# The Radiological Spectrum of Orbital Pathologies that Involve the Lacrimal Gland and the Lacrimal Fossa

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CT and MRI are utilized to differentiate between different types of masses and to determine the extent of lesions involving the lacrimal gland and the fossa. Although many diseases that affect the lacrimal gland and fossa are specifically diagnosed by imaging, it is frequently very difficult to differentiate each specific disease on the basis of image characteristics alone due to intrinsic similarities. In lacrimal gland epithelial tumors, benign pleomorphic adenomas are seen most commonly with a well defined benign appearance, and a malignant adenoid cystic carcinoma is seen with a typical invasive malignant appearance. However, a malignant myoepithelial carcinoma is seen with a benign looking appearance. Lymphomatous lesions of the lacrimal gland include a broad spectrum ranging from reactive hyperplasia to malignant lymphoma. These lesions can be very difficult to differentiate both radiologically and pathologically. Generally, lymphomas tend to occur in older patients. The developmental cystic lesions found in the lacrimal fossa such as dermoid and epidermoid cysts can be diagnosed when the cyst involves the superior temporal quadrant of the orbit and manifests as a non-enhancing cystic mass and, in case of a lipoma, it is diagnosed as a total fatty mass. However, masses of granulocytic sarcoma and xanthogranuloma, as well as vascular masses, such as a hemangiopericytoma, are difficult to diagnose correctly on the basis of preoperative imaging findings alone. A careful clinical evaluation and moreover, a pathologic verification, are needed. In this pictorial review, the various imaging spectrums of pathologic masses involving the lacrimal gland and fossa are presented, along with appropriate anatomy and pathology reviews.

**Index terms:**
Orbit, lacrimal gland
Magnetic resonance (MR)
Computed tomography (CT)

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Clinically, the lesions of the lacrimal gland and fossa are found as palpable masses in the superior lateral aspect of the orbit, and these lesions constitute about 5–13% of all of the orbital masses confirmed by biopsy (1). Many different pathological entities arise from the lacrimal gland and fossa, and as each of them requires a different therapeutic approach, the radiological characterization of each lesion is important. Approximately, half of the lacrimal gland masses are tumors of epithelial origin and the rest are lesions that arise from lymphoid or inflammatory diseases. Pseudotumors, metastatic masses and developmental cysts may also occur in the lacrimal fossa (2). This paper aims to discuss the radiologic findings of the lesions that are found in the lacrimal gland and fossa, and to aid physicians in the differential diagnoses of these various diseases.

## NORMAL ANATOMY

The lacrimal gland is located in the superior lateral aspect of the orbit, within the lacrimal fossa adjacent to the superior and lateral rectus muscles (Fig. 1). It is roughly

the same size and shape as an almond and extraconal in position and it extends deep into the orbital septum. It consists of the anterior palpebral and deeper orbital lobes, which are demarcated by the lateral horn of the aponeurosis of the levator palpebrae superioris.

The anterior palpebral lobe is approximately one-third to one-half the size of the deeper orbital lobe, and most of the lacrimal gland tumors arise in the orbital lobe.

## PATHOLOGICAL ABNORMALITIES

Half of the masses found in the lacrimal gland and fossa are epithelial tumors, of which half are pleomorphic adenomas and the rest are carcinomas. Adenoid cystic carcinomas are the most common malignancy found, and others seen, in order of decreasing frequency are as follows: pleomorphic, mucoepidermoid, adenocarcinoma,

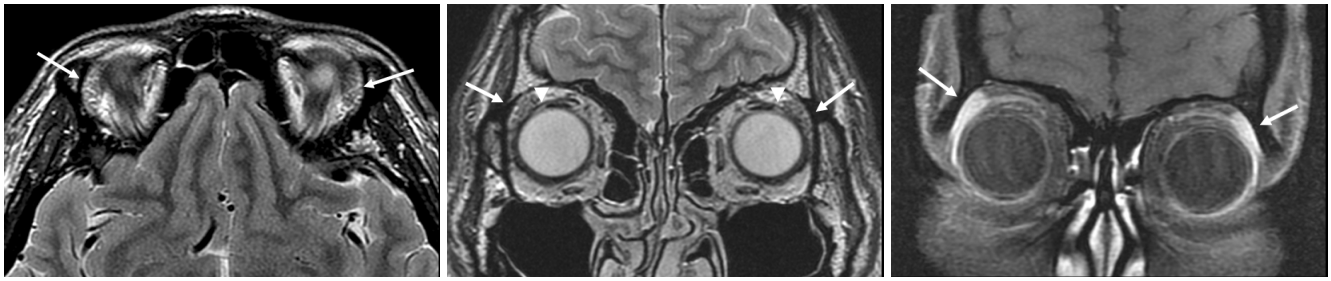


**Fig. 1.** The normal anatomy of the lacrimal gland and fossa.
**A.** A transverse T2-weighted MR image at the level of the lacrimal gland shows almond-shaped lacrimal glands at the superior lateral aspect of both eyeglobes (arrows).
**B.** A coronal T2-weighted MR image at the level of the lacrimal gland and aponeurosis of the levator palpebrae muscle shows continuous extension of the aponeurosis into the lacrimal gland, which divides the gland into anterior palpebral and deeper orbital lobes (arrowheads).
**C.** A coronal enhanced T1-weighted MR image shows well-enhanced lacrimal glands (arrows).

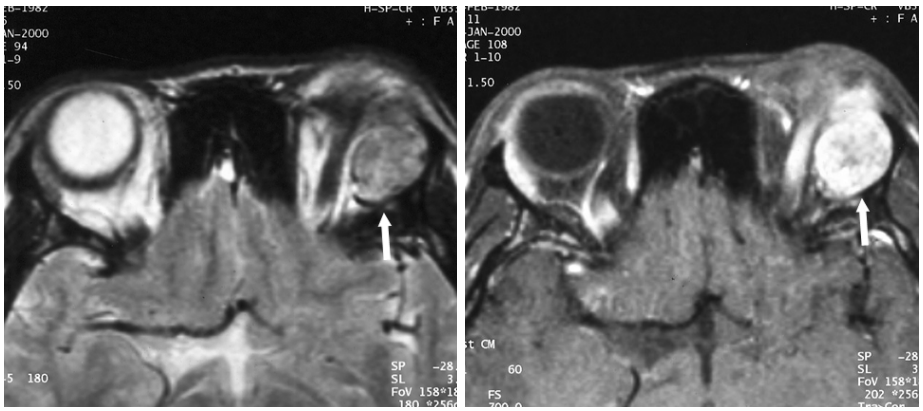


**Fig. 2.** A pleomorphic adenoma in a 44-year-old man.
**A.** A well-circumscribed, spherical tumor is seen in the left lacrimal fossa with heterogeneous intermediate signal intensity on a T2-weighted image (arrow).
**B.** On a gadolinium enhanced T1-weighted image, strong enhancement is noted in the mass (arrow).

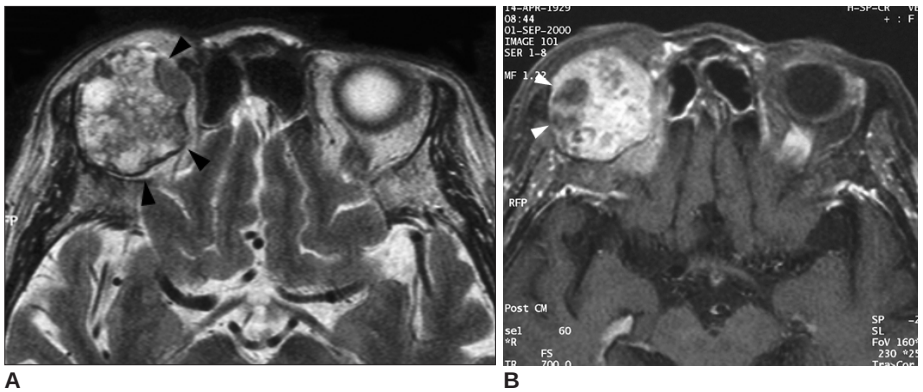


**Fig. 3.** A pleomorphic adenoma in a 63-year-old woman.
**A.** A well-circumscribed mass is seen in the right lacrimal fossa with heterogenous high signal intensity on T2-weighted images (arrowheads).
**B.** On a gadolinium enhanced T1-weighted image, the tumor shows strong enhancement with poorly enhanced areas (arrowheads), representing cystic changes.

squamous cell and undifferentiated carcinoma. In many cases, the carcinoma will occur secondary to a pleomorphic adenoma as a carcinoma ex pleomorphic adenoma.

The remaining half of the masses includes lymphoid and inflammatory lesions, and it is possible to have a wide spectrum of diseases ranging from benign dacryoadenitis to malignant lymphomas. The other lesions seen are dermoid cysts, which arise from the epithelial rest of the lacrimal fossa, and intrinsic epithelial cysts, which are formed by dilatation of the lacrimal duct. Metastases from breast or lung cancers may involve the lacrimal gland, although this is rare. Pseudotumors are nonspecific inflammatory mass lesions of the orbital tissue, and they may also be found in the lacrimal region (3).

## LACRIMAL GLAND EPITHELIAL TUMORS

### *Pleomorphic Adenoma (Benign Mixed Tumor)*

A pleomorphic adenoma is the most common tumor of the lacrimal gland, comprising 53% of all epithelial tumors. On CT, these lesions appear as a variety of attenuations depending on the composition and cellularity of the tumors. Highly cellular masses are homogeneous, while less cellular tumors tend to show non-homogeneous attenuation due to mesenchymal stroma, cystic degeneration, necrosis, or serous or mucous collection (4). Small tumors appear as relatively homogeneous lesions of low signal intensities on T1-weighted images and appear as high signal intensities on T2-weighted images, while the large tumors with hemorrhage or necrosis exhibit heterogeneous signal intensities on T1- and T2-weighted images (Figs. 2, 3).

### *Adenoid Cystic Carcinoma*

Adenoid cystic carcinoma (ACC) is the most common malignant epithelial tumor of the lacrimal gland, and it accounts for 25–40% of all epithelial tumors. The histologic findings for lacrimal gland ACC are similar to those seen for a salivary gland ACC. Lacrimal gland ACC is characterized by frequent bone and perineural invasion or extraorbital spread, and it is known to be associated with a poorer prognosis than salivary gland ACC (Fig. 4). According to Mizokami and colleagues, the 10-year survival rate of salivary gland ACC is 40–50%, whereas the lacrimal gland ACC survival rate is 20%, despite extensive surgery and postoperative radiation therapy (5).

### *Myoepithelial Carcinoma*

Myoepithelial cells are mesodermal cells of ectodermal derivation. They assist lacrimal gland secretion by contraction of intracytoplasmic myofibrils. Myoepithelial cells are found in many benign and malignant tumors, but very few tumors are composed solely of these cells. Myoepithelial carcinomas present as slow growing, painless masses and exhibit clinical findings similar to those of pleomorphic adenomas. On CT, this tumor appears as an enhancing mass with an indistinct margin and destruction of the surrounding bone is seen (Fig. 5), and on MRI, it exhibits nonspecific low T1- and high T2-signal intensities (4).

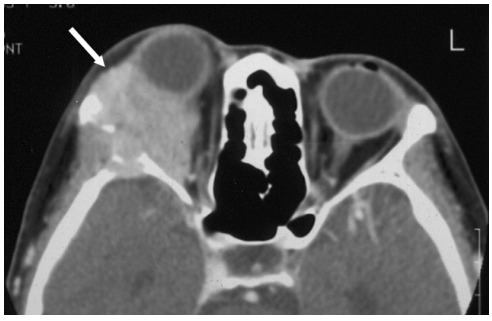



**Fig. 5.** A myoepithelial carcinoma in a 32-year-old man.
A contrast enhanced CT scan shows a well-enhancing tumor (arrow) in the right lacrimal fossa with bony destruction of zygomatic and sphenoid bone.

**A**

**B**

**Fig. 4.** An adenoid cystic carcinoma in a 55-year-old man.
**A.** Contrast enhanced CT scan shows a well-enhancing soft tissue mass (arrow) in the left lacrimal fossa with destruction of the lateral orbital wall.
**B.** A gadolinium enhanced T1-weighted image shows intracranial extension into the middle cranial fossa with dural enhancement (arrowheads).

## LACRIMAL GLAND LYMPHOID TUMORS

Primary orbital lymphomas represent approximately 5 – 14% of the extranodal lymphomas. Within the orbit, these tumors arise from the conjunctivae, eyelids, or retrobulbar tissue; lacrimal gland involvement is rare (Fig. 6). An orbital lymphoma usually occurs in older people, and the average age for occurrence of this tumor is in the sixth decade of life. Lymphoid tumors are part of a disease spectrum that includes benign lymphoid hyperplasia (Fig. 7), atypical lymphoid hyperplasia and malignant lymphoma. There is a tendency towards bilateral involvement. Whereas most lacrimal gland tumors develop in the deeper orbital lobe, lacrimal lymphomas involve the entire lacrimal gland diffusely, including the palpebral lobe. This tumor can surround the eye globe and can have a concave pancake appearance (6).

## LACRIMAL GLAND INFLAMMATORY DISEASES

### *Acute and Chronic Dacryoadenitis*

Inflammatory diseases of the lacrimal gland may be categorized into acute and chronic inflammation. Acute dacryoadenitis commonly develops in children and young adults. It is mostly unilateral and responds rapidly to treatment.

Inflammatory diseases of the lacrimal gland can cause diffuse oblong enlargement of the gland. On imaging, acute dacryoadenitis lesions show marked contrast enhancement that includes the surrounding tissue, and this may be accompanied with myositis of the lateral rectus muscle. In the presence of scleritis, the findings may include fluid collection in Tenon's space and uveoscleral enhancement (6).

Chronic dacryoadenitis may occur after an acute

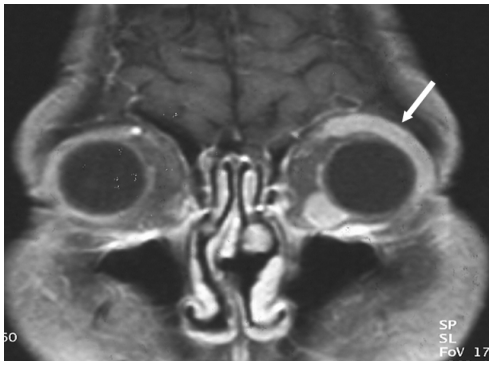



**Fig. 6.** A malignant lymphoma in a 41-year-old woman.
A gadolinium enhanced T1-weighted coronal image shows an elongated soft tissue mass (arrow) along the superior lateral aspect of the left eye globe. This lesion shows homogeneous and strong enhancement. Another enhancing nodule is noted at the inferior medial aspect of the left eye globe.

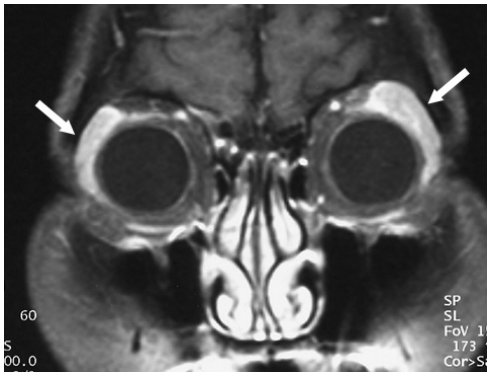



**Fig. 8.** Chronic dacryoadenitis in a 39-year-old woman.
A gadolinium-enhanced T1-weighted coronal image shows diffuse and elongated enlargement of both lacrimal glands with contrast enhancement (arrows).

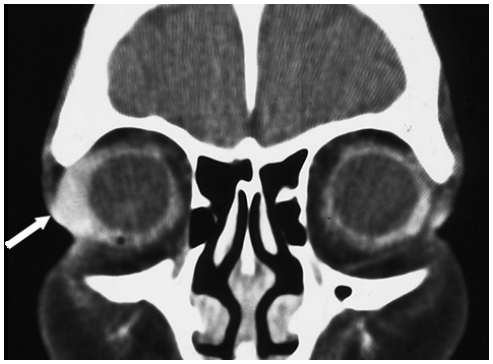


**Fig. 7.** Lymphoid hyperplasia in a 43-year-old man.
A contrast enhanced coronal CT scan shows a homogeneously-enhancing elongated mass involving the right lacrimal gland (arrow).

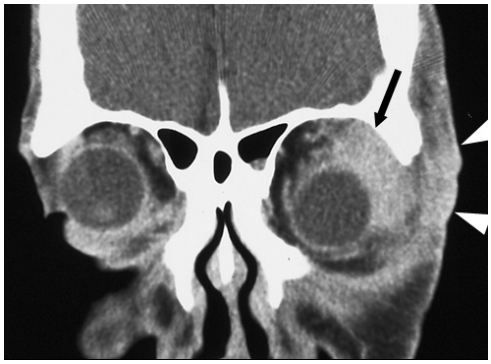


**Fig. 9.** An inflammatory pseudotumor in a 54-year-old woman.
A contrast-enhanced CT scan shows an ill-defined enhancing mass in the superior lateral aspect of left eye globe (arrow). Subcutaneous infiltrations (arrowheads) are accompanied in the lateral outside of the left orbit.

infection, or it may appear in various conditions such as sarcoidosis, tuberculosis, thyroid ophthalmopathy, Mikulicz's syndrome, sclerosing pseudotumor and Wegener's granulomatosis. It manifests as bilateral symmetric or asymmetric lacrimal gland enlargement (Fig. 8).

### *Inflammatory Pseudotumor*

An inflammatory pseudotumor is a peculiar idiopathic inflammatory process characterized by a polymorphous lymphoid infiltrate with various degrees of fibrosis (7). On MR images, inflammatory pseudotumors are usually isointense to hypointense relative to muscle on T1-weighted images with a relatively hypointense T2 signal as compared with most other tumors. Variable contrast enhancement has been reported (Fig. 9).

## MISCELLANEOUS LESIONS INVOLVING THE LACRIMAL FOSSA

### *Developmental Orbital Cysts (Dermoid and Epidermoid Cysts, Teratoma)*

Common developmental cysts found in the orbit and periorbital structures are dermoid cysts, epidermoid cysts and teratomas. Such lesions arise from an inclusion of an ectodermal element during neural tube closure, in which dermal elements become trapped in the suture line, diploe, meninges or scalp. Dermoid and epidermoid cysts are most frequently found in the superior temporal quadrant of the orbit adjacent to the zygomaticofrontal suture line.

Most of these lesions are slow-growing, but some may show rapid growth in adults. Dermoid and epidermoid cysts are seen as non-enhancing low-density masses on CT images. Dermoid cysts may show fat densities and calcification (Fig. 10). On MRI, they are seen as low signal intensities on T1-weighted images and high signal intensities on T2-weighted images, and they can exhibit characteristic fat signal intensity (4).

Epidermoid cysts may exhibit similar imaging features, but calcification is not a feature of epidermoid cysts (Fig. 11).

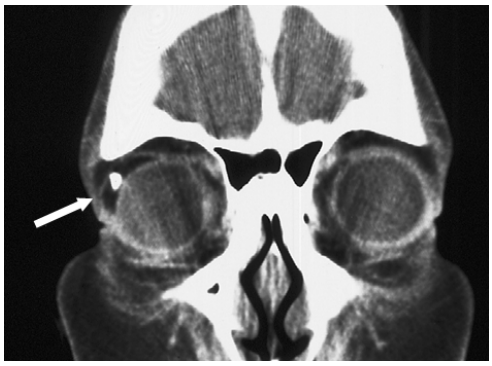


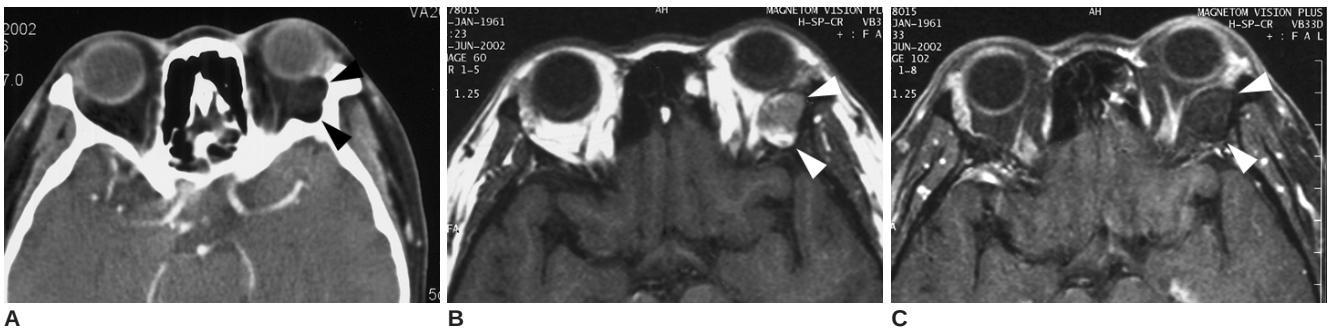
**Fig. 10.** A dermoid cyst in a 49-year-old woman.
A coronal CT scan shows a fat-containing mass with nodular calcification in the superior lateral aspect of right orbit (arrow).

### *Hemangiopericytoma*

Hemangiopericytomas are rare vascular tumors that develop from the pericytes of Zimmermann, which normally envelop the capillary and postcapillary venules of tissues. They are malignant in about 50% of cases, and although infrequent, hematogenous or lymphatic metastases may be present. They tend to recur without complete excision, and wide surgical excision is necessary. In contrast to cavernous hemangiomas, the margins of orbital hemangiopericytomas are indistinct and the tumors appear to invade the surrounding tissue (4). Erosion of adjacent bony structures may be observed, and hemangiopericytomas display marked contrast enhancement (Fig. 12).

### *Lipoma*

A lipoma of the orbit is a rare entity. The most common soft tissue sarcoma, the liposarcoma, may involve the



**A** **B** **C**

**Fig. 11.** An epidermoid cyst in a 35-year-old man.
**A.** A contrast enhanced axial CT scan shows a fat-containing mass with bony scalloping at the lateral orbital wall (arrowheads).
**B.** On a T1-weighted axial image, this mass shows heterogeneous intermediate to high signal intensity (arrowheads).
**C.** On a contrast-enhanced, fat-suppressed T1-weighted image, no enhancement is noted in the mass (arrowheads).

orbit. MRI is specific in the diagnosis of a simple lipoma when a grossly fatty mass is observed, which has few or no septa, and minimal or no areas of enhancement. A lipoma shows characteristic findings on CT with fat attenuation (Fig. 13).

*Granulocytic Sarcoma*

The orbital granulocytic sarcoma is a localized tumor composed of cells of myeloid origin. This tumor may cause progressive proptosis before or after the onset of systemic leukemia. The patterns of orbital involvement have been described in one study (8).

The uvea, choroids, retina and optic nerve may first be involved, and this involvement appears as a diffuse or localized thickening on the images. In a case where orbital soft tissue is involved, this tumor primarily involves the orbital fat and it can extend to the lacrimal gland. On MRI imaging, it shows almost the same intensity as muscle on a T1-weighted image, while it is observed with greater intensity on a T2-weighted image. For our case, the granulocytic sarcoma involved the lacrimal gland and adjacent fat (Fig. 14).

*Xanthogranuloma*

Periorbital xanthogranulomas are rare entities that occur both in children and adults. The diagnosis may be inferred from the characteristic macroscopic appearance of diffuse, yellow, plaque-like masses in the eyelid. Xanthogranuloma may be associated with a wide spectrum of hematological abnormalities including paraproteinemia, plasmacytosis, leukopenia, cryoglobulinemia or complement deficiency. There is also a recognized association between xanthogranulomas and malignancies such as multiple myeloma and non-Hodgkin's lymphoma.

Among the nine cases of periorbital xanthogranuloma five cases involved the lacrimal gland, but only one case showed bilateral enlargement of the lacrimal glands (9) (Fig. 15). It is difficult to distinguish xanthogranuloma from other diseases by the use of imaging modalities only, as its pathologic feature of granuloma formation is distinguished by prominent histioxanthomatous infiltration.

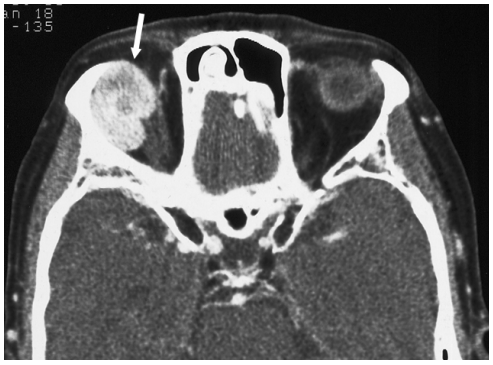


**Fig. 12.** A hemangiopericytoma in a 65-year-old man.
A contrast-enhanced axial CT scan shows a strongly enhancing mass with a lobulated contour in the superior lateral aspect of the right orbit (arrow).

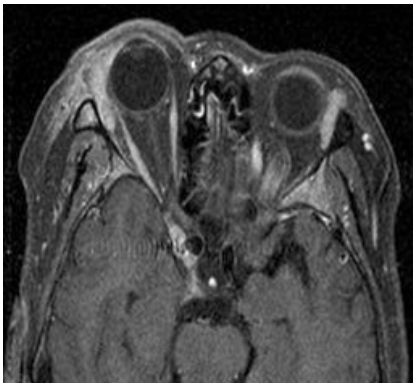


**Fig. 14.** A granulocytic sarcoma from chronic myeloid leukemia in a 40-year-old man. A gadolinium-enhanced T1-weighted axial image shows bilaterally-enlongated masses with strong enhancement in the superior lateral aspect of both orbits. Soft tissue infiltrations are noted around the right orbit mass.

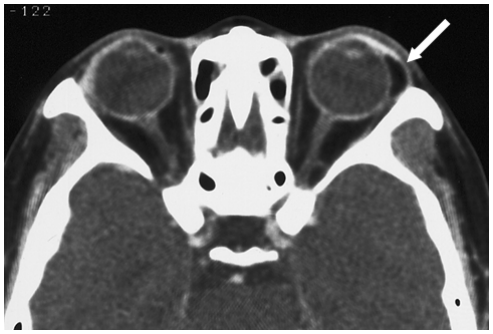


**Fig. 13.** A lipoma in a 64-year-old man.
An axial CT scan shows a fat-containing mass at the superior lateral aspect of the left orbit (arrow).

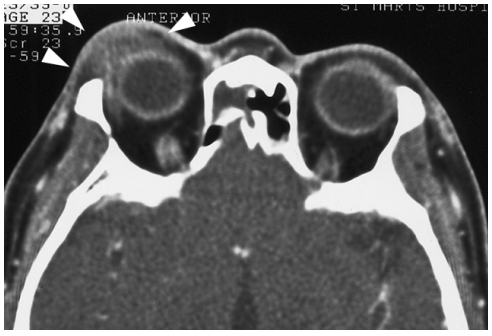


**Fig. 15.** Xanthogranuloma in a 62-year-old woman.
A contrast enhanced axial CT scan shows poorly enhancing, soft tissue density infiltrations at the anterior lateral aspect of the right orbit (arrowheads).

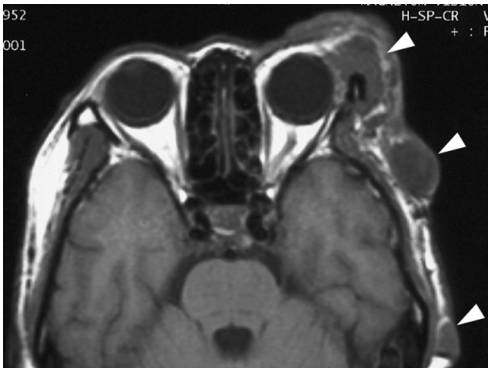


**Fig. 16.** Plexiform neurofibromatosis in a 66-year-old man.
The T1-weighted axial image shows multiple lobulating masses involving the left lacrimal fossa and the subcutaneous layer of the left temporal region (arrowheads).

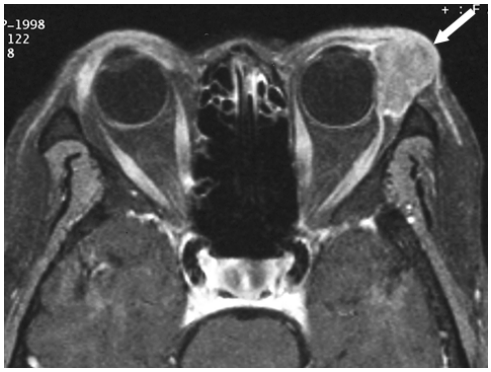


**Fig. 17.** Squamous cell carcinoma of the conjunctiva in an upper eyelid of a 61-year-old man.
A gadolinium enhanced T1-weighted axial image shows a well-enhanced lobulated mass in the superior lateral aspect of the left eye globe (arrow). The thickened skin is not separable from the mass.

*Neurofibroma*

Head and neck neurofibromas are usually plexiform tumors, and are a characteristic feature of neurofibromatosis type I. They are poorly delineated, diffusely infiltrating masses that show bone erosion and foraminal extension (4). On a T1-weighted image these lesions are mostly isointense signal intensities and they appear as hyperintense lesions on T2-weighted images. After the administration of contrast material, the lesions show moderate enhancement (Fig. 16).

*Squamous Cell Carcinoma of Conjunctiva*

Squamous cell carcinoma arising from the conjunctiva commonly occurs in the interpalpebral portion or limbus, and extends to the cornea (Fig. 17). Infrequently, it may extend to the eye and orbit, mimicking a lacrimal gland mass.

## CONCLUSION

In the lacrimal gland, various epithelial tumors are found with the most common being a pleomorphic adenoma and a spectrum of lymphoid tumors can also develop. As an inflammatory disease, acute and chronic dacryoadenitis and an inflammatory pseudotumor can develop. Surrounding the lacrimal gland, in the lacrimal fossa various diseases can develop including dermoid and epidermoid cysts as well as a developmental cyst, a vascular mass such as a hemangioma and a hemangiopericytoma, lipomas, granulocystic sarcomas, xanthogranulomas, and neurogenic tumors. Many diseases affecting the lacrimal gland and fossa display disease-specific characteristics on images which are helpful in diagnosing these diseases. Nevertheless, we needed pathologic proof for a correct diagnosis, due to the fact that the disease spectrum involving the lacrimal gland and lacrimal fossa can show similar characteristics on imaging. By reviewing the various disease categories and the radiologic findings involving the lacrimal gland and lacrimal fossa, we hope to contribute our experiences and expertise to the radiologist and physician to assist them in making more accurate diagnoses.

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