

Review article

Pleomorphic adenoma of the lacrimal gland: A review with updates on malignant transformation and molecular genetics

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Abstract

Pleomorphic adenoma (benign mixed tumor) is the most common epithelial neoplasm of the lacrimal gland. It is usually a slow growing, well-circumscribed, mass that is identical to its salivary gland counterpart. Patients generally have an excellent prognosis for vision and long-term survival after complete surgical excision. There is a tendency to reoccur, especially if there is an incomplete excision, and rarely, malignant transformation to carcinoma ex pleomorphic adenoma can occur, which has a much poorer prognosis. The molecular genetics of lacrimal gland pleomorphic adenomas have only recently been studied, but appear to display similar genetic aberration found in the salivary gland counterparts.

Keywords: Pleomorphic adenoma, Lacrimal gland, Malignant transformation, Molecular genetics

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Introduction

The lacrimal gland, an almond shaped, bi-lobed, eccrine secretory gland, approximately 2 cm long that lies in the lacrimal fossa in the supero-lateral part of each orbit, secretes a watery physiologic fluid which lubricates and provides nutrients for the eye and contains the bactericidal enzyme lysozyme.¹ Many different types of neoplasms, including metastases,² can arise within the lacrimal gland, which account for 5% to 25% of all orbital neoplasms.³ Primary lacrimal gland neoplasms often arise in the orbital lobe where the gland attached to the orbital rim about the lacrimal fossa. The most common is the benign mixed tumor or pleomorphic adenoma (PA), a benign mixed tumor consisting of epithelial and mesenchymal components.² These neoplasms characteristically presents with slow, painless enlargement of the lateral portion of the upper eyelid in the 3rd and 4th decades of life, with no clear gender predominance.⁴ Although

benign, these epithelial tumors have a propensity to recur and undergo malignant transformation if incompletely excised, leading to increased morbidity in these patients.⁵

The purpose of this review is to summarize in a multi-faceted manner the data available in the literature regarding the clinicopathologic features, radiographic findings, treatment, malignant transformation, and advances in molecular genetics in patients with lacrimal gland pleomorphic adenomas.

Clinical features

Pleomorphic adenomas (PA) are the most common epithelial tumor to arise within the lacrimal gland. They are usually slow growing and patients commonly present with slight fullness in the temporal upper lid, to those with frank proptosis, diplopia, visual impairment, and eyeball displacement.⁶ Due to the location of the tumor, at or near

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the lacrimal fossa bone, the enlarging tumor characteristically displaces the eye downward and nasally. The duration of symptoms is a very important clinical indicator of a PA. The vast majority of patients with lacrimal gland PAs have had some degree of the aforementioned symptoms for over a year, which correlates with the usual slow growing nature of the tumor. The suspicion for malignancy increases when the symptoms have been less than 10 months.⁶ Additionally, sensory loss and pain are not common symptoms of lacrimal gland PA and when present, should raise concern for a malignant neoplasm such as adenoid cystic carcinoma or mucoepidermoid carcinoma.^{6,7} However, it should be noted that very rare atypical presentations have been reported in the literature and include patients with PAs having presenting symptoms mimicking inflammatory lesions such as orbital cellulitis or painful subcutaneous nodules.^{7,8}

Radiographic imaging

Diagnostic imaging is critical for the clinical diagnosis of lacrimal gland neoplasms. Computed tomography (CT) and magnetic resonance imaging (MRI) scans are ideal, with MRI as the preferred method for visualization of the surrounding bone and examination for intracranial infiltration.⁶ On MRI, pleomorphic adenomas appear as isointense lesions with regular margins and angles.⁴ For a PA, a well circumscribed mass identified within the lacrimal fossa is usually seen (See Fig. 1A and 1B).⁶ Certain radiographic findings may illicit concern for a more aggressive malignancy and these include irregular shaped mass, bone invasion or erosion, molding of the mass to the globe or lateral orbital wall, and calcification.^{6,7} If the radiographic findings and clinical

findings are suggestive of PA, a diagnostic biopsy is not recommended as it may result in tumor recurrence later. A recurrence rate of up to 30% over five years has been reported when biopsy specimens had been taken.⁶ Fortunately, in recent years, the rate of inadvertent biopsy of PAs has dramatically decreased as a result of improved radiologic evaluation.^{7,9}

Treatment

Surgical intervention by lateral orbitotomy is the mainstay of treatment with complete resection of an intact capsule.^{6,10,11} Incomplete capsule removal or defect in the capsule at the time of surgery can lead to significantly high rate of recurrence caused by the displacement of the myxoid component of PA into the orbital cavity.^{2,12} Incisional or needle core biopsies are strictly contraindicated.¹² Radiation, while not commonly used, may be considered for rare pleomorphic adenomas that are inoperable or for recurrent/residual lesions. The prognosis is excellent when the lesion is completely excised with an intact capsule, with a less than 3% recurrence rate after 5 years. Recurrence is even more difficult to treat as they are often multiple and infiltrate normal orbital structures, leaving orbital exenteration as the only therapeutic option in some cases. In very rare instances recurrence in the frontoparietal areas of the brain have been reported in patients with incompletely resected PAs.¹³

Pathological findings

On gross examination, PAs typically have a pseudocapsule surrounding the mass lesion. It is imperative that the capsule

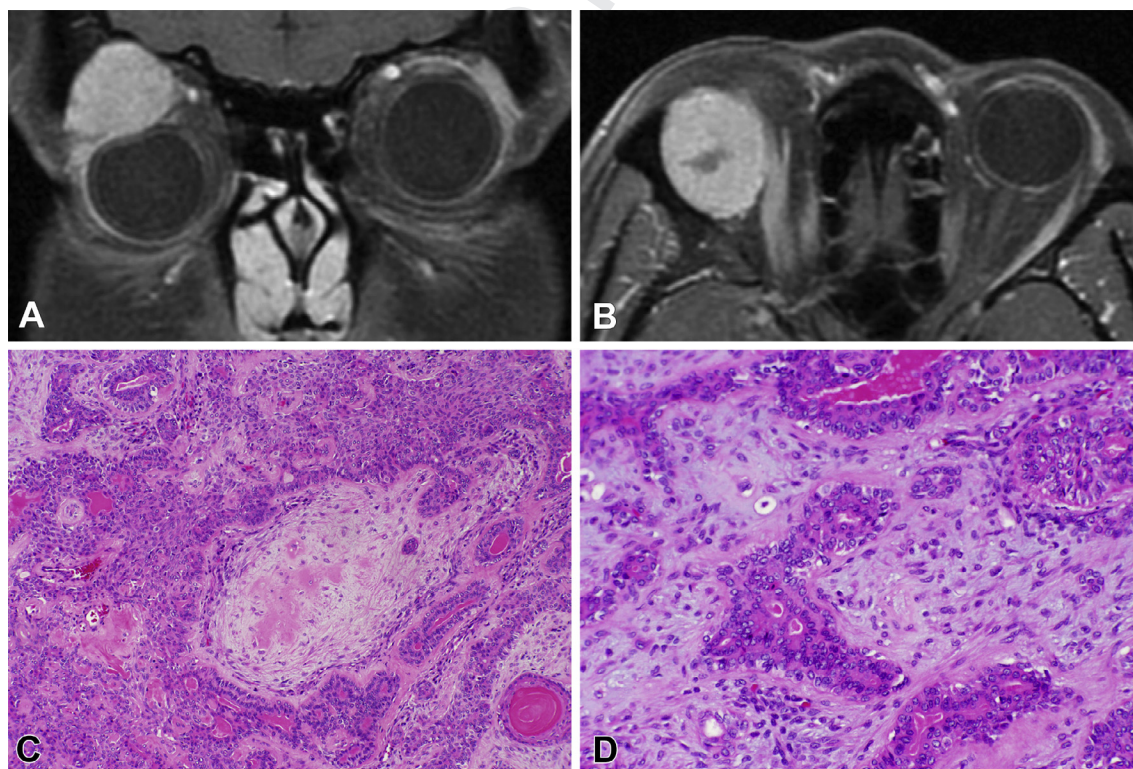


Fig. 1. Images A and B show T1-weighted MRI images demonstrating a well circumscribed superior orbital mass that has mass effect on the right eye. Images C and D are photomicrographs which highlight the biphasic nature of the neoplasm with an epithelial component forming cord and tubules and a chondromyxoid mesenchymal component.

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