Lacrimal Gland Masses

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Lacrimal gland lesions generally present as palpable masses in the superolateral aspects of the orbits. Approximately 50% of lacrimal gland masses are inflammatory lesions, 25% are lymphoid lesions or lymphoma, and the other 25% are salivary gland type tumors. Although there are overlaps and exceptions, features such as laterality, portion of gland involvement, presence or absence of bony findings, enhancement pattern, and clinical presentation are valuable in differentiating among lacrimal gland lesions (Table 1). In general, epithelial neoplasms predominantly involve the orbital lobe and are unilateral. Lymphoproliferative and inflammatory lesions tend to involve both the orbital and palpebral lobes. Bony scalloping may be seen with lymphoproliferative lesions, but most benign epithelial neoplasms or inflammatory lesions do not cause significant bone change. Malignant neoplasms may result in bone destruction or erosion although this is not a common finding in our experience. As a rule, if clinical presentation is atypical, imaging findings must be correlated with tissue diagnosis to definitely exclude malignancy.

Normal Lacrimal Gland

The lacrimal gland is an almond-shaped, eccrine secretory gland for tear production. It is located in the superolateral aspect of the orbit, abutting the superior rectus and lateral rectus muscles (Fig. 1C). The lacrimal gland consists of an orbital lobe (Figs. 1A and 2A) and a palpebral lobe (Figs. 1B and 2B), which are separated anatomically by the lateral horn of the aponeurosis of the levator palpebrae muscle (Fig. 2C). The orbital lobe is located posterior and superior to the levator palpebrae aponeurosis, and the palpebral lobe is situated anterior and inferior to it. The orbital lobe is larger and the site of most lacrimal gland epithelial neoplasms. Infiltrative and inflammatory processes tend to have a diffuse pattern, typically involving both the orbital and palpebral lobes of the gland. The palpebral lobe is the portion of the lacrimal gland that is usually visible when the upper eyelid is everted on physical examination. The lacrimal gland normally measures approximately $20 \times 12 \times 5$ mm. Although the size of a normal lacrimal gland may vary from person to person, the glands are usually symmetric, and asymmetry is an important indicator of abnormality.

On CT, the lacrimal gland is isodense to the muscle. The medial border is outlined by orbital fat and the lateral border by orbital bone (Fig. 1C). Calcifications and bony changes are well seen on CT (Fig. 2), and normal glands show symmetric contrast enhancement. The superior resolution of MRI permits better assessment of the extent of glandular and periglandular involvement. Normal lacrimal glands have intermediate (sometimes heterogeneous) signal on both T1-weighted and T2-weighted imaging and enhance symmetrically after gadolinium administration (Fig. 2).

Epithelial Neoplasms

Primary lacrimal gland tumors arising from epithelial cells are relatively common, accounting for 20% of all solid lacrimal gland masses. They typically present as unilateral lacrimal gland enlargement with inferomedial displacement of the eye. This causes proptosis and, if enlargement of the gland is rapid, may even produce diplopia. Epithelial tumors typically arise from and involve the orbital lobe of the lacrimal gland, although nearly 20% of epithelial tumors may involve the palpebral lobe. About half of epithelial neoplasms are benign and half malignant. Benign neoplasms include pleomorphic adenoma and oncocytoma. The most common malignant epithelial neoplasms are adenoid cystic carcinoma and mucoepidermoid carcinoma.

TABLE 1: Common Patterns of Lacrimal Gland Masses

Cell Type	Lesion	Laterality	Lobe Involved	Bone	Pain
Epithelial	Pleomorphic adenoma	Unilateral	Orbital	Scalloping	No
	Adenoid cystic carcinoma	Unilateral	Orbital	Destruction	Yes, with or without paresthesia
	Mucoepidermoid carcinoma	Unilateral	Orbital	Scalloping, destruction	Yes
Lymphoid	Lymphoid hyperplasia	Bilateral	Orbital and palpebral	None, with or without occasional remodeling	No
	Lymphoma	Variable	Orbital and palpebral	None, with or without occasional remodeling	No
Leukemic	Chloroma	Variable	Orbital and palpebral	Variable	Yes
Metastasis	Metastases	Variable	Variable	Variable	Variable
Inflammatory	Infectious dacryoadenitis	Unilateral	Orbital and palpebral	None	Yes
	Sarcoidosis	Bilateral	Orbital and palpebral	None	Yes
	Pseudotumor (inflammatory dacryoadenitis)	Variable, unilateral more than bilateral	Orbital and palpebral	None	Variable
	Sjögren syndrome	Bilateral	Orbital and palpebral	None	Yes (dryness)
Others	Sickle cell disease	Variable	Orbital and palpebral	None	Yes

Pleomorphic Adenoma

Pleomorphic adenoma is the most common benign epithelial tumor of the lacrimal gland. Patients usually present between 20 and 40 years old with unilateral and painless proptosis. On CT, the tumor typically appears as a well-circumscribed unilateral mass that involves the orbital lobe of the lacrimal gland (Figs. 3A and 3C), although up to 10% of these lesions may occur solely in the palpebral lobe. Slow growth of a pleomorphic adenoma often results in smooth bony scalloping (Fig. 3B). Moderate contrast enhancement is common, but calcification within the tumor is rare. On MRI, a pleomorphic adenoma appears as a unilateral orbital lobe mass that has low-to-intermediate signal compared with muscle on T1-weighted imaging and intermediate signal compared with brain cortex on T2-weighted imaging. The lesion shows moderate contrast enhancement after gadolinium administration. Although benign, up to 20% of pleomorphic adenomas undergo malignant degeneration, especially after incomplete excisional biopsy. Therefore, treatment consists of complete surgical removal without rupture of the tumor capsule. If preoperative incisional biopsy is performed, subsequent complete surgical removal plus excision of adjacent periorbital tissues almost completely eliminates the chance of recurrence.

Adenoid Cystic Carcinoma

Adenoid cystic carcinoma is the most common malignant epithelial tumor of the lacrimal gland. Patients typically present in the fourth decade of life with a painful unilateral lacrimal gland mass. The propensity of this tumor for perineural spread results in severe pain, which is sometimes associated with paresthesia.

On CT, adenoid cystic carcinoma appears as a well-defined, homogeneous, and unilateral solid mass involving the orbital lobe of the lacrimal gland. There is typically associated bone destruction (Fig. 4), and there may be calcifications within the lesion. The tumor may extend along perineural pathways (cranial nerve branches of V1 and V2) through the superior orbital fissure into the cavernous sinus. After contrast administration, the tumor usually shows diffuse enhancement.

On MRI, an adenoid cystic carcinoma is hypointense to muscle on T1-weighted imaging and intermediate-to-high in signal compared with brain cortex on T2-weighted imaging. There is diffuse enhancement after gadolinium administration. MRI has superior resolution in showing perineural spread and involvement of subjacent soft tissues.

Adenoid cystic carcinoma has a poor prognosis. Perineural extension of the lesion is so common that complete surgical removal is difficult. Therefore, brachytherapy with ^{125}I is sometimes used as adjuvant treatment. Local recurrence is common, and distant metastasis most often involves the lung. Current treatment regimens primarily focus on local disease control.

Mucoepidermoid Carcinoma

Mucoepidermoid carcinoma of the lacrimal gland is rare, representing only 1% of all lacrimal gland neoplasms. Like most malignant neoplasms of the lacrimal glands, patients with mucoepidermoid carcinoma typically present with painful proptosis due to rapid growth of this infiltrative lesion with bony scalloping or destruction and occasional invasion into adjacent structures (Fig. 5). Distant metastases to the brain have been reported. The imaging features may be indistinguishable from adenoid cystic carcinoma, and treatment includes resection, radiation, and chemotherapy.

Lymphoproliferative Lesions

Reactive Lymphoid Hyperplasia

Reactive lymphoid hyperplasia is benign and represents part of the spectrum of lymphoid diseases of the orbit. Reactive lymphoid hyperplasia has a tendency to involve bilateral lacrimal glands. The disease is diffuse and involves both orbital and palpebral lobes arising from intrinsic lacrimal lymphoid tissues. Similar to lymphoma, lymphoid hyperplasia usually has a concave inner margin and molds to the globe. Although lymphoid hyperplasia is, in general, not readily differentiated from lymphoma on CT, it is more heterogeneous compared with lymphoma, which is typically homogeneous in density. On MRI, it appears T1 hypointense to the brain cortex (Fig. 6C), and shows slightly heterogeneous contrast enhancement (Fig. 6D).

Lymphoma

The lacrimal gland contains intrinsic lymphoid as well as epithelial tissue. Primary lymphoproliferative disease of the orbit, however, including that of the lacrimal gland, is relatively rare. Most lymphoid disease arises elsewhere and involves the orbit secondarily. The most common lymphoma affecting the

lacrimal gland is of the mucosa-associated lymphoid tissue type, which has a much better prognosis than the less common secondary follicular lymphoma of the orbit. Lymphoma tends to occur in older patients who typically present with painless lacrimal gland masses.

CT shows bilateral or, less commonly, unilateral diffuse involvement of the lacrimal glands, which involves both the palpebral (Fig. 7A) and orbital (Fig. 7B) lobes. Lacrimal gland lymphoma is well defined and homogeneous and conforms to the globe and subjacent osseous structures. Lymphoma is isodense to muscle and shows mild enhancement after contrast administration (Fig. 7A). There is typically no infiltrative bone destruction, although bone remodeling and/or sclerosis may be present.

On MRI, lacrimal gland lymphoma is homogeneous and isointense to muscle on T1-weighted imaging (Fig. 8A) and isointense-to-slightly hyperintense to brain cortex on T2-weighted imaging (Fig. 8C). There is moderate enhancement after gadolinium administration (Fig. 8B). Lymphoma is hypercellular and shows slowed diffusion on diffusion-weighted imaging (Fig. 8D). The various subtypes of lymphoma have similar appearances and cannot be differentiated by imaging criteria.

The treatment of lacrimal gland lymphoma depends on the histologic classification. Local radiotherapy is used to treat lesions confined to the orbit, whereas chemotherapy is used for more widespread disease.

Leukemic Lesions

Leukemia-Chloroma or Granulocytic Sarcoma

Orbital granulocytic sarcoma, also known as chloroma because of its green color related to high myeloperoxidase content, can rarely secondarily involve the lacrimal gland. This leukemia-related tumor of myeloid cell origin represents a collection of these cells outside the bone marrow. Children and young adults are most likely to be affected. Patients typically present with progressive painful proptosis, which may develop either before or after the onset of systemic leukemia (most commonly in acute myelogenous leukemia). The uvea, choroids, retina, optic nerve, and orbital fat are more commonly primarily involved, with secondary disease extension to one or both of the lacrimal glands.

On CT, a chloroma of the lacrimal gland is homogeneous, well-defined, isodense or hypodense to muscle, and can be associated with bone involvement. Contrast-enhanced images show varying degrees of homogeneous or heterogeneous enhancement. On MRI, the lesion is hypointense to isointense to muscle on T1-weighted imaging (Figs. 9A and 9B) and isointense to brain cortex on T2-weighted imaging (Figs. 9C and 9D). Marrow involvement is common, and the degree of contrast enhancement is variable. The treatment of chloroma includes local disease control, chemotherapy, and bone marrow transplantation.

Metastasis

Rare metastatic involvement of the lacrimal gland cannot be distinguished from a primary lacrimal gland tumor on the basis of any specific imaging feature. Patients are generally older and present with a unilateral lacrimal mass or bilateral lacrimal masses without any specific pattern of lobe involvement, and there may be associated bone involvement. Primary neoplasms that most commonly metastasize to the lacrimal gland include carcinomas of the breast, prostate, kidney, thyroid, and melanoma. In the pediatric population, Ewing sarcoma or neuroblastoma may extend from adjacent bony structures to secondarily involve the lacrimal gland. Treatment focuses on local control and eradication of the primary malignant disease.

Inflammatory Lesions

Infectious Dacryoadenitis

Dacryoadenitis is inflammation of the lacrimal gland, which may be acute or chronic, infectious, or inflammatory. Acute infectious dacryoadenitis is more common in children and young adults, who typically present with unilateral painful proptosis. On CT, there is unilateral involvement with diffuse enlargement of the lacrimal gland that affects both the orbital and palpebral lobes (Fig. 10A). There is avid contrast enhancement in the acute phase of disease representing hyperemia, often with periglandular inflammatory changes and occasionally complicated by abscess formation (Figs. 11A and 11B). Inflammation may extend to subjacent muscles, causing superior and lateral rectus myositis, but there is no bone involvement. MRI shows similar unilateral diffuse enlargement and hyperenhancement of the lacrimal gland, with surrounding inflammatory changes but no bone involvement. Acute infectious dacryoadenitis responds well to antibiotic therapy.

Sarcoidosis

Sarcoidosis is the most common inflammatory disease involving the lacrimal gland. Ocular involvement, most commonly uveitis, occurs in 80% of patients with sarcoidosis. Sarcoidosis of the lacrimal gland is typically bilateral and diffuse, and it may cause painless enlargement of the orbital and palpebral lobes. On CT, sarcoidosis produces diffuse bilateral lacrimal gland enlargement with hyperenhancement on contrast-enhanced images (Figs. 12A and 12C) and no bone involvement (Figs. 12B and 12D). Concurrent imaging of the chest usually shows prominent mediastinal and hilar lymphadenopathy. On MRI, there is also diffuse bilateral enlargement of the lacrimal glands, which appear hypointense to isointense on T1-weighted imaging and heterogeneously hyperintense on T2-weighted imaging, with avid enhancement after gadolinium administration. The mainstay of treatment of sarcoidosis is corticosteroids. More aggressive forms of disease may require immunosuppressants, such as methotrexate and cyclophosphamide.

Pseudotumor

Pseudotumor (also known as inflammatory dacryoadenitis and idiopathic orbital inflammation) is an idiopathic fibroinflammatory process that can involve the lacrimal glands and is thought to be related to autoimmune derangement. Patients typically present with long-standing occasionally painful unilateral more often than bilateral enlargement of the lacrimal glands, which may have a relapsing remitting course. Mass effect in severe disease may lead to compression of the globe or extraocular muscles, but visual acuity usually remains intact. The diagnosis is made only when other diseases causing lacrimal gland enlargement have been excluded. Biopsy shows nongranulomatous inflammation, excluding neoplasm. Lacrimal gland pseudotumors have been recognized as part of an emerging rare multiorgan system disorder known as IgG4-related disease, which presents with pseudotumors and fibrosclerosing disease involving multiple organs.

On CT, pseudotumor appears as diffuse unilateral or bilateral enlargement of the lacrimal glands involving both the orbital and palpebral lobes. There is variable contrast enhancement and no bone involvement. On MRI, these lacrimal gland masses are usually isointense to hypointense to muscle on T1-weighted imaging (Fig. 13A) and isointense to hypointense to brain cortex on T2-weighted imaging (Figs. 13D and 13E) and show variable contrast enhancement. When compared with most lacrimal gland tumors, pseudotumors typically are relatively more

T2 hypointense (Figs. 13D and 13E). Treatment response of pseudotumor to corticosteroid therapy is usually excellent, although more resistant and recurrent disease may require treatment with immunosuppressants.

Sjögren Syndrome

Sjögren syndrome is an autoimmune disease that targets the lacrimal glands and salivary glands, producing the sicca syndrome of dry eyes and dry mouth. Diagnosis relies on lacrimal gland biopsy, lacrimal flow rate measurement, and rose Bengal staining. Involvement tends to be bilateral and diffuse. Early in the disease process, there is enlargement of the glands, then progressive fatty deposition, and subsequent atrophy. On CT, there is bilateral lacrimal gland enlargement or atrophy without bone involvement (Figs. 14A and 14B). On MRI, unenhanced T1-weighted imaging shows bilateral enlargement or atrophy of lacrimal glands, with patchy areas of relative T1 hyperintensity (Fig. 14C), reflecting components of fatty replacement, with subtle heterogeneous appearance after contrast administration (Fig. 14D). Treatment includes artificial tears and avoidance of low-humidity environments.

Others

Chronic inflammatory dacryoadenitis may develop in a variety of other entities, including tuberculosis, amyloidosis, thyroid ophthalmopathy, and anti-neutrophil cytoplasmic antibody–associated granulomatous vasculitis (formerly known as Wegener granulomatosis). Involvement in these conditions may be unilateral or bilateral, and treatment is targeted to the underlying disease. Lacrimal gland involvement has also been reported in patients with sickle cell disease, which presents as lacrimal gland edema, with glands appearing enlarged and homogeneously enhancing or hyperenhancing on CT and MRI.

Miscellaneous neoplastic causes of lacrimal gland masses include rhabdomyosarcoma and solitary fibrous tumor. Lacrimal gland involvement has also been described in the setting of post transplantation lymphoproliferative disease, which is exceedingly rare.

Suggested Readings

1. Jung WS, Ahn KJ, Park MR, et al. The radiological spectrum of orbital pathologies that involve the lacrimal gland and the lacrimal fossa. *Korean J Radiol* 2007; 8:336–342
2. Som PM, Curtin HD. *Head and neck imaging*, 5th ed. St. Louis, MO: Mosby Elsevier 2011
3. Kostick DA, Linberg JV. Lacrimal gland tumors. In: Tasman W, Jaeger EA, eds. *Duane's ophthalmology*, 2006 ed., vol. 2. Philadelphia, PA. Lippincott Williams & Wilkins, 2006:chapter 40 ed. Vol. 2, Chapter 40

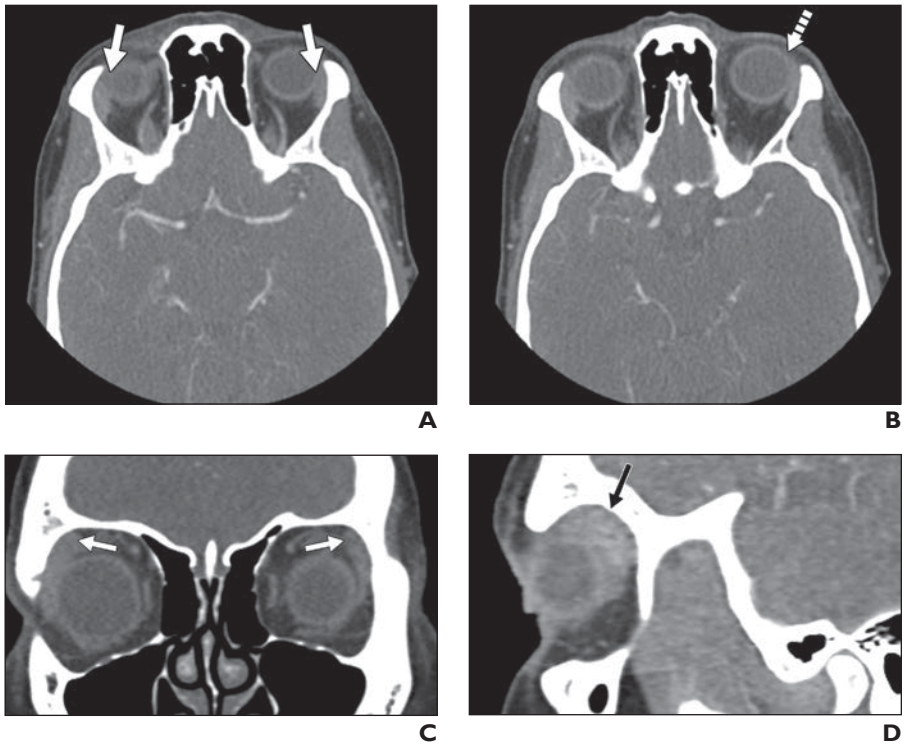

Fig. 1—Normal CT of lacrimal glands.
A and **B,** Unenhanced axial CT images show orbital (*solid arrows*, **A**) and palpebral (*dashed arrow*, **B**) lobes of lacrimal glands.
C and **D,** Coronal (**C**) and sagittal (**D**) CT images show normal lacrimal glands (*arrows*), which are isodense to muscle, in superolateral aspects of orbits.

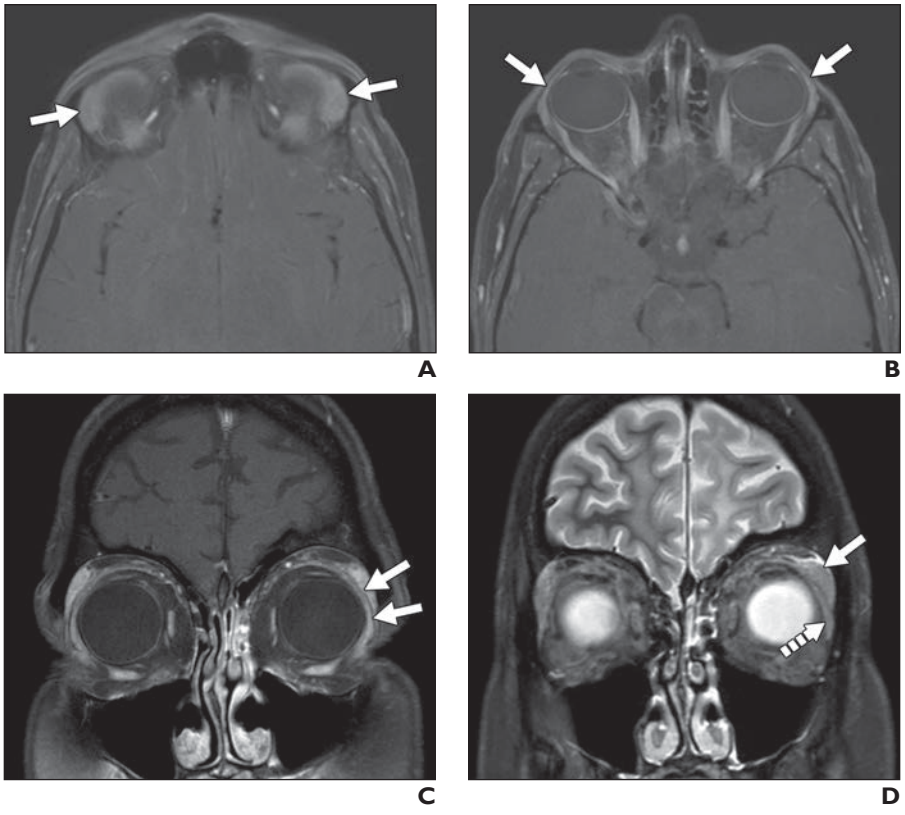

Fig. 2—Normal MRI of lacrimal glands.
A and **B,** Gadolinium-enhanced T1-weighted fat-saturated axial MR images show orbital (**A**) and palpebral (**B**) lobes of normal lacrimal glands (*arrows*), which have homogeneous enhancement and are isointense to adjacent extraocular muscles.
C and **D,** Gadolinium-enhanced T1-weighted fat-saturated and T2-weighted coronal images show location of levator palpebrae muscle aponeurosis (*arrows*, **C**), which divides lacrimal gland into posterior-superior orbital lobe (*solid arrow*, **D**) and anterior-inferior palpebral lobe (*dashed arrow*, **D**).

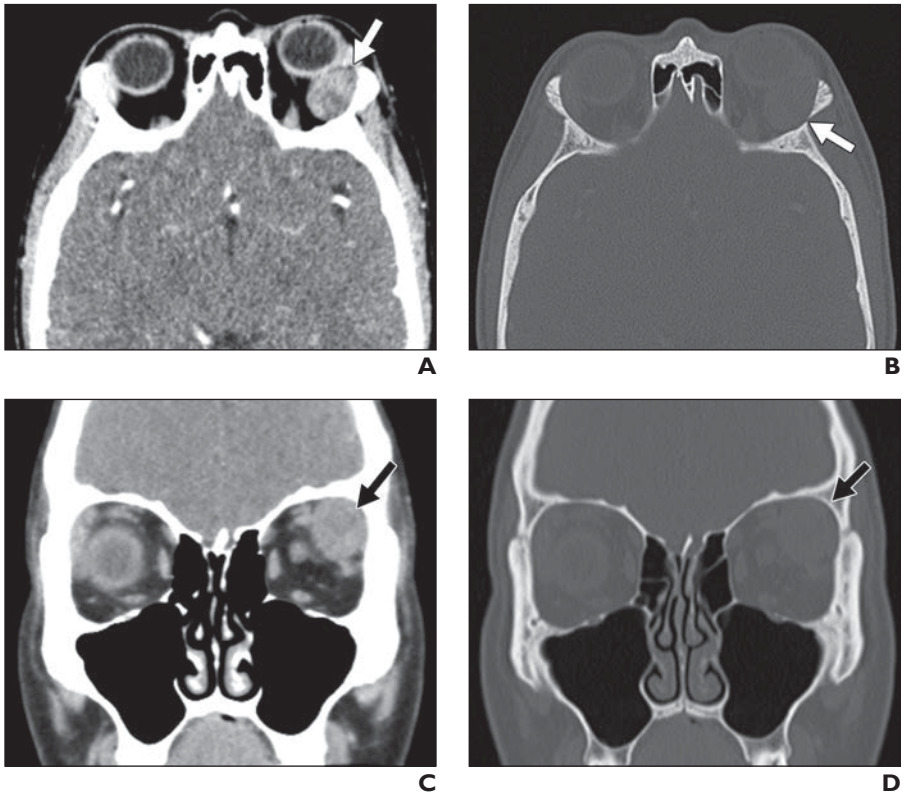

Fig. 3—Pleomorphic adenoma.
A–D, Axial (**A** and **B**) and coronal (**C** and **D**) contrast-enhanced CT images show rounded heterogeneously enhancing mass in orbital portion of lacrimal gland (*arrows*, **A** and **C**). Note subtle associated bone remodeling (*arrows*, **B** and **D**).

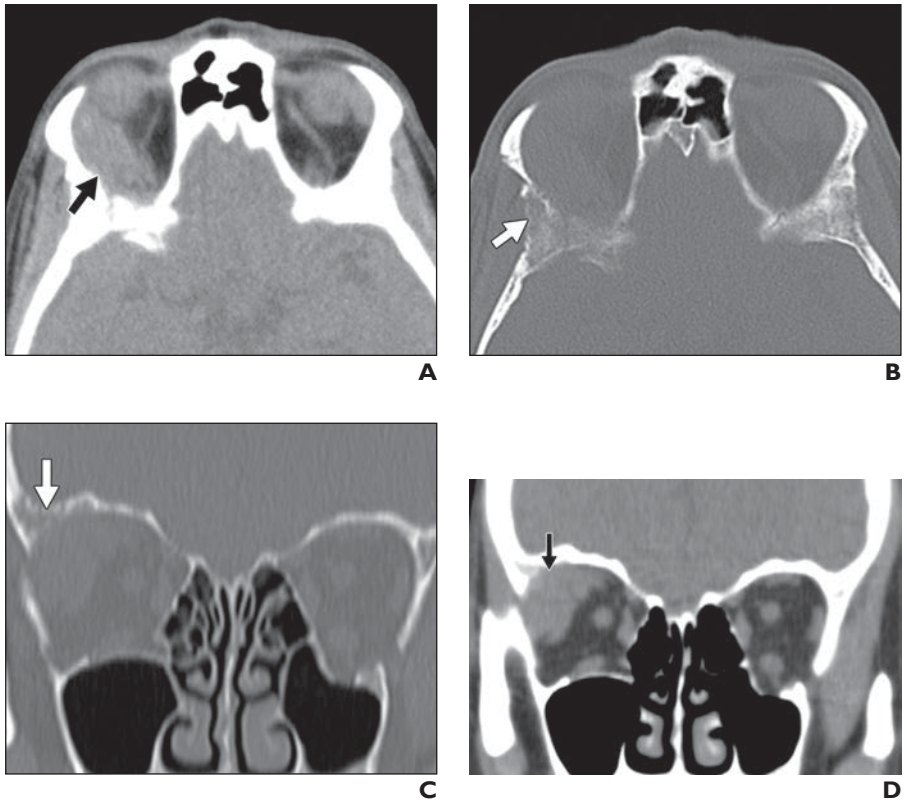

Fig. 4—Adenoid cystic carcinoma.
A–D, Axial (**A** and **B**) and coronal (**C** and **D**) unenhanced CT images show right orbital lobe lacrimal gland mass (*black arrows*, **A** and **D**) with permeative destruction of right sphenoid bone (*white arrows*, **B** and **C**).

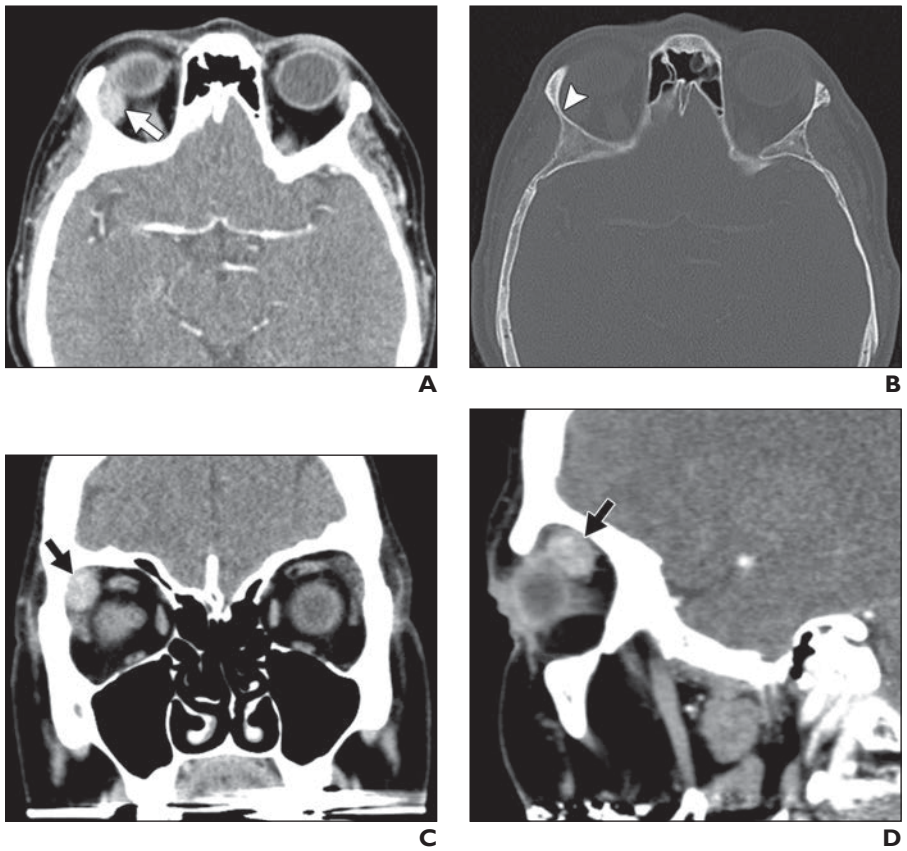

Fig. 5—Mucoepidermoid carcinoma.
A–D, Contrast-enhanced axial (**A**), coronal (**B**), and sagittal (**D**) CT images show homogeneously enhancing mass in orbital lobe of right lacrimal gland (*arrows*), with associated subtle bone remodeling (*arrowhead*, **B**).

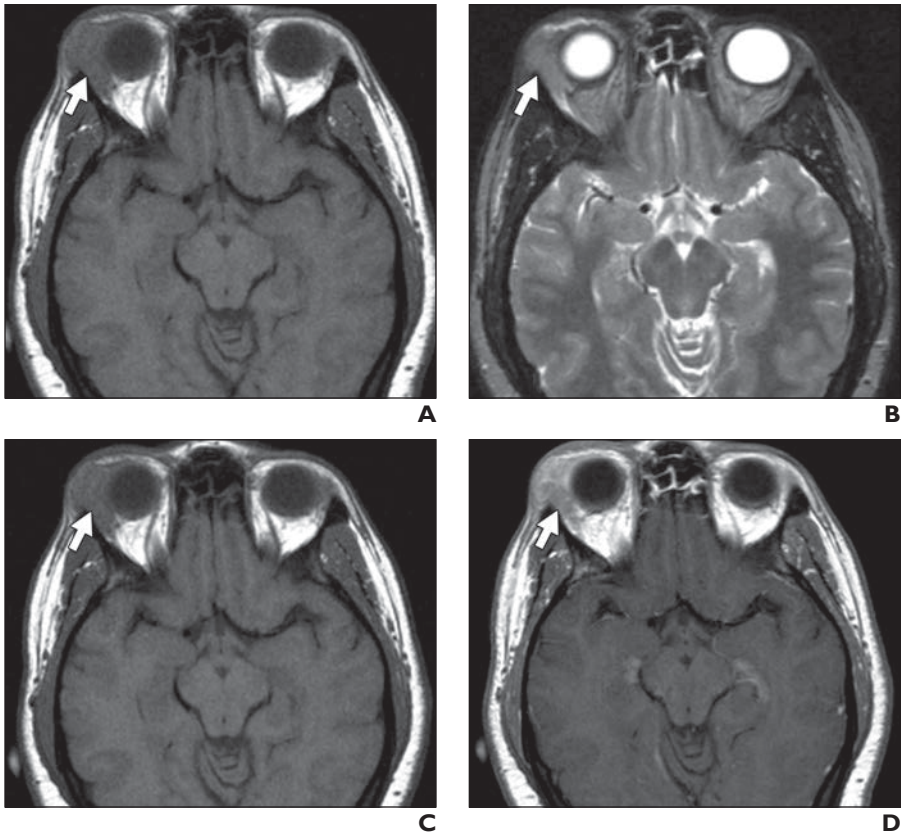

Fig. 6—Lymphoid hyperplasia.
A–D, Axial unenhanced T1-weighted (**A**) and T2-weighted (**B**) and axial unenhanced (**C**) and gadolinium-enhanced (**D**) images show right greater than left bilateral lacrimal gland masses (*arrows*) that mold to globe and show slightly heterogeneous contrast enhancement.

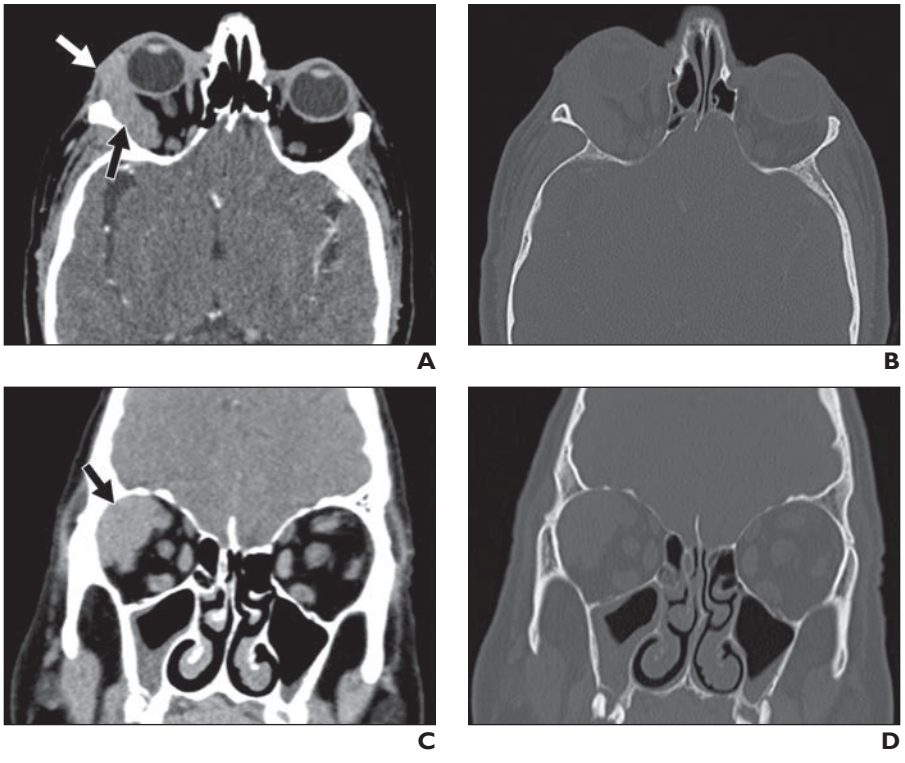

Fig. 7—Lymphoma.
A and **B,** Contrast-enhanced axial CT images show diffuse infiltration and enlargement of right lacrimal gland. There is involvement of both orbital (*black arrow*, **A**) and palpebral lobes (*white arrow*, **A**) without bony change.
C and **D,** Contrast-enhanced coronal CT images show lymphoma to be isodense to extraocular muscles with mild enhancement (*arrow*, **C**) and no bone changes (**D**).

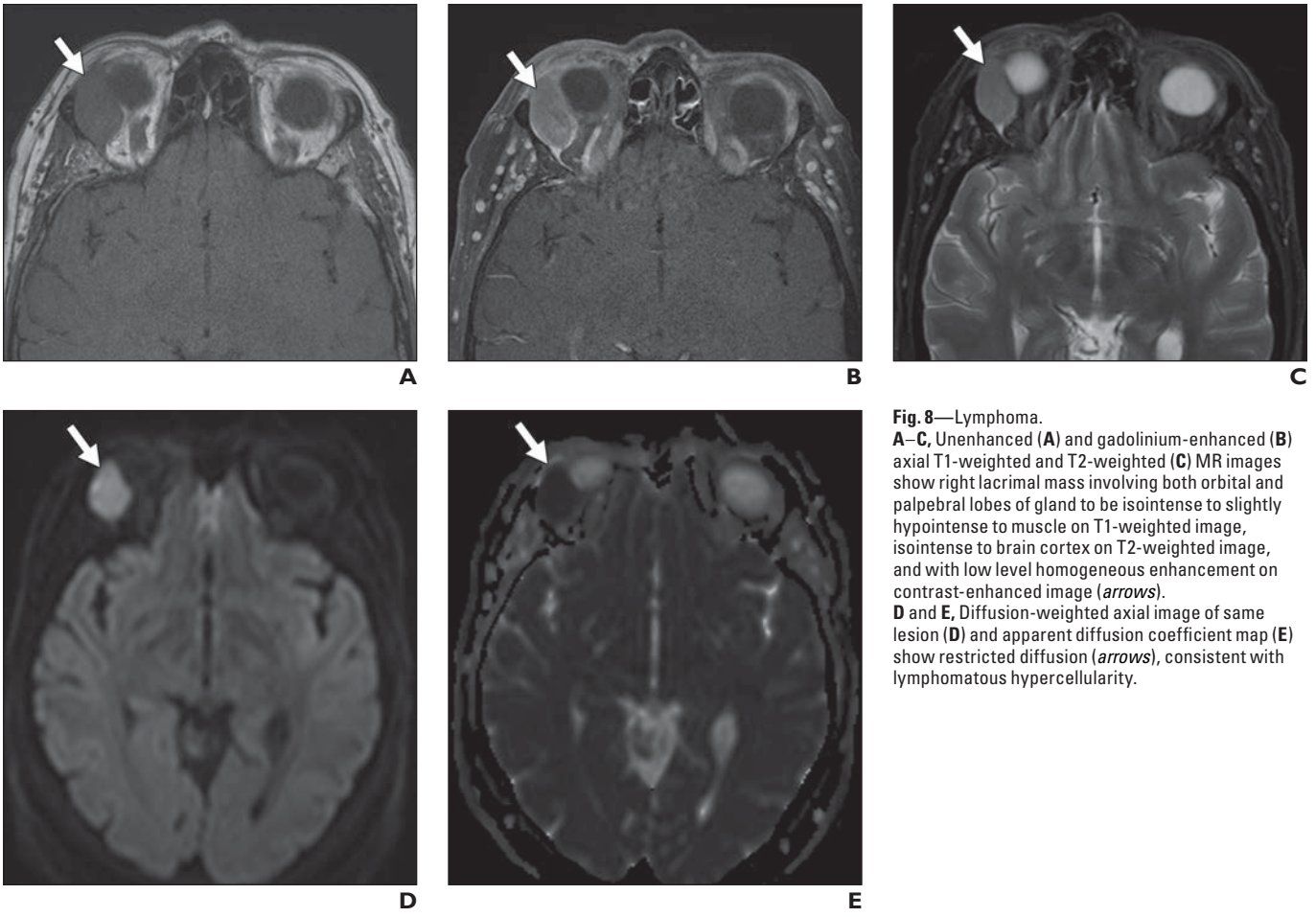

Fig. 8—Lymphoma.
A–**C,** Unenhanced (**A**) and gadolinium-enhanced (**B**) axial T1-weighted and T2-weighted (**C**) MR images show right lacrimal mass involving both orbital and palpebral lobes of gland to be isointense to slightly hypointense to muscle on T1-weighted image, isointense to brain cortex on T2-weighted image, and with low level homogeneous enhancement on contrast-enhanced image (*arrows*).
D and **E,** Diffusion-weighted axial image of same lesion (**D**) and apparent diffusion coefficient map (**E**) show restricted diffusion (*arrows*), consistent with lymphomatous hypercellularity.

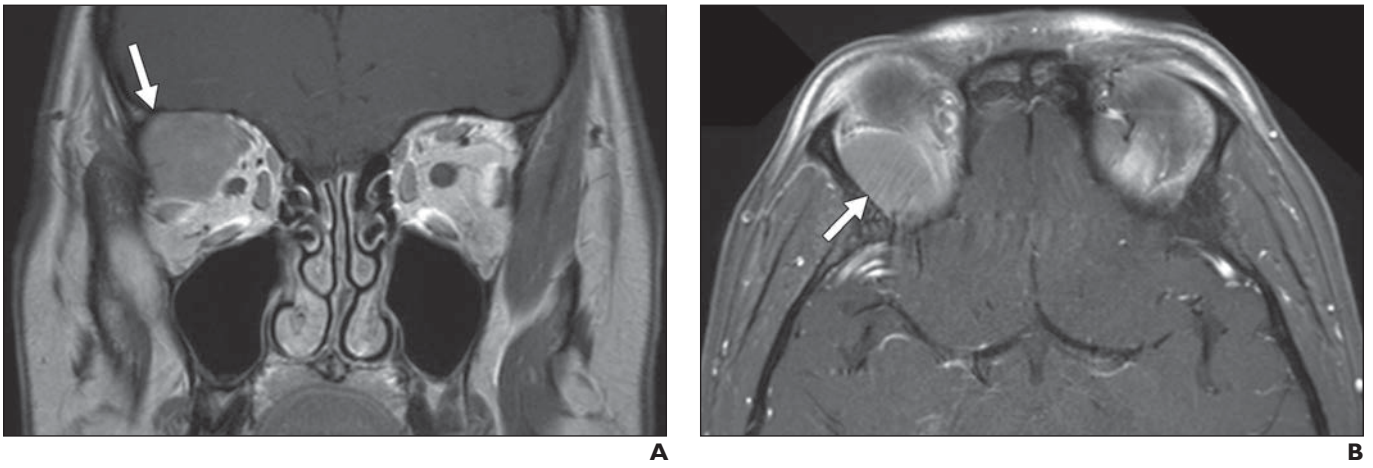

Fig. 9—Chloroma.
A and **B,** Coronal (**A**) and axial (**B**) gadolinium-enhanced T1-weighted MR images show homogeneous mildly enhancing mass hypointense to isointense to muscle in orbital lobe of right lacrimal gland (*arrows*).

(Fig. 9 continues on next page)

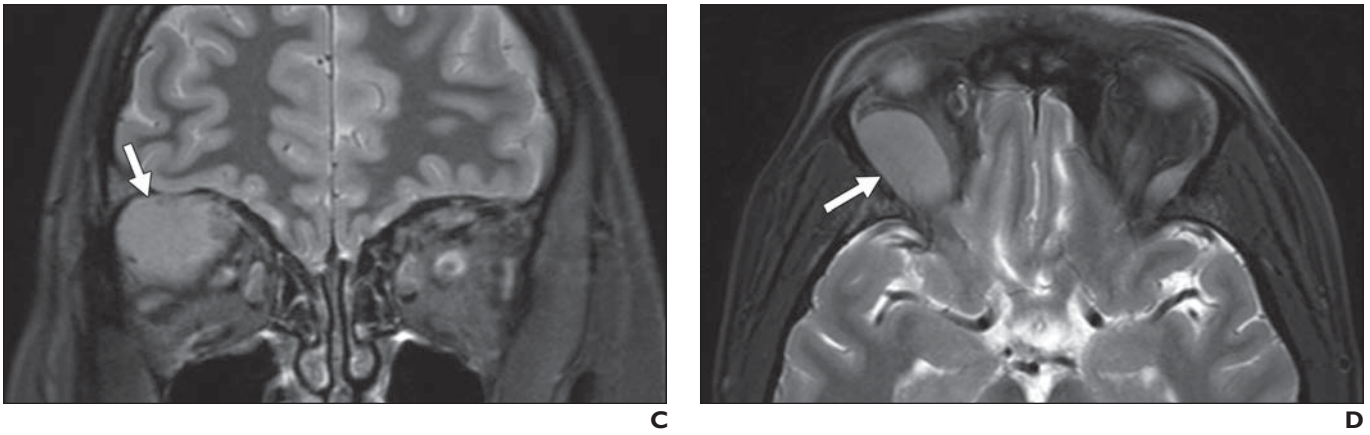

Fig. 9 (continued)—Chloroma.
C and **D,** Coronal (**C**) and axial (**D**) T2-weighted images show mass to be isointense to brain cortex (*arrows*).

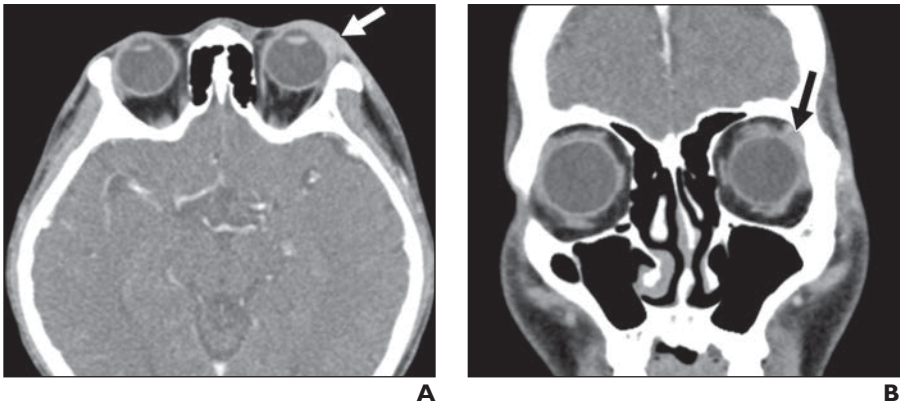

Fig. 10—Infectious dacryoadenitis.
A and **B,** Contrast-enhanced axial (**A**) and coronal (**B**) CT images show diffusely enlarged and enhancing left lacrimal gland (*arrows*). Patient presented with significant left eye pain and was diagnosed with bacterial dacryoadenitis. There are no bony changes as expected in setting of acute inflammation.

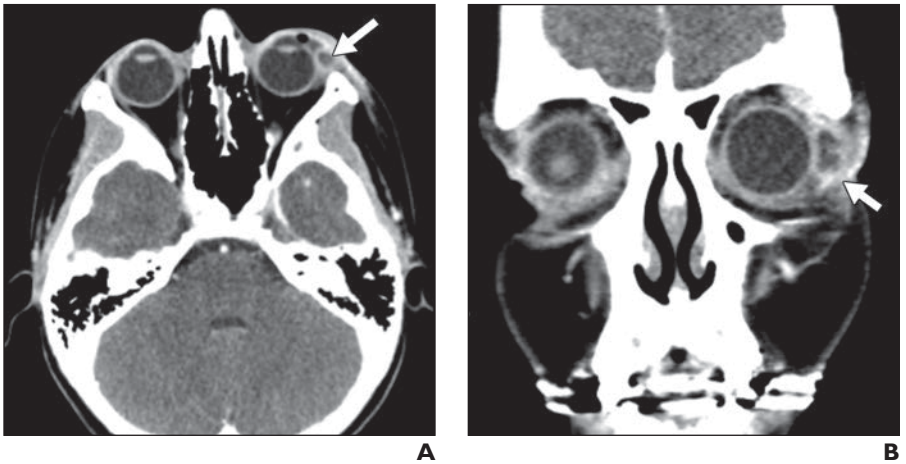

Fig. 11—Abscess.
A and **B,** Contrast-enhanced axial (**A**) and coronal (**B**) CT images show rim-enhancing lesion within edematous and hyperenhancing left lacrimal gland (*arrows*).

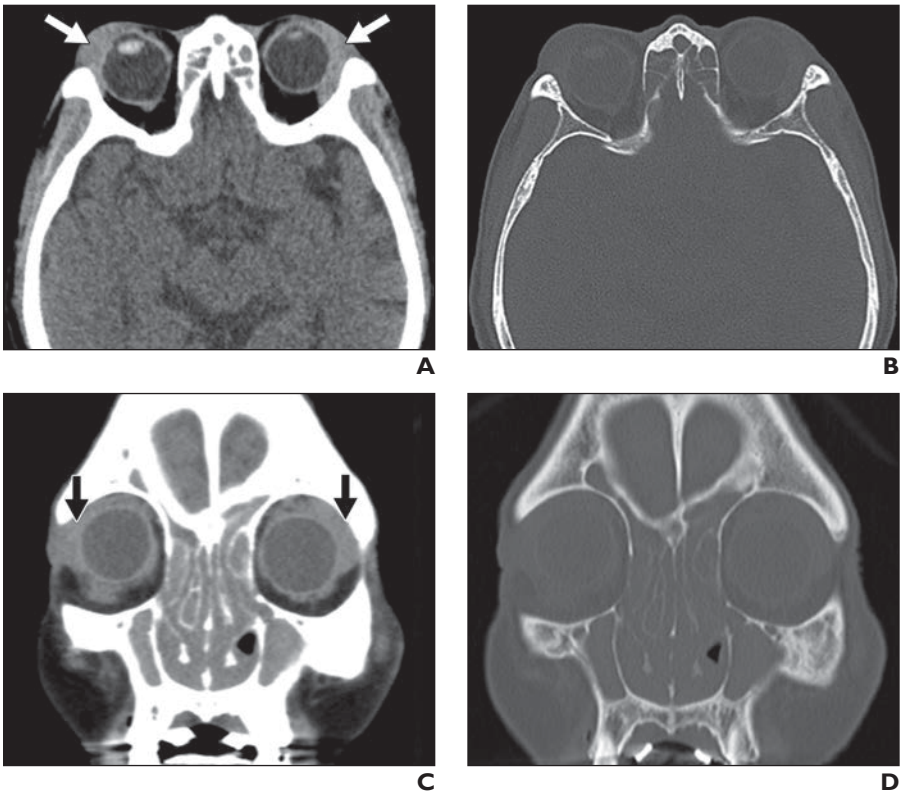

Fig. 12—Sarcoidosis.
A–D, Unenhanced axial (**A**) and coronal (**C**) CT images show diffuse bilateral lacrimal gland enlargement involving both palpebral and orbital lobes (*arrows*). Axial (**B**) and coronal (**D**) bone window images show no bone changes.

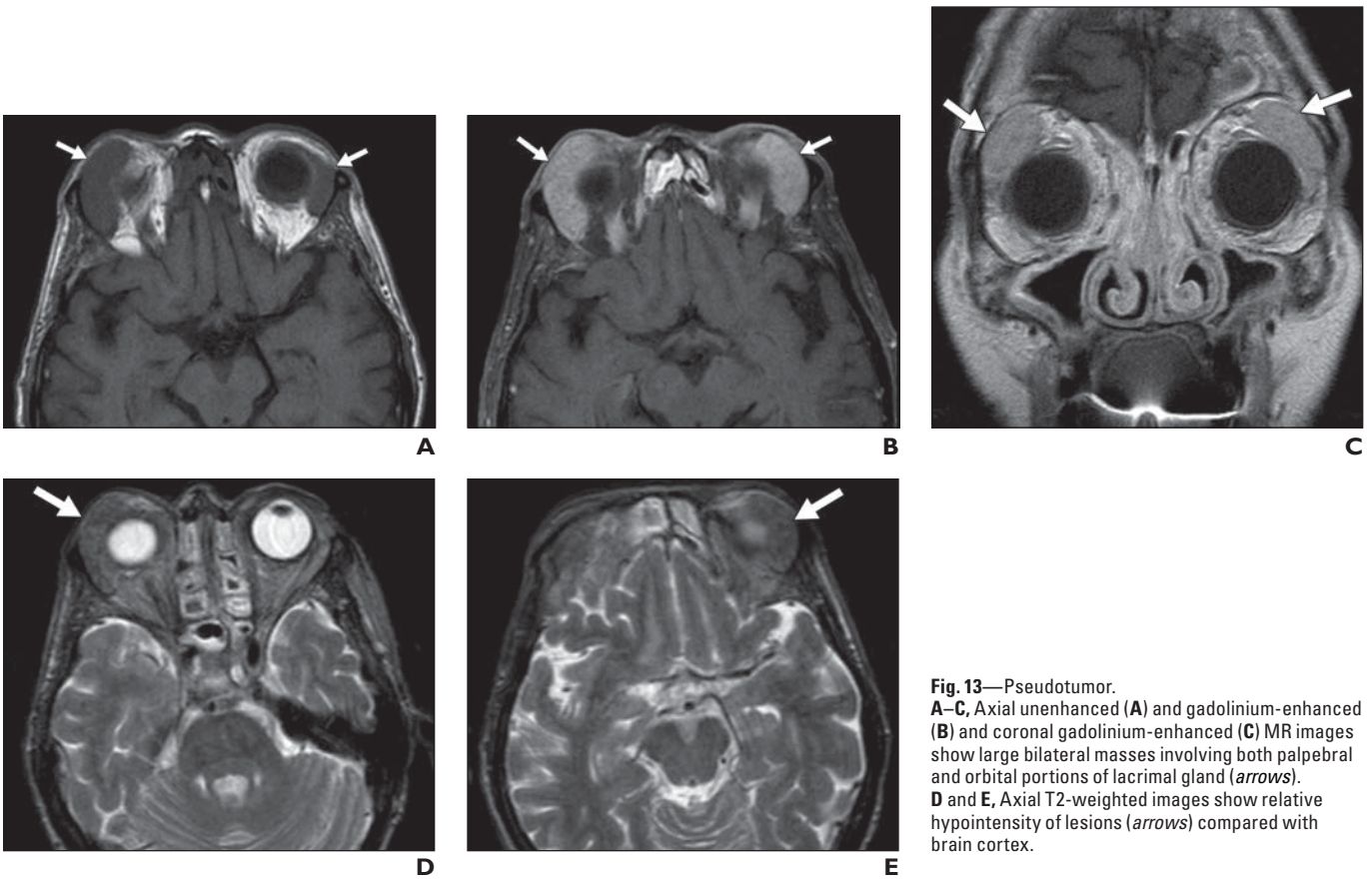

Fig. 13—Pseudotumor.
A–C, Axial unenhanced (**A**) and gadolinium-enhanced (**B**) and coronal gadolinium-enhanced (**C**) MR images show large bilateral masses involving both palpebral and orbital portions of lacrimal gland (*arrows*).
D and **E,** Axial T2-weighted images show relative hypointensity of lesions (*arrows*) compared with brain cortex.

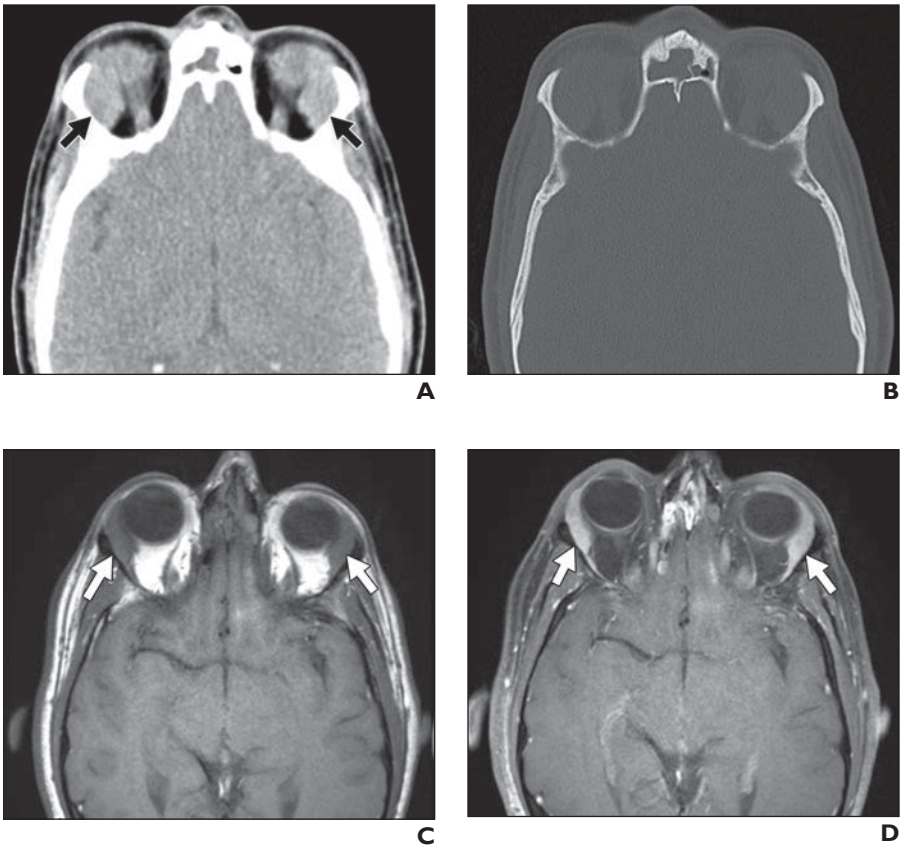

Fig. 14—Sjögren syndrome.
A and **B,** Axial unenhanced CT images show bilateral lacrimal gland enlargement without bony involvement (*arrow*, **A**).
C and **D,** Axial unenhanced (**C**) and gadolinium-enhanced (**D**) T1-weighted MR images show bilateral enlarged glands with faint patchy intrinsic T1 hyperintensity reflecting fatty deposition and mildly heterogeneous enhancement after contrast administration (*arrows*).