Primary chondromyxoid fibroma of the orbit: An orbital mass with calcification

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Primary orbital chondromyxoid fibroma is a rarely reported entity. A 34-year-old lady presented with painless, non-axial proptosis of the left eye of 6 months duration. Orbital imaging showed a supero-temporal mass with calcific foci and bone erosion. The mass caused globe compression resulting in choroidal folds. Anterior orbitotomy with complete mass excision was performed. The histopathology revealed a chondromyxoid fibroma. At 12-months follow-up, the patient is doing fine with no clinical recurrence. Chondromyxoid fibroma is an important differential diagnosis for bony orbital tumors.

Key words: Chondromyxoid fibroma, orbital calcification, orbitotomy, primary orbital tumor

Primary orbital bone tumors constitute approximately 2% of all orbital masses.[1] Amongst these, the chondromyxoid fibromas (CMFs) arising from orbital bones are even rarer. The reported overall incidence of CMFs is <1% of all bone tumors, which commonly arise from the long bones of the limbs.[2,3] Secondary orbital involvement due to the extension of craniofacial CMFs has been infrequently reported in the literature.[4,5]

To the best of our knowledge, the primary orbital CMFs have been reported only thrice in the English literature (summarized in Table 1).[6-8] In 2009, the first case of primary orbital CMF was reported by Heindel *et al*.[6] We describe the clinical features, radiology, histopathology, and outcomes in the first report of primary orbital CMF from our country.

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Case Report

A 34-year-old lady had a 6-month history of painless, progressive, protrusion of the left eye. It was associated with diminution of vision for the last 3 months. On examination, the best-corrected visual acuity of the right and the left eye was 20/20 and 20/60, respectively. The left eyeball showed infero-medial displacement, 4 mm of proptosis, and limitation of elevation [Fig. 1a and b]. The fundus evaluation revealed choroidal folds at the posterior pole. A non-tender, firm-hard mass was palpable in supero-temporal quadrant of left orbit with raised retro-bulbar resistance.

A computed tomography (CT) scan showed a well-defined mass in supero-temporal orbit, showing irregular hyperdense foci with surrounding frontal bone erosion [Fig. 1c and d]. The mass was not separately visualized from the lacrimal gland. After obtaining informed consent, a transcutaneous anterior orbitotomy with mass excision was performed via a sub-brow Benedict's incision. Intraoperatively, a defect measuring 10mm × 12mm was identified in the supero-temporal orbital roof which was filled with bone wax.

The gross specimen measured 3 × 2.5 × 2.5 centimeters [Fig. 2a and b]. The histopathology showed the presence of a spindle/stellate type of cells in a chondroid matrix and hyaline cartilage with focal areas of ossification and few bony trabeculae [Fig. 2c and d]. The final diagnosis of chondromyxoid fibroma was established.

Her postoperative course was uneventful with complete resolution of proptosis, significant improvement in visual acuity and restoration of elevation, although the choroidal folds persisted for 4 months. At 12-months follow-up, no clinical features of local recurrence were noted [Fig. 3a and b].

Discussion

Our case is a primary orbital CMF arising from the frontal bone with no extraorbital extension. CMFs are rare tumors presenting in 2nd to 3rd decade with a male preponderance.[2,3] The differential diagnosis for orbital bone tumors includes both benign (osteomas, fibrous dysplasia, chondroma, osteoblastoma and giant cell tumor) and malignant lesions (osteosarcoma, chondrosarcoma and Ewing's sarcoma).[2-5]

Any expansile and locally destructive lesion in the craniofacial skeleton must be evaluated and CMF should be thought of as a differential.[2-8] The CT scan features include an osteolytic lesion, well-defined lobulated margins with cortical erosion and septation (in long bones).[9] Foci of calcification

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may be noted in 13% cases.[2,3] Calcification is more common in skull-base CMFs and the younger population.[3,9,10] On MRI, the lesion is isointense (T1W) to muscle and homogenously hyperintense (T2W) with characteristic contrast enhancement.[9,10]

Histopathology provides the confirmative diagnosis and helps in planning further management. The histopathological differentials include both benign and malignant soft tissue, fibrous and fibro-histiocytic tumors like enchondromas, chondroblastoma, chondrosarcoma, chordoma, and myxofibrosarcoma [Table 2].[2-8] CMF is benign cartilage-forming tumors consisting of a collagenous to a myxoid matrix with the characteristic presence of stellate cells. On immunohistochemistry (IHC), they stain positive for smooth muscle antigen (SMA) and negative for keratin AE1/AE3.[2,3] Although Mullen *et al*. reported that a tailored IHC might not be very helpful for diagnosing CMF.[8]

Enchondromas show a lobulated architecture but the presence of hyaline cartilage in these tumors is a good differentiating feature. Both CMF and chondrosarcoma stain positive for S-100 and vimentin, but the absence of fibrous stroma in orbital chondrosarcomas differentiate it from CMFs.[2,3] Chordomas have infiltrative margins and are composed of nests/cords of large epithelial cells with eosinophilic or vacuolated cytoplasm, which may secondarily invade the orbital bone. On IHC, the chordomas express keratin and epithelial membrane antigen.[1-3] In myxofibrosarcoma, a multi-nodular proliferation of stellate fibroblasts within a myxoid stroma with curvilinear blood vessels is noted. On IHC, the myxofibrosarcoma stains positive for vimentin.[1-3]

Given the overlap of features, incisional biopsies or curettages may yield inadequate tissue leading to a possible erroneous diagnosis of malignant lesions like chondrosarcoma. This may change the further treatment course in the form of undue radiation and an unnecessary orbital exenteration. Moreover, an incomplete removal of a CMF can lead to a local recurrence. The reported recurrence rates range from 28% to 33% in the craniofacial sites.[2,6] Hence complete excision is recommended for both better histopathological identification and to reduce the risk of recurrence.

Conclusion

In conclusion, primary orbital chondromyxoid fibroma is an extremely rare orbital bony tumor having a few typical

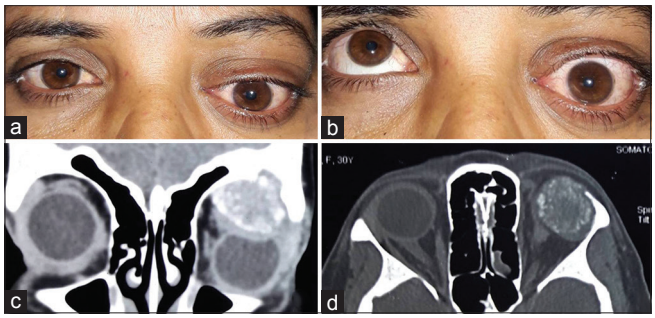

Figure 1: (a) Abaxial proptosis of the left eye with infero-medial displacement, (b) Limitation of left elevation, (c) CT scan (coronal sections) showing mildly enhancing, well-circumscribed mass in supero-temporal orbit with hyperdense foci and erosion of the adjacent frontal bone, (d) The bony window of CT scan (axial, superior sections) shows the same lesion with prominent hyperdense specs of calcification

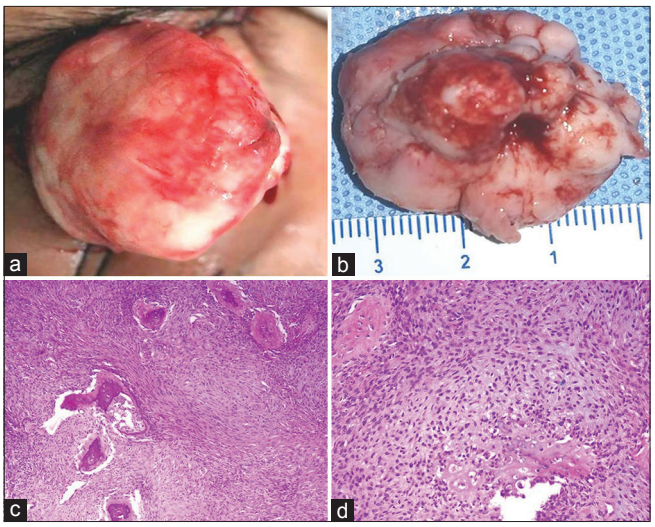

Figure 2: (a) Intraoperative picture showing expression of large mass with the smooth orbital surface, (b) The orbital bone side of the same mass shows irregular, bosselated surface, (c) (H and E, 40×) The section shows a lobular configuration with myxoid background and interspersed cartilage, (d) Higher magnification (H and E, 200×) shows spindle/stellate cells with few cartilaginous areas undergoing ossification

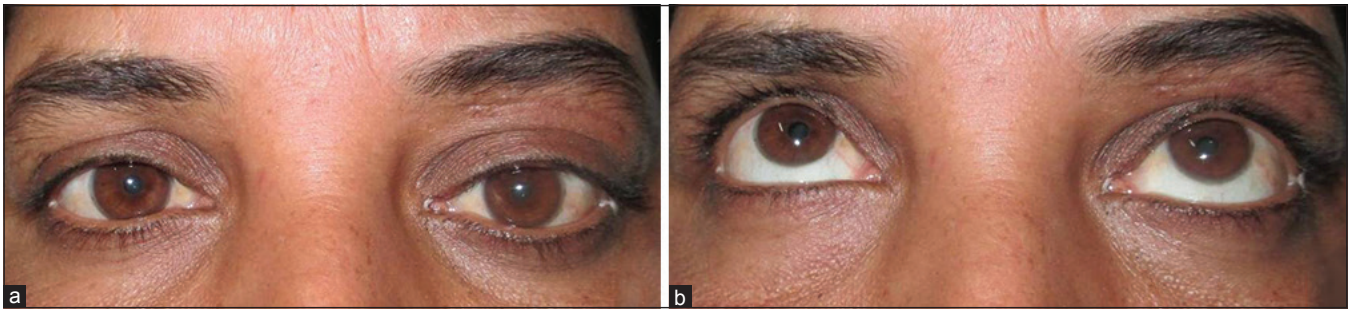

Figure 3: (a) At 12 months follow-up, the left globe appears to be in normal position with a residual sub-brow scar, (b) The left elevation has also been restored to near normal

Table 1: Primary orbital chondromyxoid fibromas: a review of literature

Author/ year	Age/ gender	Duration	Chief complaints	Evaluation	Radiology	Management	Follow-up/ outcome
Heindl LM/2009	37y/F	3 years	Left supero-temporal orbital mass, proptosis	Visual acuity: 20/20 OU, non-axial proptosis=4 mm, choroidal folds +	CT orbits: non-infiltrative mass with erosion of adjacent frontal bone	Transcutaneous extra-periosteal orbitotomy with complete mass excision	2 years/no recurrence (clinical)
Ditta LC/2012	51y/F	Incidental	History of migraine; right orbital wall mass as an incidental finding on imaging	Visual acuity: 20/20 OD & 20/30 OS; proptosis=3 mm; increased resistance to retropulsion	MRI: circumscribed lobulated intraosseous mass at the zygomatico-sphenoid junction	Surgical excision of extraosseous component	Recurrence at 5 months, re-surgery with the reconstruction of lateral orbit
Mullen MG/2017	56y/M	Incidental	History of chronic sinusitis; Left orbital mass detected on imaging; Previous incisional biopsy-myxofibrosarcoma and advised an exenteration	Visual acuity: 20/20 OU, no proptosis	CT: 3cm supero-temporal osteolytic lesion, before incisional biopsy MRI: multi-lobulated intensely enhancing expansile mass, 8 mm hypointense foci suggestive of the biopsy site	Combined neurosurgical and ophthalmologist approach for complete excision and histopathological confirmation	Follow up not mentioned
Our Case	34y/F	5 months	Left non-axial proptosis of 5 months duration	Visual acuity: OD 20/20 & OS 20/60, choroidal folds +,	CT: superotemporal mass lesion with dense foci of calcification and effacement of orbital roof	Transcutaneous anterior orbitotomy with mass excision, bony defect sealed with bone wax	No recurrence at 8 months follow up

Table 2: Differentiating clinical, radiological, histopathological and immunohistochemical features of similar orbital lesions

	Chondromyxoid Fibroma	Chondroma	Osteoma	Mesenchymal chondrosarcoma
Clinical features	Benign tumor 2nd to 3rd decade Male preponderance	Benign cartilaginous tumor No gender/age predilection Arise from trochlear/ sphenoid bone	Benign skeletal neoplasm 2nd decade Males > females Arise from paranasal sinuses	Recurrent tumor±distant metastasis 2nd to 3rd decade Female preponderance
Radiology (CT scan)	well-defined mass, lobulated osteolytic lesion with bony cortical erosion & foci of calcification	well-defined mass, minimally enhancing lesion with bony scalloping and erosion	well-circumscribed mass, dense cortical sclerosis around a radiolucent nidus	well defined mass, heterogeneous enhancement, bony expansion without destruction with areas of mottled and fine calcification
Histopathology	Gross- smooth to bosselated surface Microscopy- cartilage-forming benign tumour with collagenous to myxoid matrix and characteristic stellate cells	Grossly- glistening smooth surface Microscopy- multi-lobulated lesion with benign spindle cells resembling hyaline cartilage, no cellular atypia	Grossly- knob like protuberances over a glistening white-pink lesion Microscopy- central nidus of loose fibrovascular tissue surrounded by irregular trabeculae of bone and osteoid matrix	Grossly- lobulated surface, soft to firm consistency Microscopy- sheets/ clusters of spindle- shaped mesenchymal cells; areas of cartilaginous tissue with atypia, focal calcification/ cartilaginous changes
Immunohisto-chemistry	S-100, SMA and SOX 9	S-100 and vimentin	S-100, EMA, NSE+ve	S-100 (cartilaginous area) Vimentin & CD99 (cellular component)

*CT: Computed tomography; SMA: Smooth muscle antigen; EMA: Epithelial membrane antigen; NSE: Neuron specific enolase

radiological characters, and total excision with proper histopathology should be the aim for complete management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has given her consent for her 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.

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Conflicts of interest

There are no conflicts of interest.

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