Case Report

Orbital intraconal abducens nerve schwannoma: An interdisciplinary approach for management

ABSTRACT

Schwannomas are rare benign tumors arising from Schwann cells of the nerve sheath. Although the head-and-neck region accounts for a large percentage of extracranial schwannomas, those located within the orbit are infrequent. This paper presents an extremely rare case of orbital schwannoma arising from the terminal branch of abducens nerve in a 37-year-old female. The lesion presented as a large intraconal mass causing proptosis and weakness to abduct the left eye. Imaging showed a well-defined, solid-cystic lesion, measuring 2.7 cm and displacing the lateral rectus muscle laterally and the optic nerve medially. A multidisciplinary approach was used for the surgical management of the tumor. Access was attained through lateral rim osteotomy. Histopathological evaluation was diagnostic for schwannoma. A search of English literature revealed only five previously published cases of abducens nerve schwannomas. A review of these cases has also been discussed along with the present report.

Keywords: Access osteotomy, neurogenic, orbit, tumor

INTRODUCTION

Schwannomas are rare benign tumors that arise from the Schwann cells of neural sheath of a motor or sensory nerve. It may involve either the cranial, intraspinal, or peripheral nerves and present as a well-localized encapsulated swelling.[1] Head-and-neck region accounts for 20%–45% of extracranial schwannomas.[2] However, a lesion within the orbit is rare and contributes to about 1%–2% of all the orbital neoplasms.[3]

Based on its anatomical location, orbital schwannomas can be intraconal or extraconal. Extraconal lesions are located in the periphery of orbital cavity and usually arise from the terminal branches of trigeminal nerve, most commonly the supraorbital and infraorbital nerves.[2] Intraconal lesions are extremely rare. They are located within the rectus muscle cone and originate from the peripheral branches of oculomotor, trochlear, or abducens nerves.[4] This paper reports a case of intraconal abducens schwannoma. The clinical presentation, imaging findings, and management of the pathology have been highlighted.

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CASE REPORT

A 37-year-old systemically healthy female patient presented with a chief complaint of increase in size of left globe of eye with pain for the past 2 months. On clinical examination, proptosis with partial limitation in abduction of the left eye was seen [Figure 1]. There was no loss of vision or diplopia.

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Ophthalmological evaluation revealed normal visual acuity on using the Snellen chart eye examination. No other neurological abnormalities were detected. No neurocutaneous markers of neurofibromatosis were identified.

Magnetic resonance imaging of the left orbit revealed well-defined solid-cystic lesion, between optic nerve and lateral rectus muscle (LRM), causing stretching of the muscle and extending up to the aponeurosis [Figure 2]. The lesion showed heterogenous iso-intensities on T1 and hyper-intensities on T2-weighted contrast-enhanced images. The lesion measured 2.27 cm × 2.07 cm × 2.69 cm, with brightly enhancing solid component and nonenhancing cystic component. There was loss of fat plane between the mass and the LRM. However, the belly of LRM was of normal caliber. The lesion compressed the optic nerve, displacing it medially. Lesion caused anterior displacement of globe, producing proptosis [Figure 2].

Based on the clinical presentation and imaging findings, a provisional diagnosis of benign orbital tumor was made. Differential diagnosis included abducens nerve schwannoma, meningioma, and lymphoma. However, its characteristic location, relation to LRM, and abducens nerve weakness were strongly suggestive of isolated schwannoma arising from terminal branch of abducens nerve.

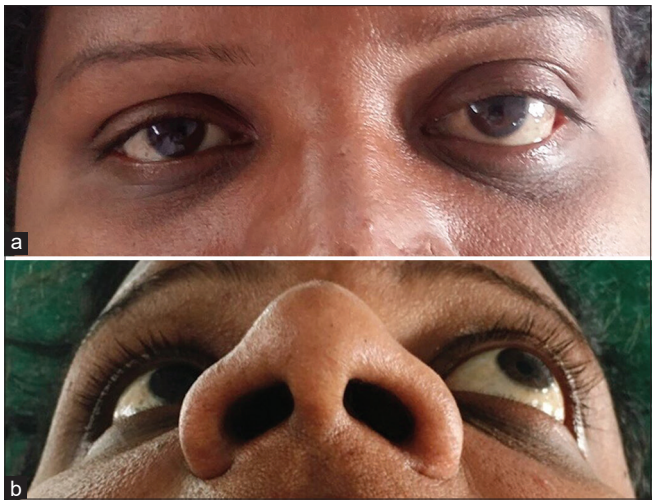

Figure 1: Preoperative photographs showing proptosis (a) and limitation of lateral movement of left eye (b)

Surgical excision of the lesion was planned under general anesthesia. Access to the lesion was achieved using lateral orbital rim osteotomy. An upper eyebrow incision with lateral crow feet extension was used to reach the lateral orbital rim [Figure 3a]. The periosteum over the lateral orbital rim was stripped and the temporalis muscle was retracted posteriorly [Figure 3b]. The osteotomy cuts were marked at the level of frontozygomatic suture above and at the base of the lateral rim below [Figure 3c]. The rim along with lateral wall was removed after completion of the cuts and stored temporarily in isotonic saline solution. Exposure to the intraconal content was attained by sharply incising the tenons capsule [Figure 3d]. Through careful blunt dissection, the lesion was freed from surrounding tissues. The lesion was found abutting the LRM. The plane of dissection was kept medial to the inner surface of the muscle and in toto excision of the lesion was done [Figure 3e]. Complete hemostasis was attained using low-power bipolar cautery. Sutures were placed to close the tenons capsule. The osteotomized lateral orbital rim was repositioned and fixed using thin profile 1.5 mm titanium microplate and screws [Figure 3f].

The excised specimen was submitted for histopathological analysis, which predominantly showed spindle-shaped cells arranged in Antoni A pattern around verocay bodies, along with less organized Antoni B tissues at some places, confirming the diagnosis of schwannoma [Figure 4].

The patient had uneventful postoperative period. There was gradual recovery of abducens nerve palsy with improvement of abduction of the left eye. The proptosis of eye was completely resolved at 2-month follow-up with no visual deficit [Figure 5]. The patient was kept on periodic recall and was free of any clinical and radiological evidence of recurrence at 2 years of follow-up.

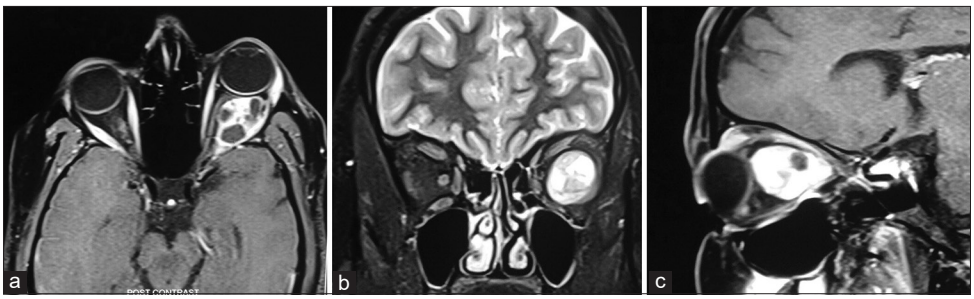

Figure 2: Magnetic resonance imaging of the lesion presenting as oval-to-round solid-cystic mass measuring 2.27 cm × 2.07 cm × 2.69 cm, located between optic nerve and lateral rectus muscle, causing stretching of the muscle and medial deviation of optic nerve; axial (a), coronal (b) and sagittal (c) views

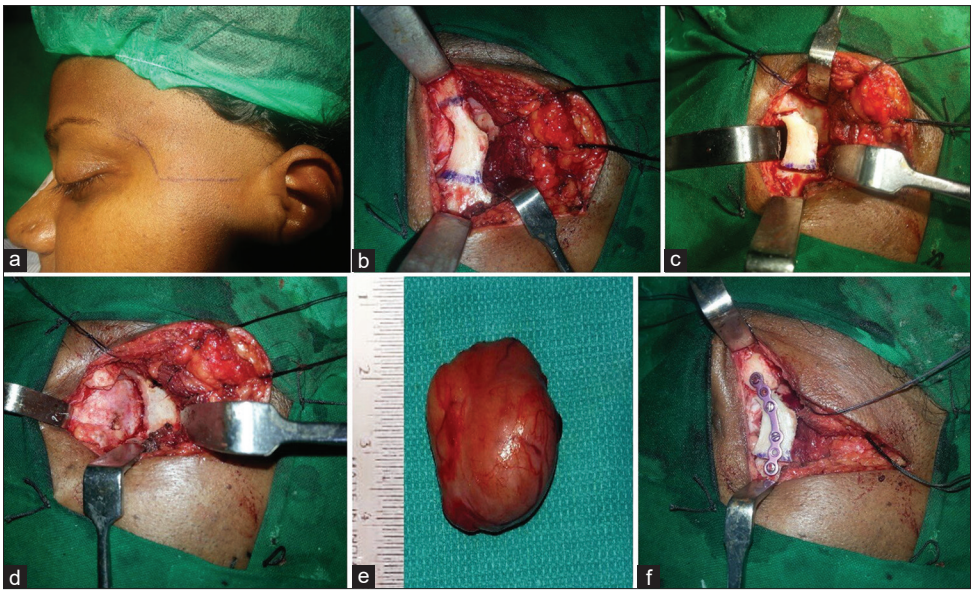

Figure 3: Intraoperative images showing; incision (a), access lateral orbital rim osteotomy cuts marked (b) and completed (c), exposure of the orbital content (d), excised lesion (e), and fixation of lateral orbital rim (f)

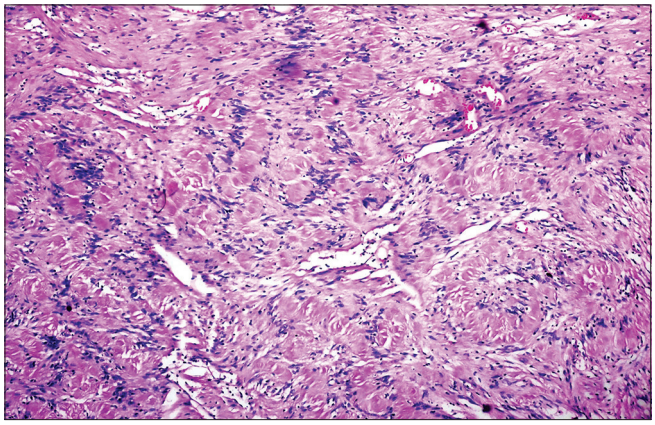

Figure 4: Photomicrograph of excised specimen showing spindle-shaped cells predominantly arranged in Antoni A pattern around verocay bodies along with less organized Antoni B tissue (H and E stained section, ×10 magnification)

DISCUSSION

Schwannomas are benign tumor arising from Schwann cells of the nerve. They are also known as neurilemmomas, perineural fibroblastomas, and neurinomas.[5] Schwannomas can present in three clinical forms; as an isolated lesion, as part of von Recklinghausen syndrome, or as multiple lesions (schwannomatosis).[6] The tumor arises from the nerve sheath and present as a well capsulated, eccentrically growing mass. It can involve the nerve in its intracranial course or arise from the terminal branches, located extracranially. Schwannomas are known to be associated with all the cranial nerves, except optic and olfactory nerves. It predominantly involves VIII cranial nerve.[7]

Although head-and-neck region accounts for large percentage of extracranial schwannomas, those arising within the orbit

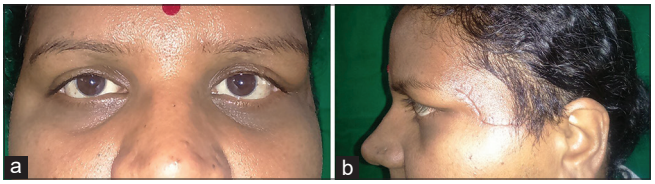

Figure 5: Postoperative clinical photographs showing resolution of proptosis (a) and surgical scar (b)

are extremely rare. Intraorbital tumors can arise from the sensory terminal branches of trigeminal nerve and are located outside the cone of rectus muscles. These extraconal orbital schwannomas may involve the infraorbital nerve and present as swelling in the inferior compartment of the orbit or over the cheek below the orbital rim.[2] Those arising from the terminal branches of ophthalmic nerve presents as swelling in superior compartment.

Intraconal schwannomas may arise from peripheral branches of oculomotor, trochlear, or abducens nerve. Although abducens nerve has the longest intracranial course, schwannoma involving the abducens nerve is extremely rare. In its intracranial course, the abducens nerve schwannomas are usually located within the cavernous sinus and in the brainstem region.[8,9] The terminal branches of abducens nerve lies within the intraconal compartment of the orbit and innervates the LRM. Its action results in the outward gaze (abduction) of the eye. Schwannomas arising from these terminal branches are extremely rare. Search of English literature in PubMed database, using terms; orbital, schwannoma, and abducens nerve showed 14 papers. On further review of the papers and going through their reference, only five case reports of isolated schwannoma

involving the terminal branch of abducens nerve were identified[10-14] [Table 1].

Isolated intraconal abducens nerve schwannomas have been reported from across the globe. Lesions were exclusively seen in adults, between the age range of 32–55 years. Both males and females and left and right eyes were equally affected. Clinical presentation of abducens nerve schwannoma is related to the pressure effect of tumor on the surrounding vital structures. These lesions typically presents as a mass lateral to the LRM and medial to the optic nerve. Smaller lesion may be asymptomatic. Iida *et al*. reported the smallest lesion of about 1 cm in size, which was accidentally discovered during medical checkup of the brain.[14] Symptomatic lesions predominantly present as proptosis,[10-13] diplopia,[10-12] and pain or discomfort in the eye.[11,13] Pressure atrophy of optic nerve from mass lesion can cause progressive vision loss.[11,12] Loss of innervation to the LRM manifests as inability to move the affected eye laterally (abduction) and is a clinical indication of involvement of the abducens nerve. Pain and proptosis, along with lack of abduction of left eye, were the clinical features of the present case.

Imaging is an essential tool in the diagnosis and treatment planning of orbital schwannomas. Magnetic resonance imaging provides information about the exact location, extent, nature, and size of the lesion. Intraconal abducens nerve schwannoma typically presents as a well-defined mass lesion, lateral to the LRM. The lesion is often seen abutting or displacing the optic nerve medially and superiority. However, infiltration into the optic nerve has never been reported.

Degenerative changes have been reported in schwannomas, in the form of calcification, hemorrhage, hyalinization, and cyst formation.[15] Cystic degeneration is believed to occur when a large tumor outgrows its blood supply. Such lesions are capable of rapid growth due to the enlargement of its cystic component.[11] The reported size of orbital abducens nerve schwannomas varied from 4.5 cm to 1 cm.[12,14] The lesions were predominantly solid, with only one cystic tumor described by Rato *et al*.[11] In our case, the lesion measured 2.7 cm and had cystic component to it. The LRM was displaced laterally with no fat plane between the mass and the muscle. Optic nerve was medially displaced, however showed normal signal intensity.

Surgical excision remains the mainstay for management and provides specimen for confirmatory histopathological diagnosis of orbital schwannomas. Although frontal craniotomy has been used to approach the lesion,[13] lateral orbital rim osteotomy (lateral orbitotomy) is more popular and provides direct access to the lateral orbital compartment. The lateral orbitotomy approach was first described by Krönlein in 1889 and later modified by Berke in 1954.[16,17] The surgical procedure involves temporary removal of the lateral orbital rim to gain access to the tumors in the lateral compartment of orbit. Both coronal[13] and S-shaped eyebrow incisions[10] have been described for lateral orbitotomy and tumor removal. In our case, sufficient access was attained using eyebrow incision with

Table 1: Review of orbital intraconal abducens nerve schwannomas

Author, publication year	Country	Age/ sex	Side	Clinical features	Tumor size (cm)	Tumor type	Incision	Surgical approach	Extent of removal	VI nerve function postoperative	Follow-up
Irace *et al.*, 2008[10]	Italy	55/ male	Left	Proptosis. diplopia	ND	Solid	S-shape eyebrow incision	Lateral orbitotomy	Total	Partial recovery	2 months
Rato *et al.*, 2012[11]	Portugal	42/ male	Left	Pain, proptosis, diplopia, progressive vision loss, focal abducens nerve palsy	2.2	Cystic	ND	Lateral orbitotomy	Total	Preserved	6 months
Feichtinger *et al.*,2013[12]	Austria	53/ female	Right	Proptosis, diplopia, optic atrophy	4.5	Solid	ND	Lateral orbitotomy	Partial	Partial recovery	1 year
Bhaganagare *et al.*, 2015[13]	India	32/ female	Right	Discomfort, proptosis	2.1	Solid	Coronal	Frontal craniotomy and superior orbitotomy	Total	Preserved	6 months
Iida *et al.*,2016[14]	Japan	51/ male	Left	Asymptomatic	1.2	Solid	ND	Lateral orbitotomy	Total	Preserved	Immediate postoperative
Present case	India	37/ female	Left	Proptosis, pain, focal abducens nerve palsy	2.7	Cystic	Eyebrow incision with crow-feet extension	Lateral orbitotomy	Total	Preserved	2 years

ND: Not described

lateral crow-feet extension. The crow-feet extension allowed wider field of access, facilitating the surgical dissection.

Knowledge of surgical anatomy and innervation of the extraocular muscles is essential to avoid its damage and limit the postoperative morbidity. The terminal branch of abducens nerve lies on the medial surface of the LRM. There are usually 3 terminal branches which innervate the muscle at its middle third.[18] Schwannomas arising from the terminal branches are believed to originate at the point of entry of nerve fiber into the muscle (myoneural junction).[10]

Although complete excision is recommended treatment, however proximity to vital structures within the orbit may not permit it at times. Feichtinger *et al*. reported enucleation while leaving the capsule behind, to prevent injury to the abducens nerve.[12] Blunt dissection around the capsulated lesion and use of low-power bipolar cautery provided tissue plane for complete removal of the lesion in the present case. Further attempts were not made to identify the terminal branches of the nerve, to minimize the risk of surgical trauma and possible damage to the nerve. The typical location of the tumor and presence of abducens nerve weakness was diagnostic of its involvement with schwannoma. Recurrence after extracapsular excision of schwannoma is very rare.[19,20] Although recurrence after intracapsular dissection is not well documented, recent study found no recurrence after intracapsular dissection within their follow-up periods.[21] No recurrence of orbital abducens nerve schwannomas were reported in literature, including the present report.

CONCLUSION

Orbital schwannomas arising from the terminal branch of abducens nerve have been previously reported in English literature in only five patients. These lesions typically present as well-defined solid or cystic mass located lateral to the LRM. It causes proptosis, pain, abducens nerve palsy, and diplopia. Magnetic resonance imaging helps to define the size, extent, and nature of the tumor. Management is purely surgical, with both complete excision and enucleation reported with satisfactory outcome. In the present case, lesion was approached with eyebrow incision with lateral extension. Lateral orbital rim access osteotomy provided adequate exposure for complete removal of the pathology. The patient was asymptomatic with resolution of symptoms at 2-year follow-up.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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