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CASE REPORT

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Unilateral orbital schwannoma arising from the supraorbital nerve: Report of a rare case

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Key Clinical Message

Schwannomas are rare tumors in the orbit, typically originating from various nerves and presenting diagnostic challenges. We present a unique case of a unilateral orbital schwannoma arising from the supraorbital nerve. A 55-year-old female presented with a painless, slowly growing mass in the right superior orbit, causing proptosis. Visual acuity remained unimpaired, and clinical examination revealed a well-defined mass in the superior orbit. A provisional diagnosis of an orbital dermoid or cyst was made, leading to excision biopsy. The histopathological examination confirmed a diagnosis of benign schwannoma. Schwannomas in the orbit, particularly those arising from the supraorbital nerve, are uncommon and often challenging to diagnose. Early surgical intervention is crucial to prevent complications associated with tumor growth. This case underscores the need to consider schwannomas as a differential diagnosis for slow-growing orbital masses in adults and emphasizes the importance of timely management to prevent vision-threatening complications.

KEYWORDS

histopathological examination, orbital tumor, proptosis, schwannoma, supraorbital nerve

1 | INTRODUCTION

Orbital tumors present a diagnostic challenge to neurosurgeons and have long been a subject of intrigue. Schwannomas in the orbit are exceptionally rare, accounting for only 1%–6% of all orbital tumors.[1,2] In the orbit, schwannomas typically manifest unilaterally and can originate from various nerves, including the supraorbital, infraorbital, supratrochlear, ciliary, oculomotor, trochlear, or abducens nerves. The diverse clinical presentation and locations of these tumors often make diagnosis difficult, with confirmation often requiring histopathological examination. This report describes an unusual case of a solitary, benign schwannoma in the superior orbit, diagnosed through histopathology.

2 | CASE REPORT

2.1 | Patient presentation

A 55-year-old female presented with a painless, gradually enlarging mass in the right superior orbit that had been evolving over the past 4 years. Initially, she did not report

any visual acuity issues, diplopia, or field defects. However, upon further inquiry, the patient mentioned occasional diplopia experienced specifically in up gaze over the last 6 months. Upon examination, her visual acuity in both eyes was 20/20 without corrective lenses. The right eye displayed a solid, non-tender, freely mobile mass situated in the superior orbit, resulting in proptosis. The dimensions of the mass measured approximately 20 mm × 15 mm. Notably, pupillary reactions, color vision, and visual fields were within normal limits. Sensation in the supraorbital, infraorbital, and corneal regions appeared intact. Moreover, a comprehensive assessment of cranial nerves and neurological functions revealed no abnormalities. To supplement these findings, additional ophthalmological details were incorporated. This included slit lamp and fundus examinations of the eye, which demonstrated no noticeable irregularities or optic nerve involvement. The patient reported subjective diplopia in up gaze, correlating with observed inferior dystopia caused by the mass. Notably, despite the diplopia, there were no compensatory head positions or evident limitations in ocular motility during routine eye movements. Examination of the left eye and a general physical examination revealed unremarkable findings consistent with the initial assessment.

2.2 | Radiological evaluation

A contrast-enhanced MRI scan revealed a non-enhancing, well-defined, homogenous mass with a right-shaped appearance. The mass measured 20 mm × 12 mm and was in the right superior orbit. The presence of this mass was causing displacement of the right eye (Figure 1).

2.3 | Diagnosis and surgical intervention

Based on the clinical presentation and radiological findings, a provisional diagnosis of an orbital dermoid or cyst was made. Consequently, the patient was scheduled for an excision biopsy. The surgical procedure involved a bicoronal incision and right frontal supraorbital craniotomy (Figure 2). Intraoperatively, a smooth, well-encapsulated mass was identified, which was firmly attached to the supraorbital nerve. The mass was carefully removed en bloc while preserving the integrity of the capsule (Figure 3).

2.4 | Postoperative recovery

The patient experienced an uneventful recovery following the surgical procedure, and the proptosis subsided.

2.5 | Histopathological examination

A histopathological examination of the excised specimen revealed a gross tissue piece with a circumscribed, globular appearance measuring 2.5 cm in diameter. Microscopically, the sections displayed spindly cells with wavy nuclei, and evidence of myxoid degeneration was observed. This examination led to the final histological diagnosis of a benign schwannoma (Figure 4).

2.6 | Outcome

The patient's proptosis showed significant improvement within a day following the surgical intervention. A

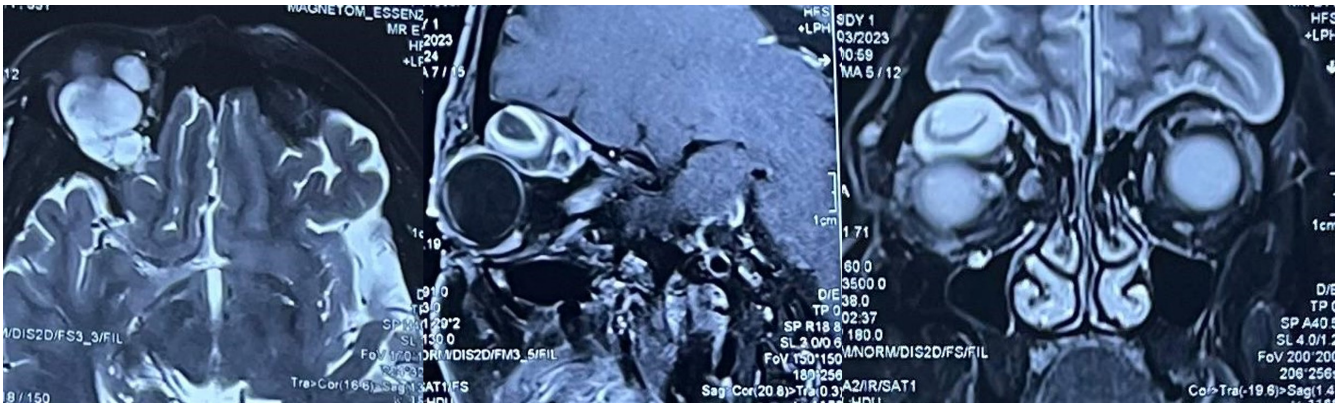

FIGURE 1 Brain MRI (T2-weighted axial, sagittal and coronal images) showing a well-defined, homogenous mass with a right-shaped appearance measuring 20 mm × 12 mm in the right superior orbit. The presence of this mass is resulting in the displacement of the right eye.

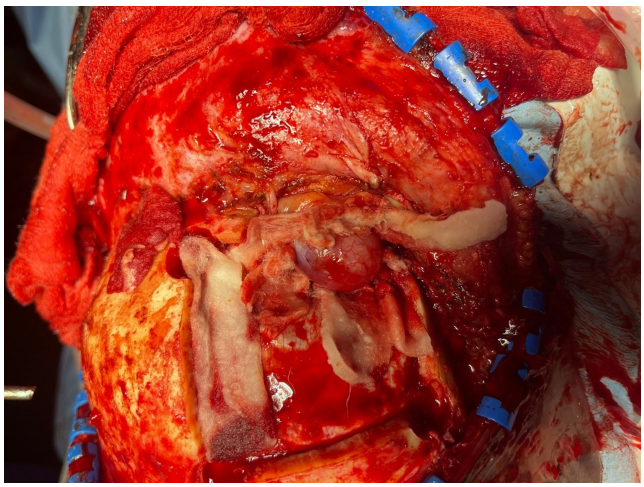

FIGURE 2 Intraoperative image during the surgical procedure. A bi-coronal incision and right frontal supraorbital craniotomy were performed to access the right superior orbit. The image shows the non-enhancing, well-defined, homogenous orbital mass (20 mm × 12 mm) responsible for right eye displacement.

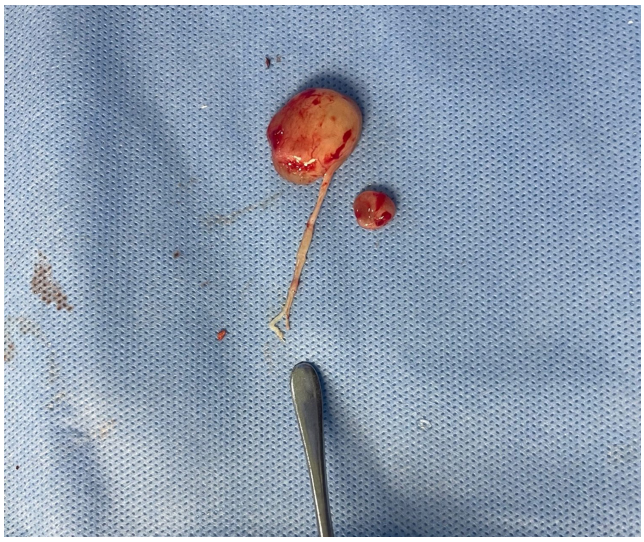

FIGURE 3 Intraoperative image depicts a surgical scene with a focus on the removed tumor mass. The tumor, previously situated in the right superior orbit, is now displayed on the operating table. It appears as a well-defined, non-enhancing mass, measuring approximately 20 mm × 12 mm. The surgical procedure was successful in excising the tumor en bloc while preserving the integrity of its capsule. The image captures a pivotal moment in the surgical procedure, highlighting the meticulous work of the surgical team.

postoperative CT scan confirmed the complete removal of the mass and the resolution of proptosis (Figure 5).

3 | DISCUSSION

Orbital masses encompass a wide spectrum of conditions, from benign growths to potentially life-threatening

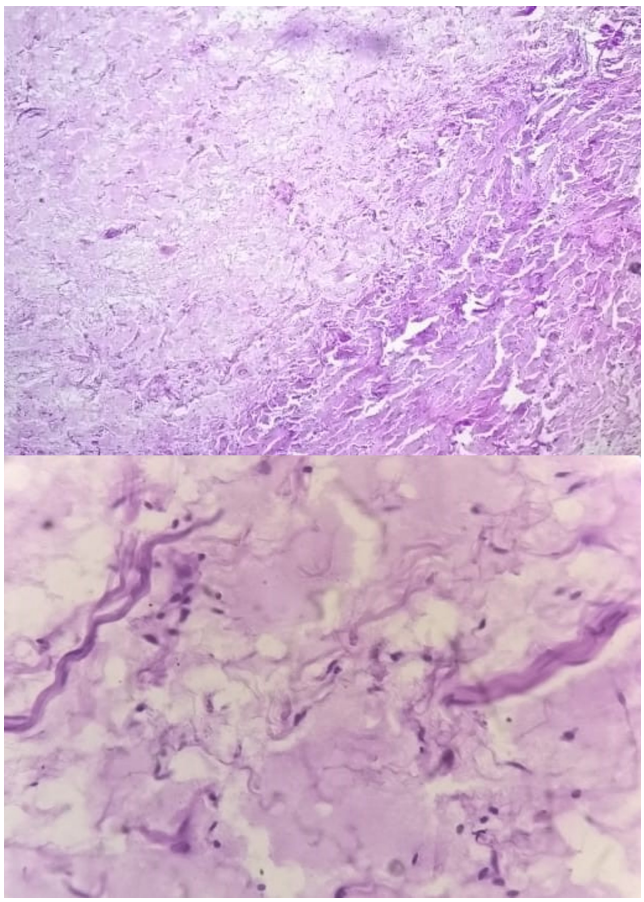

FIGURE 4 In the histological sections observed at both low and high magnifications, spindle-shaped cells with elongated and fusiform nuclei are arranged in a swirling pattern within a mesenchymal tissue. The matrix displays abundant mucin, depicted as eosinophilic amorphous aggregates. No cellular atypia, mitosis, or necrosis is evident. The lesion appears well-defined and is consistent with the diagnosis of a schwannoma with a myxoid component.

tumors, necessitating a comprehensive assessment to distinguish and manage them effectively. These masses arise from various origins, presenting a diverse array of clinical manifestations that emphasize the critical need for accurate differentiation and precise intervention.[1,2]

Optic nerve sheath meningiomas predominantly affect middle-aged women and typically present with insidious, painless vision loss and proptosis.[3] In contrast, schwannomas, although rare in the orbital region, primarily manifest in adults aged 20–70 years.[4] Initially, these tumors may remain asymptomatic when small but gradually result in painless proptosis as they expand within the orbit. The clinical presentation of schwannomas varies significantly based on their origin and location. Arising commonly from branches of the supraorbital or supratrochlear nerves, these tumors induce downward displacement of the globe.[5,6] Larger schwannomas might lead to diplopia, notably if they emerge from the orbital portion of the

third, fourth, or sixth cranial nerves. Lesions near the orbital apex might imitate retrobulbar neuritis, complicating diagnosis.[7]

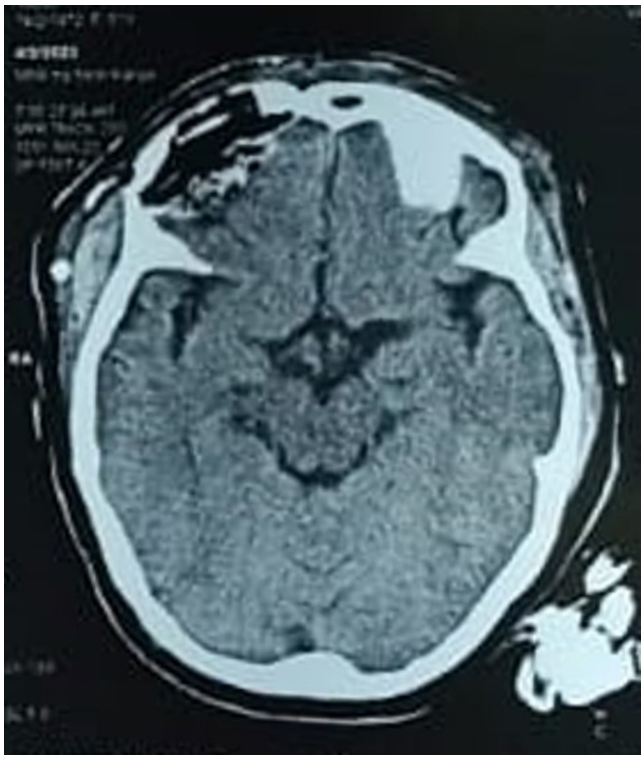

FIGURE 5 Postoperative CT scan demonstrating the successful complete removal of the schwannoma from the right superior orbit, resulting in the resolution of proptosis, and confirming the absence of residual mass.

Additionally, other benign nerve sheath tumors like neurofibromas can produce lid swelling and proptosis and are associated with type 1 neurofibromatosis. They share clinical similarities with schwannomas but have distinguishing features, including occasional extension into the superior orbital fissure. Imaging findings may overlap with schwannomas, making differentiation crucial.[7]

In contrast, malignant tumors such as metastases from various primary sites, lymphomas, rhabdomyosarcomas, and optic pathway gliomas pose significant challenges in diagnosis and treatment. Metastatic orbital lesions present with diverse symptoms such as limited ocular motility, globe displacement/proptosis, blepharoptosis, changes in vision, pain, visible mass, enophthalmos, and diplopia.[8] Lymphomas, both malignant and reactive, exhibit features of mild unilateral proptosis, limited eye motility, ptosis, changes in visual acuity, diplopia, and chemosis/edema.[9] Rhabdomyosarcomas present rapidly with proptosis and orbital displacement, potentially causing distant metastases and local invasion of orbital bones with intracranial extension.[10] Optic pathway gliomas, while relatively uncommon, affect the anterior visual pathway and involve the optic nerve, chiasm, tract, and optic radiations.[11]

Literature reviews unveil limited documentation on orbital schwannomas originating specifically from the supraorbital nerve (Table 1) [12–18]. Successful management in patients often hinges on the expedited and comprehensive removal of the tumor mass to mitigate potential complications and reduce the risk of recurrence.

Precise differentiation of these orbital masses remains pivotal. Imaging modalities such as CT and MRI aid in

TABLE 1 Reported cases of supraorbital nerve schwannoma.

Authors and year	Sex	Age	Symptoms	Location	Surgical approach	Outcome
Grinberg et al. 1974[12]	47	M	Tingling and numbness over left eyebrow and forehead, anesthesia in the distribution of the left supraorbital nerve	Left orbit	Supraorbital	No tumor recurrence
Horie et al. 1982[13]	41	F	Painless swelling of the right eye	Right orbit	Subfrontal craniotomy	Normal eye appearance
Barbagallo et al. 2004[14]	65	M	Progressive, painless proptosis of the right eye	Right orbit	Fronto-orbitozygomatic	Absent visual disturbance or field defects nor intra or periorbital pain
Boustred et al. 2007[15]	5	F	Enlarging subcutaneous mass in the right frontal region	Bilateral	Endoscopic transorbital	Normal forhead appearance
de Jong et al. 2010[16]	44	M	Anesthesia in the left supraorbital region	Left orbit	Superolateral orbitotomy	Persistence of anesthesia
Izumo et al. 2012[17]	67	M	Diplopia at lower gaze and mild hypesthesia at right frontal	Right orbit	Transcranial extradural anterior orbitotomy	Improved diplopia

Note: M: male; F: female.

characterizing their features and extent, but an accurate diagnosis frequently mandates biopsy and histopathological evaluation, guiding subsequent management strategies.

The significance of a precise differential diagnosis in orbital masses cannot be understated. Accurate discrimination between benign and malignant masses, with a keen emphasis on the distinct features and tailored management approaches for orbital schwannomas, is paramount for prompt and optimal treatment. This underscores the necessity of a collaborative multidisciplinary approach involving radiologists, ophthalmologists, oncologists, and pathologists to ensure meticulous care and favorable outcomes for individuals grappling with these complex and diverse orbital conditions.

4 | CONCLUSION

Orbital schwannoma originating from the supraorbital nerve is a rare occurrence, and clinical diagnosis is often challenging. Our patient presented with a tumor in this uncommon location. Although solitary schwannomas are rare, they should be considered as a preoperative differential diagnosis for a unilateral, slowly growing orbital mass in adults, and their prompt management is imperative to prevent vision-threatening complications.

AUTHOR CONTRIBUTIONS

Shikhil Uppal: Conceptualization; data curation; methodology. **Vineet Saggar:** Investigation; methodology; resources. **Gianluca Scalia:** Supervision; validation; visualization; writing – review and editing. **Giuseppe Emmanuele Umana:** Supervision; validation. **Manisha Sharma:** Supervision; validation; visualization. **Bipin Chaurasia:** Supervision; validation; visualization.

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DATA AVAILABILITY STATEMENT

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CONSENT

Written informed consent was obtained from the patient.

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