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Case Report

# Angiomatoid Schwannoma of the Optic Nerve: A Rare Orbital Tumor

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**Received:** December 09, 2025; **Published:** January 30, 2026

## Abstract

Schwannomas are benign peripheral nerve sheath tumors arising from Schwann cells and represent a small proportion of orbital neoplasms. Their occurrence along the optic nerve is exceptionally rare, given the absence of Schwann cells within the nerve itself. The angiomatoid variant is even less frequent and is characterized by prominent vascular spaces and hemorrhagic components, features that may complicate radiologic interpretation. Although typically slow-growing, these tumors may lead to progressive visual impairment and exophthalmos secondary to compressive effects on the optic nerve and adjacent orbital structures. Imaging-particularly MRI-plays a fundamental role in identifying lesion characteristics, assessing optic canal involvement, and guiding surgical planning, while definitive diagnosis relies on histopathological confirmation.

We report the case of a 30-year-old woman presenting with progressive right-sided exophthalmos and visual loss. MRI demonstrated a well-defined, fusiform intraconal lesion centered on the optic canal, hyperintense on T2-weighted sequences and intensely enhancing following gadolinium, with internal susceptibility artifacts suggestive of microhemorrhages. Histopathology revealed a spindle cell proliferation with pronounced angiomatoid features, confirming an angiomatoid schwannoma.

This case underscores the importance of considering optic nerve schwannoma-even in its rare angiomatoid form-in the differential diagnosis of unilateral progressive exophthalmos with visual decline. Awareness of its imaging characteristics is essential for accurate diagnosis and appropriate treatment planning.

***Keywords:*** *Optic Nerve Schwannoma; Angiomatoid Schwannoma; Orbital Tumor; Exophthalmos; MRI*

## Introduction

Schwannomas are benign tumors originating from Schwann cells and account for 2-5% of orbital neoplasms, though their occurrence along the optic nerve is exceedingly rare due to the absence of intrinsic Schwann cells within the nerve sheath [1]. These tumors are believed instead to arise from sympathetic fibers contained within the perivascular connective tissue surrounding the optic nerve [2]. Their slow growth and nonspecific clinical presentation often delay diagnosis. We present a rare case of angiomatoid schwannoma of the optic nerve with detailed radiologic and histopathologic correlation.

## Case Report

A 30-year-old woman presented with several months of progressive right-sided exophthalmos and declining visual acuity. Ocular examination showed a non-reducible exophthalmos with restricted globe motility. Visual acuity measured 0/10 in the right eye and 9/10 in the left. The patient reported no history of trauma, systemic illness, or familial ocular or neurological disorders.

Orbital MRI demonstrated a well-circumscribed, fusiform, intraconal mass centered on the right optic canal, with associated widening of the canal and posterior extension abutting the cavernous sinus. The lesion appeared hyperintense on T2-weighted, FLAIR, and STIR images (Figure 1), and hypointense on T1-weighted sequences. T2* imaging revealed internal signal voids compatible with septations or microhemorrhages. Post-contrast fat-suppressed T1 sequences showed intense, heterogeneous enhancement (Figure 2). The mass produced Grade II exophthalmos and displaced adjacent orbital structures, without evidence of intracranial extension.

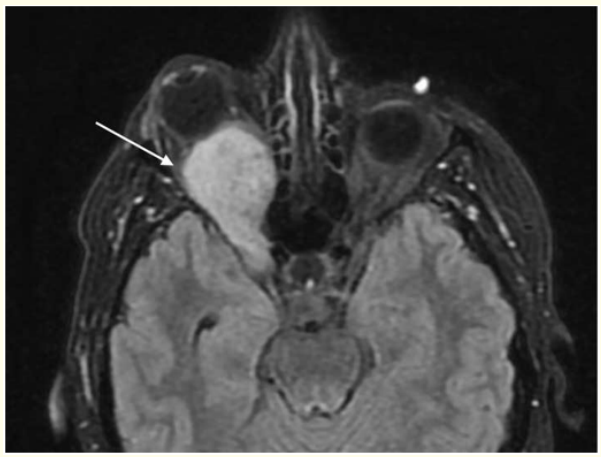

***Figure 1:*** *Axial FLAIR image showing a hyperintense, well-defined intraconal mass centered on the right optic canal.*

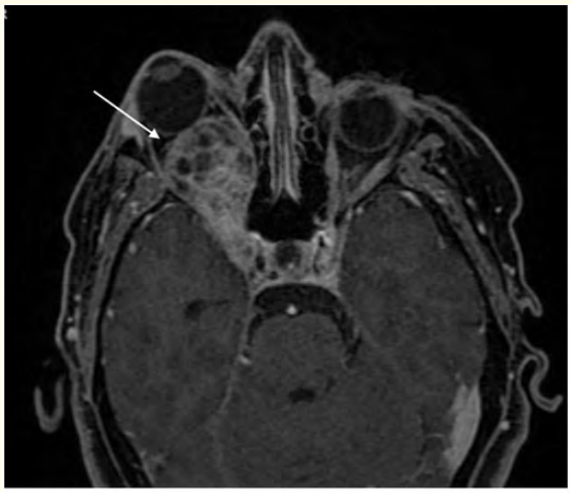

***Figure 2:*** *Axial post-contrast fat-suppressed T1-weighted MRI image showing intense, heterogeneous enhancement of the lesion.*

Histopathology demonstrated spindle-cell proliferation in a richly vascularized stroma, consistent with an angiomatoid schwannoma (Figure 3).

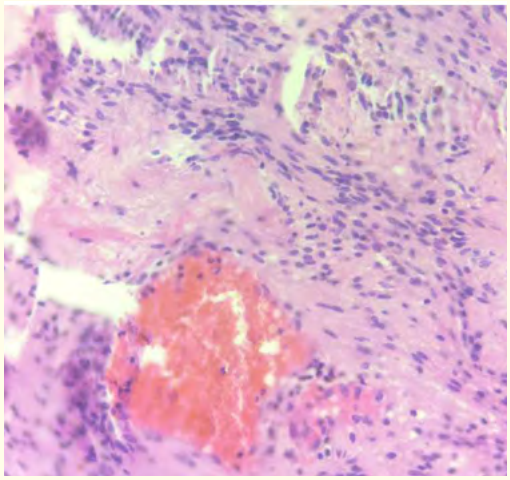

***Figure 3:*** *Histopathological section demonstrating spindle cell proliferation within a vascularized stroma with angiomatoid features (H&E, x100).*

## Discussion

Schwannomas are benign peripheral nerve sheath tumors originating from Schwann cells and typically arise along cranial, peripheral, or autonomic nerves. Within the orbit, they represent approximately 2 - 5% of primary tumors; however, schwannomas occurring in association with the optic nerve are exceptionally rare. This rarity reflects the embryologic and structural characteristics of the optic nerve, which-unlike peripheral nerves-is myelinated by oligodendrocytes and therefore normally devoid of Schwann cells. As a result, true intraneural optic nerve schwannomas are considered anatomically unlikely. Current theories propose that these tumors arise instead from sympathetic nerve fibers situated within the perivascular connective tissue surrounding the optic nerve sheath, providing the only local Schwann-cell population capable of giving rise to such neoplasms [1,2].

Clinically, optic nerve sheath schwannomas typically present with a slowly progressive, painless unilateral exophthalmos, consistent with the indolent growth of the tumor. Visual impairment is common and results from chronic compressive neuropathy. Limitation of ocular motility may be seen when the mass occupies the intraconal space or extends toward the orbital apex. Fundoscopic findings depend on chronicity and may evolve from normal appearance to optic disc pallor, edema, or choroidal folds. Because these clinical manifestations are nonspecific and progress gradually, diagnosis is often delayed, and patients are frequently evaluated first for more common orbital pathologies such as cavernous hemangioma, optic nerve glioma, or inflammatory pseudotumor [1,3].

Imaging plays a pivotal role in characterizing these lesions and guiding management. CT generally demonstrates a well-demarcated, homogeneous soft-tissue mass that may remodel or widen the optic canal when the tumor is juxtaposed to the optic nerve sheath. Although these features are not pathognomonic, they provide valuable diagnostic clues when integrated with the patient's clinical history. MRI, however, is the modality of choice due to its superior soft-tissue resolution and ability to delineate tumor-nerve relationships. Typical schwannomas appear iso- to hypointense on T1-weighted sequences and markedly hyperintense on T2-weighted and STIR images, with intense, often heterogeneous post-contrast enhancement. These findings reflect the classic histologic components of schwannomas, including Antoni A and B areas, cystic degeneration, and variable vascularity [2,4].

The angiomatoid variant, as illustrated in this case, deserves particular emphasis. Angiomatoid schwannomas exhibit prominent vascular spaces, microhemorrhages, and blood-filled pseudocystic areas. On MRI, these correspond to internal susceptibility artifacts on T2* sequences, irregular T2 hyperintensity, and heterogeneous enhancement following gadolinium injection. Recognition of this subtype is important because its imaging appearance may resemble that of vascular orbital tumors or hemorrhagic lesions, posing diagnostic challenges. MRI not only characterizes internal architecture but also evaluates potential extension toward the cavernous sinus or intracranial compartment-critical considerations for surgical planning [4].

A thorough understanding of the differential diagnosis is essential due to significant clinical and imaging overlap with other orbital tumors. Optic nerve meningioma represents a key differential: it typically demonstrates the classic "tram-track" sign on axial post-contrast imaging, with linear enhancement surrounding but sparing the optic nerve itself. Optic nerve glioma, more frequent in children and often linked to neurofibromatosis type 1, shows fusiform enlargement of the optic nerve with relatively homogeneous T2 hyperintensity and less heterogeneous enhancement. Cavernous hemangioma, the most common intraconal mass in adults, appears as a sharply marginated lesion with progressive, centripetal enhancement rather than the diffuse enhancement seen in schwannoma. Other considerations include metastatic lesions, which tend to present with more aggressive, infiltrative features, and idiopathic orbital inflammation, which is typically painful, ill-defined, and responsive to corticosteroids. High-resolution ultrasonography may support differentiation by showing a solid, well-limited mass with possible internal cystic components, although MRI remains the key differentiating tool [4,5].

Despite advances in imaging, definitive diagnosis relies on histopathologic evaluation. Classic schwannomas show alternating Antoni A (cellular, palisading) and Antoni B (loose, myxoid) areas, often with Verocay bodies. The angiomatoid subtype is distinguished by abundant vascular channels, intralesional hemorrhage, and hemosiderin deposition, accounting for its heterogeneous imaging appearance. Immunohistochemistry is confirmatory, with strong and diffuse S-100 protein expression supporting Schwann-cell origin [2,5].

Complete surgical excision remains the treatment of choice. The surgical approach-transorbital, transcranial, or combined-depends on tumor location, optic canal involvement, and proximity to the cavernous sinus. Prognosis after total excision is generally excellent, with rare recurrence reported in the literature [1,3]. However, postoperative visual outcome is variable and closely linked to the severity and chronicity of preoperative optic nerve compression. Early recognition and timely intervention are therefore crucial to optimize visual prognosis.

## Conclusion

Angiomatoid schwannoma of the optic nerve is a rare orbital tumor that presents significant diagnostic challenges due to its atypical origin and overlapping imaging features with other orbital masses. MRI is central to characterization and surgical planning, while histopathology remains indispensable for definitive diagnosis. Clinicians and radiologists should consider this entity when evaluating progressive unilateral exophthalmos accompanied by visual decline, even though its occurrence is rare.

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

## Bibliography

1. Quigley C., *et al*. "Clinico-radiological features of optic nerve sheath schwannoma: Review and illustrative case". *European Journal of Ophthalmology* 35.2 (2025): 456-465.

2. Lune A., *et al*. "Optic nerve schwannoma: a report of a rare case from India and literature review". *Cureus* 16.5 (2024): e59824.

3. Shapira Y., *et al*. "Orbito-cranial schwannoma: a multicentre experience". *Eye (London)* 37.1 (2023): 48-53.

4. Roelofs KA., *et al*. "Radiologic features of well-circumscribed orbital tumors with histopathologic correlation". *Ophthalmic Plastic and Reconstructive Surgery* 40.4 (2024): 380-387.

5. Lefebvre G and Le Corroller T. "Ultrasound and MR imaging of peripheral nerve tumors: the state of the art". *Skeletal Radiology* 52.3 (2023): 405-419.

**Volume 17 Issue 2 February 2026**