10. Tumors and Pseudotumors

**Intraocular:** Nevi · Hemangioma · Angiomatosis · Malignant melanoma · Retinoblastoma · Metastases · Pseudotumors

**Optic nerve:** Glioma · Meningioma

**Orbital:** Hemangioma · Cyst · Lymphoma · Metastases · Pseudotumors

The eye and its adnexae may be the primary site of a variety of benign and malignant ectodermal, neuroectodermal, and mesenchymal tumors. Tumors of the lid and conjunctiva are not mentioned here. Tumors of the eye and the orbit usually cause fairly early symptoms and/or signs. Visual acuity, visual fields, and the normal position of the eye are often affected. Highly malignant intraocular and orbital tumors must be differentiated from benign space-occupying lesions. The indirect ophthalmoscope, fluorescein angiography, and ultrasonography are the most important diagnostic tools for differentiating intraocular masses. Ultrasonography and CT scanning have facilitated the examination of patients with suspected orbital tumors. The macroscopic appearance of malignant orbital tumors such as rhabdomyosarcoma adds very little to the understanding or orbital disease. They are therefore not included here.

Secondary ocular and orbital tumors may arise from adjacent structures or metastasize to the eye and orbit from distant primary tumors.

Nonneoplastic space-occupying lesions (inflammatory, degenerative, developmental) are often called pseudotumors because their symptoms and/or signs mimic a malignant growth.
Primary Benign Intraocular Tumors

Common benign intraocular tumors are uveal nevi and Fuchs’ adenoma of the ciliary body. Less common are angiomomas of the choroid or retina, as well as glial tumors of the retina and optic nerve head. Benign intraocular neural, osseous, ciliary epithelial, or muscle tumors are very rare. All of them may play a role in the differential diagnosis of malignant intraocular tumors.

Uveal Nevi

Pigmented flat nevi of 1–2 disc diameters are a common finding throughout the entire uvea, but especially in the choroid. They usually do not cause symptoms and are an incidental finding during routine ophthalmoscopy. Larger nevi may be difficult to differentiate from small malignant melanomas.

Fig. 10.1. Pigmented uveal nevus at the ora serrata. Autopsy eye without clinical history.

Fig. 10.2. Cross section through a pigmented choroidal nevus at the posterior pole. The nevus fills the entire thickness of the choroid. The overlying retina is unaffected. M 379/82: Autopsy eye of a 41-year-old man without clinical history.

Fig. 10.2A. Choroidal nevus. A-scan echogram (a) and contact B-scan echogram (b) of a choroidal nevus. Note the high reflectivity and mostly homogeneous structure (thin arrow) in the A-scan (S = sclera) and the flat to slightly dome-shaped appearance in the B-scan.