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Orbital schwannoma arising within inferior rectus muscle: A rare orbital tumor

Pedram Afshar [a], Seyed Mohsen Rafizadeh [a,*], Nasim Eshraghi [b], Sajjad Mansourian [a], Amirhossein Aghajani [a], Fahimeh Asadi Amoli [c]

[a] Department of Orbital and Oculoplastic Surgery, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
[b] Vali-E-Asr Reproductive Health Research Center, Family Health Research Institute, Tehran University of Medical Sciences, Tehran, Iran
[c] Pathology Department, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran

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ABSTRACT

Purpose: To report a rare orbital schwannoma arising within inferior rectus muscle in a young woman.
Observations: In this case report, we describe a young patient with slowly progressive proptosis for one year. Preoperative imaging was suggestive of a benign tumor arising within inferior rectus muscle, and histopathologic evaluation revealed an orbital schwannoma with cystic degeneration.
Conclusion and importance: This case report highlights the importance of including schwannoma in the differential diagnosis of any orbital tumor arising within muscle.

1. Introduction

Schwannomas (neurilemmomas) are benign nerve sheath tumors that originate from schwann cells which are responsible for producing myelin sheath in peripheral nerves.[1,2] Orbital schwannomas are rare neoplasms that account for about 1–6.5 % of all tumors in the orbit.[3] They are well-defined and encapsulated tumors that usually arise from the sensory branches of cranial nerve V.[4] Orbital schwannomas have a slow rate of progression and their most common presentation is painless proptosis.[5] In this article, we report and discuss the radiological diagnosis, surgical management, and histopathologic characteristics of a rare orbital schwannoma arising within inferior rectus muscle.

2. Case report

A 32-year-old woman was referred to the orbit clinic of our hospital with a slowly progressive proptosis in her right eye that started approximately one-year prior to presentation. The patient had no eye pain and complained of diplopia when looking up and down. In her medical history, the patient did not have any significant systemic or ocular problems. On ophthalmologic examination, visual acuity was 20/20 in both eyes. Hertel exophthalmometry was 25 mm in the right eye and 19 mm in the left eye (Fig. 1A and B). The margin reflex distance 1 (MRD 1) was 3 mm in both eyes, and the margin reflex distance 2 (MRD 2) was 7 mm in the right eye and 5 mm in the left eye. Movements of the right eye were limited in upgaze (−2) and downgaze (−1). Other ophthalmic examinations, including pupillary reaction, RAPD, intraocular pressure, and fundus findings, were normal in both eyes.

Orbital CT scan revealed enlargement of the right inferior rectus muscle (Fig. 2, A). Magnetic resonance imaging (MRI) with contrast of the orbit showed a well-rounded lesion measuring 5× 15 × 25 mm in the right inferior rectus muscle, with heterogeneous enhancement, especially in the peripheral region. The other extraocular muscles and soft tissues were normal (Fig. 2, B, C, and D). The preoperative diagnosis was a benign tumor arising from the inferior rectus muscle, such as a neurogenic tumor, and the patient was scheduled for surgery.

Using an inferior transconjunctival approach, dissection was carried out to the inferior orbital rim. The periosteum was incised along the orbital rim, and the periorbita was separated from the orbital floor bone. The periorbita was then incised with fine scissors, and after gentle dissection, the enlarged inferior rectus muscle was exposed. The encapsulated creamy round cystic mass was identified, carefully dissected from the surrounding tissue, and finally resected by dissection between the inferior rectus muscle fibers. The integrity of the inferior rectus muscle was then checked, hemostasis was achieved and the conjunctiva was sutured. The eye was bandaged with antibiotic ointment, and the patient was examined the day after surgery. During the examination, the patient demonstrated 20 prisms of hypertropia and 10

* Corresponding author.
E-mail address: mohsen_raf1354@yahoo.com (S.M. Rafizadeh).

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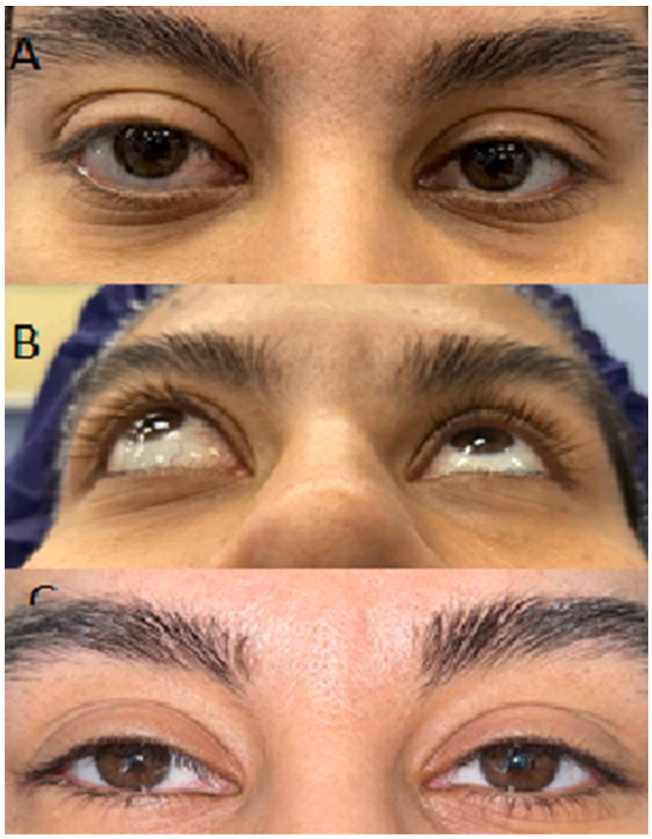

Fig. 1. The facial photographs (A and B) show right eye axial proptosis before surgery and (C) shows the same patient one year after surgery.

prisms of exotropia in the right eye. One month after the surgery, the patient's vision was 10/10, the deviation had resolved and she did not have diplopia. No signs of tumor recurrence were observed during the one-year postoperative follow-up (Fig. 1, C).

The lesion was composed of spindle cells with narrow elongated wavy nuclei with occasional nuclei showing large bizarre nuclei consistent with degenerative change. The tumor contained hypercellular areas with palisading nuclei, consistent with Antoni A areas; and hypocellular regions with myxoid stroma, consistent with Antoni B areas. Prominent central cystic degeneration areas and focal cellular atypia were seen. Neoplastic cells were narrow, elongated, and wavy with tapering ends interspersed with collagen fibers. There were no mitoses or capsular invasion. Immunohistochemical (IHC) staining of the tumor showed that tumor cells were strongly positive for S-100 protein (Fig. 3).

3. Discussion

Primary orbital schwannoma is a benign, slowly progressive, encapsulated, and usually unilateral tumor.[6] Orbital schwannomas usually arise from sensory branches of the V1 division of the trigeminal nerve, and more frequently originate from branches of supraorbital or supratrochlear nerves, which explains why superior orbit is commonly involved.[7,8] Less commonly, an orbital schwannoma may arise from motor nerves, and rarely from orbital extraocular muscles. There are very few published reports of orbital schwannomas arising from the extraocular muscle's nerve, mimicking an intramuscular tumor, in the literature.[9–13] In this article, we report a rare presentation of orbital schwannomas, a schwannoma arising from the inferior rectus muscle. Schwannomas account for 1–6.5 % of orbital tumors. Orbital schwannomas typically occur during the second to fifth decades of life and pediatric cases are very rare.[3] However, they should be considered as a differential diagnosis of orbital tumors at any age.

The most common clinical presentations of orbital schwannomas include progressive painless proptosis, globe displacement, eyelid swelling, diplopia, and pain.[14,15] In this case, the patient presented with progressive proptosis and diplopia in the absence of ocular pain.

The use of various imaging modalities plays a key role in the diagnosis of schwannoma before surgical excision. Orbital schwannoma appears as a homogenous hypodense to hyperdense well-defined lesion on CT scan. Previous studies have shown a correlation between the histologic properties of schwannomas and CT scan findings; Antoni A

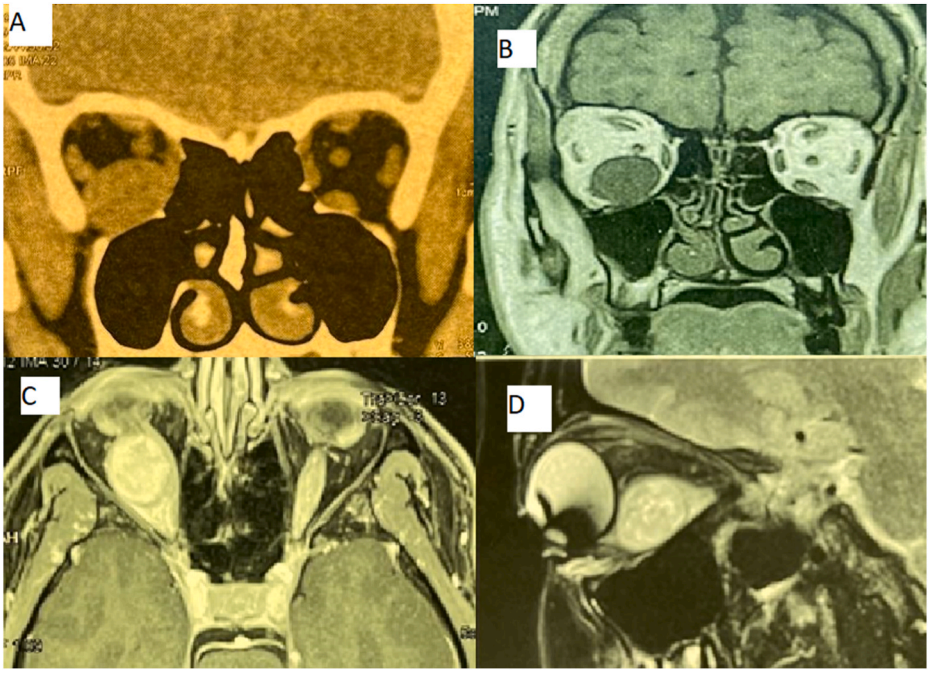

Fig. 2. Coronal orbital CT scan (A) and coronal T1 weighted orbital MRI (B) show right inferior rectus enlargement. T1 weighted orbital MRI with gadolinium and fat suppression, axial (C), and T2 weighted, sagittal (D) display a well-defined round lesion in the right inferior rectus with heterogeneous components.

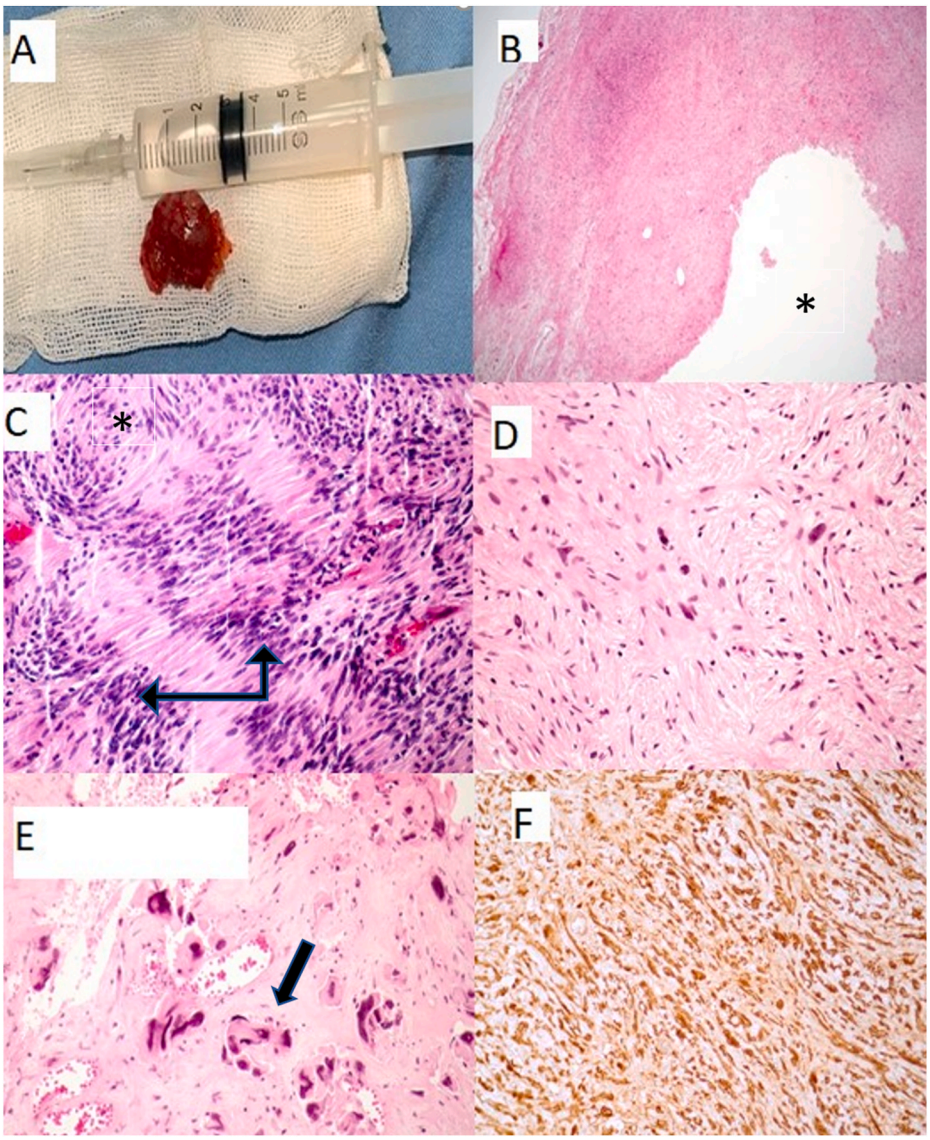

Fig. 3. Macroscopic view of the tumor at the end of the surgery, which was removed entirely (A). An encapsulated spindle cell neoplasm with central cystic degeneration (asterisk) (hematoxylin & eosin staining, X100) (B). Antoni A area with palisading nuclei (double-head arrow), and small Antoni B area at the margin of photomicrograph (asterisk) (H&E staining, X400) (C). Antoni B area; some neoplastic cells show degenerative nuclear atypia (H&E staining, X400) (D). Schwannoma which shows cellular changes and marked large pleomorphic and hyperchromatic nuclei (arrow) (H&E staining, X400) (E). Immunohistochemical examination of the tumor revealed that the tumor cells were strongly positive for S-100 protein (F).

and Antoni B areas show hyperdensity and hypodensity on CT scan imaging, respectively. Contrast enhancement on MRI demonstrates homogeneous to heterogeneous enhancement of the tumor. Variations in tumor enhancement patterns reflect the varying proportions of Antoni A and Antoni B areas in tumors.[16,17]

For typical orbital schwannomas arising from the intraconal region, precise preoperative diagnosis is not always possible, because the enhancement of the tumor is not always constant, making it difficult to differentiate it from cavernous hemangioma and meningioma.[16] There are a few reports on MRI findings of orbital schwannomas and their correlation with histologic patterns of the tumor. Wang et al. described five patterns of MRI in orbital schwannomas based on T1 and T2-weighted images and patterns of enhancement, which reflect some histologic features of the tumor.[8]

Schwannomas are well-defined encapsulated tumors with possible cystic degeneration. Previous studies have shown a correlation between histopathology and the clinical features of the tumor. Tumors with predominant Antoni B areas or cystic degeneration were more likely to have a marked cystic component.[18,19] Cellular atypia could be detected in benign lesions but there are no mitoses.

Schwannoma arising in extraocular muscle is extremely rare and most cases of orbital schwannomas are found in the extraconal or intraconal areas.[20] Only a few cases of orbital schwannomas arising from the extraocular muscle have been reported in the literature, which are listed in Table 1[9–13] Among the reported cases, there are 4 male and 2 female patients, with ages ranging from 8 to 71 years (median age 32 years). The tumor in three cases was located in the medial rectus, one in the superior oblique, and one in the inferior oblique. Therefore, our case is the first reported schwannoma in the inferior rectus muscle.

Several orbital tumors resemble schwannoma on imaging as the histology is totally different such as meningioma, cavernous hemangioma, dermoid cyst and neurofibroma.[21] Since schwannoma has a wide variety of clinical features and there is no pathognomonic clinical presentation or radiologic finding for this lesion, the diagnosis can only be confirmed by histopathologic examination.[18] Consequently, this case report revealed that schwannoma could be a differential diagnosis for

Table 1
Case Reports of orbital intramuscular schwannomas: Clinical and imaging characteristics (including the current case)
(RAPD: relative afferent pupillary defect, EOM: extraocular muscle; M: male; F: female; N/A: not assigned; IHC: immunohistochemistry; MR: medial rectus; IO: inferior rectus; SO: superior rectus; IR: inferior rectus).

Author	Clinical Presentation								Tumor size	Imaging	Involved muscle	Sex	Age
	RAPD	Diplopia	Pain	Proptosis	Motility limitation	Presenting Complaint	Visual acuity	Laterality					
Capps DH, 1990	No	No	No	Yes	No	Proptosis	20/20	OD	N/A	T1 MRI: a mass that diffusely enlarged medial rectus muscle	MR	F	8
Colapinto, 2007	No	Yes	No	No	Limitation in depression	Lower eyelid lesion and diplopia	20/20	OD	12 × 17 × 18 mm	MRI: A well-defined lesion, isointense on T1, hyperintense on T1	IO	M	68
Li J, 2015	No	No	No	Yes	Limitation in elevation	Progressive upper lid swelling	20/20	OS	8 × 18 × 32 mm	MRI: a well-defined mass in the superior orbit, with low-to-moderate signal on T1 and high T2 signal	SO	M	27
Young, 2018	Yes	No	Yes	Yes	No	Proptosis, decreased vision, lid edema, disc swelling	20/200	OS	N/A	MRI: A well-encapsulated mass in the medial aspect of the left orbit. The mass indented and displaced the globe	MR	M	71
Van Horn AN, 2021	N/A	N/A	N/A	Yes	No	Progressive proptosis	N/A	OD	N/A	T1 MRI with GAD: diffuse contrast enhancement. T2 MRI: initially isointense to muscle, followed by avid hyperintensity	MR	M	32
Afshar, 2024	No	Yes	No	Yes	Limitation in elevation and depression	Painless progressive Proptosis and diplopia	20/20	OD	5 × 15 × 25 mm	CT and MRI revealed a well-defined round lesion with heterogeneous enhancement inside the IR muscle	IR	F	32

any orbital tumor arising in extraocular muscles.

4. Patient consent

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

CRediT authorship contribution statement

Pedram Afshar: Writing – review & editing, Writing – original draft, Supervision, Methodology, Investigation, Formal analysis, Conceptualization. **Seyed Mohsen Rafizadeh:** Writing – review & editing, Writing – original draft. **Nasim Eshraghi:** Writing – review & editing, Writing – original draft, Data curation. **Sajjad Mansourian:** Writing – original draft. **Amirhossein Aghajani:** Writing – review & editing. **Fahimeh Asadi Amoli:** Investigation.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

The authors have no conflict of interest.

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