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Orbital Wall Ossifying Fibroma: Case Report

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Abstract **Case Report**

The case concerns 28-year-old patient, presented with gradual painless prominence of the right eye noted for 02 years. The ophthalmological examination of the right eye found a decreased visual acuity, non-axial proptosis, limited ocular motility examination and stage 1 papilledema on fundoscopy. CT scan showed an enhanced extra-conal mass of the right orbit with compression of the right optic nerve was noted. An excisional biopsy was done and histopathological examination confirmed a ossifying fibroma. ***Discussion:*** Ossifying fibroma is a benign fibro-osseous lesion rarly located in orbital cavity. Clinical presentation and complications vary according to its location. Imaging, based in CT scan and MRI, shows clearly demarcated, round or ovoid lesions with osteoblastic and lytic areas in the central matrix, and concentrically expanding solitary mass with a thin sclerotic margin. ***Conclusion:*** Ossifying fibroma is a rare bony neoplasm of orbit which needs multidisciplinary approach to be differentiated from other orbital tumors. Radiological and histopathological correlation are mandatory for an appropriate diagnosis to ensure complete excision to prevent recurrences.

Keywords: Ossifying fibroma, orbit, Computed Tomography.

INTRODUCTION

The orbit is the cavity containing structures essential for ocular function and the bony architecture that encases them. Orbital ossifying fibroma is an extremely rare condition, especially in adults. Its a benign fibro-osseous neoplasm. It has a more aggressive course in children compared to adults. Gradual proptosis is the most common clinical finding. CT scan is the modality of choice for bony exploration and histopathological correlation is mandatory to diagnose bony orbital neoplasms.

CASE REPORT

28-year-old patient, presented with gradual painless prominence of the right eye noted for 02 years. The ophthalmological examination of the right eye found a decreased visual acuity 6/10, not improved after optical correction and non-axial proptosis. Ocular motility examination showed limited elevation. Stage 1 papilledema is found on fundoscopy. However there was no palpable mass or postural variation of proptosis. The rest of the ocular examination, was within normal limits. CT scan showed an enhanced extra-conal mass (2,5x4x6 cm) in the superior aspect of the right orbit, with osteoblastic and osteolytic areas in the central matrix, surrounded by a thin sclerotic margin. Compression of the right optic nerve was noted (Figures 1, 2).

Patient underwent an excisional biopsy using an orbito-cranial approaches. Histopathological examination showed fibroblast rich stroma with bony trabeculae. Osteoblastic rimming without mitotic activity suggested benign fibro-osseous neoplasm. Postoperative computed tomography (CT) scan showed no residual lesion (Figure 3).

DISCUSSION

Primary tumors of orbital bone constitute 0.6% to 2% of all orbital tumors [1,2]. Selva and al, over a 24-year period, found 62 (1.9%) cases from a total of 3,340 orbital tumors, and only 1 patient (1.6%) had ossifying fibroma. IT may be classified on a clinicopathologic basis into benign fibro-osseous or cartilaginous, reactive, neoplastic and vascular disorders [3].

Clinical findings include painless progressive proptosis, ptosis, diplopia and ocular motility restrictions, rarely optic nerve compression and inflammatory signs are present [4].

Diagnostic criteria for conventional ossifying fibroma include: usually solitary, well demarcated radiographically with smooth, sclerotic borders. Centrifugal growth pattern with increasing maturity of bone as one moves to the periphery. "Shells-out" from surrounding bone intact/large pieces. Relatively avascular cellular brous stroma. Retiform bone trabeculae, with some osteoblastic rimming and/or cementum-like spherules [5].

Computed tomographic (CT) shows a round or ovoid well-defined margins lesion, is, with expansion of the involved orbital bone. The central matrix shows a mixed pattern of osteoblastic and osteolytic areas in the central matrix, surrounded by a thin sclerotic margin [2]. OF can produce a lytic deformity, simulating mucocele or epidermoid cyst [6]. Other radiographic features of OF include low signal intensity on T1-weighted and high signal intensity on T2-weighted magnetic resonance imaging MRI and high Technetium-99 radioisotope uptake due to its high osteoblastic activity [7].

Complete excision is the surgical goal. For small anterior tumors, this may be achieved via an anterior orbitotomy. Large tumors (5 cm diameter) require orbito-cranial or orbito-rhinological approaches [3]. Enucleation, or extensive resection, may be required where aggressive features are apparent [8]. Radiation is generally not preferred in children due to the risk of malignancy. Systemic therapy with interferon alpha has been reported to reduce the recurrences. Recurrence rates are reported from 30% to 56%, due to the infiltrative nature of the lesion [9].

CONCLUSION

Ossifying fibroma is a rare bony neoplasm of orbit which needs multidisciplinary approach to be differentiated from other orbital tumors. Radiological and histopathological correlation are mandatory for an appropriate diagnosis to ensure complete excision to prevent recurrences.

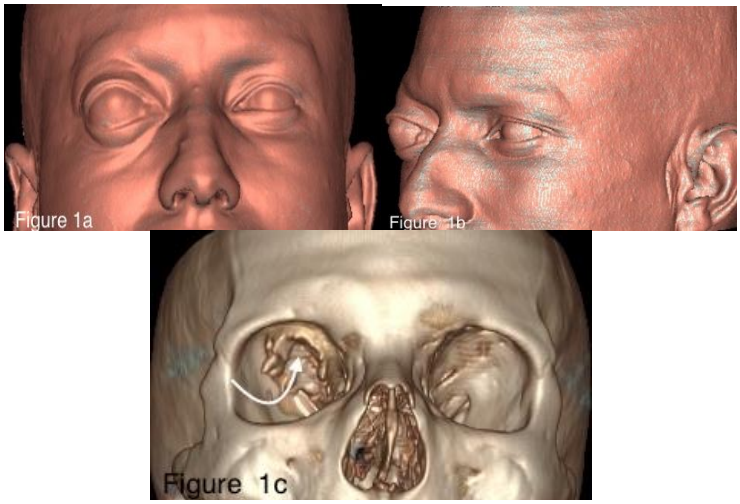

Fig-1 (a,b,c): Multiplanar reconstruction which shows exophthalmos from the front (a) and from the side (b). (c) 3D bone reconstruction showing bone anomalies in the orbital surface of the right frontal bone (curved white arrow)

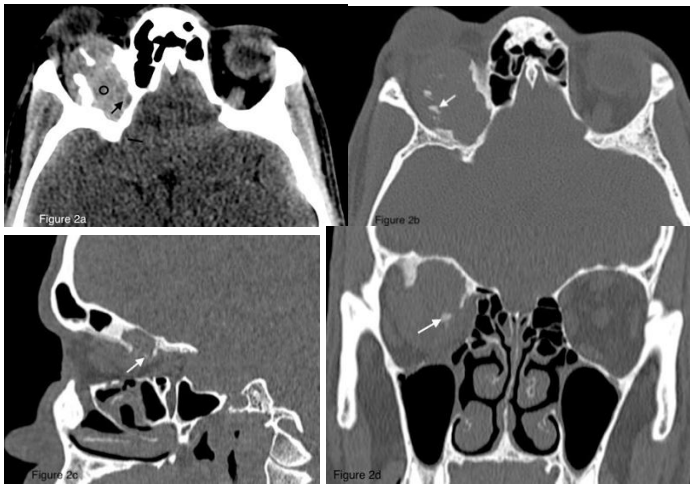

Fig-2 (a, b) Axial CT: images of the right orbit, displayed in a brain window (a) and a bone window (b), (c) sagittal and (d) coronal reconstruction in a bone window: showing an ovoid expansile mass with internal septa (black arrow), fluid- fluid levels (black cercle). There is multiple foci of calcification (white arrows) seen within the mass

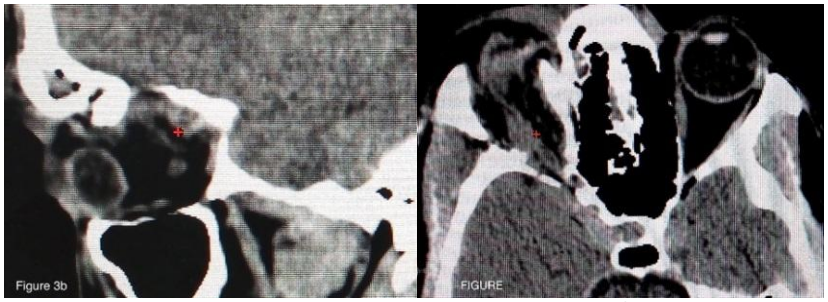

Fig-3: (a) axial and (b) sagital post operative CT scan showing surgical modifications wthout residual lesion

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