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Case Report

Intraorbital Ancient Schwannoma with Intracranial Extension: A Case Report

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Abstract

Introduction: Intra-orbital ancient schwannomas are exceedingly rare nerve sheath tumors characterized by degenerative changes such as cystic necrosis, hyalinization, and calcification. These benign neoplasms often present diagnostic challenges due to their nonspecific clinical manifestations and potential for extensive local growth. A 57-year-old male farmer presented with a 20-year history of progressive swelling in the right upper eyelid, which had enlarged to the size of a golf ball.
The lesion was associated with proptosis, inferior globe dystopia, exposure keratopathy, and complete vision loss in the affected eye. Systemic symptoms included intermittent headaches, low-grade fever, and weight loss. Magnetic resonance imaging revealed a large, heterogeneously enhancing mass arising from the superior aspect of the right orbit, with intracranial extension into the anterior cranial fossa and involvement of adjacent paranasal sinuses. Histopathological examination of the excised tumor demonstrated features consistent with an ancient schwannoma, and immunohistochemistry confirmed the diagnosis.
This case highlights the indolent yet locally aggressive nature of intra-orbital ancient schwannomas, emphasizing the importance of early diagnosis and multidisciplinary management. Surgical excision remains the mainstay of treatment, with a favorable prognosis following complete resection. However, intracranial extension and prolonged symptom duration, as seen in this case, underscore the need for heightened clinical suspicion and timely intervention.
Conclusion: Intra-orbital ancient schwannomas are rare but should be considered in the differential diagnosis of orbital masses. Early imaging and histopathological confirmation are critical for optimal management and preservation of visual function.

Keywords: Ancient Schwannoma, Intracranial Extension, Orbital Tumor, Proptosis, Vision Lost, Rare

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Introduction

Schwannomas are benign, encapsulated, slow-growing peripheral nerve sheath tumors derived from Schwann cells and account for approximately 1% of all orbital tumors.[1] They most commonly arise from sensory nerves within the orbit, including branches of the trigeminal nerve, oculomotor nerve, trochlear nerve, or sympathetic fibers. Orbital schwannomas typically present in middle-aged adults and exhibit an indolent course, often leading to delayed presentation due to their slow growth and initially minimal symptoms.

Ancient schwannoma is a rare histological variant of schwannoma characterized by long-standing degenerative changes resulting from prolonged tumor growth. These changes include cystic degeneration, hyalinization, calcification, hemorrhage, and hypocellular areas, as well as marked nuclear pleomorphism with hyperchromasia. Despite the presence of atypical nuclei, mitotic activity is absent or minimal, distinguishing ancient schwannomas from malignant peripheral nerve sheath tumors. First described by Ackerman and Taylor in 1951, ancient schwannomas are most frequently reported in the head and neck region, retroperitoneum, and extremities, with orbital involvement being exceptionally uncommon.

Intracranial extension of orbital schwannomas is rare, owing to the confined anatomy of the orbit and the typically well-encapsulated nature of these tumors. When present, intracranial extension suggests long-standing disease and may occur through the superior orbital fissure, optic canal, or erosion of adjacent bony structures. Reports of intra-orbital ancient schwannomas with intracranial extension are exceedingly scarce, with only a handful of cases documented in the literature, underscoring the unusual and locally aggressive behavior of this otherwise benign entity.

From a diagnostic perspective, ancient schwannomas pose significant challenges. Radiologically, they may demonstrate heterogeneous signal intensity on magnetic resonance imaging due to cystic degeneration, hemorrhage, and fibrosis, occasionally mimicking malignant orbital tumors or inflammatory lesions. Similarly, the presence of nuclear atypia on histopathology can lead to misdiagnosis as sarcoma if the degenerative nature of the changes is not recognized. Therefore, accurate diagnosis relies on careful clinicoradiological correlation, histopathological evaluation, and confirmatory immunohistochemistry, particularly diffuse S-100 protein positivity.

This case report describes a rare intra-orbital ancient schwannoma with intracranial extension, highlighting its clinical presentation, imaging characteristics, and histopathological features. The report aims to expand existing literature on this rare entity, emphasize common diagnostic pitfalls, and reinforce the importance of early recognition and multidisciplinary management to prevent extensive local extension and irreversible visual loss.

Case Presentation

A 57-year-old male farmer who presented with a 20-year history of progressive proptosis of the right eye. He was apparently in good health until he noticed a small swelling in the right upper lid, initially about the size of a groundnut. Over time, this swelling gradually increased in size, eventually reaching the size of a golf ball, with proptosis and extending to involve the right side of his face.

The swelling has remained painless and mobile but is associated with intermittent itching, discharge from the eye, headache, low-grade fever, and progressive weight loss. There has been no history of loss of consciousness or seizures. In addition, the patient reports a non-productive cough and occasional chest pain. He denies any abdominal pain or distension, yellowish discoloration of the eyes, bone pain, or similar swellings elsewhere in the body.

Following the onset of symptoms, the patient initially sought medical attention at a tertiary health facility in his state and was referred to this hospital. However, he did not follow through with the referral immediately. Instead, he visited a private hospital in Gombe, where a biopsy and computed tomographic (CT) scan were performed, although the results are currently unavailable.

He has no history of using corrective lenses, ocular trauma, or previous eye surgery. There is no family history of blindness or similar illness. He does not have any known drug allergies and has no history of hypertension or diabetes.

On examination, the patient was a middle-aged man, conscious and alert, afebrile, and in no apparent distress. He was not pale, anicteric, acyanosed, and showed no signs of dehydration. There was no peripheral lymphadenopathy and no pedal edema.

Visual acuity assessment revealed no perception of light (NPL) in the right eye and 6/5 vision in the left eye.

Right Eye:
Examination of the right eye revealed a large, non-tender swelling involving the upper eyelid, measuring approximately 10 cm by 8 cm, associated with proptosis and inferior dystopia. The overlying skin was freely mobile with no differential warmth. There was restriction of movement in all directions of gaze.

The conjunctiva was hyperemic with patchy areas of dryness. The cornea showed signs of exposure keratopathy, and further evaluation of the posterior segment was not possible due to corneal haziness.

Figure 1: The patient is a 57-year-old male with a 20-year history of a progressively enlarging, painless swelling on the forehead. The mass is firm, well-circumscribed, and appears to be slowly growing without signs of inflammation or tenderness.

Left Eye:
The left eye appeared entirely normal. The lids were normal, the conjunctiva was quiet, and the cornea was clear. The anterior chamber was of normal depth and quiet, with a round, briskly reactive pupil. Extraocular muscle movements were full in all directions of gaze.

These findings were consistent with a diagnosis of a right orbital tumor leading to significant globe displacement and visual loss.

Medical therapy was initiated with oral cocodamol for analgesia and topical chloramphenicol ointment to the right eye (twice daily) to prevent secondary infection and keep the eye moist. A neurosurgical consultation was requested in view of the suspected intracranial involvement.

As part of the systemic and radiological workup, the patient underwent a full blood count with differential and a contrast-enhanced Magnetic Resonance Imaging (MRI) of the brain and orbits.

Magnetic Resonance Imaging revealed a large, irregularly shaped, heterogeneously enhancing mass arising from the superior aspect of the right orbit. The lesion appeared hypointense on T1-weighted images, iso- to hyperintense on FLAIR, and hyperintense on T2-weighted sequences.

The mass demonstrated extension through the orbital roof into the anterior cranial fossa, with involvement of the inferior portion of the right frontal lobe, as well as infiltration of the ethmoidal and maxillary sinuses. The lesion caused expansion of the orbital cavity, resulting in proptosis of the right globe and compression and stretching of the right optic nerve.

The total dimensions of the mass were approximately 9.4 cm (anteroposterior) × 6.8 cm (transverse) × 4.5 cm (craniocaudal). There was associated perilesional edema, with mass effect noted on the frontal horn of the ipsilateral lateral ventricle. [Figures 2a and 2b]

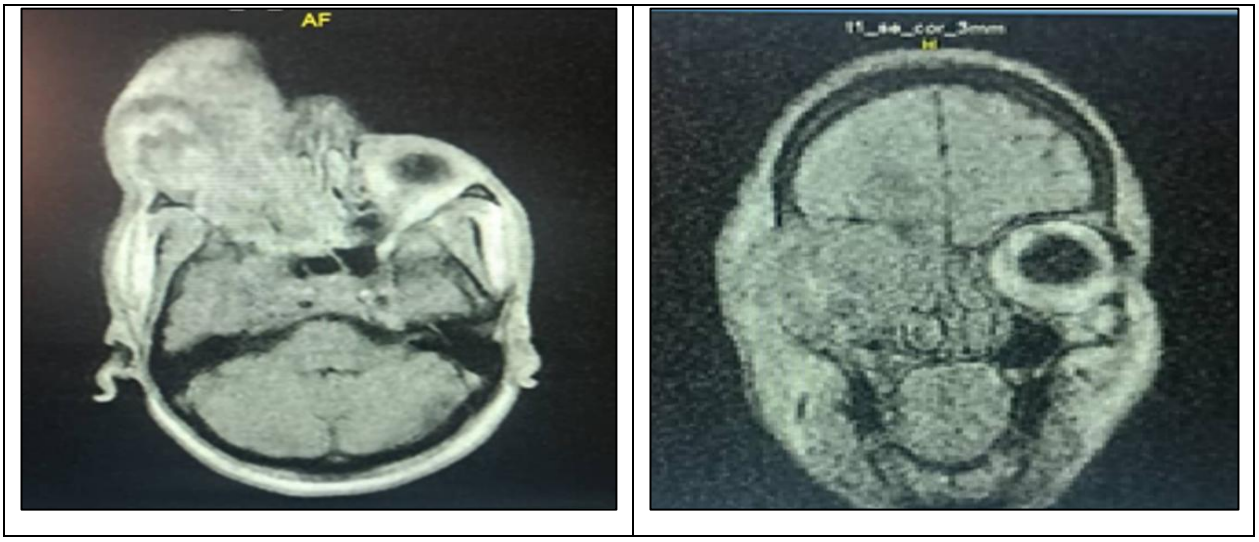


Figure 2[a & b]: Magnetic Resonance Imaging Features

Legend for Figure 2[a & b]

(a) Post-contrast T1-weighted axial MRI shows a large, heterogeneously enhancing mass in the left craniofacial region with suspected intracranial extension and mass effect on the left temporal lobe, cerebellum, and adjacent structures. Features suggest an aggressive neoplasm, warranting further CT assessment for bone involvement and histopathological confirmation.

(b) Coronal T1-weighted MRI reveals a large, well-defined, heterogeneously enhancing mass in the right orbit, causing proptosis and anterolateral globe displacement, with extension into adjacent maxillofacial structures and possible soft tissue infiltration.

Abbreviations: MRI- Magnetic Resonance Imaging; CT- Computed Tomography

The left orbit and its contents appeared normal. The remaining cerebral and cerebellar hemispheres, including the brainstem, basal ganglia, thalami, basal cisterns, and ventricular system, were preserved.
A right superior intra-orbital mass with intracranial extension, involving adjacent paranasal sinuses and the inferior frontal lobe, with mass effect on adjacent cerebral structures.

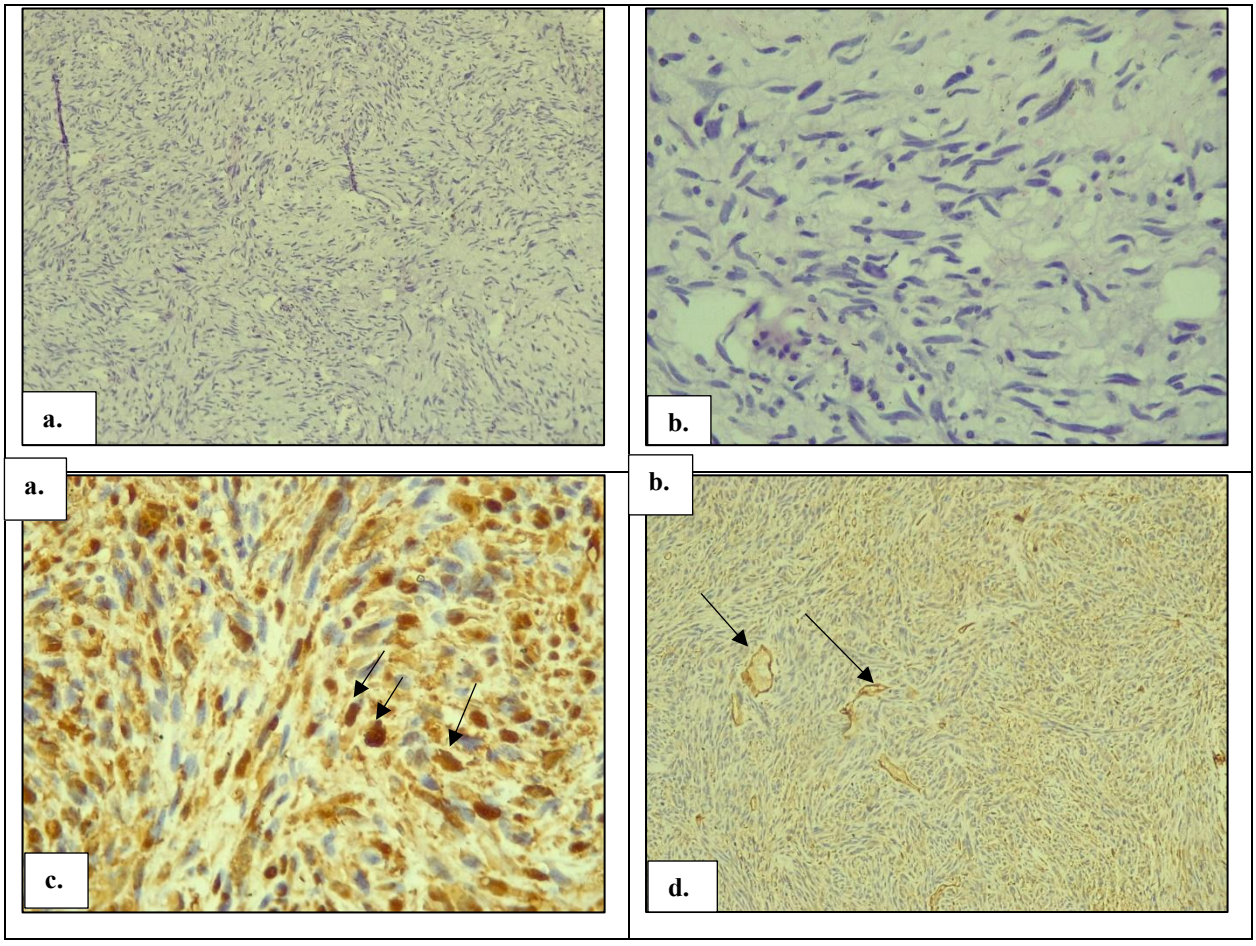
The patient was admitted for further evaluation and management. He was counselled regarding the poor visual prognosis in the affected eye. A decision was made to proceed with excisional biopsy under general anesthesia, followed by histopathological analysis to establish a definitive diagnosis.

At surgery, a mass extending from the right upper lid through the roof of the orbit to the anterior cranial fossa posteriorly, and the ethmoidal sinus medially.

Under general anaesthesia, a brow incision was made, and dissection was done posteriorly till the mass was dissected out and excised. Haemostasis was achieved, and a drain was inserted. The globe and upper lid were preserved.

A mass extending from the right upper eyelid through the ethmoidal sinus through the roof of the orbit into the anterior cranial fossa was excised and sent for histology.

The microscopic examination revealed a fairly circumscribed spindle cell tumour with focal areas of encapsulation. The tumor is disposed in short fascicles, sheets, vague herringbone, and whorling patterns. The comprising cells are bland, wavy, buckled to oval cells with hyperchromatic nuclei and moderate amphophilic cytoplasm. Numerous areas showed random degenerative nuclear atypia, and mitosis was infrequent, 3 per 10 high-power fields. The stroma is moderately loose, displaying extensive hyalinization, myxoid changes, and cystic degeneration. Areas showing hyalinized thick-walled vascular channels, with some having perivascular lymphocytic cuffing noted. Differential diagnoses of a solitary intra-orbital schwannoma, solitary fibrous tumour, and microcystic meningioma were entertained based on the histological features. Immunohistochemical stains employed included: S100, Progesterone Receptor (PR), and Cluster of Differentiation 34 (CD34). Tests were valid and revealed strong diffuse cytoplasmic and nuclear immunopositivity to S100. PR antibody was negative, while CD34 showed focal positivity mainly in the myxoid areas and vascular channels, and negativity in the neoplastic cells. [Figures 3a & 3b show the haematoxylin and eosin microscopical features, while Figures 3c, 3d, 3e & 3f show the immunohistochemical features]



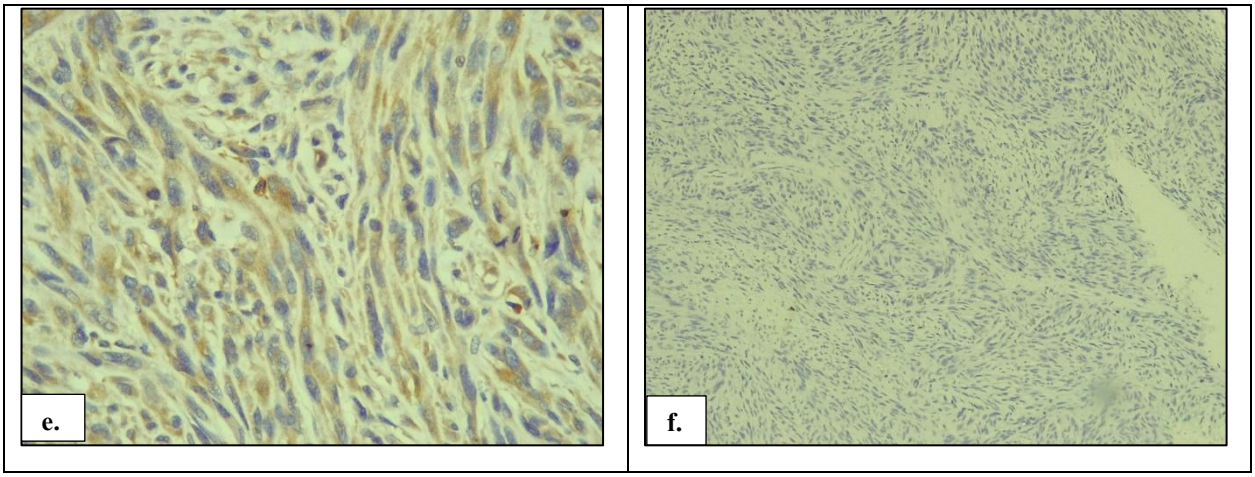


Figure 3[a-f] Microscopical Features (H&E and Immunohistochemical)

Legend for Figure 3[a-f]

(a) Photomicrograph showing a hypocellular bland spindle cell tumour with a loosely textured myxoid stroma, H&E x100. (b) Photomicrograph showing bland wavy to buckled spindle cells having darkly stained nuclei, H&E x400. (c) Photomicrograph with thin black arrows highlighting neoplastic schwann cells displaying diffuse strong cytoplasmic and nuclear immunopositivity to S100 antibody, S100-PBM x400. (d) Photomicrograph with thin black arrows highlighting vascular channels in the Antoni B areas showing CD34 immunopositivity, CD34-PBM x100. (e) Photomicrograph of the neoplastic Schwann cells showing CD34 immunonegativity, CD34-PBM x 400. (e) Photomicrograph showing PR immunonegativity in the neoplastic schwann cells, PR-PBM x100.

Abbreviations: CD34- Cluster of Differentiation 34; H&E- Haematoxylin and Eosin; PBM- Polymer Based Method; PR- Progesterone Receptor; S100- Solubility in 100%

Discussion

Orbital schwannomas are uncommon benign peripheral nerve sheath tumors, accounting for approximately 1% of all orbital neoplasms. Ancient schwannoma represents a rare histopathological variant resulting from long-standing tumor evolution, and intra-orbital localization of this subtype is exceedingly rare. Fewer than 20 cases of intra-orbital ancient schwannomas have been reported globally, with only a minority demonstrating intracranial extension, making the present case particularly unusual.[1,2]

Most published cases describe tumors measuring less than 4 cm, presenting after symptom durations ranging from 3 to 10 years.[1,3] In contrast, the current patient presented after a 20-year disease course with a massive lesion measuring approximately 10 × 8 cm and intracranial extension into the anterior cranial fossa.[4,5] Similar cases reported by Sharma et al. and Kim et al.[6,7] involved combined orbital and intracranial disease and required multidisciplinary surgical approaches. However, none demonstrated such prolonged neglect with complete visual loss at presentation, highlighting the exceptional nature of this case in both size and duration.

The term *ancient schwannoma* reflects chronicity rather than biological aggressiveness. Prolonged tumor growth leads to relative ischemia, metabolic stress, and repeated microhemorrhages within the lesion, resulting in secondary degenerative changes such as cystic cavitation, hyalinization, calcification, and nuclear atypia. These atypical nuclei represent degenerative alterations rather than malignant

transformation, as evidenced by low mitotic activity, absence of necrosis, and preserved encapsulation. In this patient, the extraordinary duration of disease likely contributed directly to the pronounced degenerative histological features observed.[8,9]

Magnetic resonance imaging is central to preoperative assessment but may be misleading in ancient schwannomas. Typical schwannomas demonstrate the "target sign" on T2-weighted images—central hypointensity (Antoni A tissue) surrounded by peripheral hyperintensity (Antoni B tissue)—as well as the "fascicular sign."[8,10] In ancient schwannomas, these features are often obscured by extensive cystic degeneration, hemorrhage, and fibrosis, resulting in heterogeneous signal intensity and enhancement patterns. The heterogeneous enhancement and T2 hyperintensity observed in this case correlate well with the histologic predominance of Antoni B areas, cystic degeneration, and hyalinized vascular channels.[11]

The atypical imaging appearance and nuclear pleomorphism of ancient schwannomas frequently complicate differentiation from other orbital tumors. Meningiomas may show a dural tail and homogeneous enhancement; cavernous hemangiomas typically demonstrate progressive centripetal enhancement; lacrimal gland tumors often present with bone remodeling and characteristic anatomic localization; and neurofibromas lack encapsulation and are often associated with neurofibromatosis.[12,13] Histologically, nuclear atypia may raise concern for malignant peripheral nerve sheath tumor, solitary fibrous tumor, or microcystic meningioma. In this case, diffuse S-100 positivity, low mitotic index, PR negativity, and CD34 staining confined to Antoni B areas supported the diagnosis of ancient schwannoma and excluded malignant or meningeal neoplasms.[14]

Complete surgical excision remains the definitive treatment for orbital schwannomas. When total resection is achieved, prognosis is excellent, and recurrence is rare. However, incomplete excision—often due to proximity to critical neurovascular structures or intracranial extension—is associated with recurrence rates of approximately 10–15%. Schwannomas are generally radioresistant, and adjuvant radiotherapy is reserved for residual or unresectable disease. In the present case, irreversible visual loss underscores the functional consequences of delayed presentation, despite the tumor's benign histology.[15,16]

The 20-year delay in presentation was a pivotal factor contributing to extensive local invasion, intracranial extension, optic nerve compression, and permanent visual loss. Early diagnosis and intervention could likely have preserved visual function and prevented intracranial spread. This case emphasizes the need for heightened clinical suspicion and early imaging in patients presenting with slowly progressive orbital masses.

Conclusion

Intra-orbital ancient schwannoma is a rare benign entity that may masquerade as a malignant or aggressive orbital tumor due to its degenerative histologic features and atypical imaging appearance. Prolonged disease duration can result in massive tumor growth, intracranial extension, and irreversible visual loss. Accurate diagnosis requires careful radiologic–pathologic correlation and immunohistochemical confirmation. Early detection and complete surgical excision are essential to optimize outcomes and prevent long-term complications.

References

1. Rose GE, Wright JE. Isolated peripheral nerve sheath tumours of the orbit. *Eye*. 1991;5(6):668–73.
2. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax: a clinicopathological evaluation of forty-eight cases. *Cancer*. 1951;4(4):669–91.
3. Pushker N, Meel R, Sharma S, Bajaj MS, Kashyap S, Sen S. Giant orbital schwannoma with intracranial extension. *Orbit*. 2011;30(2):114–6. https://doi.org/10. 1136/bjo.2009.170829

4. Gupta A, Prabhakaran VC, Dodd T et al. Orbital schwannomas: clinical and imaging features. *Clin Exp Ophthalmol*. 2008;36(7):632–7.
5. Lee JH, Kim CY, Kim N, et al. Ancient schwannoma of the orbit: a case report. *Korean J Ophthalmol*. 2012;26(5):388–91.
6. **Sharma R, Sharma K, Parmar D, Tanwar V, Thakur S, Asotra S.** *Excision of massive orbital schwannoma through anterior transconjunctival approach: A case report.* Indian Journal of Ophthalmology Case Reports. 2023;3(3):814-816. doi:10.4103/IJO.IJO_3099_22.
7. **Kim KS, Jung JW, Yoon KC, Kwon YJ, Hwang JH, Lee SY.** *Schwannoma of the Orbit.* Archives of Craniofacial Surgery. 2015;16(2):67-72. doi:10.7181/acfs.2015.16.2.67.
8. Chen KT. Ancient schwannoma. A clinicopathological study. *Cancer*. 1980;45(9):2283–7.
9. Rennert J, Doerfler A. Imaging of peripheral nerve sheath tumors with pathologic correlation. *Eur J Radiol*. 2007;62(2):214–28.
10. Sharma MC, Arora R, Mahapatra AK, et al. Intracranial schwannoma: a clinicopathological study. *Neurol India*. 2003;51(3):353–5.
11. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. 6th ed. Elsevier; 2014.
12. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. *Acta Neuropathol*. 2012;123(3):295–319. https://doi.org/10.1007/s00401-012-0954-z.
13. Kalouch S, Chakir N, El Kacemi H, et al. Ancient schwannoma of the orbit: a case report. *Pan Afr Med J*. 2017; 28:23.
14. . Rootman J. Diseases of the Orbit: A Multidisciplinary Approach. 2nd ed. Lippincott Williams & Wilkins; 2003.
15. Gosk J, Gutkowska O, Zimmer K, et al. Results of surgery for schwannomas of peripheral nerves. *Chir Narzadow Ruchu Ortop Pol*. 2006;71(3):203–7.
16. Knight DM, Birchall MA, Hilton CJ, et al. Evaluation of the management of schwannoma: a single centre experience. *Br J Oral Maxillofac Surg*. 2007;45(1):35–8.

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Declaration of Patient's Consent
We authors certify that we have obtained all appropriate patient consent forms. In the form, the patient has given his consent for all clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity.

Declaration of Helsinki
The study was conducted according to the principles of the Helsinki Declaration.

Availability of Research Data
Authors are available and ready to supply the data upon any requests through the corresponding author.