

## Pleomorphic Adenoma of the Eyelid with Apocrine Gland Origin; an Atypical Location

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### Abstract

**Purpose:** To describe the clinical presentation and treatment of a patient with a cutaneous pleomorphic adenoma of the eyelid.

**Case Report:** A 73-year-old male patient presented with a nodular mass on the lateral third of his right upper eyelid, which had slowly enlarged over 10 years. Radiologic features were of an extra-conical mass, with no invasion of adjacent structures. An excisional biopsy of the lesion was performed. The histopathological examination revealed a biphasic tumor, composed of tubules with a double layer of epithelial cells arranged in a chondromyxoid stroma. The inner epithelial cells were positive for pancytokeratins AE1/AE3 and carcinoembryonic antigen. The outer epithelial cells and stromal component expressed vimentin and S100 protein. These pathologic findings were consistent with a palpebral pleomorphic adenoma, with an apocrine gland origin.

**Conclusion:** Pleomorphic adenomas of the skin are rare tumors, and even less frequent as tumors of the ocular adnexa. These lesions should be considered in the differential diagnosis of palpebral nodular masses, and complete excision should be attempted due to the possibility of malignant transformation.

**Keywords:** Pleomorphic Adenoma; Apocrine Gland Tumor; Ocular Adnexal Tumor

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### INTRODUCTION

Pleomorphic adenoma (PA), also known as chondroid syringoma, is a tumor that arises most often in the lacrimal or salivary glands.<sup>[1]</sup> Occasionally, PA can arise from the eccrine or apocrine glands of the skin, particularly on the cheek, nose or skin above the lip and, rarely, in the periocular region.<sup>[2-4]</sup> PA is one of the rare neoplasms

occurring in the adnexal glands of the eyelids.<sup>[5]</sup> Of the series of 228 benign adnexal ocular tumors reviewed by Ozdal et al, no case of PA in the eyelid was reported.<sup>[6,7]</sup> Also in the extensive revision of 5504 eyelid skin tumors performed by Deprez and Uffer, of the 579 adnexal skin tumors observed, no PA were found.<sup>[8]</sup> This tumor has no specific clinical manifestations, and may be impossible to differentiate clinically from other subcutaneous eyelid lesions.<sup>[1]</sup> Radiologic features are also nonspecific, although they can accurately show the extent and location of the lesion.<sup>[2,9]</sup> Histopathological and immunohistochemical analyses are essential for a definite diagnosis since they are characteristic for skin PA and permit a differential diagnosis with more frequent PA of the lacrimal gland.

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We report a case of pleomorphic adenoma which originated from the ocular adnexa with important clinical implications for management and follow-up.

## CASE REPORT

A 73-year-old male patient presented with a firm, subcutaneous nodule in the right upper eyelid [Figure 1a]. The lesion was present for 10 years, slowly growing and painless. The mass was round, relatively hard and moved freely under the skin, without pain on palpation or mobilization. The overlying skin showed no inflammatory signs or abnormal pigmentation. There was no restriction in ocular movements, and the remaining ophthalmological examinations were unremarkable.

The patient underwent an orbital CT scan that revealed an extra-conical mass, adjacent to the lateral rectus insertion, with no distortion of the globe or invasion of the bone or muscular structures. The mass had a heterogeneous density, and two cysts were present in its superoanterior portion [Figure 1b].

An excisional biopsy was performed over the upper eyelid dermis, through a 15 mm horizontal incision in the lateral third of the eyelid crease. The tumor was well circumscribed with no adhesions and was easily dissected and removed in its entirety [Figure 1c]. The lacrimal gland was found to be independent of the mass, with a normal appearance. The incision was closed with a 6.0 polypropylene suture. The postoperative appearance can be observed in [Figure 1d].

On gross pathologic examination, the nodule was well-defined with dimensions 20 mm × 15 mm × 12 mm. A smooth and tan cutting surface was found, with tiny cavitated areas containing a pasty whitish material. The entire specimen was submitted to histologic examination. The tumor was totally encapsulated and presented a biphasic pattern, consisting of a chondromyxoid stroma and an epithelial component [Figure 2a]. The epithelium was arranged in nests, cords, and tubes, or glandular branching structures layered by a double epithelium

in which the inner epithelial layer exhibited apocrine features, such as decapitation [Figure 2b and 2c]. There was no significant atypia. Numerous cysts were present in the tumor, lined by metaplastic stratified squamous epithelium and containing keratinized epithelial plugs in the lumen [Figure 2d]. No areas of necrosis, interstitial hemorrhage or appreciable mitotic activity were found. No satellite nodules were identified. Immunohistochemistry revealed a variable expression of the inner epithelial cells to pancytokeratins AE1/AE3 and to carcinoembryonic antigen (CEA) [Figure 2e and 2f]. The outer epithelial cells and stromal component expressed vimentin and S100 protein [Figure 2g and 2h]. There was no evidence of lacrimal gland parenchyma or signs of malignant changes. These findings defined the tumor as a palpebral pleomorphic adenoma and were consistent with apocrine differentiation.

The systemic work-up was negative for metastasis; thus the patient was kept under vigilance with periodic observations. In 2 years of follow-up, he has remained asymptomatic without the signs of local recurrence.

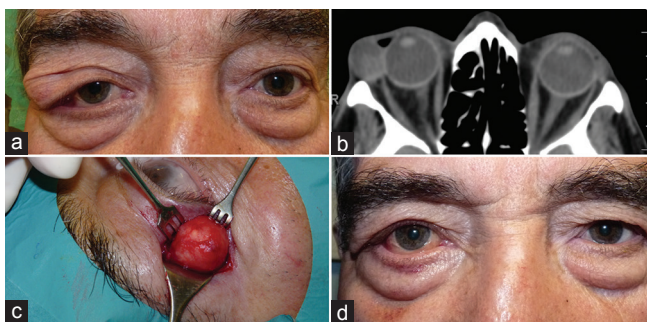
## DISCUSSION

Pleomorphic adenoma is a rare mixed tumor of the skin, composed of both epithelial and mesenchymal stromal derived elements.<sup>[6,10]</sup> The classification of these tumors into an apocrine type or eccrine type relies on the histopathological appearance of the glandular component. Comparable to our case, the tumor may originate from the apocrine sweat glands, where the glandular epithelial component is lined by a double layer of cuboidal epithelial cells arranged in islands, chords or tubules. The lesions with an eccrine type morphology consist of small tubular lumina lined by a single layer of cuboidal epithelial cells.<sup>[11,12]</sup> Immunohistochemical markers are of limited value to differentiate apocrine from eccrine sweat gland types of PA. CEA is positive on the secretory lumen of the secretory cells and the luminal surface of the duct in eccrine sweat glands, while on the apocrine secretory cells, it may also label the luminal membranes.<sup>[12]</sup>

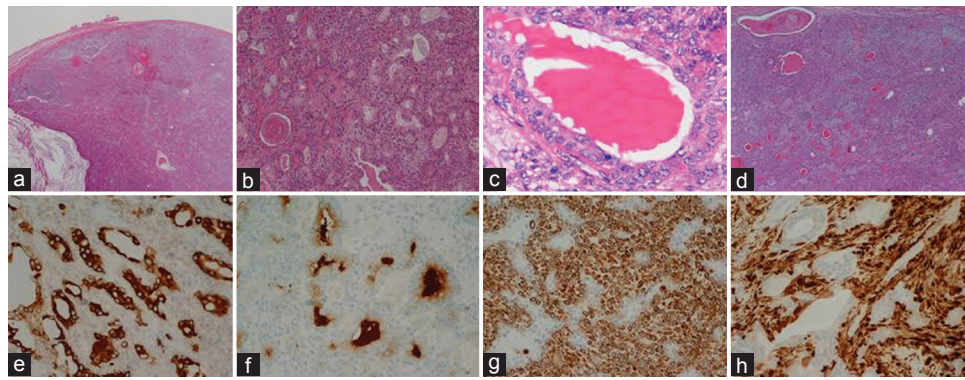
The stromal component in PA may be myxoid, chondroid, adipocytic or fibrous. In our case, the stromal cells showed myoepithelial differentiation, illustrated by being positive to S100 and vimentin.<sup>[11,13]</sup>

No features suspicious of malignancy such as cytological atypia, increased mitotic figures, infiltrative margins, satellite tumor nodules or tumor necrosis were found on histology. Furthermore, there was no reported recurrence or metastases to date. As none of these criteria were observed, we excluded the diagnosis of atypical or malignant PA.<sup>[14,15]</sup>

The most important differential diagnosis of PA of the eyelid is the much more frequent PA arising from the lacrimal gland, which accounts for 10% of all lacrimal gland lesions.<sup>[16]</sup> On the histopathological examination, the borders of the tumor must be carefully examined for



**Figure 1.** (a) Preoperative presentation of the lesion on the right upper eyelid. (b) Cranial CT scan, axial view showing an extra-conical mass, adjacent to the lateral rectus insertion. (c) Excisional biopsy of the tumor through the lateral third of the eyelid crease. (d) Postoperative presentation.



**Figure 2.** (a) Well-defined biphasic tumor composed of an epithelial component set in a stromal component (H and E,  $\times 20$ ). (b) Epithelial cells arranged in nests, cords or tubes (H and E,  $\times 100$ ). (c) Duct with a double layered epithelium showing “decapitation” secretion (H and E,  $\times 400$ ) (d) Keratinous cysts visible within the stroma (H and E,  $\times 40$ ). (e) AE1/AE3 positivity identifies the epithelial cells of the lesion (AE1/AE3,  $\times 200$ ). (f) CEA is present in the secretory lumen of secretory cells and luminal surface (CEA,  $\times 200$ ). (g and h) Vimentin and S100 positivity is present in the basal layer and in the stromal cells (Vimentin,  $\times 200$ , S100,  $\times 200$ ). HE, Hematoxylin & eosin staining; CEA, Carcinoembryonic antigen.

normal lacrimal gland tissue.<sup>[6]</sup> In our case, the sections of the tumor were observed independently by two pathologists who found no evidence of lacrimal gland parenchyma, excluding a lacrimal gland PA.

The treatment of choice for an eyelid PA is complete surgical removal, with a few millimeters safety margin. PAs are generally capsulated and well circumscribed and can be totally removed. However, these patients must be monitored due to the possibility of local recurrence and the risk of malignant transformation.

In conclusion, although PA of the eyelid is a rare tumor, it should be considered in the differential diagnosis of palpebral masses. Management of these patients includes complete excision of the lesion and long-term follow-up. Although PA is primarily benign, malignant forms have been reported.<sup>[17-20]</sup>

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## Conflicts of Interest

There are no conflicts of interest.

## REFERENCES

- Shields J, Shields C, editors. Eyelid pleomorphic adenoma (benign mixed tumor). Eyelid, Conjunctival, and Orbital Tumors. 2<sup>nd</sup> ed. Philadelphia: Lippincott Williams & Wilkins; 2008. p. 72.
- Belfquih H, El Mostarchid B, Oukabli M, Akhaddar A, Boucetta M. Benign chondroid syringoma of the orbit: A rare cause of exophthalmos. *Head Face Med* 2012;8:8.
- Sungur N, Uysal A, Gümüs M, Koçer U. An unusual chondroid syringoma. *Dermatol Surg* 2003;29:977-979.
- Bekerecioglu M, Tercan M, Karakok M, Atik B. Benign chondroid syringoma: A confusing clinical diagnosis. *Eur J Plast Surg* 2002;25:316-318.
- Palioura S, Jakobiec FA, Zakka FR, Iwamoto M. Pleomorphic adenoma (formerly chondroid syringoma) of the eyelid margin with a pseudocystic appearance. *Surv Ophthalmol* 2013;58:486-491.
- Gündüz K, Demirel S, Heper AO, Günalp I. A rare case of atypical chondroid syringoma of the lower eyelid and review of the literature. *Surv Ophthalmol* 2006;51:280-285.
- Ozidal PC, Callejo SA, Codère F, Burnier MN Jr. Benign ocular adnexal tumours of apocrine, eccrine or hair follicle origin. *Can J Ophthalmol* 2003;38:357-363.
- Deprez M, Uffer S. Clinicopathological features of eyelid skin tumors. A retrospective study of 5504 cases and review of literature. *Am J Dermatopathol* 2009;31:256-262.
- Nicolaou S, Dubec JJ, Munk PL, O’Connell JX, Lee MJ. Malignant chondroid syringoma of the skin: Magnetic resonance imaging features. *Australas Radiol* 2001;45:240-243.
- Mandeville JT, Roh JH, Woog JJ, Gonnering RS, Levin PS, Mazzoli RA, et al Cutaneous benign mixed tumor (chondroid syringoma) of the eyelid: Clinical presentation and management. *Ophthalm Plast Reconstr Surg* 2004;20:110-116.
- Obaidat NA, Alsaad KO, Ghazarian D. Skin adnexal neoplasms – Part 2: An approach to tumours of cutaneous sweat glands. *J Clin Pathol* 2007;60:145-159.
- Saga K. Histochemical and immunohistochemical markers for human eccrine and apocrine glands: An aid for histopathologic differentiation of sweat gland tumors. *J Investig Dermatol Symp Proc* 2001;6:49-53.
- Fuertes L, Santonja C, Kutzner H, Requena L. Immunohistochemistry in dermatopathology: A review of the most commonly used antibodies (part I). *Actas Dermosifiliogr* 2013;104:99-127.
- Elder D, Elenitsas R, Johnson B, Murphy G. Lever’s Histopathology of the Skin. 9<sup>th</sup> ed. Philadelphia: Lippincott Williams & Wilkins; 2005. p. 893-894.
- Weedon D. Weedon’s Skin Pathology. 3<sup>rd</sup> ed. London: Churchill Livingstone Elsevier; 2010. p. 791.
- Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: The 2002 Montgomery Lecture, part 1. *Ophthalmology* 2004;111:997-1008.
- Barnett MD, Wallack MK, Zuretti A, Mesia L, Emery RS, Berson AM. Recurrent malignant chondroid syringoma of the foot: A case report and review of the literature. *Am J Clin Oncol* 2000;23:227-232.
- Metzler G, Schaumburg-Lever G, Hornstein O, Rassner G. Malignant chondroid syringoma: Immunohistopathology. *Am J Dermatopathol* 1996;18:83-89.
- Ishimura E, Iwamoto H, Kobashi Y, Yamabe H, Ichijima K. Malignant chondroid syringoma. Report of a case with widespread metastasis and review of pertinent literature. *Cancer* 1983;52:1966-1973.
- Harrist TJ, Aretz TH, Mihm MC Jr., Evans GW, Rodriguez FL. Cutaneous malignant mixed tumor. *Arch Dermatol* 1981;117:719-724.