

Solid variant of orbital angioleiomyoma: An unusual tumor at an unusual site

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We describe the clinicopathological features of a solid variant of orbital angioleiomyoma. A review of clinical records, diagnostic, and radiographic studies combined with histopathological evaluation with standard histochemical staining and

immunohistochemistry was conducted. A 22-year-old male patient presented with a mass in the region of the left lacrimal gland that was gradually increasing over the past 2 years. Radiological and clinical examinations showed no signs suspicious of a malignancy and fine needle aspiration cytology was inconclusive. Therefore, an excision biopsy was performed. On histopathological examination, the picture was consistent with a benign spindle cell tumor. Immunohistochemistry showed positivity for CD 34 and CD 31 (markers for vascular endothelium). The tumor also showed positivity for smooth muscle actin and Ki-67 proliferative index was low. Angioleiomyomas are rarely encountered in the orbit and has features seen in leiomyoma as well as some vascular tumor elements. In most cases, surgical excision is usually curative.

Key words: Angioleiomyoma, hemangioma, lacrimal gland, orbit, proptosis, tumor

Angioleiomyoma is a benign subset of leiomyoma, a commonly encountered smooth muscle tumor. However, only 8.5% of angioleiomyomas are seen in the head and neck region, of which

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only a handful of reports have documented its occurrence in the periorbital region.^[1,2] Angioleiomyomas are characterized by endothelium-lined vascular channels and background of smooth muscle.^[3] Wolter, in 1965, and Henderson and Harrison, in 1970, have previously described orbital angioleiomyomas.^[4,5] More recently, Korn *et al.* have reported a case of angioleiomyoma of the lacrimal sac.^[3] Taxonomically, Morimoto, in 1973 classified angioleiomyoma into three separate histologic types: solid, venous, and cavernous.^[6] Here, we present the clinical, radiological, and histopathological features of a solid variant of orbital angioleiomyoma in a young Indian male and review the available literature. A systematic search was performed on PubMed using suitable keywords. Non-English language articles that had abstracts translated into English were also reviewed.

Case Report

A 22-year-old-male patient presented with a painless swelling over the left eye along with mechanical ptosis of the upper lid. Proptosis of 2 mm was also noted. The patient had observed a gradual increase in size over 2 years [Fig. 1a]. No diplopia or pain was reported. On examination, the mass was located anterior to the orbital rim in the superolateral part of the orbit; with the posterior edge not palpable as it was within the orbit. The mass was firm to hard in consistency with a smooth surface and nontender. There was no globe displacement, motility restriction, or diplopia. The patient gave no history suggestive of a waxing and waning course. Anterior segment evaluation, intraocular pressure, and fundus examination of both eyes were normal. A computed tomography showed an iso-to-hypodense mass lesion in the superotemporal aspect of the left orbit, arising possibly from the lacrimal gland since the gland could not be clearly delineated from the mass. No obvious change to the bony orbit could be noted on the scans [Fig. 1b]. Fine needle aspiration cytology smears drawn showed no malignant cells; however, a conclusive diagnosis could not be made. A working diagnosis of a pleomorphic adenoma of the lacrimal gland was made, and an excision biopsy was performed. Intraoperatively, the mass was pink, vascular, and well circumscribed but not encapsulated. The lacrimal gland was seen in close relation to the mass, compressed between the mass and the superolateral orbital bony rim. The mass was excised completely and processed for histopathological examination. It measured 20 mm × 20 mm × 7 mm.

The cut surface was homogeneous and did not have any distended vascular channels, lacrimal tissue, or cystic dilatations. On microscopic examination, spindle cells in compact sheaves closely opposed to thin walled vessels were seen in a fibrocollagenous background [Fig. 2a]. The nuclei were open, oval, and contained small, uniform nucleoli [Fig. 2b]. No necrosis was noted, and a few engorged vessels were seen in the outlying area. No lymphoid aggregates or germinal centers were identifiable. Immunohistochemical studies showed that the tumor cells stained positive for CD 34 and CD 31, which are markers for endothelium [Fig. 3a and b]. Furthermore, the tumor stained positive for smooth muscle actin and negative for CD68 [Fig. 4a and b]. HMB-45 immunostaining was negative, and the Ki-67 proliferation index was low with <5% of the cells staining positive [Fig. 4c]. Ki-67 is a cellular marker for proliferation. Thus, the tumor had no conclusive features of

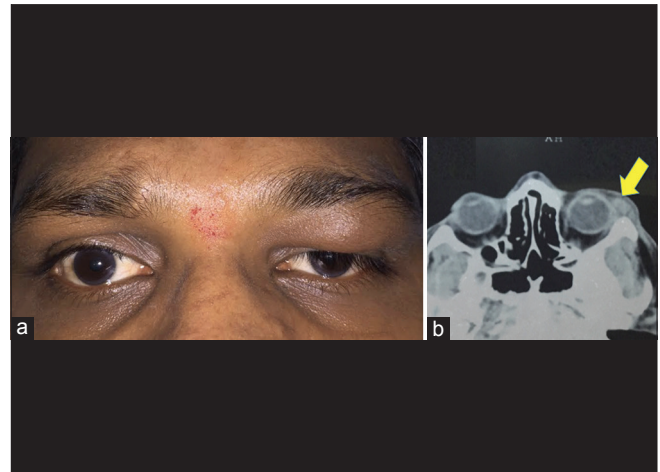


Figure 1: (a) External clinical photograph shows the mass located in the superolateral quadrant of the orbit. (b) Axial slice of the computed tomography scan showing the mass in the vicinity of the lacrimal gland (yellow arrow)

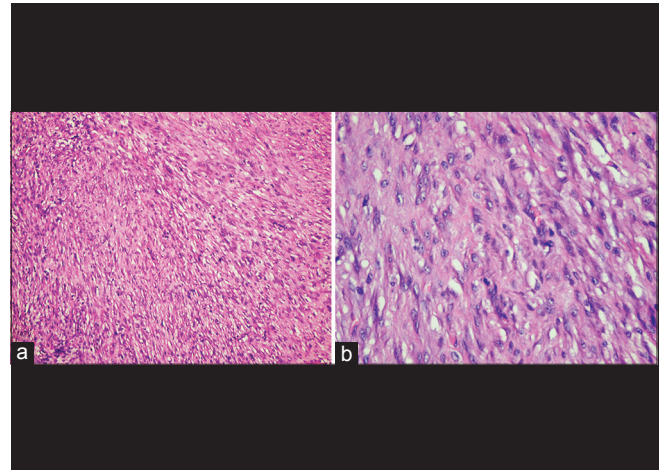


Figure 2: (a) Low power view showing spindle cells in a background of fibrocollagenous tissue (H and E, ×10). (b) High power view demonstrating uniform oval nuclei (H and E, ×40)

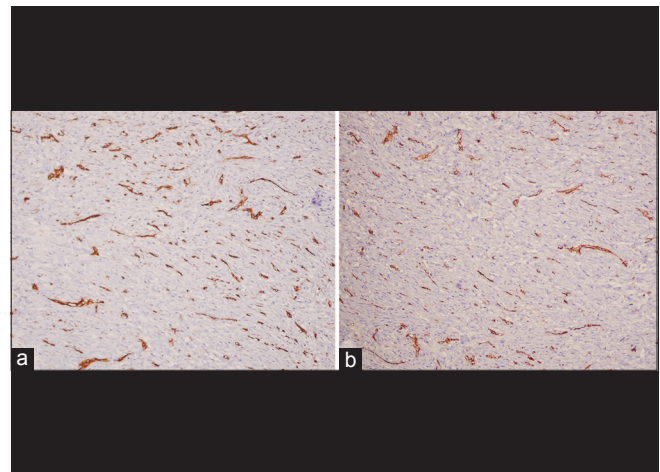


Figure 3: (a and b) Immunohistochemical studies showed that the tumor cells stained positive for CD 34 (endothelium) and CD 31 (×10)

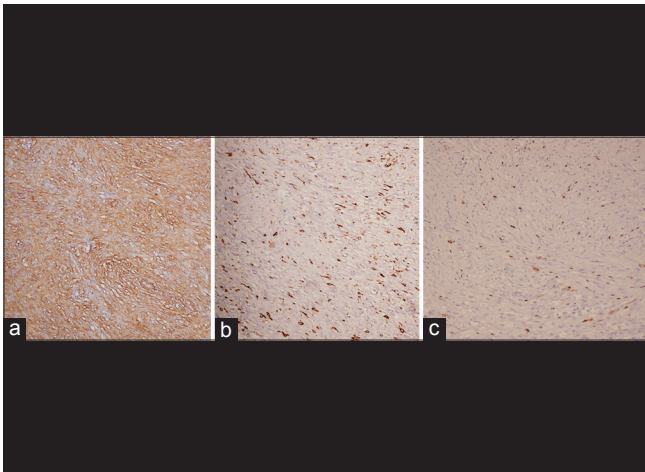


Figure 4: (a and b) Immunohistochemical studies positivity for smooth muscle actin and immunonegativity for CD68 ($\times 10$). (c) The Ki-67 proliferation index was low with $<5\%$ of the cells staining positive ($\times 10$)

a malignancy and the final diagnosis based on its microscopic appearance, and immunohistochemical characteristics was that of a solid variant of angioleiomyoma of the orbit. As it had been excised completely, no further treatment was warranted. At 1-year follow-up, no recurrence was observed.

Discussion

The differentials for such an orbital tumor could be cavernous hemangioma, angiomyofibroma, or complex orbital angiomyoma.^[7-9] The tissue of origin of angioleiomyoma is considered to be smooth muscle; so the possible native tissues from which the tumor could have arisen include blood vessels, pericytes, Müller's muscle or the capsulopalpebral muscle of Hesser.^[3] Angioleiomyoma is well recognized within the spectrum of vascular lesions of the soft tissues and has been subdivided into three types – solid, venous, and cavernous.^[7,10] Since there were no dilated cavernous spaces within our tumor, we consider our case to be a solid variant. Immunohistochemistry helps to differentiate from similar orbital tumors, namely angiomyoma and angiomyofibroma.^[6-8]

Morimoto, who classified angioleiomyomas, observed that solid variants are often painful and seen in the extremities. In contrast to his comments, our case was a solid variant which was painless.^[6,10] Jakobiec *et al.* described an angiomyofibroma of the orbit as a hybrid tumor exhibiting characteristics of a vascular leiomyoma and cavernous hemangioma.^[7] The lack of the cavernous venous channels, the unusual location, and absence of the classical purplish hue rule out cavernous hemangioma and angiofibroma in our case. Jakobiec has also described in detail the characteristics of a complex orbital angiomyoma, which had features of a lymphangiohemangioma.^[9] Our case lacked any lymphoid tissue with no overlapping features of a lymphangioma.

Lin *et al.* have described a series that included six orbital angioleiomyomas.^[11] Three of them were located in the muscle cone, two of them were located in the superotemporal orbit,

and one was located in the inferior orbit. Two other eyelid angioleiomyomas were also described in the same series. Of these eight cases, six of the cases had a complete fibrous capsule. Based on their histological classification, however, only one case was the same type as our case: the solid variant; five cases were of the cavernous type and two cases were venous type. Alam *et al.*, have also reported a similar tumor from the anterior orbit, which was found to be of the cavernous subtype.^[12]

Preoperative magnetic resonance imaging may provide some clues: Magnetic resonance findings of peripheral angioleiomyomas were relatively nonspecific, but T2-weighted images show a mass with mixed areas that are both hyper- and iso-intense relative to the skeletal muscle and a hypointense rim.^[11-13] Angioleiomyomas of the orbit are rare tumors with good prognosis, and the treatment of choice remains complete surgical excision.^[3]

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Conflicts of interest

There are no conflicts of interest.

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