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Pleomorphic Adenoma with Prominent Clear Cell Myoepithelioma Component of the Lacrimal Gland

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Abstract

We report a case of a pleomorphic adenoma of the lacrimal gland with a prominent clear cell myoepitheliomatous component. An 81-year-old Caucasian woman experienced a 2 month history of right supraorbital swelling and proptosis. Excisional biopsy revealed a multi-component lesion including a stromal component featuring glandular structures made of small epithelioid and spindle cells and a trabecular component with small islands of vacuolated cells, displaced nuclei, and clear cytoplasm. Immunohistochemical analysis revealed strong cytokeratin AE1/3 reactivity and focal smooth muscle actin positivity. The pathologic findings including immunohistochemistry results were consistent with a pleomorphic adenoma with prominent clear cell myoepithelioma component.

Myoepitheliomas are glandular or soft tissue tumors rarely found in the orbit and ocular adnexa. They comprise only 1-1.5% of all salivary gland tumors and may develop *de novo* or from a pleomorphic adenoma.¹ An unusual case of a lacrimal gland pleomorphic adenoma with a prominent myoepithelioma component having clear cell features is described, with clinicopathologic correlation and a brief review of the literature. This report is in full compliance with the Declaration of Helsinki and current Health Insurance Portability and Accountability Act regulations.

Case Report

An 81-year-old Caucasian female presented with a right-sided palpable, superotemporal orbital mass. The patient reported rapidly increasing right, superotemporal swelling beginning 2 months prior to evaluation. Past medical history was significant for breast carcinoma with mastectomy.

Clinical examination revealed a palpable, firm right superotemporal mass and associated right-sided proptosis (Fig. 1A). The mass displaced the globe inferonasally. A computed tomography (CT) scan revealed a well-circumscribed mass of heterogeneous density in the right lacrimal gland fossa with no signs of bony erosion (Fig 1B & C). Due to the unusual appearance on imaging and unusually rapid growth, an incisional biopsy was performed. Microscopic examination showed duct-like structures bordered by epithelial cells

surrounded by darker, myoepithelial cells (Fig. 2A & B). The diagnosis of pleomorphic adenoma was made. Subsequently, total excision was performed via right anterior orbitotomy. Prior to excision, examination showed her visual acuity to be 20/20 in both eyes.

Gross examination revealed a red to pink, encapsulated mass measuring 2.5×1.6×1.4 cm. Microscopically, the specimen was polymorphic with two distinct components. The first component comprising 40% of the entire tumor contained glandular and stromal elements, similar to the biopsy. The second component comprising 60% of the entire tumor was made up of a trabecular proliferation of cells. The first component had duct-like structures lined by low cuboidal epithelial cells surrounded by myoepithelial cells (Fig 2C & D). This component blended seamlessly into the stroma, which contained a mixture of small epithelioid and spindle-shaped cells within a collagenous matrix. There were occasional areas of cartilage present within the stromal component. The second component was trabecular with small islands of cells with large vacuoles, displaced nuclei and clear cytoplasm (Fig 2E & F). Sparse (< 10%) ductal architecture lined by low cuboidal epithelium was present in this component of the tumor. The clear cells appeared contiguous with the myoepithelial lining of the ductal epithelial cells. The first component, consistent with a typical pleomorphic adenoma, was surrounded by a fibrous pseudocapsule. The second component, having clear cell myoepitheliomatous features, was encased by its own pseudocapsule but was adjacent to the pleomorphic adenoma component. Immunostaining was strongly positive for cytokeratin AE1,3 (Fig. 2G) and vimentin. There was focal positivity for smooth muscle actin (SMA) (Fig. 2H) and MAK6, with rare positivity for S100. Approximately 10% of nuclei were positive for Ki67. Based on the histoarchitecture and immunohistochemical findings, a diagnosis of pleomorphic adenoma with prominent clear cell myoepithelioma component was made. There have been no signs of recurrence 5 months after total excision.

Discussion

Myoepitheliomas are extremely rare in the lacrimal gland with few cases reported in the English literature (see Table). Two previous studies have reported cases of clear cell myoepithelial carcinomas arising in pleomorphic adenomas of the lacrimal gland^{2, 3}. Our case was a pleomorphic adenoma with prominent myoepitheliomatous features and lacked the nuclear pleomorphism and mitotic activity found in myoepithelial carcinoma.

Myoepitheliomas arise in salivary glands including the lacrimal gland.⁴ Dardick et. al.⁵ classified myoepitheliomas by both cellular features and architecture. Those authors classified myoepithelioma variants to include spindle-shaped, hyaline, epithelioid, clear, or mixed cytology. The clear cell was the least common comprising ~2.5% of the lesions while the spindle cell variant represented 32.5% of samples⁵.

Some authors believe that epithelial tumors of salivary glands exist on a continuum from pleomorphic to monomorphic with myoepitheliomas at one end of the spectrum representing a pure proliferation of myoepithelial cells and a tumor composition of less than 5-10% ductal structures. On the other end of the spectrum are monomorphic adenomas composed

mainly of ductal epithelial cells. In the middle of the spectrum are pleomorphic adenomas (mixed tumors) containing both epithelial and myoepithelial components.^{5, 6} Others suggest that pleomorphic adenomas and myoepitheliomas are distinct and do not share a common cell precursor. A common etiology for myoepitheliomas and pleomorphic adenomas is supported by genetic analysis showing a shared chromosomal aberration (12q) between these two entities⁷. An apparent clone of myoepithelial cells may be present within benign or malignant mixed tumors of lacrimal glands, with the myoepithelial clonal population staining for muscle actin and variably for keratin⁶, as in our case.

Our case is clinically important for two reasons. First, pleomorphic adenomas may exhibit myoepithelial prominence, or a myoepithelial carcinoma may arise from a previously excised pleomorphic adenoma and complete tumor sampling is required for a correct diagnosis. In this case, the radiologic imaging revealed a heterogeneous mass with both solid and cystic components. Initial incisional biopsy of the more anterior, solid area demonstrated features of a typical pleomorphic adenoma. It was only upon complete excision of the tumor that the myoepithelioma component was identified. Fine needle aspiration biopsy has been shown to be useful tool in confirming the diagnosis of myoepithelial carcinoma.⁸ Second, the clear cell variant of myoepithelioma may be more aggressive than other forms of myoepithelioma and likely more aggressive than pleomorphic adenomas, increasing the importance of correct characterization⁹. Ostrowski et. al.² suggested that myoepitheliomas with clear cells should always be considered a low-grade malignancy making the difference between a diagnosed “myoepithelioma” vs “myoepithelial carcinoma” academic. For this reason, myoepithelial carcinoma cannot be definitively excluded in our case. Complete excision of intact pseudocapsule via lateral orbitotomy is recommended in the cases of myoepithelioma of the lacrimal gland.

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FIG. 1.

(A) Clinical appearance prior to surgery demonstrated a right sided, superotemporal mass. (B) Coronal CT showed a well-circumscribed mass superotemporal to the right globe causing inferior displacement. (C) Axial CT reveals a mass posterolateral to the right globe. The white asterisk denotes a hyperdense area which was the site of the initial incisional biopsy and was histologically consistent with a pleomorphic adenoma. The black asterisk indicates a hypodense area corresponding to the myoepitheliomatous part of the tumor seen after complete excision.

FIG. 2.

(A) Initial incisional biopsy revealed pleomorphic adenoma; (B) epithelial cells line duct-like structures which are surrounded by myoepithelial cells (C) Subsequent excisional biopsy again revealed a pleomorphic adenoma component; (D) epithelial cells lining duct-like structures surrounded by myoepithelial cells; (E) clear cells within trabecular component of the tumor; (F) higher magnification shows large vacuolated cells with displaced nuclei and clear cytoplasm; (G) strong immunoreactivity for cytokeratins AE1,3 in clear cells; (H) focal smooth muscle actin immunoreactivity in clear cells. (hematoxylin and

eosin, A. 25X, B.100X, C. 25X, D. 100X, E. 100X, F. 100X; peroxidase anti-peroxidase G. 100X, H. 100X)

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Table 1**Cases of Myoepithelioma of Lacrimal Gland**

Author (year)	Age/Sex	Cell Type	Diagnosis	Treatment	Outcome
Present	81/F	Clear cell	Myoepithelioma	Total excision	No recurrence at 4 months
Heathcote et. al. (1990) ¹⁰	"middle aged"/F	Spindle cell	Myoepithelioma	Total excision	No recurrence at 15 months
Font et. al. (1992) ¹¹	23/F	Spindle cell	Myoepithelioma	Total excision	No recurrence at 12 years
Ni et. al. (1992) ¹²	Not reported	Spindle cell	Myoepithelioma	Not reported	Not reported
	Not reported	Spindle cell	Myoepithelioma	Not reported	Not reported
Grossniklaus et. al. (1997) ¹³	76/F	Mixed	Myoepithelioma	Total excision	No recurrence at 10 months
Okudela et. al. (2000) ¹⁴	34/M	Mixed	Myoepithelioma *	Total excision	No recurrence at 36 months
Bolzoni et. al. (2005) ¹⁵	46/M	Plasmacytoid	Myoepithelioma	Total excision	No recurrence at 22 months
Pasquale et. al. (2005) ¹⁶	57/F	Epithelioid	Myoepithelioma	Total excision	No recurrence at 24 months
Weis et. al. (2009) ¹⁷	Not reported	Mixed	Myoepithelioma	Not reported	Not reported
Eldesouky et. al. (2014) ¹⁸	Not reported	Not reported	Myoepithelioma	Not reported	Not reported

* possible malignant transformation