



A Rare Presentation of Intraconal Schwannoma and Internal Ophthalmoplegia

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Case Presentation

- Thirty-four-year-old healthy female:
- Asymptomatic evaluation for Lasik vision correction.
- Screening evaluation demonstrated Right Exophthalmos and Optic Nerve Head Swelling OD prompting Orbital referral.
- No pain, irritation, diplopia, or blurry vision. Visual acuity was 20/20 on both sides, color vision symmetric and no relative afferent pupillary defect was noted. No resistance to retropulsion was noted. Right exophthalmos measuring 27 on the right compared to 21 on the left side present. No lagophthalmos, clear corneas, 2+ optic nerve head swelling OD.

Testing

- MRI revealed a large well-circumscribed intraconal lesion on the right measuring 2.2 x 2.1 x 1.9 cm with low T1 signal. The mass was noted to displace the right optic nerve medially, the lateral rectus muscle laterally, and it nearly abutted the back of the globe.

Surgery

- Lateral orbitotomy with bone window and intraoperative navigation undertaken.
- Large intraconal tumor noted moderately encapsulated superficially and deeply infiltrating but not attached to the optic nerve. The tumor was decompressed to facilitate adequate removal and visualization of tumor capsule removal from the space just behind the globe. A portion of the lesion was sent for frozen section analysis and was determined to be a low-grade spindle cell tumor. During the procedure, the right pupil was noted to be enlarged.

Post-Op Course

- At eight hours post-op, Vision was 20/30, pupil fixed at 7mm, no RAPD by reverse, EOMs full and no ptosis.
- During the 1 week postoperative office follow-up, 1+ edema and erythema was present without any significant ptosis. Exophthalmometry measurement improved by 6-7 mm. The tumor was determined to be a benign schwannoma. Persistent mydriasis OS was present secondary to the peripheral nerve sheath tumor. Vision 20/25 at distance, J5 at near OD.
- At 6 months post-op, vision was 20/20 at distance, EOMs full, eyes symmetric, scar nicely healed. Pupil OD remains 7mm fixed and vision subjectively blurred at near.

Figures



Figure 1. Worms eye view depicting preoperative exophthalmos.



Figure 2. 2 months postoperative, persistent mydriasis OD.



Figure 3. Transverse plane view of orbital schwannoma with impingement on lateral rectus muscle visible.



Figure 4. Parasagittal view of orbital schwannoma depicting near-abutment of posterior globe.

Discussion

Presence and removal of intraconal orbital schwannoma with resulting Internal Ophthalmoplegia.

- **Orbital Schwannoma:**
 - Typically benign, slow-growing peripheral nerve tumor with a favorable outcome
 - Reported in CN III-VI and associated sympathetic and parasympathetic fibers
 - Associated with exophthalmos, diplopia, optic neuropathy due to compression
 - Treatment includes:
 - Incisional biopsy to confirm histopathologic nature of tumor
 - Complete surgical excision to prevent nerve compression, intracranial spread, and malignant transformation
 - Radiation may be used for recurrent cases
 - Recurrence is attributed to:
 - Incomplete or piecemeal excision - often due to inability to remove tumor without damaging surrounding structures
 - Tumor seeding
 - Possible involvement of NF2 or schwannomatosis
- **Internal Ophthalmoplegia:**
 - Oculomotor nerve (CN III) is responsible for major extraocular movements of the eye
 - Parasympathetics course from Edinger-Westphal nucleus via inferior division of CN III, synapse in ciliary ganglion, then travel as short ciliary nerves to:
 - Sphincter pupillae muscle
 - Constricts pupil in pupillary light reflex and accommodation
 - Ciliary muscle
 - Acts on suspensory ligaments and lens in accommodation
 - Intact extraocular movements with impaired accommodation and persistent mydriasis suggest involvement of parasympathetics coursing with CN III

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