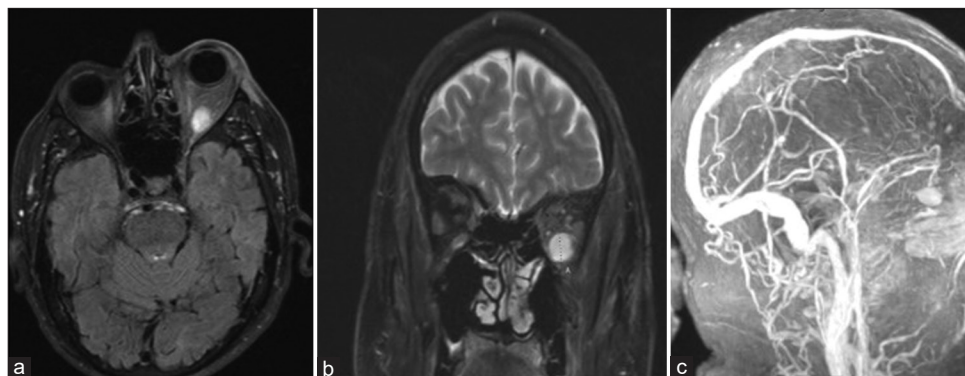


Figure 3: Magnetic resonance imaging angiogram axial (a), coronal (b), sagittal, and (c) showing hyperdense left inferior intraconal hyperdense well-circumscribed mass in the left orbit.

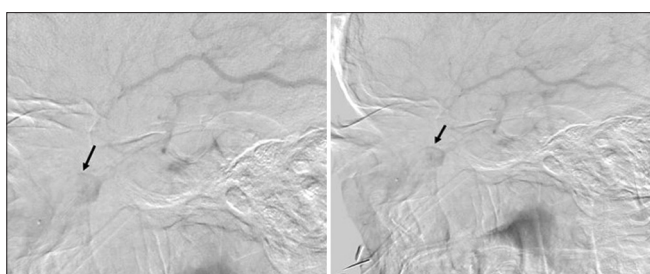


Figure 4: Diagnostic cerebral angiogram showing intraconal venous varix (arrows point) measuring 8 mm in diameter.



Figure 5: Photograph of the patient at the follow-up visit showing the absence of chemosis and scleral injection with mild proptosis in the left eye.

oral antibiotic course and was tapering his prednisone course. A 1-year orbital MRI was conducted and showed a slight increase in the enhancing intraconal mass in the left orbit [Figure 7]. The patient had no symptoms. Since the mass size had not significantly increase and the patient's condition was stable, no additional imaging or clinical follow-up was required per standard of care.



Figure 6: Two weeks follow-up computed tomography orbit with contrast showing stable soft-tissue intraconal mass in the left orbit with central enhancement in consistent with the previously diagnosed thrombosed orbital varix.

DISCUSSION

Orbital varices are rare vascular malformations that account for only 0–1.3% of all histologically identified orbital masses.^[3] Reports of a thrombosed orbital varix are scarce in scientific literature. Few cases have been described,^[1,2,7] which makes it challenging for clinicians to identify and appropriately manage. Our case is one of very few to describe progressive unilateral proptosis caused by a thrombosed varix of the inferior ophthalmic vein. The etiology of orbital varices is mostly unknown, and minimal literature about its pathophysiology is available.^[7] Symptoms can range from mild, painless proptosis to more severe symptoms, such as diplopia and variceal hemorrhage, by which the latter can result in optic nerve compression and visual field defects.^[7] A thrombosed orbital varix can arise as sequelae



Figure 7: One-year orbital magnetic resonance imaging follow-up without contrast, showing a slight increase of the intraconal mass size in the left orbit.

of inflammatory conditions such as orbital cellulitis, orbital infiltration, and compression of venous outflow by primary nerve sheath tumor, metastatic tumors, or lymphoproliferative disease. Differential diagnosis includes cavernous venous malformation, carotid-cavernous fistula, and dural arteriovenous fistula.

Orbital varix can be diagnosed clinically through observation of worsening proptosis proceeding the Valsalva maneuver. However, confirmation with imaging is required, especially if hemorrhage or thrombosis are suspected, and is instrumental in excluding differential diagnoses (i.e., – soft-tissue neoplasm or arteriovenous fistula). CT scan with contrast is the preferred imaging modality, on which a varix appears as a smooth or heterogeneous enhancing soft-tissue mass.^[7] In the case of thrombosis, orbital varix may demonstrate patchy enhancement.^[7] MRI angiogram can be used as an affirmative to CT contrast studies. Advantageously, it also allows for the identification of a draining varix vein. Catheter angiography can play a role in intervention or diagnosis by showing engorgement of the varix mass (especially with a provocative Valsalva maneuver), demonstrating the draining vein, and excluding other orbital vascular anomalies that may mimic orbital varices, such as an arteriovenous fistula or cavernous malformation.

Given the potential significant complications of an orbital varix, such as hemorrhage, blindness, and challenging surgical exposure associated with surgical excision, conservative management with observation is typically recommended.^[5,7] Surgery is usually reserved for recurrent painful proptosis, cosmetic disfigurement, or optic nerve compression.^[5,7] Transvenous endovascular coiling and transvenous sclerotherapy have been recently adopted in symptomatic cases as a feasible and safe substitute for

surgical excision.^[6] Some cases may resolve with solely medical management, as experienced in our reported case. A 5-day course of intravenous methylprednisolone followed by a course of oral antibiotics and tapered oral prednisone led to a profound improvement in our patient's condition.

CONCLUSION

An orbital varix may present with mild to severe symptoms with potentially fatal sequelae if not promptly and adequately treated. Management, depending on case severity, ranges from medical treatment to escalated surgical innervation. Our case is one of few progressive unilateral proptosis caused by a thrombosed varix of the inferior ophthalmic vein described in the literature. We encourage further investigation into the causes and epidemiology of orbital varices.

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Authors' contributions

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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