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PICTORIAL REVIEW

The forgotten lacrimal gland and lacrimal drainage apparatus: pictorial review of CT and MRI findings and differential diagnosis

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ABSTRACT:

The lacrimal gland is a bilobed serous gland located in the superolateral aspect of the orbit. Lacrimal system pathologies can be broadly divided into pathologies of the lacrimal gland and those of the nasolacrimal drainage apparatus. These include distinct congenital, infectious, inflammatory, and benign, indeterminate, and malignant neoplastic lesions. Trauma and resultant fractures affecting lacrimal drainage apparatus is not part of this review; only non-traumatic diseases will be discussed. CT is the initial modality of choice because of its ability to delineate lacrimal system anatomy and demonstrate most lacrimal drainage system abnormalities and their extent. It also assesses bony architecture and characterizes any osseous changes. MRI is helpful in further characterizing these lesions and better assessing involvement of the surrounding soft tissue structures. In this pictorial review, we will review the anatomy of the lacrimal system, describe CT/MRI findings of the common and uncommon lacrimal system abnormalities and discuss relevance of imaging with regards to patient management.

LACRIMAL SYSTEM ANATOMY

The lacrimal gland is a bilobed serous gland measuring approximately 20 × 12 × 5 mm located in the superolateral aspect of the orbit containing both epithelial and lymphoid tissue.[1] Tears secreted by the lacrimal gland are mixed with mucous secretions produced by the conjunctival and eyelid glands such as the meibomian glands which spread as an oily evaporation-resistant tear film across the globe by the upper and lower eyelids. The drainage of tears occurs through an active pumping mechanism facilitated by the lacrimal drainage system (Figure 1):

(1) The contraction of the orbitalis oculi muscle during eyelids closure leads to the production of positive pressure in the lacrimal sac which subsequently causes closure of the valve of Rosenmüller, resulting in tears being forced down the nasolacrimal duct.

(2) The opening of the eyelids produces negative pressure in the lacrimal sac, causing the opening of the valve of Rosenmüller, which results in tears being drawn through the puncti down into the canaliculi and into the lacrimal sac.

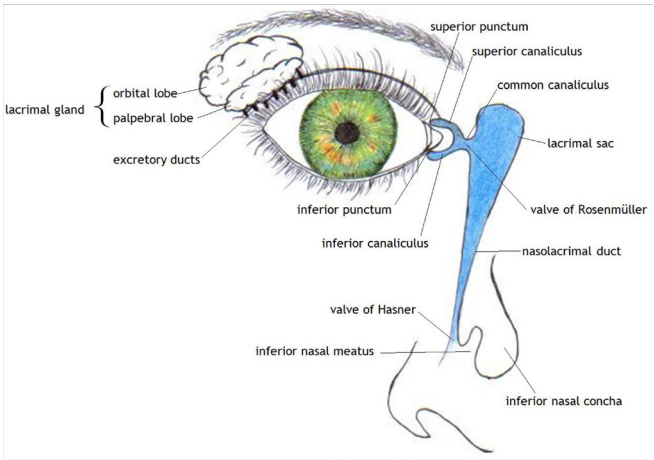
PATHOLOGY

Lacrimal system pathologies can be broadly classified into pathologies of the lacrimal gland and pathologies of the nasolacrimal drainage apparatus (Table 1). We will discuss some of the entities further below, including a variety of common and uncommon lacrimal system pathologies.

A) Pathologies of the Lacrimal Gland

a) Infectious/inflammatory pathologies

Figure 1. Illustration showcasing the lacrimal drainage system and its key structures including the upper and lower puncti, canaliculi, lacrimal sac, and nasolacrimal duct.



i) Dacryoadenitis:

Dacryoadenitis refers to inflammation of the lacrimal gland and affects about 1 in 10,000 ophthalmic patients. Acute infections typically cause diffuse enlargement of the lacrimal gland that often leads to compression of the globe.[2] On CT and MRI, there is enlargement of the lacrimal glands along the superolateral aspect of the orbit with concomitant compression of surrounding orbital structures (Figure 2). Depending on the underlying etiology, potential treatment options include empiric antibiotics, conservative management, or steroids.

ii) Amyloidosis:

Amyloidosis refers to a group of disorders characterized by the extracellular deposition of insoluble protein fibrils.[3] Amyloidosis of the lacrimal gland is rare with about seven cases reported in the literature. On CT, lacrimal gland infiltration typically appears as a homogenous soft tissue mass with occasional areas of calcification in the lacrimal gland. On imaging, extraocular muscle involvement may also be seen (Figure 3). The management of amyloidosis is complex with surgical debulking being the primary modality for the treatment of localized amyloidosis.

b) Benign neoplastic pathologies

i) Dermoid cyst:

Orbital dermoid cysts represent the most common congenital orbital masses comprising closed sacs lined by epithelium derived from the ectoderm.[4] They account for between 3–9% of all orbital tumours. The presence of fat, rim calcification, and scalloping of bone are the common imaging findings on CT (Figure 4). Treatment varies depending on the size, location, and involvement of surrounding orbital structures. Superficial lesions typically require cosmetic excision while deeper ones require a more invasive approach.

ii) Pleomorphic adenoma of the lacrimal gland:

Pleomorphic adenomas represent the most common epithelial neoplasm to originate from the lacrimal gland accounting for 12–25% of all lacrimal tumours.[5] It is slow-growing and well-circumscribed with the enlarging tumour typically displacing the eye downward and nasally. On CT, the tumour appears as a well-circumscribed mass with contrast enhancement (Figure 5). Complete surgical excision of the tumour is recommended as recurrence is high with an incompletely excised tumour.

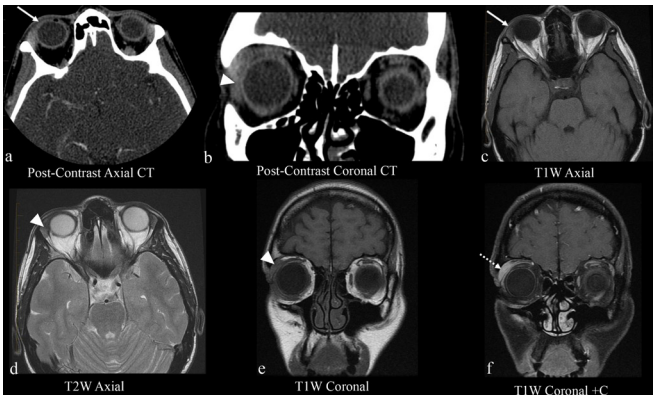
iii) Plexiform neurofibroma:

Plexiform neurofibroma represents an uncommon variant of neurofibroma characterized by a benign tumour of the peripheral nerves, virtually pathognomonic of neurofibromatosis Type 1.[6] Its incidence is estimated to be 1/2500 births per year. On MRI, a deforming mass is typically identified with a varying degree of involvement of surrounding craniofacial structures (Figure 6). Surgical resection of the deforming masses represents the mainstay of treatment.

Table 1. Lacrimal system pathologies

Lacrimal system component	Lacrimal gland	Lacrimal drainage apparatus
Congenital pathologies		Dacryocystocele
Infectious/inflammatory pathologies	Dacryoadenitis Amyloidosis Sarcoidosis Sjögren syndrome	Dacryocystitis Sclerosing orbital pseudotumour Granulomatosis with polyangiitis
Benign neoplastic pathologies	Dermoid cyst Pleomorphic adenoma Plexiform neurofibroma	Capillary hemangioma Dermoid Oncocytic papillary cystadenoma
Malignant neoplastic pathologies	Adenoid cystic carcinoma Follicular lymphoma Leukemic deposits	Lymphoma Leukemic deposits Lacrimal sac squamous cell carcinoma Basal cell carcinoma Metastasis

Figure 2. (a–f) Post-contrast CT (a–b) and MRI (c–f) in a 36-year-old female with dacryoadenitis. Diffuse enlargement of the right lacrimal gland (arrowheads) with homogeneous post-contrast enhancement (dotted arrow) and inflammatory fat stranding (arrows) extending into the soft tissues of the upper eyelid.



c) Malignant neoplastic pathologies:

i) Adenoid cystic carcinoma of the lacrimal gland:

Adenoid cystic carcinoma is the most common form of malignant epithelial lacrimal gland tumour accounting for 60% of all malignant lesions with collective incidence rate of 0.43/million person/year. It represents an extraconal malignant disorder originating usually from the orbital lobe of a lacrimal gland.[7] On CT, adenoid cystic carcinoma typically appears as a homogeneous, unilateral solid mass with associated bone destruction and variable degree of focal dystrophic calcification (Figure 7), reported in 20% of the patients. Current treatment modalities focus on local disease control.

Figure 3. (a–d) Post-contrast CT in a 65-year-old female: Bilateral asymmetric enlargement of the lacrimal glands (dotted arrows) associated with internal calcifications (arrows) caused by amyloidosis.

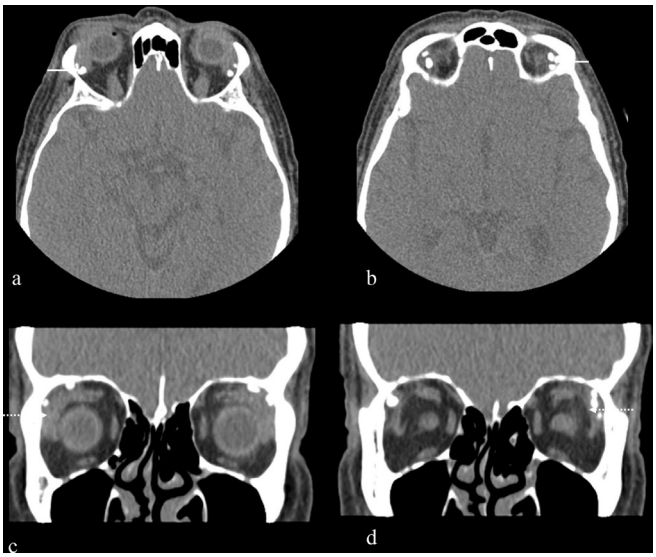
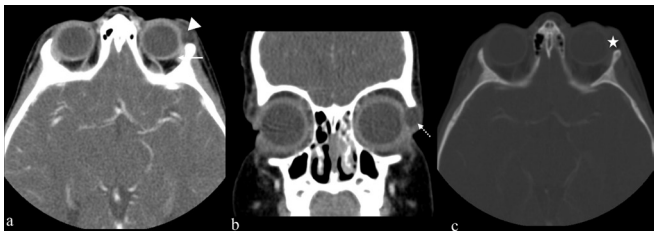


Figure 4. (a–c) Post-contrast CT in 31-year-old female with dermoid cyst. Hypodense round lesion (arrowhead) with subtle peripheral enhancement (dotted arrow), exerting mass effect over the lateral aspect of the left globe and abutting the left lacrimal gland (arrow) with pressure erosion (asterisk) of the adjacent orbital osseous structures.



B) Pathologies of the Lacrimal Drainage Apparatus

a) Congenital

i) Dacryocystocele:

Dacryocystoceles are caused by obstruction of both the proximal and distal ends of the nasolacrimal duct with an incidence of 1 in 3884 live births.[8] Distal nasolacrimal duct obstruction at the valve of Hasner is caused by an imperforate Hasner membrane whereas obstruction more proximally occurs at the valve of Rosenmüller, the cause being less clearly understood. CT is the imaging modality of choice. On CT, nasolacrimal duct dilatation is often seen along with a homogenous, well-defined mass involving the medial canthus or nasal cavity with mild enhancement of lesion wall on post-contrast images and adjacent soft tissue (Figure 8). Treatment of dacryocystoceles ranges from applying manual pressure or probing with irrigation to endoscopic resection in more severe cases.

ii) Dacryocystitis:

Dacryocystitis is inflammation of the nasolacrimal sac related to impaired drainage through the lacrimal drainage system and superimposed infection.[9] It has an incidence of 15/10,000 patients. On CT, they are well-circumscribed

Figure 5. (a, b) Post-contrast CT in a 64-year-old female with pleomorphic adenoma (biopsy proven). Enhancing soft tissue mass lesion (arrows) in the superolateral aspect of the left orbit arising from the lacrimal gland.

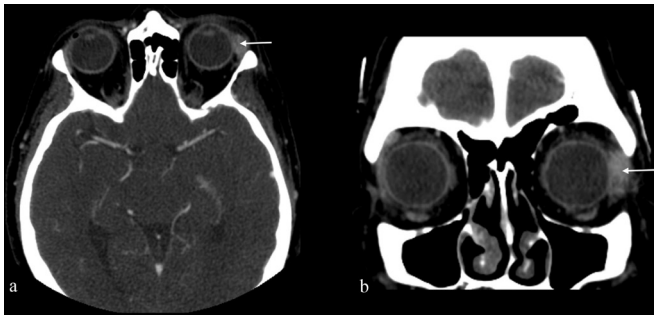


Figure 6. (a–c) MRI in a 29-month-old with plexiform neurofibroma. Fusiform mass lesion (arrows) extending into the right orbit along the lateral wall, involving the right lateral rectus muscle, superior, and oblique muscles as well as the lacrimal gland.

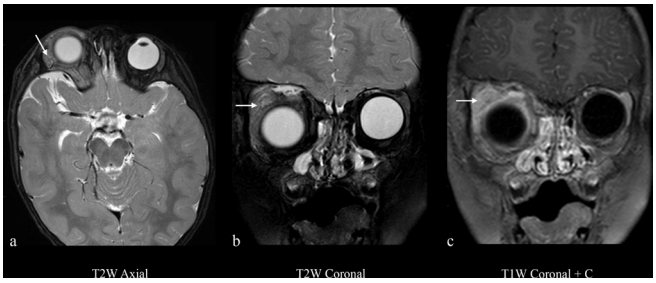
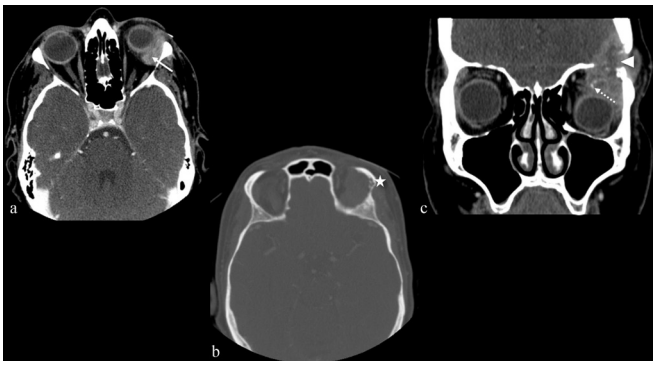


Figure 7. (a–c) Post-contrast CT in a 53-year-old female with adenoid cystic carcinoma of the lacrimal gland. Large locally invasive (arrowhead) solid partially calcified (dotted arrow) enhancing (arrow) lacrimal gland lesion associated with bone destruction (asterisk).



round lesions with peripheral enhancement around the inner canthus with adjacent soft tissue thickening and fat stranding with associated complications like pre/post septal cellulitis (Figure 9) and abscess formation. Imaging allows us to distinguish between dacryocystitis with pre-septal cellulitis (treated medically with oral antibiotics), post-septal

Figure 8. (a, b) Post-contrast CT in an 18-day-old infant with dacryocystocele: Rounded, well-defined, rim-enhancing lesion (dotted arrow) abutting the medial preseptal right orbit, extending down the lacrimal canal into the inferior meatus (arrow), and obstructing the nasal cavity.

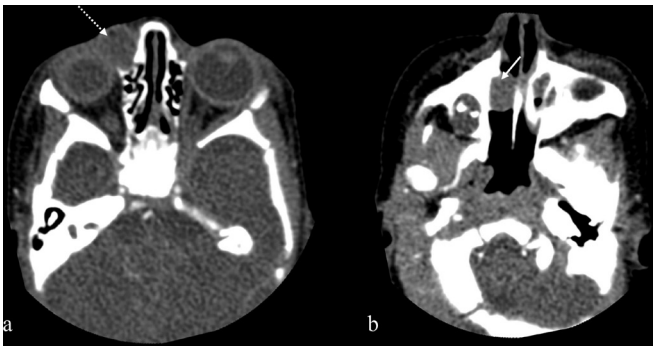
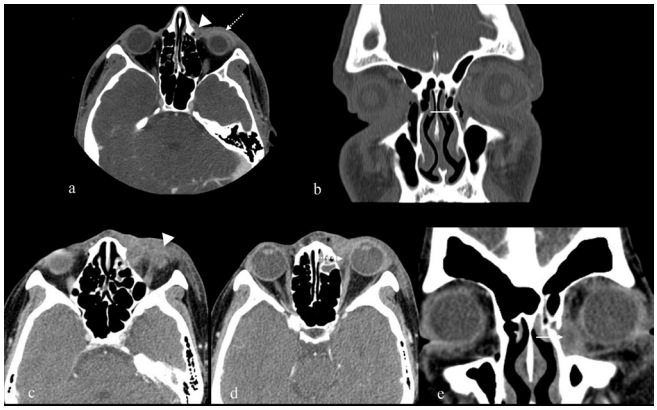


Figure 9. (a, b) Post-contrast CT in an 18-year-old male with dacryocystitis. Soft tissue swelling (arrowhead) centered over the medial canthus of the left orbit associated with preseptal fat stranding (dotted arrow) and inflammatory changes in the left nasolacrimal duct (arrow), in keeping with dacryocystitis and preseptal cellulitis. (c, d, e) Post-contrast axial and coronal CT in a 47-year-old female with left preseptal (arrowhead) and post septal extraconal cellulitis (dotted arrow) and dacryocystitis (arrow).



orbital inflammation (treated more aggressively medically with IV antibiotics), and abscess (managed surgically). b) Infectious/Inflammatory pathologies

i) Sclerosing orbital pseudotumour:

Orbital pseudotumour, accounts for 8–11% of all orbital tumours. It is an idiopathic inflammatory condition that commonly involves the extraocular muscles, mostly associated with autoimmune conditions.[10] On MRI, pseudotumours are iso- to hypointense on T_2 weighted images with

Figure 10. (a–e) MRI in a 37-year-old female with sclerosing orbital pseudotumour. Homogenously enhancing (arrowhead) soft tissue mass lesion in the medial canthus of the right orbit with expansion of the nasolacrimal duct. The mass extends into the nasal cavity, obliterating the right osteomeatal complex (dotted arrow) and into the intraconal orbital compartment (arrow), with thickening of the medial and inferior rectus muscles.

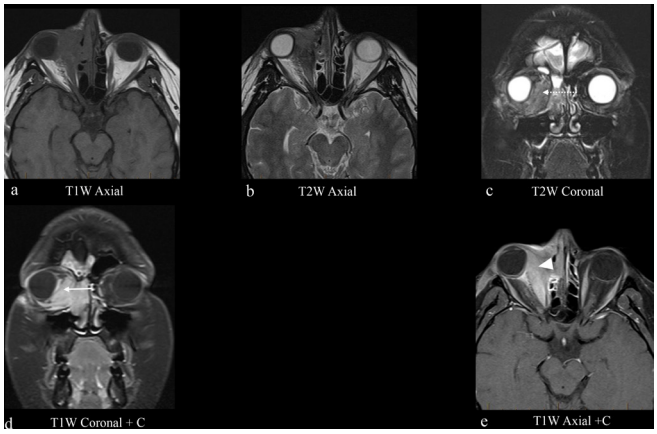


Figure 11. (a–c) MRI in a 26-year-old female with granulomatosis with polyangitis. Soft tissue thickening of the left medial canthus (arrows) with extensive changes in the nasal cavity and paranasal sinuses, including septal perforation (dotted arrow), provide clues to the diagnosis.

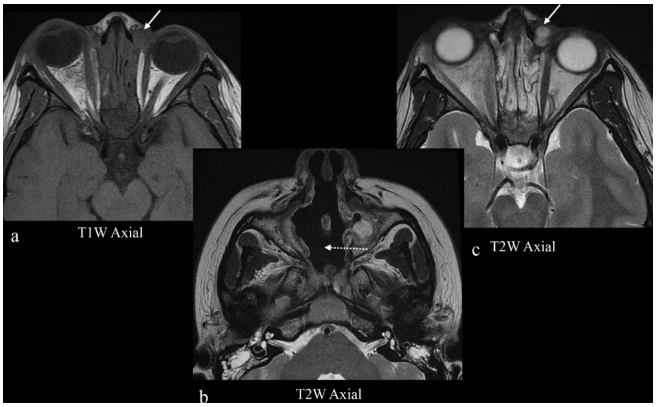
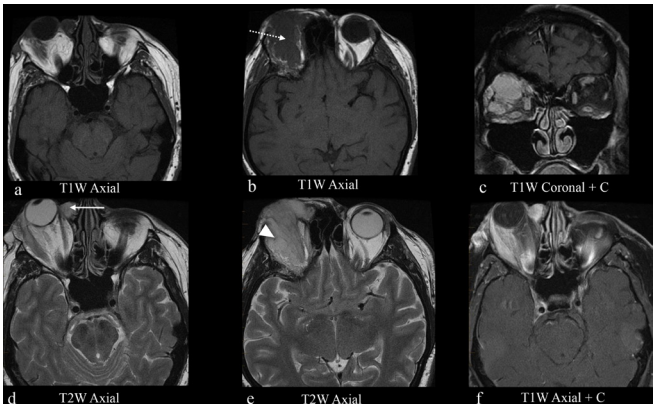


Figure 12. (a–f) MRI of the orbit in a 54-year-old male with capillary hemangioma. Avidly enhancing (arrowhead) large right orbital mass (dotted arrow) lesion involving the extraconal orbital compartment as well as the lacrimal sac and lacrimal duct (arrow).

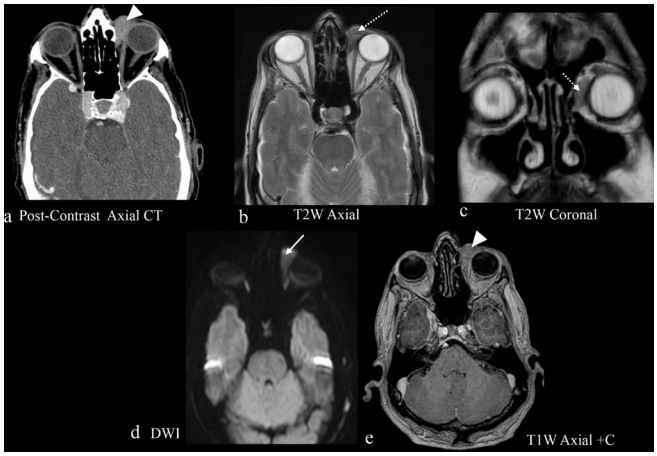


loss of fat between the involved muscles and periosteum of the involved orbital wall (Figure 10). Restricted eye movements due to pain, diplopia, and proptosis are common clinical manifestations. Differential diagnosis includes Grave's disease, lymphoma, rhabdomyosarcoma, and metastasis. Most cases resolve rapidly after treatment with corticosteroids.

ii) Granulomatosis with polyangiitis:

Granulomatosis with polyangiitis represents a form of necrotizing, non-caseating granulomatous vasculitis that affects the small-to-medium-sized blood vessels in the body which results from an immune-mediated response to vascular injury.[11] It has an incidence of 8–10 cases/million with orbital involvement reported in about 50% of patients. On MRI, granulomatosis with polyangiitis involvement of the lacrimal gland appears hypointense on T_2 weighted images with associated soft tissue thickening (Figure 11).

Figure 13. (a–e) CT (a) and MR (b–e) in a 68-year-old male with orbital lymphoma. Well-defined homogenously enhancing (arrowheads) soft tissue mass with low signal on T_2W images (dotted arrows) and restricted diffusion on DWI (arrow) in the left medial canthus.



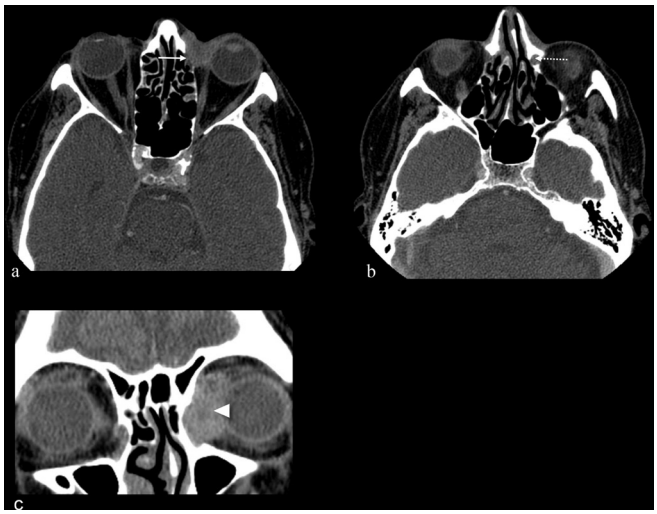
Additional MRI findings including osseous erosion and septal perforation are important findings that aid in the diagnosis Treatment entails systemic immunosuppressive therapy with cyclophosphamide, methotrexate, and/or steroids along with biologics such as rituximab.

c) Benign neoplastic pathologies

i) Capillary hemangioma:

Capillary hemangiomas of the orbit represent the most common orbital tumours of infancy. They are typically

Figure 14. (a–c) Post-contrast CT in a 75-year-old male with squamous cell carcinoma of the lacrimal sac. Heterogeneously enhancing soft tissue (arrowhead) along the medial wall of the left orbit with destruction of the lamina papyracea (arrow), widening of the nasolacrimal duct (dotted arrow), and infiltration of the left globe along the insertion of superior oblique and medial rectus muscles.



located anterior to the globe within the eyelid, manifesting at birth or shortly thereafter.[12] On MRI, the lesions appear mildly T1 hypointense and T2 iso- to hyperintense and demonstrate homogeneous enhancement with enhancing intratumoural vessels (Figure 12). The lesions tend to be self-limiting and injection or systemic administration of corticosteroids is usually reserved for cases with compromised vision.

d) Malignant neoplastic pathologies

i) Lymphoma:

Orbital lymphoma represents one of the most common orbital tumours and originates from mucosa-associated lymphoid tissue as non-Hodgkin B-cell lymphoma.[13] They constitute about 50% of all primary orbital malignancies in adults. On contrast-enhanced CT, there is mild to moderate homogenous enhancement of the lesions. On MRI, lesions appear iso- to hypointense on T_1 and iso- to hyperintense on T_2 weighted images with restricted diffusion on DWI (Figure 13). Treatment entails a combination of surgical resection, radiotherapy, and chemotherapy.

ii) Lacrimal sac squamous cell carcinoma:

Lacrimal sac squamous cell carcinoma represents the most common histological subtype of nasolacrimal tumours which as a group are relatively uncommon.[14] About 775 cases have been reported in the literature. On CT, squamous cell carcinomas of the lacrimal sac appear as soft tissue lesions involving the medial canthus region with bony destruction, widening of nasolacrimal duct and infiltration of the globe (Figure 14). Treatment typically entails wide local resection followed by chemotherapy and/or radiotherapy.

CONCLUSION

The lacrimal apparatus can be involved by a wide range of pathologies. Knowledge of disease entities and careful examination of the images is key to successful patient management.

This pictorial review from our institutions familiarizes radiologists with the anatomy of the lacrimal apparatus and CT/MRI imaging features of common and uncommon non-traumatic pathologies of the lacrimal gland and lacrimal apparatus.

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