Pleomorphic adenoma of the lacrimal gland in a teenager

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Abstract
Lacrimal gland pleomorphic adenoma, accounts for more than half of the epithelial forms of lacrimal gland tumours. The commonest age group affected is the fifth decade. We report this tumour in a teenaged girl which is an unusual age group to present with this tumour. To conclude lacrimal gland lesions of slow growing nature with well-defined border and those which have an intact capsule observed per operatively should be excised intact to prevent malignant transformation and recurrence. The typical location and radiological findings of pleomorphic adenoma renders itself to easy detection and such presentation even in a young patient should not mislead the treating surgeon to plan an incision biopsy.

Keywords: Pleomorphic adenoma, Young patient.

Introduction
Tumours of the lacrimal gland are rare, constituting 7-9% of all orbital tumors.[1,2] Among all of them, the most common epithelial tumor is the lacrimal gland pleomorphic adenoma, accounting for more than half of the epithelial forms.[3] 0.6% of all orbital cases of tumours[4] and 12% of all lesions of the lacrimal gland.[5] We report this tumour in a teenaged girl which is an unusual age group to present with this tumour.

Case report
An 13 year old girl, presented to our hospital with chief complaint of protrusion of right eye since 1 year. This was gradual, painless and not associated with any other ocular or systemic complaint. Her past, family and personal history were unremarkable. On examination her general and systemic examination were within normal limits. Her ocular examination revealed a right eye proptosis of 6 mm (hertels-26mm right eye and 20 mm left eye) with 4mm down and 2mm in dystopia. (Fig. 1) Her uncorrected visual acuity was 6/6 in both eyes. Anterior and posterior segment were within normal limits. Her proptosis evaluation demonstrated a positive resistance to retropulsion, limited superior gaze of minus three in straight, levo and dextro elevation. There was no change in proptosis with valsalva manoeuvre or posture. There was diplopia in up gaze. Colour vision was normal in both eyes. Differential applanation tonometry was not significant. There were no exposure signs on ocular surface. Lids were normal. There was no pain, pulsation or change of periocular skin.

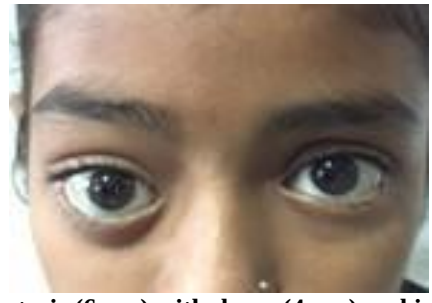

Fig. 1: Right eye proptosis (6mm) with down (4mm) and in (2mm) dystopia

A CT scan orbit, complete blood count and peripheral blood smears were ordered. Her CT revealed an extra conal well defined ovoid supero lateral orbital mass extending from anterior to mid orbit with homogeneous hyper dense texture and moderate contrast enhancement. (Fig. 2 a and b) Keeping the differentials of a peripheral nerve tumour, dacryoadenitis and vascular malformation in mind, an anterior orbitotomy with mass excision or incision biopsy was planned. The mass was approached by a lid crease incision and deepened to expose the superolateral anterior orbital extaraconal compartment. A reddish well defined oval mass was noted in the area of lacrimal gland. (Fig. 3) The mass was excised completely and incision closed in 2 layers. The mas was ovoid in shape with measuring approximately 25mm x 20mm. (Fig. 4) The Histopatholgy report confirmed the mass as pleomorphic adenoma. (Fig. 5) Her

postoperative period was uneventful. Proptosis and gaze restriction were resolved completely. (Fig. 6)

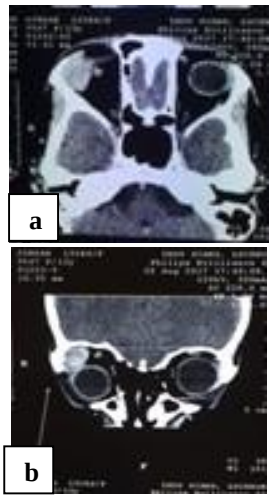

Fig. 2: (a) Axial CT scan showing hyperdense well-defined extraconal mass in lacrimal gland fossa (b) Coronal sections showing the extent of the mass till mid orbit

Fig. 3: Reddish well encapsulated mass after dissection from surrounding structures

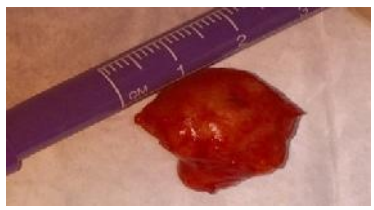

Fig. 4: Excised mass measuring 25mm x 20 mm with surface boss elations

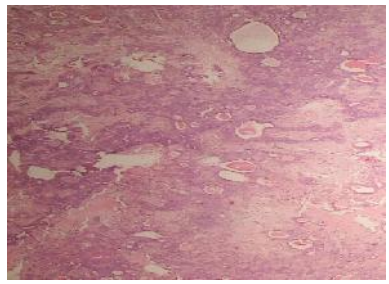

Fig. 5: Histopathology photo showing abundant stroma consisting of fibro-collagenous tissue and several small cystic structures filled with keratinized material

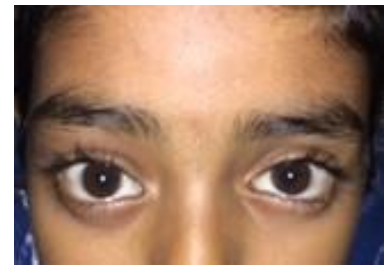

Fig. 6: Post-operative two weeks follow up photos showing reversal of proptosis and dystopia

Discussion

The term pleomorphic (benign mixed tumor) was first defined and used by Willis.[6] Pleomorphic adenoma is the most common epithelial tumour of lacrimal gland. The age group of presentation is usually fifth decade. It is rare in children. To reduce the risk of recurrence and malignant transformation, pleomorphic adenomas of the lacrimal gland should be removed intact, without prior biopsy.[7] There have been scattered case reports of occurrence of pleomorphic adenoma in young adults and children.[8-10] Malignant transformation can occur in 10–20% of pleomorphic adenomas of the lacrimal gland.[11]

Conclusion

Pleomorphic adenoma of the lacrimal gland must be among the top differentials to be considered in the presence of long-term painless exophthalmous with down and inward globe displacement. For the patients suspected to have pleomorphic adenoma, incisional biopsy should not be performed in order to prevent the likelihood of a relapse. The excision of the mass with its capsule can extend the patients' lives and improve their quality of life. Although the commonest age group is fourth to fifth decade, there have been reports of children as young as 5 year old diagnosed with this tumour. To conclude lacrimal gland lesions of slow growing nature with well-defined border and those which have an intact capsule observed per operatively should be excised intact to prevent malignant transfomation and recurrence. The typical location and radiological findings of pleomorphic adenoma renders itself to easy detection and such presentation even in a young patient should not mislead the treating surgeon to plan an incision biopsy.

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