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Case Report: Intraorbital Intraconal Schwannoma Centered on the Optic Nerve

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Abstract | **Case Report**

Orbital schwannomas are rare, encapsulated benign neural tumors representing less than 1% of all orbital masses. We report the case of a 48-year-old female presenting with a slowly progressive unilateral left-sided grade III proptosis over 3 years, associated with complete blindness. Dedicated 3T orbital MRI demonstrated a well-circumscribed, retrobulbar intraconal mass centered on the optic nerve, featuring heterogeneous T1/T2 signal intensity, peripheral wall and internal septal contrast enhancement following gadolinium administration, with no restriction on ADC mapping. Detailed semiological analysis allows accurate preoperative diagnosis and guides management strategy.

Keywords: Orbital schwannoma, Optic nerve, Orbital MRI, Intraconal tumor, Proptosis, Case Report.

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INTRODUCTION & CLINICAL BACKGROUND

Schwannomas (neurilemomas) of the orbital cavity are benign, slow-growing neoplasms arising from Schwann cells of the neural sheath of peripheral sensory or motor nerves. Direct involvement or intimate contiguity with the optic nerve is exceptionally rare, as optic nerve fibers are myelinated by oligodendrocytes rather than Schwann cells. In most orbital cases, schwannomas originate from peripheral branches of cranial nerves (V1, V2, III, IV, or VI). The typical clinical presentation includes indolent, progressive proptosis. Here, we present a remarkable case owing to its large size, strictly intraconal retrobulbar location, and significant functional impact on the optic nerve.

Clinical Case Presentation (Observation)

A 48-year-old female patient, with a surgical history of a gastric lesion 4 years prior, presented to the ophthalmology department for the evaluation of a progressive left orbito-palpebral deformation with ocular protrusion evolving over 3 years. The onset was insidious, without pain, inflammatory signs, or systemic fever.

- **Ophthalmological Examination:**

Left visual acuity was completely absent (No Perception of Light - NPL). A relative afferent pupillary defect (Marcus Gunn pupil) was noted. Extraocular motility examination revealed global restriction of abduction and elevation secondary to mechanical mass effect. Fundus examination of the left eye demonstrated total optic atrophy with complete papillary pallor. The right eye was unremarkable with a visual acuity of 20/20.

- **Neurological & Physical Examination:**

No signs of increased intracranial pressure (ICP). No cutaneous stigmata of neurofibromatosis (absence of café-au-lait spots or neurofibromas).

Detailed Semiological Characterization on MRI

High-resolution dedicated orbital MRI performed in axial, coronal, and sagittal planes (T1, T2, high-resolution T2, FLAIR, DWI/ADC, and fat-suppressed contrast-enhanced T1 sequences) demonstrated:

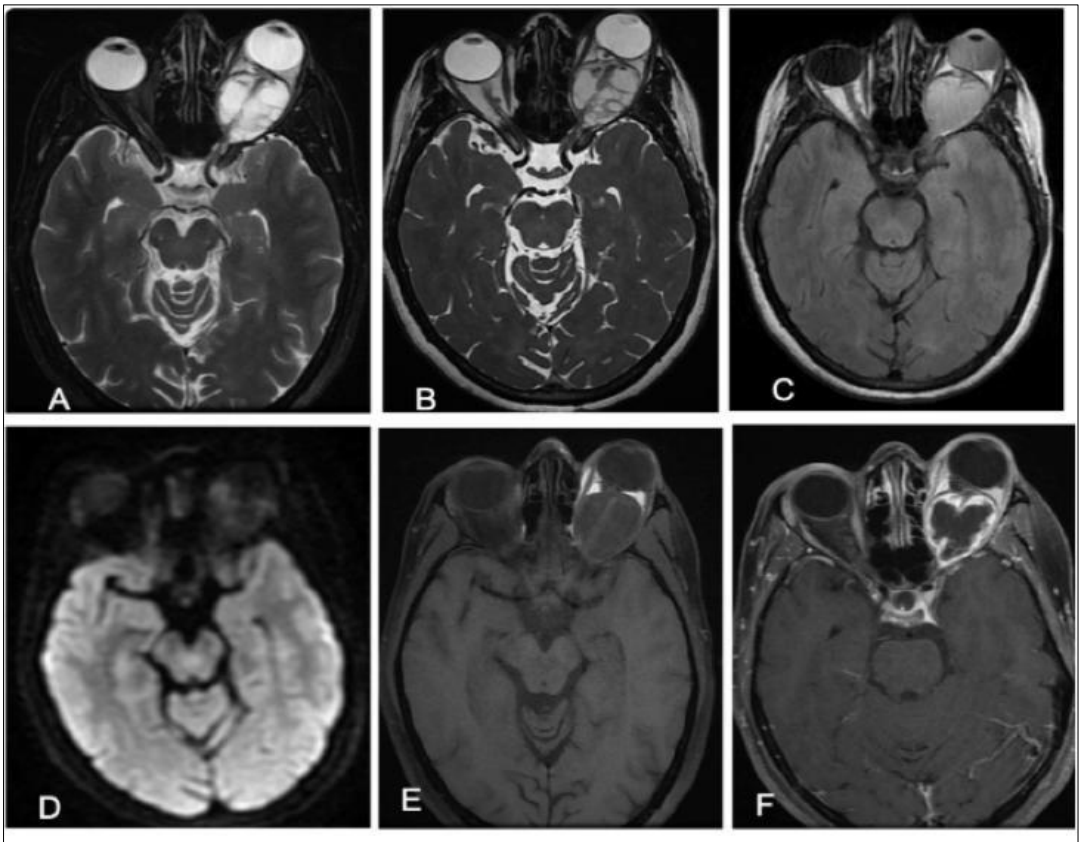
- **Morphology & Measurements:** Voluminous, retrobulbar left intraconal mass, well-demarcated with regular oval contours, measuring 37 x 27 x 25 mm.
- **Anatomical Relationships:** The mass was directly centered along the course of the left optic nerve, causing medial displacement and stretching. The globe was displaced directly forward while maintaining its anatomical sphericity, resulting in axial proptosis of grade III. Posteriorly, the lesion extended towards the superior orbital fissure (SOF). Extraocular

muscles were peripherally displaced without signs of direct muscular invasion. Intraconal and extraconal fat preserved normal intensity.

- **MRI Signal Characteristics:** Heterogeneous hypointense signal on T1-weighted images, hyperintense heterogeneous signal on T2-weighted sequences (reflecting mixed Antoni A cellular and Antoni B myxoid/cystic architecture), and intermediate signal on FLAIR.
- **Diffusion & Dynamic Enhancement:** Apparent diffusion coefficient (ADC) mapping showed no significant restriction, ruling out a hypercellular malignant lesion. Post-gadolinium T1-weighted images revealed intense, heterogeneous enhancement of a thick capsule and internal septa.

2. Radiological Differential Diagnosis

- **Optic Nerve Sheath Meningioma:** Typically displays the classic 'tram-track' sign on axial images (dural enhancement encircling a non-enhancing optic nerve core) and calcifications on CT. Schwannoma surrounds or displaces the nerve without circumferential tram-track enhancement.
- **Optic Nerve Glioma:** Predominantly affects pediatric and young populations (frequently associated with Neurofibromatosis Type 1), demonstrating more uniform, fusiform expansion without prominent cystic/septated components.
- **Cavernous Hemangioma (Cavernous Venous Malformation):** The most common intraconal mass in adults, characterized by progressive, slow centripetal filling on delayed contrast-enhanced images, distinct from the septated pattern of schwannoma.



IRM orbitaire en coupes axiales en SP T2(A), T2HR(B), FLAIR (C), diffusion (D), T1(E), T1 après injection de gadolinium (F).

DISCUSSION & MANAGEMENT STRATEGY

Definitive management of orbital schwannomas relies on complete surgical excision via an appropriate orbitotomy approach (anterior, lateral, or transcranial depending on posterior extension). The presence of a well-defined fibrous capsule facilitates enucleation and microsurgical dissection. In this patient, management considerations integrated irreversible visual loss (established optic atrophy) and the necessity to excise the expanding mass to relieve disfiguring proptosis and prevent further compression at the superior orbital fissure.

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