Author Communication

A Curious Orbital Lesion in a Young Girl

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A 15-year old girl presented with a slow growing mass in the orbit with normal visual acuity. We followed the patient for a year and closely observed the increase in size of the mass for over a year both clinically and radiologically. The mass grew gradually with no effect on her visual acuity, which was 6/6 in both eyes using Snellen chart; pupillary reactions were normal with no afferent papillary defect and her extraocular movements were full. Her anterior segment and posterior segment of the eye including the optic disc and macula and the intraocular pressure remained within normal limits. We removed the mass surgically on the request of the patient, as the mass had caused significant disfigurement. It was a multidisciplinary approach by an ENT and an eye surgeon for the effective and complete removal of the lesion. We did a frontal orbitotomy under general anesthesia and the mass recovered was sent for biopsy. The biopsy report concluded the mass to be juvenile psammomatoid ossifying fibroma.

Key Words: Orbit, psammomatoid ossifying fibroma, frontal orbitotomy.

Extraconal orbital tumors in children exhibit both a diagnostic and radiological challenge to the ophthalmologists. The Orbital Tumors or lesions in children can present as hematological malformations such as hemangiomas and lymphangiomas. Orbital varix however are found in an older age group and tend to present with intermittent proptosis; size of the lesion increasing on valsalva maneuver. Other primary tumors include fibrous dysplasia, rhabdomyosarcoma, and optic nerve Glioma. Rhabdomyosarcoma is the most common mesenchymal neoplasm of orbit in children.[1] Metastatic tumors include Neuroblastoma, Wilm's and Ewing's sarcoma. Inflammatory lesions include Pseudotumor and myositis. Dermoid cyst may also present both in a pediatric as well as the adult population. A differential diagnosis of a rapidly growing proptosis may also be infectious conditions such as Orbital cellulitis. Clinical presentation combined with the characteristic imaging features of the disease can narrow the differential diagnoses. Imaging modalities most often used to examine these masses include B scan ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) especially for lesions involving the optic nerve and orbital apex[2].

CASE STUDY

A 15-year old girl presented with painless proptosis of right eye for the past one year (Figure 1). She denied any history of increase in proptosis on bending, any history of trauma, any visual obscurations or any associated headache, nausea or vomiting. She did not have fever and had no systemic symptoms either such as loss of appetite, weight loss, lethargy, bone pain etc. On examination, her right eye was proposed with a measurement of 24 mm by Hertel exophthalmometer while left eye was 21 mm. Anterior segment was normal with no corneal edema, no chemosis or no dilated and tortuous vessels on the conjunctiva. The pupillary reactions were normal with no relative afferent pupillary defect. Posterior segment was normal in both eyes with normal disc and macula and no choroidal folds or disc/macular edema. Her extra ocular movements were normal and she had intra ocular pressure of 15 mm Hg in both eyes. On palpation, no mass was palpable and no thrill was

noted. The preauricular and submandibular lymph nodes were not enlarged and were normal on palpation.

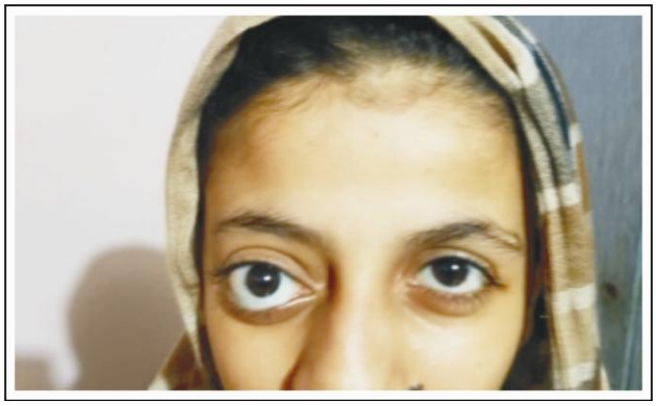

Fig. 1: Right eye proptosis.

CT scan imaging revealed a lobulated peripherally enhancing mass involving the right orbital roof causing significant bony expansion. It measured 3.8 x 3.2 × 4.9 cm. It had caused effacement of posterior ethmoidal cells with intra orbital extraconal extension causing compression and displacement of right superior and medial recti resulting in proptosis of globe (Figure 2 and Figure 3).

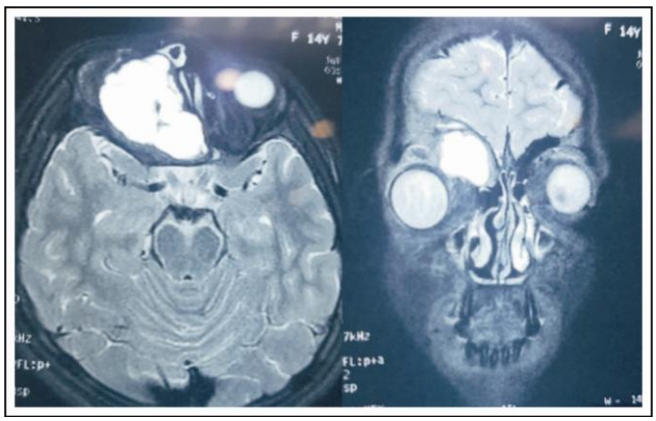

Fig. 2 & 3: Axial and Coronal CT scans of patient with contrast showing an enhancing lesion causing downward globe displacement and occupying the frontal sinus and almost all of the orbital region.

A combined orbital and ENT surgery was performed on the patient under general anesthesia. An

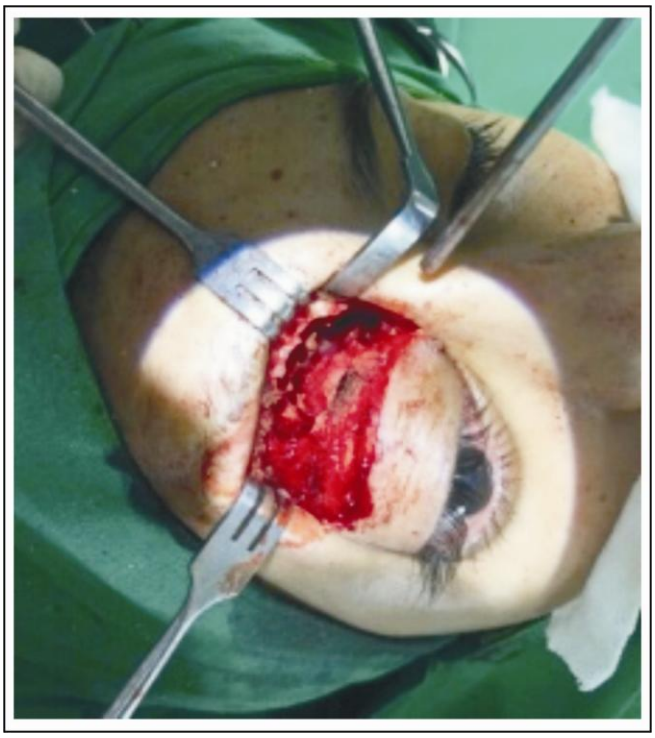

Fig. 4: Frontal Orbitotomy with incision made just beneath the eyebrow along its whole length.

incision was made just beneath the eyebrow along its length to gain access to the superior orbit and the frontoethmoidal region. Soft tissue was dissected until the periosteum was reached which was divided with No. 11 scalpel blade. The periosteum was separated from the underlying bone with a freer periosteum elevator. Frontal osteotomy was done using a round fluted burr (Figure 4). This allowed an entry to the supra orbital space and the mass was identified and removed piece meal from the orbital, frontal and ethmoidal region and was found to be pink fleshy and firm in consistency (Figure 5). The mass was sent for biopsy. The wound was closed in 3 layers: the periosteum and muscles with Vicryl 6/0 and the skin with prolene 5/0 interrupted sutures. A redivac drain was also placed in the wound and was removed after 24 hours as the fluid collected was 30 ml only. Patient was given oral broad spectrum antibiotics and steroids and an antibacterial ointment for topical use for 10 days. Skin sutures were removed after 8 days.

Histological analysis revealed the mass to be composed of fibroblastic cells arranged in sheets with prominent psammomatous bodies, favouring juvenile psammomatous ossifying fibroma.

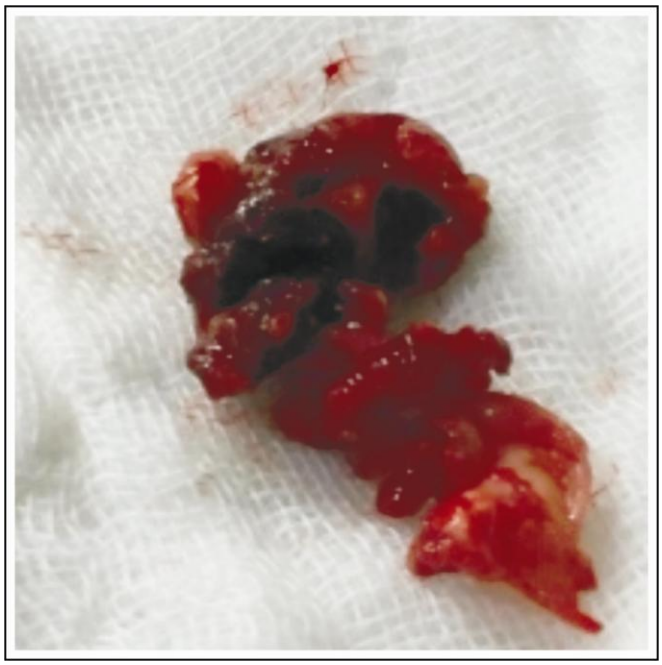

Fig. 5: Fleshy mass excised from the orbit piece meal and sent for histological analysis.

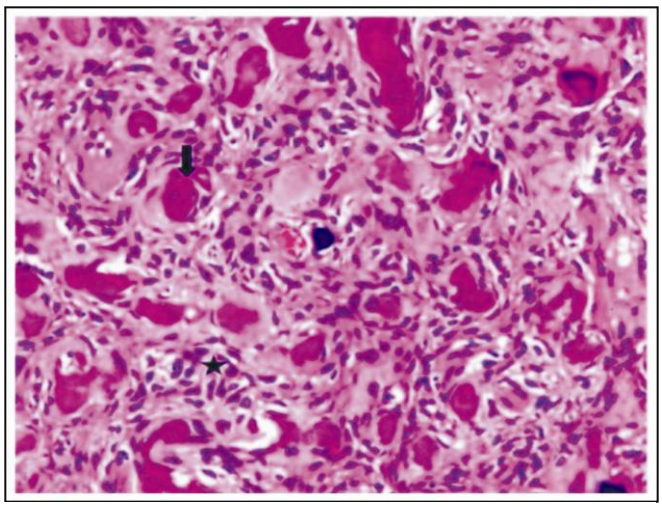

Fig. 6: Photomicrograph: Showing numerous spindle-shaped cells arranged in fascicular storiform pattern (asterix) with irregular strands of trabeculae with plump osteoblast, spheroidal ossicles with basophilic center and eosinophilic periphery resembling psammoma-like bodies (arrow). H & E × 40 × 7.

DISCUSSION

Ossifying fibroma (OF) is a benign tumor of bone that has the tendency for aggressive growth, bony destruction, and potential for regrowth[3]. Juvenile ossifying fibroma (JOF) is an atypical lesion that can be differentiated from other types of ossifying fibromas on the basis of age of presentation, common location and their aggressive growth[3]. It may exhibit vigorous growth and has a problem of recurrence[4].

Mofty explained that there are two different histological types, trabecular Juvenile ossifying fibroma and Psammomatous Ossifying fibroma, based on their histological appearance and their tendency to occur in a particular age. They are commonly found during the age of 8.5-12 years and 16-33 years. The former representing the trabecular juvenile ossifying fibroma and the latter representing the psammomatous subtype[5]. The word "psammoma" is from a greek word "psammos" which means "sand"[6]. Psammoma-like bodies are seen to have a dark border of lucent particles from which small slender sharp pointed bodies and needle-like crystalloids emanating toward the outer edge[7].

Treatment of Juvenille Psammomatous Ossifying Fibroma consists of complete surgical excision of the lesion as an inadequate excision may pose a problem with the mass reccurring locally[8,9,10].

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CONFLICT OF INTEREST

None

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Author's Contribution

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Treating physician. Preparing of manuscript. Literature search. Final review.