Comment. Small round cell tumors manifesting in the orbit should raise the suspicion for a wide variety of tumors. The differential diagnosis includes lymphoblastic lymphoma, embryonal rhabdomyosarcoma, neuroblastoma, primitive neuroectodermal tumor, Ewing sarcoma, melanoma, and ependymoma, as well as DSRCT.

Several features are consistent with the diagnosis of DSRCT in this patient. First, the majority of patients with this tumor are between the ages of 15 and 35 years with a 4:1 male-to-female preponderance. Second, immunohistochemical examination revealed positive cytokeratin and desmin staining in a characteristic perinuclear dot-like pattern, which is pathognomonic for DSRCT. In addition, although the histogenesis of DSRCT remains unknown, positive CD99 and neuron-specific enolase staining have been observed in these tumors, supporting an origin from a progenitor cell with potential for multiphenotypic differentiation. Finally, the fluorescence in situ study demonstrates an EWS gene rearrangement that is characteristic of this tumor. Primitive neuroectodermal tumor is excluded on the basis of strong positive cytokeratin and desmin staining.

The unusual feature of this clinical manifestation is the location of the tumor, which typically appears in the abdomen or involves other serosal surfaces. Only 4 cases of nonserosal DSRCT have been reported. Two cases involved soft tissues and bone. Adsay et al described a hypothenar mass in the right hand of a 34-year-old man, and Mihok and Cha reported a neck mass in a 16-year-old boy. Two other cases manifested in the head. Tison et al described an intracranial lesion located in the posterior fossa in a 24-year-old man. Finke et al reported an ethmoidal sinus lesion in a 21-year-old woman.

To our knowledge, this is the first case of DSRCT in the orbit, causing visual disturbances and eventually pain in the eye with proptosis and loss of vision. Resection followed by intensity-modulated radiation therapy has resulted in disease control.

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Optical Coherence Tomography of Optic Disc Swelling in Acute Primary Angle-Closure Glaucoma

Acute primary angle-closure glaucoma (APACG) is an ophthalmic emergency with poor outcome if initial recognition and treatment are delayed. Posterior segment signs are often difficult to detect owing to corneal edema, anterior chamber inflammation, cataract, and the use of miotics. Optical coherence tomography (OCT) is a noncontact, noninvasive imaging technology capable of obtaining high-resolution tomographic images of posterior segment ocular structures. We report a case of optic disc swelling and branch retinal vein occlusion after APACG with imaging performed with OCT and fluorescein angiography. We discuss the disc appearance on OCT imaging and a finding of a partial thrombus in the venous circulation on fluorescein angiography.

Report of a Case. A 64-year-old Chinese woman had pain, blurring of vision, and redness in the left eye for 5 days. She had no previous episodes of eye pain. Her visual acuity was 20/30 OD and hand motion OS with a left relative afferent pupillary defect and mid-dilated pupil. The left eye had conjunctival injection, corneal edema, and a shallow anterior chamber. The intraocular pressure (IOP) was 14 mm Hg OD and 54 mm Hg OS. Gonioscopy confirmed angle closure. The IOP was lowered with intravenous acetazolamide, topical timolol maleate, pilocarpine hydrochloride, and brimonidine tartrate. Topical steroids were administered. Six hours after initial examination, her IOP was 12 mm Hg.

The left cornea had cleared 12 hours after initial examination to allow a view of the left disc. This was swollen and hyperemic with peripapillary flame hemorrhages. The fundus was otherwise unremarkable. The IOP was 10 mm Hg OS. Bilateral peripheral iridotomy was performed. The OCT (OCT 3; Zeiss-Humphrey Systems, Dublin, Calif) of the left disc showed swelling with adjacent thickening of the retina in the papilla-macula region (Figure 1). The right disc showed no swelling.

One week later, the left disc was still swollen and hyperemic, with fluorescein angiography showing no central retinal vein occlusion. There was, however, a partial thrombus of the left superotemporal branch vein (Figure 2). One month later, the left disc was pale and cupped (0.7). The visual acuity was counting fingers OS.
Comment. Posterior segment pathologic findings reported after APACG include irreversible optic nerve damage, anterior ischemic optic neuropathy, vitreous hemorrhage, and ocular decompression retinopathy. Optical coherence tomography is a diagnostic imaging tool that can perform micron resolution imaging in the eye. Its high resolution permits the imaging of fine structures within the retina. In this report, OCT demonstrates that in APACG the retinal swelling involves all layers of the retina. The retinal thickness was observed to have decreased at 9 months (Figure 1) compared with initial examination. This finding could have been due to resolution of retinal swelling alone or in combination with loss of tissue. Unfortunately, no OCT retinal thickness measurements were obtained at the point of resolution of retinal swelling, which could then be used for comparison with the retinal thickness measurements at the same location 9 months after the attack of APACG. This may have demonstrated tissue losses at the posterior pole due to APACG, similar to those detected in primary open-angle glaucoma.

Retinal circulatory disturbances such as central retinal vein occlusion have been described in APACG, but branch retinal vein occlusion is rare. The partial thrombus in the superotemporal branch vein was unexpected.

The mechanism of disc swelling and subsequent optic atrophy that occurred at 1 month could be due
to marked ischemia that occurred from compression of vessels in the prelaminar region during the prolonged period of increased IOP. The resulting anoxia led to edema. With reduction in optic nerve head blood flow, there was irreversible ischemic damage to the axons. An alternative explanation is disc swelling due to blockade of rapid axonal transport during the prolonged period of increased IOP. The ganglion cells were unable to tolerate the severe blockade of transport, and axon death occurred. The presence of disc swelling at initial examination may be a prognostic marker for visual outcome, although to our knowledge this has not been reported in the literature.

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Cavitary Melanoma of Ciliary Body
Simulating a Cyst

Cystlike cavities rarely occur in uveal melanoma, and are recognized on ultrasonography as acoustically hollow regions. In such cases, diagnostic confusion between a solid or cystic lesion can lead to delay in treatment of melanoma. We report a 42-year-old man with a ciliary body mass that simulated a large cyst ultrasonographically but was later confirmed on enucleation to be a cavitary melanoma.

Report of a Case. A 42-year-old man had a 2-year history of decreased vision in his right eye. Following several unsuccessful refractions at a regional referral center, the patient was found to have a multicystic ciliary body mass inducing lenticular astigmatism and was referred to the Wills Eye Hospital Oncology Service (Philadelphia, Pa).

Visual acuity was 20/200 OD and 20/20 OS. The right eye had episcleral sentinel vessels inferiorly and superior subluxation of the cataractous lens. Fundus examination revealed a brown ciliochoroidal mass inferiorly, measuring 14 × 13 × 12 mm (Figure 1). On transillumination, a light shadow corresponding to the rim of the mass was found, but the mass transmitted light overall. B-scan ultrasonography revealed a multicystic lesion with the largest cyst measuring 11 mm in diameter, surrounded by a thin reflective wall and base measuring only 1.0 mm in thickness (Figure 2). The cavities occupied 90% of the lesion. Based on the solid basal component and the sentinel vessels, our diagnosis was cavitary melanoma of the ciliary body. Enucleation was performed (Figure 3).

Histopathological examination disclosed a ciliary body mass composed of spindle B malignant melanoma cells lining large intratumoral cavities. The cavities were filled with serous fluid and had no endothelial or epithelial lining (Figure 4 A-C). After a 1-year follow-up, the patient was healthy without metastasis.

Comment. Cystlike cavities are uncommon in uveal melanoma. In 1 report, microscopic cavitation was found by histopathological examination in 8.4% of uveal melanomas. However, clinically visible cavitation is very rare. We were able to find only 25 reported cases of cavitary uveal melanomas (Table 1 and Table 2).

Ultrasongraphy is an important diagnostic tool for uveal melanoma, particularly those with cavitary changes. On ultrasonography, cavitary melanomas can manifest single or multiple cavities, sometimes with fine echoes within the cavity. In reported cases, the mean volume of melanomas occupied by cavity was 54% (range, 30%-79%). Our case was unusual in that the cavity occupied 90% of the melanoma.