Unusual intraconal localization of orbital giant cell angiofibroma

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Giant cell angiofibroma (GCA) is a recently reported rare soft-tissue tumor that can develop in various sites including orbit. Orbital GCAs were mainly located in the eyelid or extraconal regions such as lacrimal gland and conjunctiva. We report an atypical case of a GCA arising in the intraconal area of the orbit in a 65-year-old male patient. The tumor was excised in total by lateral orbitotomy. Histological and immunohistochemical features were consistent with the diagnosis of GCA. No recurrence was observed during the follow-up of over 2 years. GCA is a rare tumor that should be considered in the differential diagnosis of intraconal orbital tumors. Complete surgical removal is the current optimal treatment option.

Key words: Giant cell angiofibroma, intraconal, orbit

Giant cell angiofibroma (GCA) is a rare solitary lesion which was originally described in the orbit 20 years ago, recently classified as a variant of solitary fibrous tumor (SFT).[1] It typically presents as a slowly growing anterior orbital mass and has a tendency to behave in a benign fashion. Since the initial report of the disease, GCA has been observed in many extraconal regions of the orbit;[2-11] yet, occurrence in the intraconal region is extremely rare. In literature, there were only two cases of GCA in which intraconal involvement was reported.[3,4]

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/ijo.IJO_755_16

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Manuscript received: 28.09.16; Revision accepted: 26.09.17

In this article, we aimed to present the clinical and histopathologic features of GCA occurring in the retrobulbar intraconal area.

Case Report

A 65-year-old male presented to our department with a 3-month history of slowly growing prominence of the right eye. On physical examination, the right eye showed painless proptosis without diplopia. Ocular motility of the right eye was mildly limited both in lateral and downward directions. Hertel exophthalmometer values of the right eye and left eye were 23 mm and 13 mm, respectively. Visual acuity measured with Snellen chart was 0.4 for the right eye and 0.8 for the left eye. Anterior segment examination revealed a clear cornea with grade 1 nuclear sclerosis in both eyes. Fundus examination showed choroidal folds in the right eye.

Orbital magnetic resonance imaging revealed a well-defined mass measuring 32 mm × 31 mm × 25 mm in intraconal region of the right eye, which was isointense relative to the adjacent soft tissue in T1 signal and hyperintense in T2 signal [Fig. 1a and b]. There was no sign of bone erosion or infiltration to adjacent soft tissues. Lateral orbitotomy through upper lid skin crease incision was performed to gain access to the posterior orbit. The dissection extended through orbicularis muscle and deep fascia to the periosteum of the orbital rim. Periosteum along the lateral orbital rim was cut and elevated from the lateral orbital wall. The bone was cut with an oscillating saw, angling the cut slightly inferiorly and parallel to the orbital roof. The cut was made into the thin bone along the sphenozygomatic suture line. A second cut was made through the orbital rim just above the zygomatic arch. Small holes were drilled on either side of each cut near the rim to facilitate later replacement of the bone. The bony rim fractured outward. The periorbita was then opened. A well-circumscribed, encapsulated, and highly vascular lesion was seen. To improve exposure,

Cite this article as: Ekin MA, Ugurlu SK, Cakalagaoglu F. Unusual intraconal localization of orbital giant cell angiofibroma. Indian J Ophthalmol 2018;66:160-3.

inferior subciliary approach for inferior orbitotomy was performed following canthotomy and cantholysis. Then, the tumor was freed up entirely with blunt dissection and removed in total [Fig. 1c]. Macroscopically, the tumor was ovoid, highly vascular, white-gray with smooth borders [Fig. 1d]. Microscopic examination revealed richly vascular, well-circumscribed, patternless spindle-shaped cells arranged in sheets. Floret-like multinucleated giant cells often lined the pseudovascular spaces [Fig. 1e]. Immunohistochemistry was performed; spindle cells as well as the giant cells showed reactivity for Vimentin, CD34, and bcl-2 [Fig. 1f]. Neural crest marker S100, desmin, KI67 and ALK-1 were all negative. The morphology and immunohistochemical profile was consistent with the diagnosis of GCA. Immediate postoperative period was uneventful [Fig. 2a and b]. Proptosis regressed completely with recovery of full extraocular motility. No recurrences were detected over 2-year follow-up with good cosmetic and functional results.

Discussion

Since its first description by Dei Tos *et al*.[2] in 1995, GCA is now well established as part of the hemangiopericytoma and SFT family, characterized by proliferation of CD34-positive fibroblast and myofibroblast.[12] Until now, there are various cases describing involvement of extraorbital sites including axillary-inguinal regions, posterior mediastinum, head and neck, hip, vulva, oral cavity, and nasolacrimal canal.[1] In this report, we present a rare case of GCA in a male patient that was unusually located within the intraconal orbit.

Few cases of orbital GCA have been reported in the literature [Table 1].[2,11] Most of these tumors have involved either anterior orbital space or the eyelid and conjunctiva.[2,11] Involvement of intraconal orbit has been reported only in two previous cases.[3,4] Keyserling *et al*.[3] presented a case with intraconal GCA which abutted and medially displaced optic nerve. They could not resect the mass totally due to profuse bleeding from the tumor during orbitotomy. In the other case, Hayashi *et al*.[4] reported a man with GCA which was located within the muscle cone near the lateral rectus and inferior oblique muscles. However, Hayashi *et al*.[4] did not give information regarding whether total resection of the mass was achieved or not. In our patient, we successfully removed the tumor without adjacent tissue damage and functional loss. Our case constituted the tumor with the largest GCA size that has been excised completely among those located in the intraconal region.

In general, orbital GCA cases presented with similar clinical characteristics of a benign, noninfiltrative, slow-growing tumor. However, they may simulate an aggressive or malignant process and grow rapidly. GCA is mostly seen in men and the age range was from 16 to 78 years. The size of the tumor in our patient (3.2 cm × 3.1 cm × 2.5 cm) was larger than previous cases which were ranged between 1.2 and 3 cm. Although GCA is an isolated lesion, it has been reported as an eyelid mass in a patient with tuberous sclerosis and a cutaneous lesion in the thigh region of a patient with dermatofibrosarcoma protuberans.[6] In cases with orbital GCA, eyelid swelling and proptosis are the most common symptoms. Other symptoms associated with GCA depend on the site of the tumor and are secondary to local mass effect such as visual disturbances and pain.[1,2] The diagnosis of GCA may be virtually impossible during preoperative management because it shares many radiological and clinical features with other soft-tissue tumors of the orbit. On computed tomography, GCA appears as a well-demarcated, homogeneous mass with intense contrast enhancement and no bone erosion. On magnetic resonance imaging, circumscribed soft-tissue mass and stippled signal intensity voids with intense contrast enhancement likely correspond to the pseudovascular spaces in a vascular lesion.[4-10] The presence of pseudovascular spaces can lead to this lesion

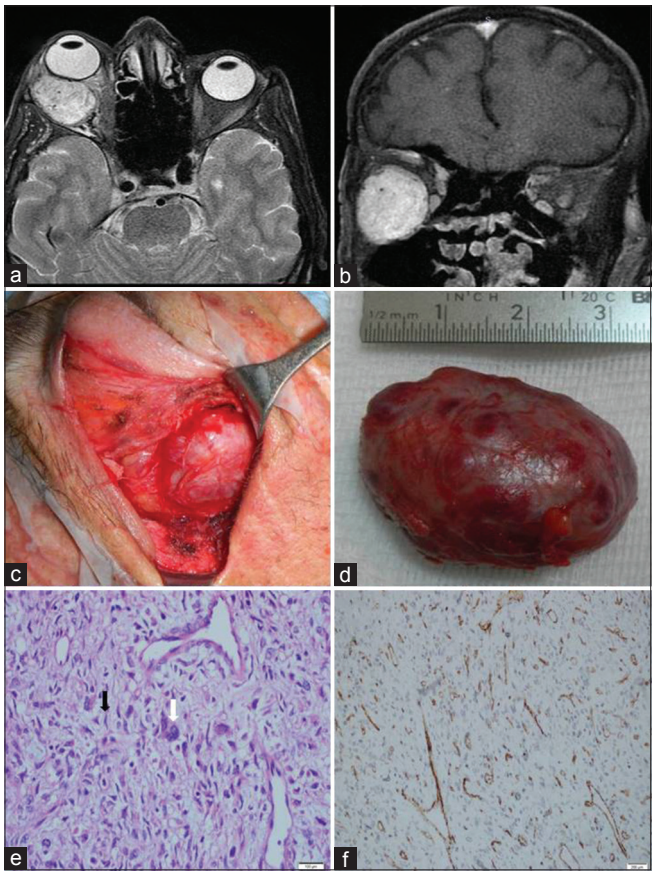

Figure 1: (a) Axial and (b) coronal MR scan demonstrating a well-circumscribed mass of relatively homogeneous consistency in intraconal region of the right eye. (c) Intraoperative identification of giant cell angiofibroma. (d) Postoperative appearance of the mass after total excision. (e) Patternless giant cell (white arrow) and spindle cell (black arrow) proliferation (Hematoxylin-eosin, ×100). (f) Diffuse CD34 positivity in tumor cells (Diaminobenzidine, ×100)

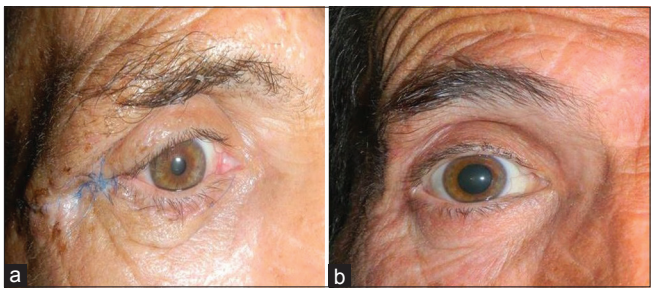

Figure 2: (a and b) Postoperative images show complete healing

Table 1: Orbital giant cell angiofibroma cases reported in the literature

Authors	Age (year)	Sex	Location	Duration (month)	Symptom	Size (cm)	Management	Recurrence
Ganesan *et al*.[9]	57	Male	Lower lid	18	Painless mass	1.2	Total excision	No
Hayashi *et al*.[4]	46	Male	Lower lid	11	Painless mass	2	Total excision	No
	78	Female	Upper lid	NA	Painless mass and ptosis	2	Biopsy	NA
	65	Male	Intraconal	NA	Painless mass	NA	Total excision	NA
	47	Male	Upper lid	8	Painless mass	1.5	Total excision	NA
	65	Male	Inferior conjunctiva	NA	Painless mass	NA	Total excision	NA
Keyserling *et al*.[3]	60	Male	Extraconal and intraconal	60	Proptosis	3	Partial resection and RT	NA
Song *et al*.[6]	24	Male	Inferior conjunctiva	12	Painless mass	1.3×1.4×0.6	Total excision	No
Farmer *et al*.[8]	57	Female	Lower lid	16	Painless mass	1.5	Total excision	No
Ereño *et al*.[11]	73	Male	Orbit	NA	Rapidly growing mass	1.5	Total excision	Yes
DeSousa *et al*.[10]	68	Male	Orbit	60	Painless mass	NA	Total excision	No
Zoumalan *et al*.[5]	21	Female	Orbit	12	Painless mass and epiphora	1.5×1.0×0.8	Total excision	Yes
Demirci *et al*.[7]	16	Female	Extraconal	18	Painless mass, ptosis, and proptosis	NA	Total excision	No

NA: Not applicable, RT: Radiotherapy

being misdiagnosed as hemangioma, lymphangioma, and metastases.[3]

On histopathology, GCA is a highly vascularized tumor with patternless spindle cell proliferation containing pseudovascular spaces and floret-like multinucleate giant cells. GCA shares morphologic features with SFT. Guillou *et al*.[1] pointed out in a retrospective review that 10 of a total 100 extrapleural SFT had at least some focal GCA-like areas, containing multinucleated giant cells, round-to-spindle cells, and small, hyalinized vessels. Zoumalan *et al*.[5] reported a case that initially presented with an orbital GCA, but on recurrence, the tumor was found to be SFT. This supports the hypothesis that GCA is a variant of SFT. Both neoplasms stain similarly immunohistochemically. Tumor cells are characteristically reactive for CD34, CD99, Vimentin, and less frequently bcl-2 but negative for CD31, CD117, S100, muscle-specific actin, and desmin.[5,8] Furusato *et al*.[12] reviewed and analyzed 41 fibroblastic orbital tumors which originally were diagnosed as hemangiopericytomas (16/41), fibrous histiocytomas (9/41), mixed hemangiopericytoma/fibrous histiocytoma (14/41), and GCA (2/41) of orbit. After histologic and immunohistochemical (CD34, CD99, bcl2, Ki67, p53) review, all cases were reclassified as SFT. The results of this study suggested that these entities share overlapping morphologic and immunohistochemical features and should be designated as SFTs.

There have been limited reports regarding long-term behavior of GCA, but complete surgical excision was considered the optimal treatment. Although GCA is typically regarded as a benign tumor, local recurrence, and persistent tumor have been described previously.[2,8] Keyserling *et al*.[3] reported a case with both extraconal and intraconal recurrence after a 9-month follow-up due to incomplete resection. Dei Tos *et al*.,[2] observed local recurrence in one of 6 patients during 60 months after local excision. Ereño *et al*.[11] reported a case of GCA in a 73-year-old man. The tumor was located in the upper-inner quadrant of the right orbit and recurred 16 months after surgical removal.[11] In the postoperative period, our case has been free of recurrence for 22 months.

Conclusion

Our study demonstrated that GCA should be included in the differential diagnosis of intraconal orbital tumors. Furthermore, surgical removal and subsequent histopathological and immunohistochemical analysis of the tumor are usually warranted for the final diagnosis.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

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