

# Orbital Oculomotor Nerve Schwannoma Extending to the Cavernous Sinus: A Rare Cause of Proptosis

Hina Kauser<sup>1</sup>, MBBS, MS; Omar Rashid<sup>1</sup>, MBBS, MS; Waseem Anwar<sup>1</sup>, MBBS, MS; Sabina Khan<sup>2</sup>, MBBS, MD

<sup>1</sup>Department of Ophthalmology, Hamdard Institute of Medical Sciences and Research, Jamia Hamdard, New Delhi, India

<sup>2</sup>Department of Pathology, Hamdard Institute of Medical Sciences and Research, Jamia Hamdard, New Delhi, India

### Abstract

**Purpose:** To report a case of orbital oculomotor nerve schwannoma extending to the cavernous sinus through the superior orbital fissure presenting with proptosis, but without any neurological sign.

**Case Report:** A 32-year-old man presented with axial proptosis of his left eye. Visual acuity and other ocular examinations were normal. Orbital magnetic resonance imaging revealed a well-defined fusiform retrobulbar lesion in the left orbit extending into the superior orbital fissure and left cavernous sinus measuring 43 mm × 21 mm × 19 mm and causing superomedial displacement of the optic nerve and axial proptosis. The patient was scheduled for surgery, and gross total excision was done. Postoperatively, the patient developed total third nerve palsy. Pre and postoperative third nerve deficit confirmed the origin of the tumor from the oculomotor nerve. Histopathological examination revealed schwannoma.

**Conclusion:** Orbital oculomotor nerve schwannoma, although rare, can be the cause of proptosis. Diagnosis can be confirmed histopathologically. It is a benign tumor; however, it can extend intracranially without any neurological symptoms. Therefore, neuroimaging is essential to rule out intracranial extension. Early surgical removal is mandatory.

**Keywords:** Cavernous Sinus; Oculomotor Nerve; Proptosis; Schwannoma

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### INTRODUCTION

Schwannomas are encapsulated tumors which grow from peripheral nerves and rarely occur in the orbital and periorbital regions. Orbital schwannoma accounts for 1–4% of all orbital tumors,<sup>[1,2]</sup> are usually benign, but malignant changes have been described in some cases.<sup>[3]</sup> They usually arise from sensory rather than motor nerves. About 24% of orbital schwannomas have been reported to origin from the first division of the trigeminal nerve; however, in the majority of orbital schwannomas, the origin may not be identified based on radiological and clinical findings. Preoperative and postoperative neurological deficits usually lead to the identification of the source of these tumors.<sup>[4]</sup> Schwannomas originating from the oculomotor nerve are extremely rare. Approximately, 45 cases of oculomotor

nerve schwannomas have been reported in the literature so far. In a recent review, only 4 cases of oculomotor nerve schwannomas located in the orbit have been described.<sup>[5]</sup>

Herein, we report a case of orbital oculomotor nerve schwannoma extending to the cavernous sinus through the superior orbital fissure presenting with proptosis. As per our knowledge, no case of orbital schwannoma with these characteristics has been reported in the literature.

### CASE REPORT

A 32-year-old man presented with axial proptosis of the left eye [Figure 1a and b]. He had noted painless protrusion of this eye since 6 months before which was

#### Correspondence to:

Hina Kauser, MBBS, MS. RZ-904/24, Gali 24, Flat 202, Tughlakabad Extension, New Delhi - 110 019, India.  
E-mail: hina3kauser@yahoo.co.in

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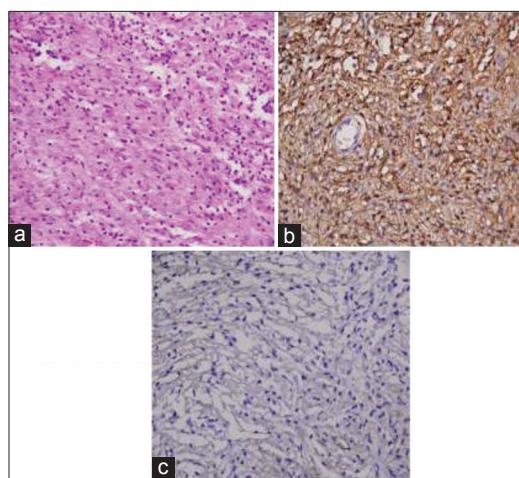
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gradually increasing. On examination, visual acuity was 20/20 in both eyes, and ocular movements and corneal sensation were normal bilaterally. Exophthalmometry revealed 3 mm of proptosis in the left eye. No abnormal findings were seen in the anterior segment and fundus examination. Systemic evaluation was done to rule out neurofibromatosis, and hematological tests were within normal limits. In magnetic resonance imaging, an antero-posteriorly oblong fusiform lesion measuring 43 mm × 21 mm × 19 mm was detected in the retrobulbar region of the left orbit extending to the left cavernous sinus through the superior orbital fissure causing superomedial displacement of the optic nerve and proptosis [Figure 2]. The patient was referred to the neurosurgery department and complete excision of the tumor was performed through left fronto-orbitotomy under general anesthesia, with careful separation and preservation of the nerve from which the tumor originated. During the operation, a greyish-yellow, soft, moderately vascular, cavitron ultrasonic aspirator suckable tumor arising from the sheath of left oculomotor nerve and extending up to the lateral wall of the cavernous sinus was seen. Total excision including the cavernous sinus portion of the lesion was done. Postoperatively, the patient developed total oculomotor nerve palsy in his left eye.



**Figure 1.** Preoperative axial proptosis of the left eye.

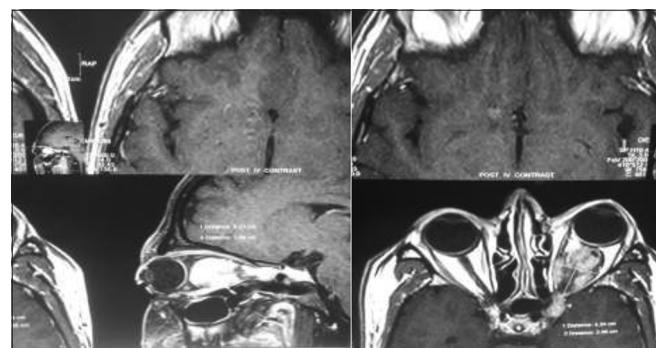


**Figure 3.** Tumor cells arranged in fascicles with elongated nuclei showing palisading (H and E stain 400X) (a) Tumor cells with S-100 positivity (b) Tumor cells showing EMA negativity (c).

Histopathological examination of hematoxylin and eosin stained sections showed Antoni type A [Figure 3a] and Antoni type B cellular patterns. Immunohistochemical examination revealed the tumor cells to be strongly positive for S-100 protein [Figure 3b] and negative for epithelial membrane antigen [Figure 3c] confirming the diagnosis of schwannoma. After 4 months, no sign of recurrence was noted except for complete third nerve palsy [Figure 4].

## DISCUSSION

Schwannomas are encapsulated, slow-growing tumors composed of differentiated schwann cells. They may occur in any location and rarely in the orbital and periorbital region accounting only for 1% of orbital tumors.<sup>[6]</sup> They are also found in patients with neurofibromatosis.<sup>[7]</sup> Schwannomas are commonly seen in the 20–50 age group but may occur at any age with no sexual preference.<sup>[8]</sup> Schwannomas are usually asymptomatic when small and may produce various signs and symptoms depending on the size,



**Figure 2.** MRI showing an antero-posteriorly oblong fusiform lesion measuring 4.3x2.1x1.9 cms in size in the retro bulbar region of left orbit extending into superior orbital fissure and the left cavernous sinus causing superomedial displacement of optic nerve and proptosis.



**Figure 4.** Complete third nerve palsy four months after operation.

origin, location, and extent of the tumor.<sup>[9]</sup> They may present as painless progressive proptosis on enlargement or may compress the optic nerve along with papilledema or optic atrophy. Patients may present with sudden or gradual visual loss and ocular motility dysfunction, but such symptoms are not specific for the schwannoma. Early surgical excision of the tumor to prevent compression of the optic nerve is the treatment of choice.<sup>[10]</sup> Thirty-eight reported cases with solitary oculomotor schwannoma including 15 male and 23 female subjects aged 8–74 years; among these subjects preoperative oculomotor dysfunction was seen in 29 patients. The tumor was situated in the orbit in only 4 cases (solitary orbital type), in the subarachnoid space in 17 subjects (cisternal type), in the cavernous sinus in 12 patients (cavernous type), and extending from the cavernous sinus to the cistern in 5 cases (cisternocavernous type).<sup>[11–14]</sup> As the growth of orbital schwannoma is slow, operation is usually delayed by 2 years from the onset of symptoms.<sup>[15]</sup> Total surgical excision is the treatment of choice for orbital schwannomas causing pain, disfigurement, diplopia, or optic neuropathy.<sup>[16]</sup>

Only 4 cases of orbital oculomotor schwannoma (solitary orbital type) have been reported in the literature and to date, no case of orbital oculomotor schwannoma extending to the cavernous sinus has been reported. In the patient presented herein, the oculomotor nerve was the origin of schwannoma but this could not be identified based on preoperative radiological findings and clinical presentation. In fact, the origin of the tumor was confirmed during surgery and by development of postoperative neurological deficit. The patient had no sign of compression even though the tumor was extending to the cavernous sinus through the superior orbital fissure. Preoperative oculomotor dysfunction (reported in 29 cases out of 38 in the literature) was not present in our case.

In summary, this case report illustrates that the absence of neurological and compressive signs and symptoms in a patient with proptosis does not exclude intracranial extension and necessitates radiological investigations. Although orbital schwannomas are benign slow-growing tumors, early surgical excision is advocated to prevent compression of the optic nerve and intracranial extension.

## REFERENCES

- Cantore WA. Neural orbital tumors. *Curr Opin Ophthalmol* 2000;11:367-371.
- Rose GE, Wright JE. Isolated peripheral nerve sheath tumours of the orbit. *Eye (Lond)* 1991;5:668-673.
- Jakobiec FA, Font RL, Zimmerman LE. Malignant peripheral nerve sheath tumors of the orbit: A clinicopathologic study of eight cases. *Trans Am Ophthalmol Soc* 1985;83:332-366.
- Cantore G, Ciappetta P, Raco A, Lunardi P. Orbital schwannomas: Report of nine cases and review of the literature. *Neurosurgery* 1986;19:583-588.
- Scheller C, Rachinger JC, Prell J, Alfieri A, Rampp S, Sel S, et al. Intraorbital oculomotor nerve schwannoma affecting only the parasympathetic fibers. *J Neurol Surg A Cent Eur Neurosurg* 2013;74:120-123.
- Dasar M, Koppel BS, Heise CW, Mazzeo MJ, Poon TP, Duffy KR. Multiple spinal intradural schwannomas in the absence of von Recklinghausen's disease. *Spine (Phila Pa 1976)* 1993;18:2556-2559.
- Pollock SC. Tumors of cranial and peripheral nerves. In: Miller NR, Newman NJ, editors. *Walsh and Hoyt's Clinical Neuro-Ophthalmology*. Baltimore: Williams and Wilkins Co.; 1998. p. 2297-2327.
- Delfini R, Missori P, Tarantino R, Ciappetta P, Cantore G. Primary benign tumors of the orbital cavity: Comparative data in a series of patients with optic nerve glioma, sheath meningioma, or neurinoma. *Surg Neurol* 1996;45:147-154.
- Rootman J, Goldberg C, Robertson W. Primary orbital schwannomas. *Br J Ophthalmol* 1982;66:194-204.
- Konrad EA, Thiel HJ. Schwannoma of the orbit. *Ophthalmologica* 1984;188:118-127.
- Barat JL, Marchal JC, Bracard S, Auque J, Martin-Beuzart S, Hepner H. Neurinoma of the oculomotor nerves. Apropos of 2 cases. *Neurochirurgie* 1992;38:183-187.
- Kurokawa Y, Uede T, Honda O, Honmou O. Successful removal of intracavernous neurinoma originating from the oculomotor nerve – Case report. *Neurol Med Chir (Tokyo)* 1992;32:225-228.
- Schluthess R, Kristof R, Schramm J. Complete removal of an oculomotor nerve neurinoma without permanent functional deficit. Case report. *Ger J Ophthalmol* 1993;2:228-233.
- Kachhara R, Nair S, Radhakrishnan VV. Oculomotor nerve neurinoma: Report of two cases. *Acta Neurochir (Wien)* 1998;140:1147-1151.
- Anegawa S, Hayashi T, Torigoe R, Iwasako K, Sakae N, Oshio Y. Orbital neurinoma presenting orbital apex syndrome. *No Shinkei Geka* 1997;25:473-477.
- Butt ZA, McNab AA. Orbital neurilemmoma: Report of seven cases. *J Clin Neurosci* 1998;5:390-393.

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