Case Report

Oculomotor nerve schwannoma of orbit with extension into cavernous sinus

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ABSTRACT

Schwannomas (neurilemmomas) are benign encapsulated, slow growing peripheral nerve sheath tumors. Primary orbital schwannomas are uncommon, accounting for about 1–4% of orbital tumors. Characteristically, they arise from the first division of the trigeminal nerve. Oculomotor schwannomas are rare tumors with only one case reported till date. Here, we report a 28-year-old male presenting with an axial proptosis of left eyeball with progressive loss of vision. Magnetic resonance imaging of orbit revealed a large left-sided orbital mass extending into cavernous sinus. Surgical excision of the mass was done, and based on clinical, histological, and immunohistochemical analysis, a final diagnosis of oculomotor nerve schwannoma with left orbital extension was made.

Key words: Magnetic resonance imaging, nerve, oculomotor, orbit, proptosis, schwannoma

INTRODUCTION

Schwannomas are encapsulated, slow growing tumors comprising differentiated Schwann cells. They may occur in any location, and orbital schwannoma accounts for 1–4% of all orbital masses.[1] The tumor has no significant sex predilection and may present at any age.[2] Yoshida and Kawase have categorized extracranial schwannomas into infratemporal, orbital, and pterygopalatine fossa components.[3] Typically, they arise from the first division of the trigeminal nerve. Oculomotor schwannomas are rare masses with only one case reported till date. The displacement of the eyeball is related to the site and extent of the tumor mass. The patients commonly present with proptosis, diplopia, limitation of eyeball movement, and neuropathy. Most schwannomas necessitate timely treatment as they grow progressively. The growth of the mass may compress the optic nerve leading to optic disc edema or optic atrophy. Surgical excision of this mass is the treatment of choice, and radiation therapy is a secondary treatment in few cases.[4,5] We present a case of oculomotor nerve schwannoma extending into cavernous sinus which is rare and discuss the differential diagnosis.

CASE REPORT

A 28-year-old male presented with painless, progressive vision loss in left eye since 2 years associated with progressive proptosis. On ocular examination, perception of light was absent in left eye and visual acuity was 6/6 in the right eye. The corneal sensation was reduced, and ocular movements were restricted on the left side due to associated third cranial nerve palsy. Slit lamp and fundus examination showed dilated nonreacting pupil in the left eye and optic atrophy. Magnetic resonance imaging (MRI) images [Figure 1] demonstrated a large well-defined fusiform mass in left orbit extending into left cavernous sinus through optic foramen and superior

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orbital fissure and causing displacement of optic nerve and proptosis. The mass was heterogenously hypointense on T1-weighted and hyperintense on T2-weighted images. There were areas of cystic degeneration appearing hyperintense on T2 and incomplete signal suppression on FLAIR. Imaging findings were suggestive of a diagnosis of orbital mass with extension into cavernous sinus. Excision biopsy was done under general anesthesia, and histopathological examination revealed areas of closely packed spindle cells having fusiform nuclei and eosinophilic cytoplasm (Antoni A) [Figure 2] and immunohistochemical analysis of the specimen was positive for S-100 protein [Figure 3] thus confirming the diagnosis. The patient was taken up for surgery, and total excision was performed. The patient had total third nerve palsy after surgery. Intraoperative evaluation as well as pre- and post-operative third nerve deficit corroborated with the origin of the tumor from the oculomotor nerve.

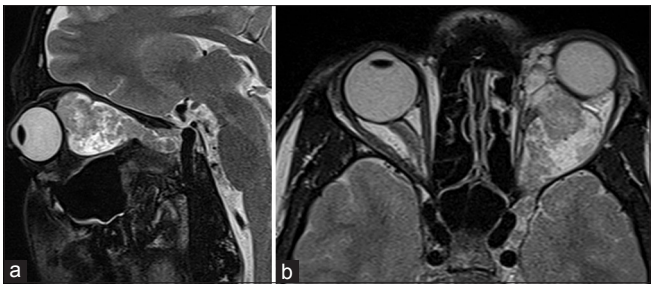

Figure 1: (a) Sagittal T2-weighted image magnetic resonance imaging showing an oblong-fusiform mass in the retrobulbar region of left orbit extending into superior orbital fissure and the left cavernous sinus causing proptosis. (b) Axial T2-weighted image showing heterogeneously hyperintense mass in orbit and extending into cavernous sinus

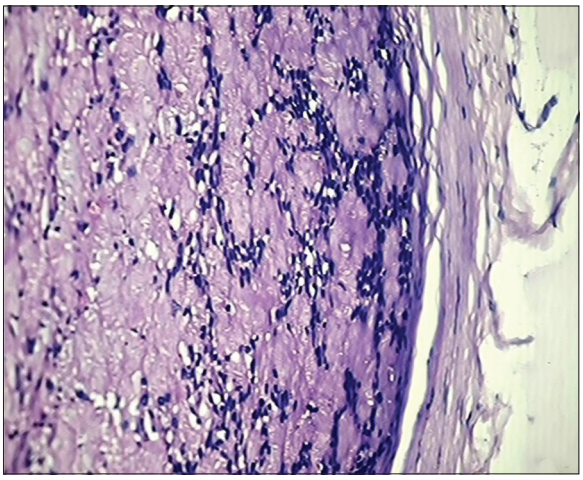

Figure 2: Hematoxylin and eosin stain depicting spindle cells in a myxoid collagenous background

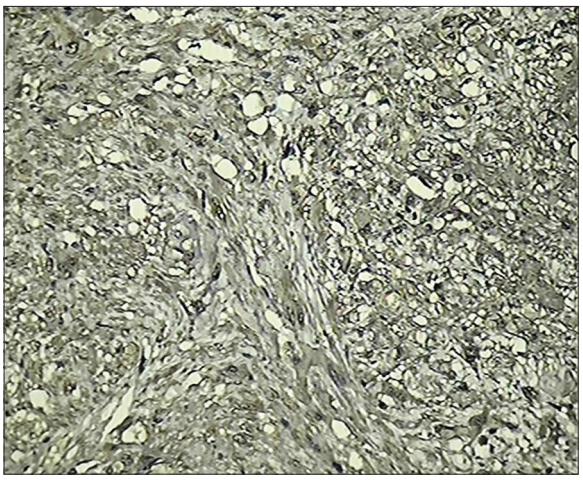

Figure 3: Immunohistochemistry revealing S-100 positivity indicating schwannoma

DISCUSSION

Schwannomas typically originate from Schwann cells and propagate along cranial, peripheral, and autosomic nerves. They predominantly originate from sensory nerves rather than motor nerves. In present case, the mass was a schwannoma affecting the third nerve with resultant restricted extraocular movements in the right eye. He also had optic atrophy due to the compression of the optic nerve by the mass. In this patient, the oculomotor nerve was the source of schwannoma, but this could not be confirmed based on preoperative MRI examination and clinical findings. The source of the tumor was established during surgery and by the occurrence of postoperative neurological deficit. The main differential diagnosis of a mass involving orbit and cavernous sinus is trigeminal schwannoma,[5] which has different clinical findings, and the bulk of the mass lies in the cavernous sinus while in our case the bulk of the mass was present in the orbit. On review of literature, approximately 24% of orbital schwannomas have been described to arise from the first division of the trigeminal nerve; yet, the source of more than half of the orbital schwannomas could not be recognized from preoperative radiological features, intraoperative anatomical judgment, and clinical manifestations. Consequently, postoperative neurological deficits, such as restriction of eyeball movement and numbness of the forehead, generally led clinicians to associate the oculomotor nerves and trigeminal nerves as the source of these masses. Histological findings can be variable, with the Antoni A and B type cells present typically but not always seen. A true capsule of perineural tissue encloses schwannomas and if in one piece confirms complete excision. Immunohistochemical analysis with S-100 protein gives complementary information when classical histology is not present. Surgical removal is the treatment of choice in symptomatic patients. The surgical approach is dependent on the site of the masses. Intraorbital schwannomas mainly occur in the superior portions of both the intraconal and extraconal spaces of orbit consequently, anterior orbitotomy or supra orbitotomy is favored in majority of cases.[6-8] To best of our knowledge, only one case of orbital oculomotor

schwannoma extending into cavernous sinus has been reported till date.[9] This is probably due to the fact that in majority of the cases, the origin of the orbital schwannoma cannot be ascertained intraoperatively. The nerve origin is generally made out from histological examination of the specimen and the postoperative neurological deficits of the patient. To conclude, schwannoma though an infrequent entity should be considered as a differential diagnosis in a patient with slow progressive painless vision loss and proptosis and timely surgical intervention is obligatory for the prevention of permanent vision loss.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Daras 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;18:2556-9.
2. Delfini R, Missori P, Tarantino R, Ciapetta 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;45:147-53.
3. Yoshida K, Kawase T. Trigeminal neurinomas extending into multiple fossae: Surgical methods and review of the literature. J Neurosurg 1999;91:202-11.
4. Goel A, Muzumdar D, Raman C. Trigeminal neuroma: Analysis of surgical experience with 73 cases. Neurosurgery 2003;52:783-90.
5. Ghosh S, Das D, Varshney R, Nandy S. Orbital extension of trigeminal schwannoma. J Neurosci Rural Pract 2015;6:102-4.
6. Cantore G, Ciappetta P, Raco A, Lunardi P. Orbital schwannomas: Report of nine cases and review of the literature. Neurosurgery 1986;19:583-8.
7. Levin LA, Jakobiec FA. Peripheral nerve sheath tumors of the orbit. In: Albert DM, Jakobeic FA, editors. Principles and Practice of Ophthalmology. Philadelphia: WB Saunders; 1994. p. 1978-2004.
8. Pollock SC. Tumors of cranial and peripheral nerves. In: Miller NR, Newman NJ, editors. Walsh and Hoyt's Clinical Neuro-ophthalmology. Vol. 2. Baltimore: Williams and Wilkins Co.; 1998. p. 2297-327.
9. Kauser H, Rashid O, Anwar W, Khan S. Orbital oculomotor nerve schwannoma extending to the cavernous sinus: A rare cause of proptosis. J Ophthalmic Vis Res 2014;9:514-6.