

CASE REPORT

A rare case of atypical pleomorphic adenoma arising from periocular ectopic lacrimal gland

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Abstract

Purpose To describe features of atypical pleomorphic adenoma, a rare clinical entity, particularly when found in ectopic periocular lacrimal gland tissue.

Methods Case report of biopsy-confirmed periocular atypical pleomorphic adenoma.

Results A 35-year-old female presented with a unique orbital lesion found to be ectopic lacrimal gland demonstrating atypical pleomorphic adenoma on formal histopathologic review. Pleomorphic adenoma is pathologically characterized as an epithelial

lesion intermixed with mesenchymal elements. It is further classified as atypical with the presence of features such as hypercellularity, regions of necrosis or hyalinization, cellular dysplasia, capsular violation, and malignant characteristics without frank local extension or distant metastases.

Conclusions Due to its rarity, the natural history and prognosis of atypical pleomorphic adenoma is unclear. Physicians need to recognize this entity, and complete surgical excision with strict follow-up regimens are likely warranted.

Keywords Pleomorphic adenoma · Atypical · Ectopic · Lacrimal gland · Periocular · Orbit

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Introduction

Pleomorphic adenoma (PA) is the most common benign epithelial neoplasm of the lacrimal gland and has potential for malignant transformation. Infrequent reports of ectopic lacrimal gland histopathologically showing PA have been described in the periocular and orbital region [1]. However, this report highlights a unique case of atypical PA within ectopic lacrimal gland, a rare clinical entity previously described in salivary glands [2–4].

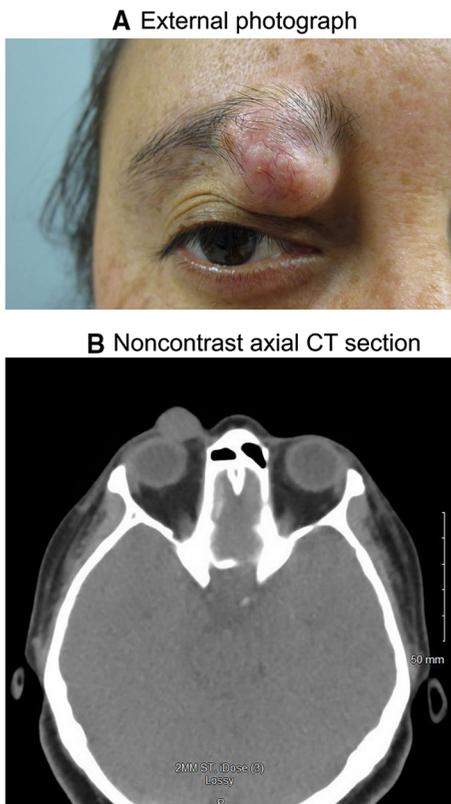


Fig. 1 Clinical Presentation of Orbital Lesion and CT Imaging. **a** External photograph demonstrates right periorbital lesion. **b** Noncontrast axial CT imaging reveals mass occupying the right anterior orbit without involvement of the globe or postseptal extension

Case report

A 35-year-old female presented with a periorbital growth localized to the right sub-brow region that gradually enlarged over 1.5 years, with a more rapid increase in size during the 8 months preceding examination.

Complete ophthalmic exam was normal with the exception of right upper eyelid mechanical ptosis secondary to the mass which was a slightly mobile, non-transilluminating solid lesion along the infero-medial brow measuring 18 mm × 22 mm with overlying telangiectasias (Fig. 1).

Computed tomography (CT) of the orbits showed a well-circumscribed 2-centimeter right periorbital soft tissue mass abutting the globe without significant mass effect, surrounding inflammatory change, postseptal extension, or regional bone erosion (Fig. 1). An encapsulated lobular mass was removed in total during excisional biopsy without capsular violation. Pathologic examination revealed atypical PA with increased hyalinization and mesenchymal features compared to ordinary PA; there was no extracapsular extension (Fig. 2).

Discussion

Pleomorphic adenoma, the most common lacrimal gland epithelial neoplasm, usually occurs in the orbital lobe and, less commonly, in accessory glands of

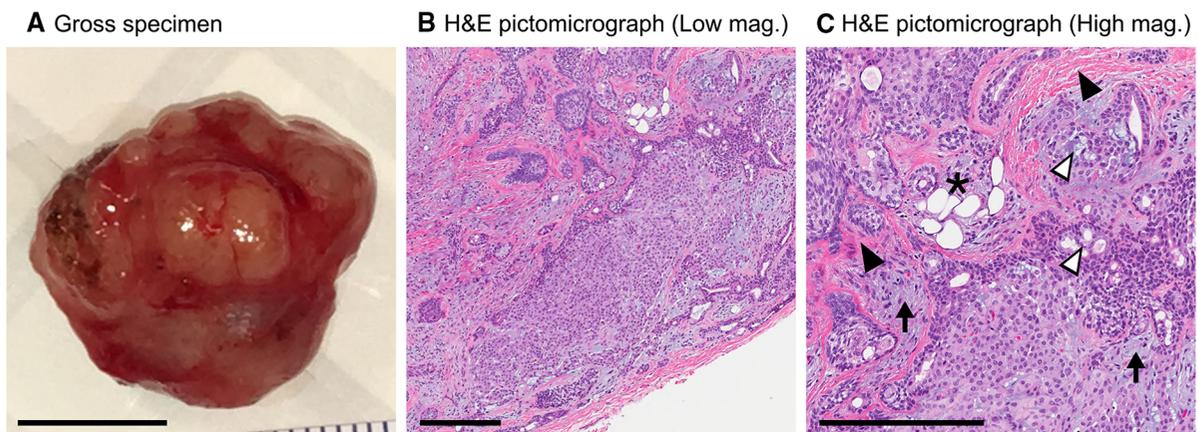


Fig. 2 Histopathology of orbital lesion. **a** Photograph of gross specimen. **b**, **c** H&E sections of the lesion demonstrate an admixture of both epithelial and mesenchymal elements, including adipose tissue (asterisk), mucinous glands (white

arrowheads), myxoid material (black arrows), and hyalinized stroma (black arrowheads); scale bars represent 1 cm (**a**) and 300 μ m (**b**, **c**)

Krause and Wolfring. Pathologic examination reveals an epithelial lesion intermixed with varying degrees of mesenchymal elements including mucoid, chondroid, osseous, adipose, and hyaline material [2, 5]. In all cases, complete surgical excision is recommended to decrease the chance for local recurrence and possible malignant transformation which has ranged from 1.9 to 23.3% in existing literature [2, 3].

Ectopic lacrimal gland is infrequently encountered, and the clinical presentation varies depending on the underlying histopathology. Review of literature shows limited reports of ordinary PA within periocular ectopic lacrimal tissue involving the eyelids, canthus, subconjunctival space, and orbit [1]. Atypical PA is an uncommon neoplastic subtype within this already limited group, and thus far, has been described in non-lacrimal major and minor salivary tissues including the submandibular and parotid glands [2–4, 6]. Initially coined by Auclair and Ellis, the features warranting atypical classification include hypercellularity, regions of necrosis and hyalinization, cellular dysplasia (i.e., pleomorphism, atypical nuclei/nucleoli, mitoses), capsular violation, and features of malignancy without frank regional extension or distant metastases [2–4]. Nearly 14% of atypical PA cases were noted to progress to invasive carcinoma [4]. Due to its rarity, the natural history and prognosis of this histopathologic variant is unclear. Nonetheless, physicians need to recognize its existence. Complete

surgical excision and more strict follow-up regimens are likely warranted for affected patients.

Compliance with ethical standards

Consent for publication The patient has consented to submission of this case report for review and publication by the journal.

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