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Case Report

Orbital leiomyoma mimicking a cavernous hemangioma

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ABSTRACT

Background: Orbital leiomyoma is a rare benign tumor of the smooth muscles of the orbit with intraconal or extraconal location. The tumor affects twice as many men as women, with an average age of 30 years.

Case Description: A 38-year-old female presented with proptosis: Axile, nonpulsatile, painless, nonreducible, Grade III, with impairment of oculomotricity, associated with a decrease in visual acuity on the affected side with an AV at 03/10, and fundus: Grade I papilledema. The computed tomography scan showed a solitary, tissue mass, well-limited, encapsulated, and usually homogeneous, with homogeneous enhancement after contrast injection. The magnetic resonance imaging showed a rounded, homogeneous lesion, isointense on T1 and hyperintense on T2, with homogeneous enhancement after gadolinium injection. Suggestive of an intraorbital cavernous hemangioma. The patient underwent surgery via a subfrontal approach, with macroscopically complete resection. Histopathological examination was in favor of an orbital leiomyoma.

Conclusion: Although very rare, orbital leiomyoma should be considered in the differential diagnosis of patients with orbital tumors.

Keywords: Cavernous hemangioma, Leiomyoma, Orbital tumors

INTRODUCTION

Leiomyoma is a benign tumor arising from the smooth muscle, but its occurrence in the orbit is rare. Although these tumors are common, their unusual orbital location can make diagnosis challenging. Surgical excision is recommended to reduce exophthalmos and confirm the diagnosis. The prognosis after surgery is generally favorable.[10]

CASE REPORT

A 38-year-old female presented with right axial exophthalmos [Figure 1], nonpulsatile, painless, and nonreducible, classified as Grade III. Oculomotor function and the photomotor reflex were preserved, accompanied by decreased visual acuity in the affected eye, with a visual acuity of 3/10 and a Grade I fundus. The left eye was unremarkable.

Computed tomography (CT) scan [Figure 2] revealed a solitary, well-demarcated, encapsulated, and generally homogeneous tissue mass, with uniform contrast enhancement following contrast injection. Magnetic resonance imaging (MRI) [Figure 3] showed a well-circumscribed 15-mm round intraorbital mass, displacing the optic nerve upward and causing Grade III exophthalmos. It was T1-isointense and T2-isointense, with homogeneous contrast enhancement after

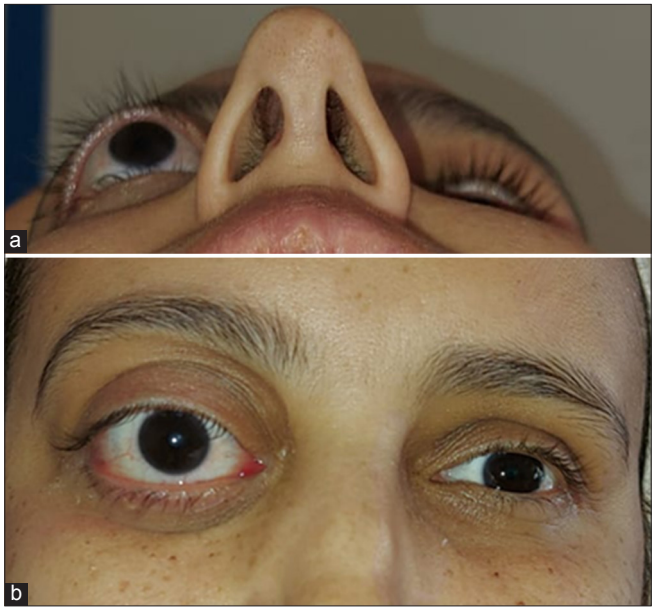

Figure 1: (a and b) Clinical photograph showing exophthalmos in the right eye.

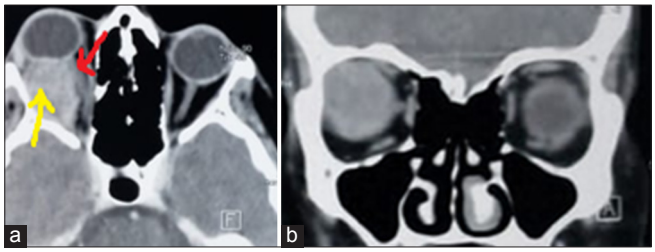

Figure 2: Intraconal leiomyoma of the orbit. (a and b) Axial and coronal images showing a well-defined intraconal mass (yellow arrow) extrinsic to the optic nerve (red arrow).

gadolinium injection, suggesting an intraorbital cavernous hemangioma. In the flair and diffusion sequences, the lesion appeared isointense.

Surgical management was chosen, and the patient underwent surgery via a subfrontal approach. During the procedure, the lesion [Figure 4] appeared as a firm, whitish, encapsulated mass, adhering to the superior aspect of the optic nerve and attached inferiorly by a fibrous band. Upon accidental perforation of the tumor, it was noted not to bleed, which is atypical for an intraorbital cavernous hemangioma. Resection was performed with a macroscopically complete excision. Histological [Figure 5] examination revealed the presence of smooth muscle cells. The findings were consistent with an orbital leiomyoma, likely originating from smooth muscle cells in the orbital muscle wall. The patient had an uneventful postoperative course, with complete resolution of the exophthalmos [Figures 6a and b], and with a satisfactory postoperative CT scan [Figure 6c].

DISCUSSION

Leiomyoma is a rare tumor in the orbit due to the limited presence of smooth muscle tissue in this region. Lodato described the first case in 1896.[8] It was initially thought to originate from the capsulopalpebral muscle of Hessar. Later, Nath and Shukla reported a case of intraconal leiomyoma and attributed its origin to the smooth muscle of blood vessels located in the orbital apex.[10,13] Among the cases reported since 1963, the majority have occurred in patients under the age of 40 years, with no observed gender predilection. Clinically, patients typically present with a progressive onset of exophthalmos and/or diplopia. Leiomyomas can be located either intra- or extraconally, with a slight preference for the former.[3,7]

MRI signal characteristics in this case were isointense in T1 and T2, with intense enhancement after gadolinium injection. The appearance was identical to that of cavernous hemangioma.[1] In this case, the lesion appears isointense on FLAIR and diffusion sequences. Due to the rarity of intraorbital leiomyomas, data concerning their characteristics on these sequences remain limited in the literature.

A tumor composed of dense smooth muscle cells and relatively sparse vascularization could display signal characteristics similar to those of uterine leiomyomas, namely a weaker signal at T2. The differential diagnosis includes several entities. Cavernous hemangioma and schwannoma should be considered. MRI can distinguish a leiomyoma from a cavernous hemangioma, the latter being T2 hyperintense. However, the differentiation between a leiomyoma and a schwannoma is more complex, as both tumors show signals of similar intensity. Nevertheless, definitive diagnosis relies primarily on histopathological examination.[2]

Moreover, various surgical approaches may be considered depending on the tumor's location and extent. Paluzzi *et al.* suggested that the orbit can be viewed as a clock, with the approach varying based on tumor position.[11]

The principle when choosing a surgical corridor is to avoid working across or around nerves. Specifically, one should avoid crossing the plane of the optic nerve. Therefore, orbital pathology lateral to the optic nerve is accessed through lateral orbitotomies, and medial pathology is accessed through medial orbitotomies.

Conventionally, external approaches to the orbit provide excellent access to tumors located superiorly and laterally to the optic nerve and orbit. These tumors are usually best approached via a pterional or fronto-orbital temporal craniotomy, with or without orbitozygomatic osteotomies.[4,11] Tumors with lateral intracranial extension can also be treated by this approach. Another variant is the lateral orbitotomy, which provides excellent access for orbital tumors lateral to the

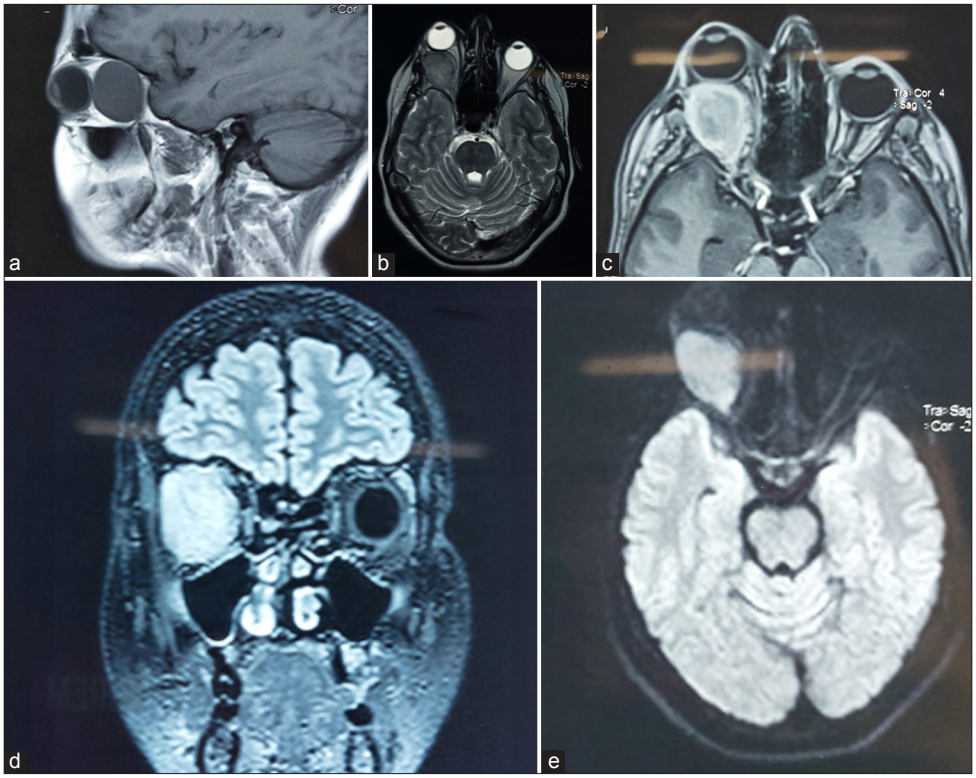

Figure 3: Magnetic resonance imaging showing a well-circumscribed right intracoronal lesion. (a) T1-weighted sagittal image and (b) T2-weighted axial image showing a well-defined isosignal area. (c) Axial T1-weighted image after gadolinium injection showing homogeneous contrast. (d) Coronal fluid-attenuated inversion recovery-weighted image and (e) axial diffusion-weighted image showing an isointense lesion.

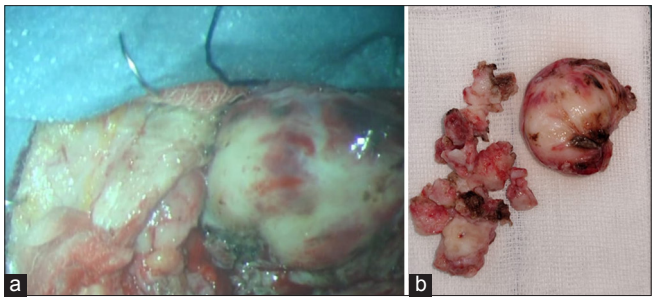

Figure 4: Intraoperative images: (a) Encapsulated tumor lesion before excision. (b) Appearance of the lesion after complete excision.

optic nerve and apex.[4,11] For tumors located anteriorly in the orbit, an anteromedial microorbitotomy or transconjunctival approach is the traditional approach for resection.

Extended endonasal approaches (EEAs) provide excellent access for intraconal and extraconal tumors that are medial and inferior to the optic nerve and can be applied to any medial intracranial extension, provided that key neurovascular structures (e.g., the optic nerve and internal carotid artery) remain lateral to the tumor. EEAs also provide access to most of the orbit, from the posterior globe to the orbital apex.[11,14] The main advantage of any EEA is its anterior and medial trajectory, which is most suitable for skull base lesions that are anteromedial to critical neurovascular structures. The key anatomic landmark is the optic nerve. Tumors that displace the optic nerve superiorly and laterally are usually excellent targets for an endonasal approach. There are three main endonasal approaches to the orbit: The medial-inferior extraconal approach, the transmaxillary extraconal approach, and the medial intraconal approach.

In our case, although a minimally invasive approach could have been considered based on the tumor's location, the initial radiological suspicion of a cavernous hemangioma with potential bleeding risk justified the choice of a subfrontal approach, offering better exposure and surgical control.

Histologically, the tumor consists of bundles of spindle-shaped smooth muscle cells interspersed with abundant endothelial sinusoids or dilated capillaries.[7] A spectrum of histological patterns exists, depending on the relative proportion of vascular and smooth muscle components. When the tumor is predominantly vascular, with smooth muscle cells scattered within the interstitial spaces, it is classified as an angiomyoma or

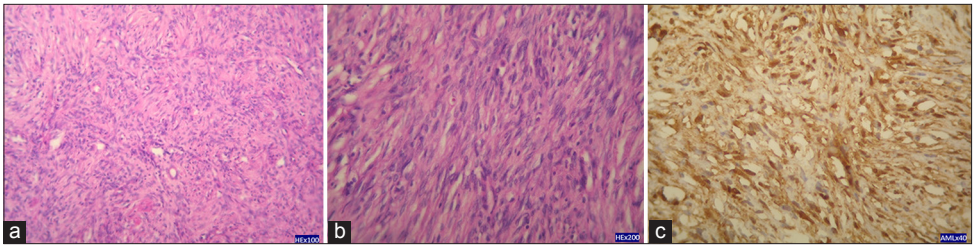

Figure 5: (a and b) Photomicrograph showing characteristic compact bundles of spindle cells with cigar-shaped nuclei. Nuclear palisading, mitotic activity, and necrosis are absent (hematoxylin–eosin, original magnification ×100). (c) Immunohistochemical analysis showing strong expression of smooth muscle actin (×40).

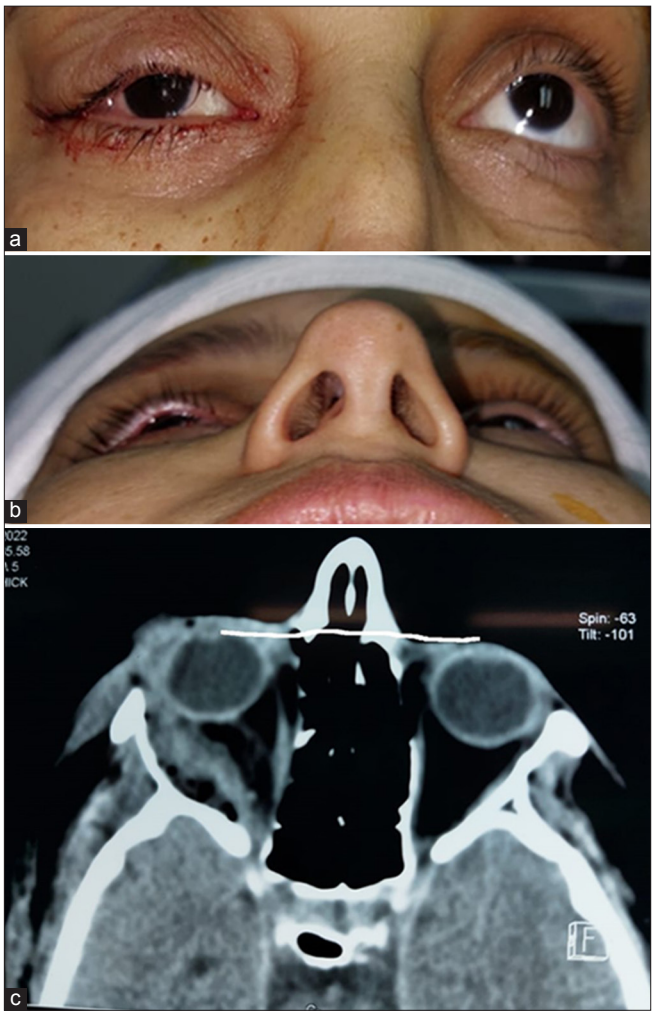

Figure 6: (a and b) Clinical photograph showing reduced exophthalmos in the right eye. (c) Postoperative computed tomography scan.

hemangioleiomyoma.[5,15] Conversely, a predominantly solid smooth muscle tumor, such as the present case, lacking apparent vascular components, resembles leiomyomas found in other anatomical sites. This variant is uncommon, as the majority of cases exhibit significant vascularization.[3] At the end of the vascular spectrum, the tumor transitions into a cavernous hemangioma. Regardless of subtype, these tumors are encapsulated within a well-defined fibrous capsule. In some instances, satellite lobulations are observed, necessitating wide excision to ensure complete removal.[6]

The definitive treatment for orbital leiomyoma is a complete surgical extraction. Leiomyoma is a rare tumor in the orbit due to the limited presence of smooth muscle tissue in this region.[6,12] Furthermore, as far as we know, orbital leiomyomas, like those found in other places, are very rare. The rarity of the condition makes our case a valuable addition to the present literature.

CONCLUSION

Leiomyoma is a well-circumscribed benign orbital mass that is usually intracanal and probably arises from the orbital smooth muscle. The MRI appearance is similar to other well-encapsulated masses, such as cavernous hemangioma and schwannoma. Although rare, leiomyoma should be considered in this differential.

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