

Orbital Angioleiomyoma: Report of a Case

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Abstract

Purpose: To report a rare case of orbital angioleiomyoma (ALM) and its management.

Methods: A 22-year-old woman presented with slowly progressive painless axial proptosis. Computed tomography and magnetic resonance imaging demonstrated a well-defined intraconal mass that was isointense in T1 and hyperintense in T2 weighted images, heterogeneously enhanced by gadolinium.

Results: The tumor, despite having significant adhesions to surrounding tissues and noticeable hemorrhage, was excised completely via the lateral orbitotomy approach. Histopathologic analysis of the specimen was consistent with an orbital ALM. The patient recovered from the operation uneventfully.

Conclusion: ALM must be considered in the differential diagnoses of orbital tumors.

Keywords: Angioleiomyoma, Axial proptosis, Orbital tumor, Smooth muscle, Vascular tumor

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INTRODUCTION

Angioleiomyoma (ALM) is an uncommon tumor that includes 5% of soft-tissue tumors, of which <10% present in the head-and-neck area, and few cases were reported in the eyelids and orbit.^{1,2}

ALM originates from smooth muscle cells that are seen in three areas in the orbital territory: i) in the structure of Muller's muscle and capsulo-palpebral muscle of the eyelids, ii) in the pulleys of the extraocular muscles, and iii) in the vessel walls as pericytes.³ Accordingly, smooth muscle tumors originate in these three sites, including eyelids,⁴ near the extraocular muscles,^{5,6} and near the orbital vessel walls in the depth of the orbit.^{4,7}

There are only few reports of leiomyoma and ALM in the orbital muscle cone.^{4,7}

Herein, the authors report a rare case of intraconal orbital ALM and its management.

CASE REPORT

A 22-year-old female was referred because of a painless progressive axial proptosis of the right eye in the past 2 years. Her medical history was negative. Visual acuity of both eyes without correction was 20/20, and there was no significant refractive error such as induced hyperopia. A 3-mm proptosis was recorded for the right eye, which did not alter by the Valsalva maneuver. Anterior and posterior segment examinations were normal [Figure 1a-c].

In computed tomography scan, an oval mass was seen in the right orbital intraconal space with extension to the extraconal space. The mass caused excavation of the lateral orbital bone without bony erosion [Figure 2]. In magnetic resonance imaging (MRI), the mass displaced the right optic nerve to the supranasal space. The mass was isointense with brain gray matter in T₁-weighted images,

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hyperintense in T₂-weighted images, and partially enhanced with gadolinium [Figure 3].

The tumor approached via lateral orbitotomy together with opening the periorbita and dissected it from the surrounding tissues through a combination of the “index finger dissection” and the “piecemeal” methods^{1,8} to avoid any damages to the optic nerve or extraocular muscles. The tumor was completely excised with a significant difficulty due to the lack of a well-defined complete capsule around the tumor. In addition, a

profound bleeding from the tumor occurred during the surgery that was controlled with head elevation, reducing systemic blood pressure.

Microscopic examinations disclosed fragments of a partially pseudo-encapsulated mass composed of different-sized vascular channels, predominantly of venous type, and intermixed with infiltration of spindle cells in the surrounding collagenous stroma. Cellular atypia, mitoses, or tissue necrosis were not evident. The spindle cells were immunoreactive for α -smooth muscle actin, and the vascular endothelial cells showed immunoreactivity for CD34 [Figure 4]. The overall histochemical and immunohistochemical features were consistent with the diagnosis of ALM.

In postoperative examinations, the patient's proptosis completely improved with no ptosis or diplopia. Her 2-year follow-up was uneventful and free of recurrence [Figure 1d-f]. Appropriate consent for publication of the report was obtained from the patient.

DISCUSSION

According to Morimoto's study,⁹ ALMs were histologically classified into three subgroups: (i) solid tumors consisting of condensed smooth muscle cells with multiple tiny vascular channels, (ii) venous tumors with prominent thick-walled vessels that do not have notable vascular spaces, and (iii) cavernous tumors that predominantly consist of vascular spaces intermixed with stromal infiltration of smooth muscle cells. In addition, based on the site of tumor occurrence, the ALMs were classified into more abundant and less abundant groups. The former is of solid type and painful, predominantly seen in extremities. The latter is of venous type and painless, and mostly seen in the head and neck area.^{3,9} The presented case in our report, according to the above classifications, was of the venous type and included in the less abundant group of the ALMs.

Angiomyofibromas, angiomyomas, and leiomyomas have been reported as benign orbital intraconal tumors,^{7,10,11} and are considered the histopathologic differential diagnoses of ALM.⁶ The histochemical and immunohistochemical features of these three tumors are quite different from the presented features in our case that was, in fact, a hybrid of leiomyoma and venous hemangioma.

Radiologic differential diagnoses of orbital ALM are other well-defined intraconal round tumors such as cavernous hemangioma, hemangiopericytoma, schwannoma, and neurofibroma, all of which are iso to hyperintense in the MRI study and enhanced with Gadolinium injection.^{1,6} However, one of the most important signs differentiating ALM from these tumors is the lack of complete capsule or pseudo-capsule in ALM. This may cause the prominent attachment of the tumor to the surrounding tissues and maybe contributed to the challenges during tumor excision such as the occurrence of intraoperative profound bleeding.

From the etiologic point of view, two theories have been proposed for the formation of this hamartomatous hybrid tumor,



Figure 1: Clinical appearance of the patient: Left column shows axial proptosis before operation from inferior (a), front (b), and superior (c) views, respectively. The right column shows improvement of proptosis after the operation from inferior (d), front (e), and superior (f) views, respectively

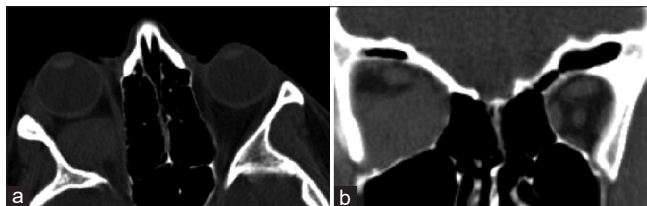


Figure 2: Axial image (a) and coronal image (b) noncontrast computed tomography show a well-defined mass lesion with apical expansion of the bony confines of the right orbit

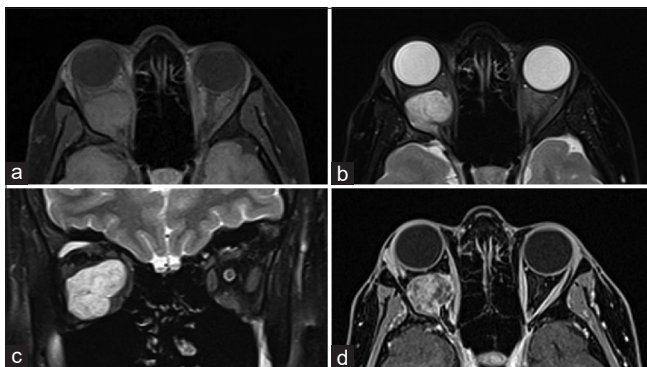


Figure 3: Magnetic resonance scans show a well-circumscribed intraconal with extraconal extension mass in the right orbit with homogeneous isointense signal on T₁-weighted axial image (a), hyperintense signal on T₂-weighted axial and coronal images (b and c), and heterogeneous enhancement with Gadolinium axial image (d)

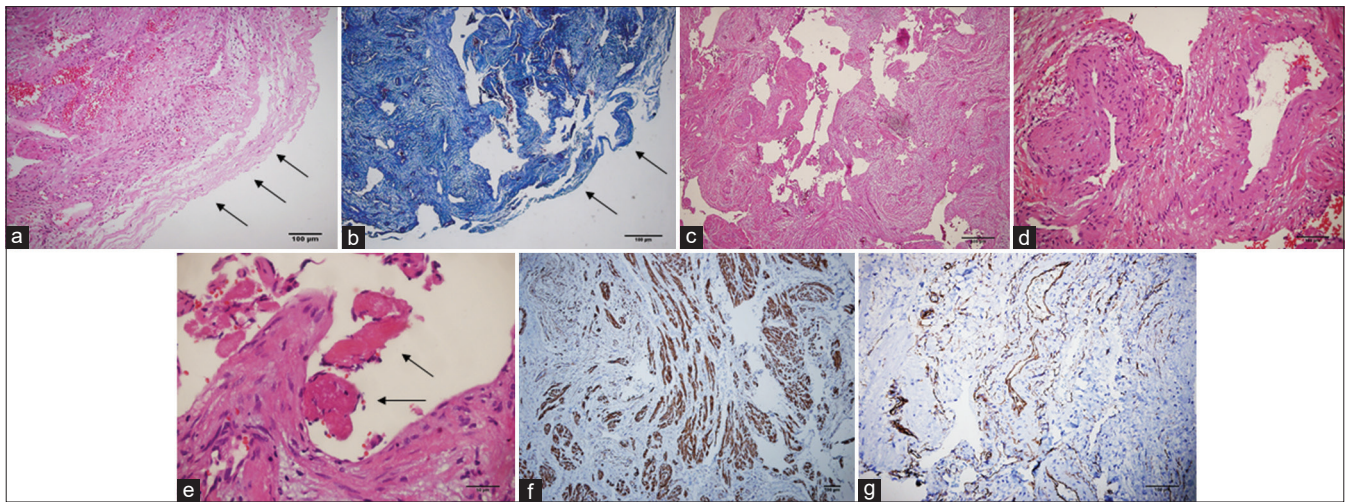


Figure 4: Histopathology of the tumor. Partial encapsulation of tumor (a: H and E), and (b: Masson's trichrome). The tumor is composed of vascular channels (c: H and E), predominantly of venous type (d: H and E) and the presence of focal outpouchings of capillary-type vessels from the sinusoidal channels (e: H and E). Clusters of spindle cells in the intervascular stroma and in the vascular walls show immunoreactivity for α -smooth muscle actin (f). Note the immunoreactivity of the vascular endothelial cells for CD34 immunostaining (g). (a, b, d: scale 100 μ m; c: scale 500 μ m; e: scale 50 μ m)

including venous stasis and changes of serum estrogen levels.³ The latter was suggested based on the more prevalence of ALMs in females than in male gender,¹ and when a nonoperable orbital leiomyoma was reported that was treated with a GnRH analog with no evidence of tumor recurrence over a 5-year follow-up.¹² Both theories cannot be excluded in our case.

Excision of cavernous hemangioma, which is the most common differential diagnosis of ALM, is very easy,⁸ while surgical excision of ALM can be quite challenging due to lack of a prominent and well-developed capsule in ALM. Insisting on complete tumor removal may cause significant hemorrhage or unfavorable damage to the optic nerve or adjacent extraocular muscles.^{1,11,12} Implementation of the gentle “index finger dissection” technique together with the “piecemeal” method, as performed in our case, can be suggested for complete excision of ALMs while avoiding intraoperative complications.^{1,8} However, given that the residual parts of the tumor have been shown that regress spontaneously, it has also been suggested to leave the apical part of the tumor behind when complete excision is not possible.^{11,12}

In conclusion, orbital ALMs are rare primary orbital vascular tumors. Because of the significant adhesions of the tumor to the adjacent tissues, gentle excision of the tumor via a lateral orbitotomy is recommended for proper management of the intraoperative bleeding and avoiding inadvertent complications.

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 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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