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LACRIMAL GLAND NEOPLASIA

The lacrimal glands are paired glandular structures that reside in the superotemporal orbit in a recess of the frontal bone called the lacrimal gland fossa. Their job is to secrete the aqueous layer of the tear film. Tumors of the lacrimal gland represent a small percentage of orbital masses and comprise 6-12% of all orbital spaceoccupying lesions. The typical symptoms of all lacrimal gland tumors are those of facial asymmetry, due to displacement of the globe; swelling of the lacrimal gland; reduced ocular motility; diplopia; and ptosis. The differential diagnosis of lacrimal gland enlargement is very diverse and may include any of the following:

- Neoplasia
 - Epithelial (Pleomorphic Adenoma, Adenoid cystic Carcinoma)
 - Non-epithelial (lymphoma, plasmacytoma, solitary fibrous tumor)
- Inflammatory
 - Sarcoid, IgG4 Disease, Idiopathic Orbital Inflammation, Sjogren Disease
 - Polyarteritis nodosa/Granulomatosis with Polyangiitis
- Infectious
 - o Viral (EBV, Mumps)
 - Bacterial (staphylococcus, gonococcus)

NEOPLASIA

Lymphoproliferative disease of the orbit spans from reactive hyperplasia to malignant lymphoma. Of malignant lymphomas in the lacrimal gland, more than 95% are B-cell origin and 80% are low grade lymphoma. The most common subtype is extranodal marginal zone lymphoma (EMZL)

of mucosal associated lymphoid tissue (MALT) type (Figure 1).

All patients with lymphoma of the lacrimal gland need a full systemic workup, and management requires a multidisciplinary team approach. Radiation therapy is the mainstay of treatment for orbital lymphoproliferative disease and if systemic lymphoma is found then chemotherapy is often initiated.

Around 25% of all lacrimal gland tumors are primary epithelial tumors. The most common tumor is the benign pleomorphic adenoma (PA) that comprises around 50% of epithelial tumors (Figure 2). The remaining 50% of the primary epithelial tumors are malignant. The most frequently encountered is adenoid cystic carcinoma (ACC). Preoperative diagnosis is based on the clinical history and imaging analyses. On CT and MRI imaging pleomorphic adenoma typically appears as a solid, well-defined, round, or oval space-occupying lesion that occasionally shows calcification and remodeling of adjacent bones. This contrasts with malignant tumors that may have irregular margins and signs of bony erosion (Figure 3). By Jeremy Clark, MD

About Jeremy Clark, M.D.

Dr. Jeremy Clark is an Oculofacial Plastic and Orbital Surgeon with UofL Physicians – Eye Specialists. Dr. Clark completed a joint fellowship in Oculofacial Plastic and Orbital Surgery at the University of Louisville and Indiana University. Dr. Clark is the Associate Residency Director for the Department of Ophthalmology and Visual Sciences at UofL School of Medicine



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The oculoplastic and orbital surgery service see patients in Louisville with satellite offices in Elizabethtown and Owensboro, Kentucky and Madison, Indiana. They diagnose and treat many periocular and orbital disease. These disease processes include aesthetic and reconstructive surgery specializing in the face, orbits, eyelids and lacrimal system, specifically: ophthalmic Graves' disease, repair of oculofacial/orbital trauma, development and reconstruction of the anophthalmic socket, correction of eyelid malposition, and tearing/lacrimal outflow issues.



Figure e 1: Extranodal marginal zone lymphoma of the Right Orbit



Figure 2: Pleomorphic Adenoma of the Lacrimal Gland



Figure 3: Imaging suggestive of Adenoid Cystic Carcinoma of the Lacrimal Gland showing lacrimal gland fossa bony erosion

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