

Figure 29-2 The extraocular muscles are greatly distended in this postmortem dissection of tissues from a patient with thyroid (Graves) ophthalmopathy. Note that the tendons of the muscles are spared. (Courtesy Dr. Ralph C. Eagle, Jr., Wills Eye Hospital, Philadelphia, Pa.)

as an acute bacterial infection or as a component of a fungal infection. This occurs most commonly in immunosuppressed individuals, in patients with diabetic ketoacidosis, or, rarely, in persons without any predisposition. Systemic conditions such as *Wegener granulomatosis* (Chapter 11) may present first in the orbit and may be confined there for prolonged periods of time, or alternatively, it may involve the orbit secondarily by extension from the sinuses.

Idiopathic orbital inflammation, also known as orbital inflammatory pseudotumor (Fig. 29-3), is another inflammatory condition affecting the orbit. This condition may be unilateral or bilateral, and may affect all orbital tissue elements or may be confined to the lacrimal gland (*sclerosing dacryoadenitis*), the extraocular muscles (*orbital myositis*), or the Tenon's capsule, the fascial layer that wraps around the eye (*posterior scleritis*). IgG4-related disease (Chapter 6) should be excluded before declaring an orbital inflammation to be idiopathic.

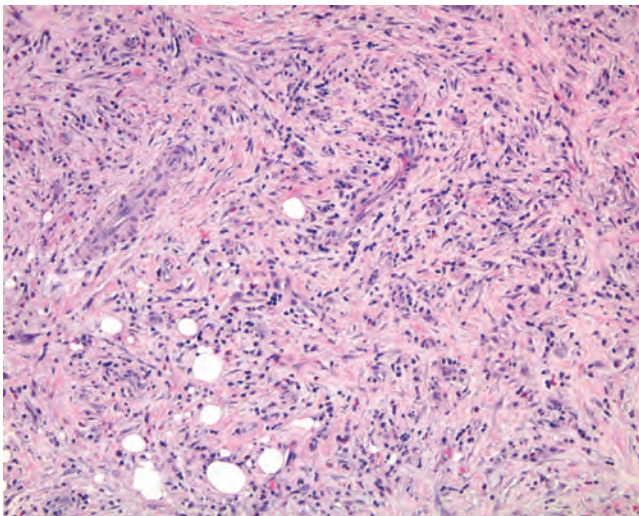


Figure 29-3 In idiopathic orbital inflammation (orbital inflammatory pseudotumor) the orbital fat is replaced by fibrosis. Note the chronic inflammation, accompanied in this case by eosinophils.

MORPHOLOGY

Idiopathic orbital inflammation is characterized histologically by chronic inflammation and variable degrees of fibrosis. The inflammatory infiltrate typically includes lymphocytes and plasma cells and occasionally eosinophils. Germinal centers, when present, raise the suspicion of a reactive lymphoid hyperplasia. Vasculitis may be present, suggesting an underlying systemic condition. The presence of necrosis and degenerating collagen along with vasculitis should raise the suspicion of *Wegener granulomatosis*. Idiopathic orbital inflammation is typically confined to the orbit but may develop concomitantly with sclerosing inflammation in the retroperitoneum, the mediastinum, and the thyroid, especially as a manifestation of IgG4-related disease.

Neoplasms

The most frequently encountered primary neoplasms of the orbit are vascular in origin: the capillary hemangioma of infancy and early childhood, and the lymphangioma (both of which are unencapsulated), and the encapsulated cavernous hemangioma found typically in adults. These are described in other chapters. Only a handful of orbital masses are encapsulated (e.g., pleomorphic adenoma of the lacrimal gland, dermoid cyst, neurilemmoma), and the recognition of encapsulation on imaging studies allows the surgeon to anticipate pathologic findings.

Non-Hodgkin lymphoma, like idiopathic orbital inflammation, can affect the entire orbit or can be confined to compartments of the orbit such as the lacrimal gland. Orbital lymphomas are classified according to the WHO classification system (Chapter 13).

Primary orbital malignancies may arise from any of the orbital tissues and are classified according to the scheme used for the parent tissue. For example, the lacrimal gland may be considered a minor salivary gland, and tumors of the lacrimal gland are classified as salivary gland tumors.

Metastases to the orbit may present with distinctive signs and symptoms that point to the origin of the tumor. For example, metastatic prostatic carcinoma may present clinically like idiopathic orbital inflammation; metastatic neuroblastoma and Wilms tumor—richly vascular neoplasms—may produce characteristic periocular ecchymoses. Neoplasms may also invade from the sinuses into the orbit.

KEY CONCEPTS

- Proptosis results from lesions or pathologic changes in tissue that occupy space in the orbit. The orbit is a compartment that is only open anteriorly and is closed in all other dimensions by bone.
- Inflammation in the orbit may develop by extension of local disease in adjacent tissues (e.g., sinusitis) or as a component of systemic disease (e.g., *Wegener granulomatosis*)
- The most common primary tumors of the orbit are vascular (e.g., capillary and cavernous hemangiomas).