



A Case Report on Unilateral Non-axial Proptosis of a Young Female: Lacrimal Gland Tumour

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ABSTRACT

Background: Lacrimal gland adenoma is a benign tumour of the lacrimal gland mostly involving the orbital part of the gland and composed of epithelial and myoepithelial components. It occurs in the third and fourth decade of life as a gradual progressive enlargement of the lacrimal gland.

Case: This is a case report of a 30-year-old female presenting with the forward bulging of the right eye causing the eye to be displaced inferiomedially over the course of one year. After careful clinical examinations and MRI, the clinical diagnosis of lacrimal gland adenoma was established and was planned for right lateral orbitotomy (without marginotomy) via eyelid crease incision and transcutaneous-transseptal approach with complete excision of the tumour under general anaesthesia.

Observations: This case was presented to the out patient department with mild painful non-axial proptosis of the right eye. After clinical examination, her MRI report showed a well-defined altered signal intensity enhancing lesion on the extraconal compartment of the right orbit in the antero-supero-lateral aspect. After complete surgical removal of the mass, it was sent for histopathological analysis and it confirmed it as a pleomorphic adenoma. The patient has been following up every 6 months for 2 years and is asymptomatic.

Conclusion: Being the most prevalent lacrimal gland tumour, pleomorphic adenoma affects the unilateral lacrimal gland causing non-axial proptosis. Complete removal of mass has an excellent prognosis with complete resolution of symptoms.

Keywords: Histopathology; lacrimal gland tumour; lateral orbitotomy; pleomorphic adenoma; proptosis.

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INTRODUCTION

The lacrimal gland, an almond-shaped, bi-lobed, eccrine secretory gland, is roughly two centimetres long and situated in the lacrimal fossa in each orbit. It secretes a watery physiologic fluid that lubricates, nourishes the eye, and contains the enzyme lysozyme for bactericidal purposes (Harrison et al., 2018).

Pleomorphic adenoma is a type of benign tumour of the lacrimal gland, which is composed of epithelium and myoepithelial components. Pleomorphic adenoma in 80-90% involves the orbital lobe and rarely involves the palpebral lobe. (Ahmad SS, Anwar R, Khan MA, Usmani N, 2021) These neoplasms typically appear in the third and fourth decades of life as a gradual and painless enlargement of the lateral area of the upper eyelid, with no obvious gender

predilection. (Harrison et al., 2018); Young et al., 2018) After complete diagnosis and imaging, the treatment procedure is complete surgical excision of the tumour.

CASE REPORT

A 30-year-old female presented to our OPD with a chief complaint of forward bulging of the right eye, which gradually progressed over the course of one year. The bulging appeared to be greater on the temporal side, causing the eye to be displaced slightly downward and medially. The patient experienced mild pain and slight restriction of motility only on dextro-elevation. There were no other associated symptoms such as double vision, watering, redness, discharge, or diminution of vision. The bulging did not vary with posture or with strain.



Figure 1: First day of presentation at OPD, showing non-axial proptosis of right eye (displaced downward and medially), Bird's eye view(B) showing proptosis of right eye and Worm eye view(C) showing proptosis of RE.

She did not give a history of trauma, medication, or any surgery in the past. She also did not give a history of chronic diseases. Proptosis of the right eye was visible on bird's eye and worm's eye view.

On clinical examination, the anteriorly palpable mass over the right superolateral orbit was non-mobile, elongated with a regular margin, and firm in consistency with mild tenderness which extended from the lateral orbital rim towards two-thirds of the superior orbital rim. The underlying attachment was non-discernible. There was no compressibility, thrill, discolouration, periorbital change, or pulsation. The regional lymph nodes were not palpable.

The anterior displacement of the right eye was 4 mm when compared to the left eye. The inferior displacement was 3 mm and the medial displacement was 2 mm in comparison to the left eye by using two ruler method.

The slit lamp examination revealed an increased size and downward bulging of the palpebral part of the lacrimal gland during levodepression.

The rest of the ocular examinations were unremarkable. Her best corrected visual acuity was also normal.

With the provisional diagnosis of a right lacrimal gland tumour, most likely a pleomorphic adenoma, a magnetic resonance imaging (MRI) of the orbits and brain was obtained and lab investigations were sent. The lab investigation reports were all normal.

Her MRI showed a well-defined altered signal intensity enhancing lesion on the extraconal compartment of right orbit in the antero-supero-lateral aspect, inseparable from the right lacrimal gland abutting superior and lateral rectus muscle, There was loss of intervening fat plane at superoposterior aspect of right globe, causing right eye proptosis, with no macroscopic invasion. There was no evidence of erosion of bony orbit or extraorbital / intraconal extension of lesion or intralesional haemorrhage. With these findings, MRI report was suggestive of neoplastic right lacrimal gland lesions, probably pleomorphic adenoma of right lacrimal gland.

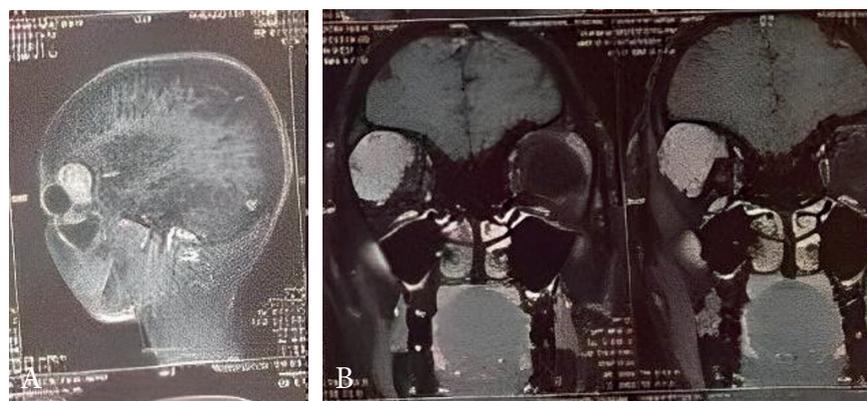


Figure 2 : (A) T1 weighted post-contrast MRI images, sagittal view showing right superior lateral orbital well-defined mass with heterogeneous signal intensity and Figure (B) : T1 weighted post contrast MRI images, coronal section showing well-defined lobulated altered signal intensity enhancing lesion of the extraconal compartment in anterior-superior-lateral aspect.

Thereafter, the clinical diagnosis of pleomorphic adenoma of right lacrimal gland was made and surgical excision of the mass was planned.

After taking the informed written consent,

right lateral orbitotomy (without marginotomy) via eyelid crease incision and transcutaneous-transseptal approach with complete excision of the tumour was performed under general anaesthesia.

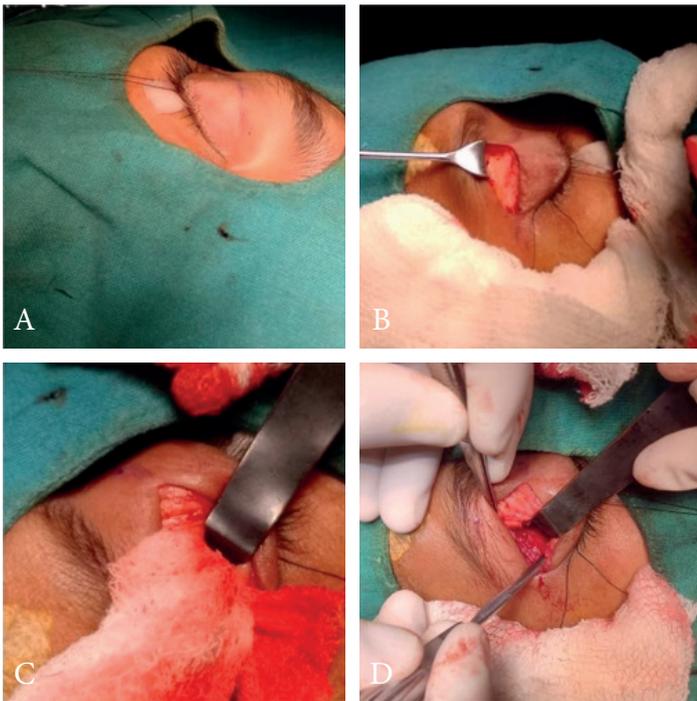


Figure 3: Tractional Suture with 4'0 silk was applied to the right upper lid and the incision site was marked over the lateral eyelid crease(A); Supero-lateral transcutaneous eyelid crease incision (B); Overlying orbicularis oculi and septum was dissected (approach- transcutaneous-transseptal)(C); Levator aponeurosis and preaponeurotic fat pad seen, separated and the mass was freed from overlying periorbita (D).

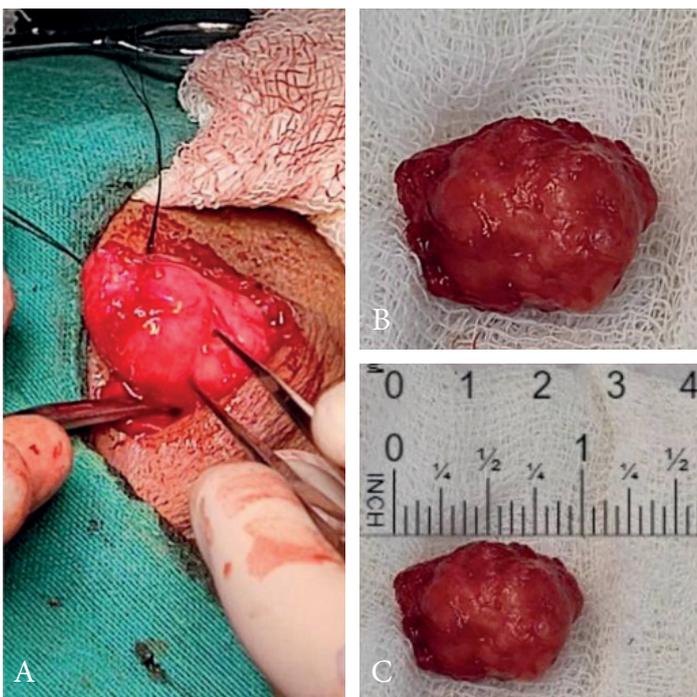


Figure 4 : In Toto removal of mass. (Encapsulated, globular, fleshy consistency mass, attached to overlying supero-temporal periorbital with posterior extension into extraconal space(A); Around 2.5 cm sized red-pinkish mass with regular margin, fleshy consistency, firm and with no abnormal vascularity (B,C).

Before and after surgical resection photographs



Figure 5: Before surgery showing RE non-axial proptosis with displacement of RE downward and medially; Figure B: 3rd week post-operative period, showing scar over the incision site, along with mechanical ptosis Figure C : Follow-up after one year of surgery (no visible proptosis and perfect alignment).

The mass was sent for histopathological examination. The report showed encapsulated mass with the proliferation of both epithelial and stromal elements. Epithelial cells were arranged in sheets admixed with multiple layers of myoepithelial cells. The cells had round to oval nuclei, and fine chromatin in a moderate amount of eosinophilic to clear cytoplasm. No atypia or mitosis was seen. It was thus concluded as pleomorphic adenoma.

The patient was post-operatively managed with antibiotics, corticosteroids, and non-steroidal anti-inflammatory drugs (NSAIDs).

She was discharged from the hospital after five days and now the patient is following up every six months and no evidence of recurrence has been noticed.

DISCUSSION

The protrusion of the eyeball may result in a wide variety of conditions such as thyroid disorders, orbital tumours, trauma, vascular disorders, infection, inflammation, and vascular disorders, which can be vision or life-threatening ((Ahmad SS, Anwar R, Khan MA, Usmani N, 2021) .

Out of these conditions in the age group of 10-30 years, lacrimal gland tumour is common.

Pleomorphic adenoma of lacrimal gland is a slow-growing, mixed tumour accounting for 20-25% of all lacrimal glands tumours and arises from epithelial cells and myoepithelial cells. It is the most common type of tumours of lacrimal glands (Ahmad SS, Anwar R, Khan MA, Usmani N, 2020). It occurs mostly in the

orbital lobe (80-90%) and the rest occurs in the palpebral lobe.

Histologically, these neoplasms are classified into two types i.e. epithelial and non-epithelial types. Twenty to thirty percent of neoplasms are epithelial types and 55% of epithelial tumours of lacrimal glands are benign (Shields et al., 2004).

Clinically, pleomorphic adenoma of lacrimal gland is defined as a gradually expanding, painless swelling that can lead to ptosis, non-axial proptosis, double vision and reduced eye movement (Perez et al., 2006). The average duration of symptoms may be up to two years. The average age of presentation is about 40 years and pain is an uncommon symptom (Perez et al., 2006; Holstein et al., 2015)

The clinical history, examination and imaging studies serve as the foundation for preoperative diagnosis. In normal MRI, lacrimal gland in gadolinium enhanced T1 weighted fat saturated images will show homogenous enhancement and are isointense to adjacent extraocular muscles.

In comparison to normal MRI, pleomorphic adenoma on MRI presents as low to intermediate T1 signal and intermediate T2 signal compared with orbit muscles. They show moderate enhancement after contrast administration (Young et al., 2018).

In computed tomography (CT) and MRI images, it shows as a solid, well-defined, round or oval space-occupying lesion, with the occasional bone remodelling and calcification. Malignant tumours, in contrast, may have uneven borders and exhibit features of bone erosion (Brown and Sampath, 2008).

The pleomorphic adenoma tumours have an average diameter of 2.1 cm and range in size from 1.3 - 5 cm upon gross examination. Their texture ranges from soft and shimmering in tumours with a predominance of mesenchymal cells to hard tumours with a predominance of epithelium cells. In a study published by Pushkar et al. (2013) on Clinicopathologic Review of Epithelial Tumours of the Lacrimal Gland, all pleomorphic adenoma tumours were completely encapsulated on both gross and light microscope inspection. All pleomorphic adenoma tumours contained different amounts of epithelial, mesenchymal, and myoepithelial cells under a light microscope. Myxomatous and chondroid regions, which are representations of the mesenchymal tissue, were visible. Microscopically, 18 (36%) patients had cystic degeneration, 16 (32%) had squamous metaplasia, six (12%) had calcification, and one (2%) had ossification (Pushker et al., 2013).

In a study done by Mendoza et al., (2013) the pleomorphic adenoma were invested with a variably thick pseudocapsule but an atypical pleomorphic adenoma was composed of double layered, epithelium lined glandular structures with small to patulous lumens and spindly outer myoepithelial cells (Mendoza et al., 2013).

Pleomorphic adenoma is pseudoencapsulated benign neoplasm primarily consisting of bland, lumen-forming and spindled myoepitheliomatous cells, each with a nucleus that is often oval, uniform, finely stippled, and either lacks a nucleolus entirely or has a tiny, punctate nucleus (Vogele et al., 2022). In pleomorphic adenoma, lumens have been demonstrated to have double-layered cellular walls and to show splaying-off or feathering of

the outer myoepithelial cells into the stroma, where they experienced diverse alterations and transformed into hyaline, myxoid, or chondroid foci. ('1. Ellis GL, Auclair PL. Tumours of the salivary glands. AFIP Atlas of Tumor Pathology, 4th series. Washington, D.C.: American Registry of Pathology'; no date).

Depending upon the location, the definite management for a pleomorphic adenoma involves complete excision. Excision can be approached via lateral canthal rhytids or upper lid crease incision (Halli et al., 2011). Intraoperative capsule rupture without complete removal may be associated with recurrence (Halli et al., 2011).

In 1889, Kronlein was the first one to suggest the lateral approach (Society, 2015), which Berke later refined. Klirtschafter and Chu (Wirtschafter and Chu, 2015) also discussed lateral orbitotomy technique to preserve the lateral rim, where they used acorn-tipped bur to straighten the external surface of the lateral wall of the orbital rim and the lateral canthus tendon. The best management now stands with complete excision of the tumour with its pseudocapsule via a lateral orbitotomy approach (Halli et al., 2011).

Lacrimal gland fossa exposure through lid crease or brow incision has been explained by Zeynel et al. (Year) in "Surgical Technique for Orbital Tumor" (Techniques and Tumours, 2019). Similar approach has been explained by

Dutton et al. in the "Textbook of Oculoplastic, Lacrimal and, Orbital Surgery" published in 1991 (Dutton, J.J., 1991). Here, a lid crease incision was given and orbital septum was opened. After retracting the fat, the lesion was identified for removal. This procedure can be combined with lateral orbital approach for better visualisation and removal of lacrimal gland tumour. (Dutton JJ, 2010) In our case, lateral orbitotomy via eyelid crease incision and transcutaneous-transseptal approach was performed without bone removal giving adequate surgical exposure with aesthetically acceptable scar, enabling the surgeon to completely excise the tumour with less post-operative complications.

Prognosis of complete excision of pleomorphic adenoma of lacrimal gland is excellent. The five-year recurrence rate of complete excision is 3% while with the incomplete excision, it could be as high as 32% (Halli et al., 2011).

CONCLUSION

Pleomorphic adenoma is the most prevalent benign lacrimal gland tumour, which usually affects unilateral lacrimal gland and causes non-axial proptosis. Complete surgical excision is usually carried out following clinical diagnosis and imaging findings. Patients are likely to have total resolution of symptoms and excellent prognosis after complete surgical removal.



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