

## Case Report

# Pleomorphic adenoma of retrobulbar ectopic lacrimal gland: a rare occurrence

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## Abstract

Ectopic lacrimal gland encompasses all lacrimal tissue that is outside the lacrimal gland fossa, excluding the accessory glands of Krause and of Wolfring. The bulbar conjunctiva and limbus are most commonly involved regions while retrobulbar region is unusual. This tissue may undergo neoplastic transformation, the commonest tumour being pleomorphic adenoma. A 30 year old male presented with painless loss of vision and mild proptosis of the right eye since 5 months. MRI disclosed a well-defined, lobulated contrast enhancing mass in the right retrobulbar region measuring 2 X 2 X 2 cm and a differential diagnosis of vascular or glial neoplasm was entertained. Excision biopsy revealed an encapsulated mass away from the lacrimal fossa. Histopathology showed features of pleomorphic adenoma which was confirmed on IHC.

**Key words:** Pleomorphic adenoma, Ectopic lacrimal gland, Retrobulbar

## Introduction

The lacrimal apparatus comprises of the main lacrimal gland and the two sets accessory glands (of Krause and of Wolfring). While the main lacrimal gland is situated in the orbital lacrimal fossa, its palpebral lobe is situated on the temporal side of the superior fornix[1]. While the eyelids [2], tarsal plate [3] and the nasal mucosa [4] have been reported to harbor ectopic lacrimal gland tissue, the commonest sites involved are

the bulbar conjunctiva and limbal area [5, 6]. Orbital involvement is relatively uncommon [7]. Pleomorphic adenoma is the commonest epithelial neoplasm to beset the lacrimal gland. [8]

We present a case of unilateral proptosis with diminished vision secondary to a pleomorphic adenoma in an ectopic lacrimal gland deep within the orbit.

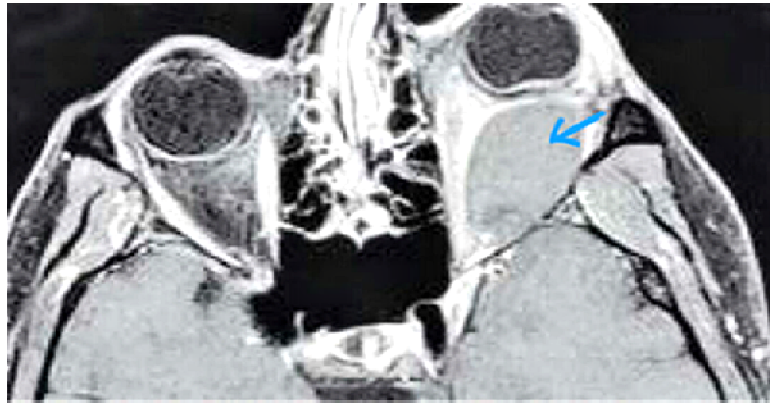
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A 30-year-old man presented the OPD with complaints of painless loss of vision in the right eye and slight protrusion of the right eyeball since 5 months. On examination, there was mild proptosis of the right eye accompanied by visual acuity limited to counting fingers at 3 feet; his best corrective visual acuity in the left eye was 6/12. There was restriction in abduction of the right eye while the left eye showed no remarkable indispositions. Ultrasound revealed a hypoechoic, predominantly solid (with small cystic area) mass superolateral to right eyeball, suggestive of epidermoid lacrimal gland tumor. Magnetic resonance displayed a well-defined, lobulated, contrast enhancing retrobulbar mass in right orbit, separate from the lacrimal gland (Figure-1). Based on these observations a differential diagnosis of cavernous hemangioma or glial tumour or connective tissue tumour was proposed. Lateral orbitotomy was done and the mass was completely excised. Gross examination exhibited a well circumscribed lobulated mass measuring 2X2X2cm (Figure2)

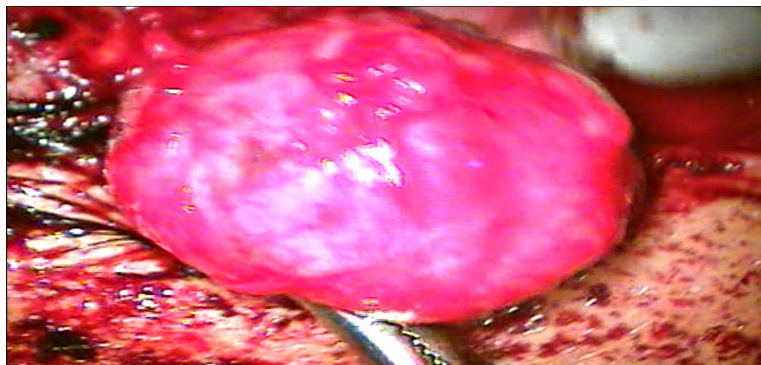
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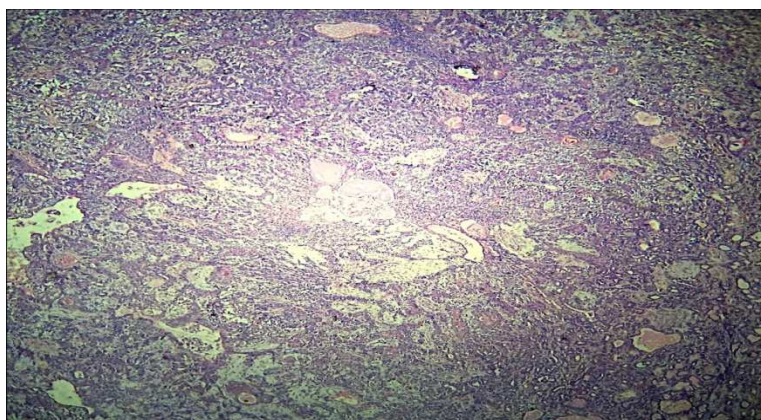
with a grayish white cut surface. Histopathology unveiled an encapsulated tumor with predominantly oval to spindle cells arranged in principally in cords, hemangiopericytoma like pattern and occasional pseudo rosettes, with microcystic areas. Abundant myxoid stroma to which the spindle cells appeared to “melt” and sparse glandular elements were also present; mitoses and necrosis were absent (Figures 3, 4 and 5). Pericapsular normal lacrimal gland tissue was also noted. In light of the morphological features pleomorphic adenoma was considered the principal diagnosis while hemangiopericytoma was entertained as a differential due to the unusually large areas of perivascular arrangement of the spindle cells. Confirmation was done through immunohistochemistry (IHC) which showed diffuse positivity for cytokeratin and S100 protein (Figure 6). The postoperative recovery was uneventful and the patient was discharged.



**Figure-1:** MRI showing a well defined, lobulated, contrast enhancing retrobulbar mass in right orbit (arrow).

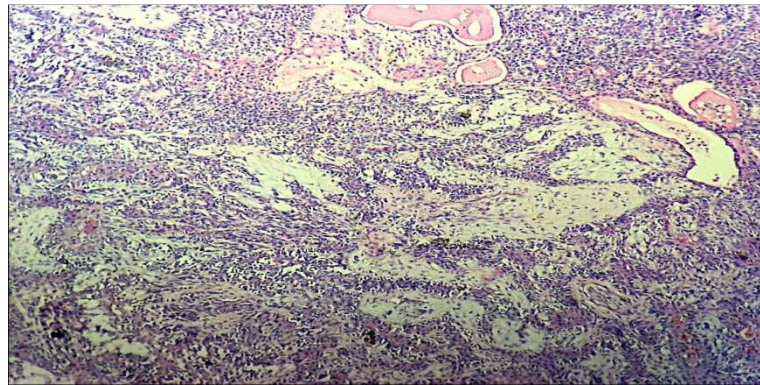


**Figure-2:** Intraoperative photograph showing an encapsulated, well defined slightly lobulated tumour mass.

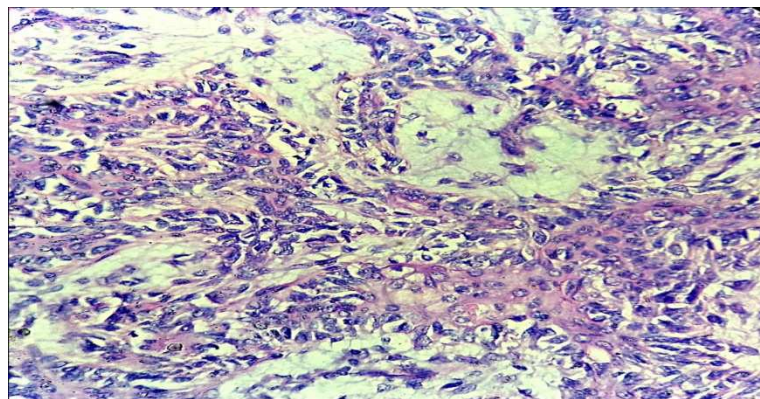


**Figure-3:** Biphasic population of spindle cells arranged in cords, few microcystic areas, fibromyxoid stroma and little glandular element at the periphery; interspersed hemangiopericytoma-like areas (H&E, 40X).

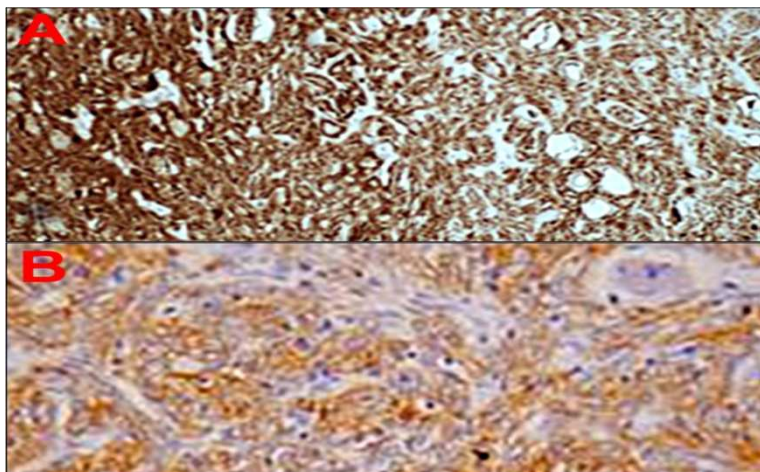
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**Figure-4:** Anastomosing cords of spindle cells, adjacent myxoid stroma with few dilated blood vessels. (H&E, 100X)



**Figure-5:** Oval to spindle cells which appear to “melt” into the adjacent myxoid stroma (H&E, 100X).



**Figure-6:** Immunohistochemistry showing positive (A) S 100 protein & (B) Cytokeratin.

**Discussion**

The orbital lobe of lacrimal gland is derived from five to six epithelial buds formed by the proliferating epithelial cells of the conjunctival fornix at around two months of gestation following which other epithelial

buds commence the formation of the palpebral lobe [9-11]. During development, a part of the gland may get secluded and develop dissociated from the main gland [12]. 30% of lacrimal gland lesions are made up by

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epithelial tumours, 12% of which are pleomorphic adenomas. [13]. Some investigators have reported the proportion to be as high as 48% [14]. The tumour has a high tendency to occur in the orbital lobe of lacrimal gland [8], however it has been reported in the palpebral lobe [15], in the upper lid [8], Krause's gland in lower lid [16], gland of Wolfring in lower tarsal conjunctiva [17] and the orbit [18].

Although cases have been reported between 6-80 years of age, the tumour usually manifests between 30-70 years [18]. Perusal of literature unearthed 4 reported occurrences of pleomorphic adenoma in an ectopic lacrimal gland situated deep within the orbit [19-22]. Complete removal, with the capsule remaining intact, is the curative treatment for pleomorphic adenoma arising from lacrimal gland tissue [8].

Gradually increasing painless proptosis of the right eye were the main findings in our patient. This growth pattern seems similar to the growth pattern of pleomorphic adenoma arising from the main lacrimal gland [23, 24]. The essential diagnostic feature of a pleomorphic adenoma is that it is composed of both epithelial and mesenchymal tissues.

The epithelial cells form characteristic ductal structures with surrounding myoepithelial cells, which trail out gradually into myxomatous mesenchyme [14,25]. IHC exhibits positivity for vimentin, S-100, calponin, p63, and cytokeratin [26]. The present case demonstrates an unusual site of occurrence of pleomorphic adenoma. Due to the anatomical location, tumours like hemangiopericytoma and glial tumours like astrocytoma were first considered in differential diagnosis. However, the representative morphology along with IHC proved confirmatory.

## Conclusion

Pleomorphic adenoma should be considered a possibility in slowly growing retro-orbital tumours which on radiological assessment are well circumscribed and devoid of any bony or soft tissue invasion. While microscopic evidence of glandular structures surrounded by myoepithelial cells "melting" into a chondromyxoid stroma is the prototypic histology, demonstration of epithelial (cytokeratin, EMA and CEA) and myoepithelial (calponin, S100 and p63) elements on IHC clinches the diagnosis in doubtful cases.

## References

1. Alyahya GA, Bangsgaard R, Prause JU, Heegaard S. Occurrence of lacrimal gland tissue outside the lacrimal fossa: comparison of clinical and histopathological findings. *Acta Ophthalmol Scand.* 2005 Feb;83(1):100-3.
2. McCulley TJ, Yip CC, Kersten RC, Kulwin DR. An ectopic site of lacrimal gland secretion mimicking epiphora. *Arch Ophthalmol.* 2002 Nov;120(11):1586-7.
3. Lee WR, Aitken DA, Kirkness CM. Diffuse hyperplasia of intratarsal ectopic lacrimal gland tissue. *Arch Ophthalmol.* 2002 Dec;120(12):1748-52.
4. Pe'er J, Ilsar M. Ectopic lacrimal gland under the nasal mucosa. *Am J Ophthalmol.* 1982 Sep;94(3):418-9.
5. Pfaffenbach DD, Green WR. Ectopic lacrimal gland. *Int Ophthalmol Clin.* 1971 Fall;11(3):149-59.
6. Pokorny KS, Hyman BM, Jakobiec FA, Perry HD, Caputo AR, Iwamoto T. Epibulbar Choristomas Containing Lacrimal Tissue: Clinical Distinction from Dermoids and Histologic Evidence of an Origin from the Palpebral Love. *Ophthalmology.* 1987; 94(10):1249-57.
7. Sakurai H, Mitsuhashi N, Hayakawa K, Nozaki M, Kurosaki H, Kishi S, Joshita T, Niibe H. Ectopic lacrimal gland of the orbit. *J Nucl Med.* 1997 Sep;38(9):1498-500.
8. Adel H. Alsuhaibani MD. Slow-growing large pleomorphic adenoma of ectopic lacrimal gland tissue in the upper eyelid. *Saudi Journal of Ophthalmology* 2012; 26(4): 453-5.
9. Duke-Elder S, Cook C. Normal and abnormal development. Part I. Embryology. In: Duke-Elder S, editor. *System of Ophthalmology*, Vol. III. London. Kimpton; 1963.
10. Jakobiec F, Iwamoto T. Ocular adnexae: introduction to lids, conjunctiva and orbit. In: Jakobiec F, editor, *Ocular Anatomy, Embryology and Teratology.* Philadelphia. Harper & Row; 1982.

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11. Ozanics V, Jakobiec F. Prenatal development of the eye and its adnexa. In: Jakobiec F, editor, *Ocular Anatomy, Embryology and Teratology*. Philadelphia. Harper & Row; 1982.
12. Nikhil, Kaushik C. Ectopic lacrimal gland in the eyelids (a case report). *Indian J Ophthalmol*. 1985 Jan-Feb; 33(1):65-6.
13. Font RL, Smith SL, Bryan RG. Malignant epithelial tumors of the lacrimal gland: a clinicopathologic study of 21 cases. *Arch Ophthalmol*. 1998 May;116(5):613-6.
14. Weis E, Rootman J, Joly TJ, Berean KW, Al-Katan HM, Pasternak S, Bonavolontà G, Strianese D, Saeed P, Feldman KA, Vangveeravong S, Lapointe JS, White VA. Epithelial lacrimal gland tumors: pathologic classification and current understanding. *Arch Ophthalmol*. 2009 Aug;127(8):1016-28. doi: 10.1001/archophthalmol.2009.209.
15. Yamada T, Kato T, Hayasaka S, Hayasaka Y, Kadoi C. Benign pleomorphic adenoma arising from the palpebral lobe of the lacrimal gland associated with elevated intraocular pressure. *Ophthalmologica*. 1999; 213(4):269-72.
16. Saini JS, Mukherjee AK, Naik P. Pleomorphic adenoma of Krause's gland in lower lid. *Indian J Ophthalmol*. 1985 May-Jun;33(3):181-2.
17. Alyahya GA, Stenman G, Persson F, Prause JU, Skjødt K, Saunte JP, Heegaard S. Pleomorphic adenoma arising in an accessory lacrimal gland of Wolfring. *Ophthalmology*. 2006 May;113(5):879-82. Epub 2006 Mar 13.
18. Patyal S, Banarji A, Bhadauria M, Gurunadh VS. Pleomorphic adenoma of a subconjunctival ectopic lacrimal gland. *Indian J Ophthalmol*. 2010 May-Jun;58(3): 245-7. doi: 10.4103/0301-4738.62656.
19. Mindlin A, Lamberts D, Barsky D. Mixed lacrimal gland tumor arising from ectopic lacrimal gland tissue in the orbit. *J Pediatr Ophthalmol* 1977; 14(1): 44-47.
20. Mueller EC, Borit A. Aberrant lacrimal gland and pleomorphic adenoma within the muscle cone. *Ann Ophthalmol*. 1979 Apr;11(4):661-3.
21. Mokkalapati PK, Mahapatra M, Satish K, Ram K. Giant pleomorphic adenoma of lacrimal gland presenting with diminished vision—a case report. *Asian Journal of Ophthalmology*. 2015; 14(2): 83-6.
22. Misra S, Bhandari A, Misra N, Gogri P, Mahajan S. Pleomorphic adenoma of a deep orbital ectopic lacrimal gland. *Orbit*. 2016 Oct; 35 (5):295-7. doi: 10.1080/01676830.2016.1193526. Epub 2016 Aug 10.
23. Lloyd GA. Lacrimal gland tumours: the role of CT and conventional radiology. *Br J Radiol*. 1981 Dec;54(648): 1034-8.
24. Rose GE, Wright JE. Pleomorphic adenoma of the lacrimal gland. *Br J Ophthalmol*. 1992 Jul; 76 (7): 395-400.
25. Barnes L, Eveson J, Reichart P, Sidransky D. *Pathology and Genetics of Tumours of the Head and Neck*. Lyon. International Agency for Research on Cancer; 2005.
26. Chan JKC, Cheuk W. Tumours of the salivary glands. In: Fletcher CD, editor. *Diagnostic Histopathology of Tumours*, Volume 1. 4<sup>th</sup> ed. Philadelphia. Elsevier Saunders; 2013.

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