the patients was 39.5 years. The mean distance from the limbus to the center of the tumor for anterior neurofibroma and schwannoma was 4 mm. The relative size and topographic location of the lesions are shown in the inset of part A of the Figure.

Comment. Episcleral neurofibromas and schwannomas likely arise from the long posterior ciliary nerves within the sclera. The ciliary nerves pierce the sclera near the optic nerve and pass anteriorly within the sclera and suprachoroidal space while branching to form a loose connection between the long and short posterior ciliary nerves, leading to 20 to 30 branches at the level of the ciliary body. The nerve loop of Axenfeld is an anastomotic interconnection of the long ciliary nerve that occasionally turns to enter the sclera before turning back again to continue anteriorly to the ciliary body. The loops occur in the area 2 to 4 mm posterior to the limbus, and no loops occur in the zones directly anterior to the median and lateral rectus tendons.

Anterior episcleral nerve sheath tumors have a topographic distribution similar to the Axenfeld nerve loop, permitting speculation of their relationship. The distances from the center of the tumor to the limbus narrowly ranged from 3 to 4 mm. None of these tumors occurred directly anterior to the horizontal rectus muscle tendons (Table). The anterior tumors were evenly distributed among the remaining quadrants as described for Axenfeld nerve loops. Two of 4 episcleral schwannomas occurred in the posterior sclera. If the distribution were random in the ciliary nerves, then some tumors would be expected to be found in the horizontal plane and at more variable distances from the limbus.

The apex of the loop can extend through the full thickness of the sclera and project above the scleral surface. Pain and tenderness have been noted when the nerve loops are located anteriorly. The sharp bend in the nerve and the ensuing stress perhaps facilitate the abnormal proliferation of nerve sheath cells and the formation of these tumors. The differential diagnosis for all of the reported episcleral nerve sheath tumors includes solitary circumcised neurona, but in our case the published cases lack sufficient encapsulation to warrant this diagnosis.

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High-resolution spectral-domain optical coherence tomography (SD-OCT) is a new exciting technology for visualization of microstructural alterations in retinal diseases. We investigated the applicability of this noninvasive method to detect in vivo early vitreous seeding of a histologically proven choroidal melanoma with transretinal tumor extension (Knapp-Ronne melanoma).

Report of a Case. A 68-year-old man had blurred vision and inferotemporal visual field defect of the right eye for 4 months. His visual acuity was 20/30 OD and 20/20 OS. On fundoscopy, a large pigmented choroidal mass with overlying hemorrhages was observed in the superonasal quadrant (Figure, A). Subretinal fluid around the tumor and inferior serous retinal detachment were present. Echography revealed a solid mass of 4.8-mm prominence, 11.2 × 10.6 mm² base, and low homogeneous reflectivity, with a more reflective apical cap, implying a uveal melanoma rupturing the Bruch membrane and covered by an apical hemorrhage (Figure, B). Ultrasound could neither exclude nor confirm tumor extension through the retina.

The patient was investigated using a commercially available SD-OCT (Spectralis HRA+ OCT; Heidelberg Engineering, Heidelberg, Germany). Discrete, irregularly spheroidal bodies were present in the vitreous (Figure, C). The retina overlying the tumor showed thinning toward the tumor apex where the retina was completely obliterated and particles 20 to 30 µm in size could be detected in the immediately adjacent vitreous, suggesting transretinal seeding of the uveal melanoma (Figure, D). Without evidence of a primary or metastatic lesion elsewhere in the body, the tumor was classified as T2a N0 M0, according to the 2009 tumor, node, metastasis (TNM) classification system. Enucleation of the right eye was performed.

Macroscopically, the sectioned globe disclosed a 5-mm-high, 12-mm-wide, gray-white mass with a prominent brownish cap and dark pigmented foci (melanin) on the
tumor surface (Figure, E). Histopathological examination revealed a choroidal mixed-cell-type melanoma rupturing the Bruch membrane and showing intratumoral and supratumoral hemorrhages (Figure, F). Severe atrophic and degenerative changes were present in the overlying retina. At the tumor apex, the thinning had progressed to obliteration of the retina with absence of the internal limiting membrane. Within the supratumoral hemorrhage, melanoma cells could be found percolated through the attenuated retina and into the immediately adjacent vitreous (Figure, F, insert). Immunohistochemically, these intravitreal cells were positive for S-100, HMB-
45, PNL2, melan-A, tyrosinase, and MITF, but negative for iron stain.

Comment. In our patient, the clinical suspicion of trans-retinal seeding by a uveal melanoma raised by SD-OCT could be confirmed histologically. Primary choroidal melanoma with retinal perforation and extension into the vitreous (Knapp-Rønne type) is a rare entity occurring in about 1 in 250 of uveal melanomas. Detection of retinal perforation is valuable owing to increased risk of recurrent vitreous hemorrhage after radiotherapy and increased likelihood of rhegmatogenous retinal detachment after transscleral local resection.

However, early recognition of Knapp-Rønne melanoma can sometimes be a clinical challenge because ophthalmoscopic assessment, and even echography with a resolution of approximately 100 µm, might sometimes be too imprecise for imaging of focal retinal perforation.

In contrast, conventional OCT produces cross-sectional images with approximately 10-µm resolution for visualization of microstructural alterations in retinal diseases as well as of the overlying retina in choroidal tumors. Recently, SD-OCT technology has improved resolution up to 3.5 µm per pixel.

Using this technique, we could detect spheroidal bodies in the vitreous and, particularly, adjacent to the tumor apex where the overlying retina was completely obliterated. Although SD-OCT allows no exact differentiation between melanoma cells, melanomacrophages, or clusters of blood cells, our clinical findings presumed a Knapp-Rønne melanoma that could be confirmed histologically.

In the future, SD-OCT might become a helpful tool for clinical detection of vitreous seeding from uveal melanomas.

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Clinical Detection of Melanoma-Associated Spongiform Scleropathy by Ultrasound Biomicroscopy and Its Correlation With Pathological Diagnosis

Ciliary body and choroidal melanoma account for greater than 90% of all uveal melanomas. Extrac- scleral extension is an important prognostic factor in uveal melanoma and has been described in 8% of eyes enucleated in the Collaborative Ocular Melanoma Study. Infiltrating solid tumors may cause cellular and degenerative changes in connective tissue surrounding the tumor, contributing to tumor cell invasion. For these reasons, characterization of scleral changes adjacent to the tumor may be relevant to tumor invasion.

Melanoma-associated spongiform scleropathy (MASS) is a histopathological entity described as an area within the sclera adjacent to a choroidal or ciliary body melanoma where collagen fibers appear to have disintegrated into loose fibers. Melanoma-associated spongiform scleropathy is observed in approximately one-third of eyes with uveal melanoma. Biochemical analyses of MASS show decreased collagen and amino acids and increased glycosaminoglycans and water uptake. This degradation process, mediated by matrix metalloproteinases, may weaken the structural barrier and facilitate local tumor invasion. Alyahya observed MASS in 91% of eyes with tumor invasion, in contrast to 23% of eyes without. We report for the first time the clinical detection of MASS by ultrasound biomicroscopy (UBM) in ciliary body melanoma, with histopathological correlation.

Report of a Case. A 44-year-old man presented with painless decreased vision in the right eye for several months. Visual acuity was 20/25, and on dilated examination a melanocytic ciliochoroidal mass was identified abutting the lens (Figure 1A). Gonioscopy showed a pigmented mass invading the anterior chamber angle (Figure 1B).

B-scan ultrasound showed a large dome-shaped mass involving the ciliary body that measured 5.4 mm in thickness. There were cystic spaces in the lesion. A-scan demonstrated medium reflectivity with a decrescendo pattern. Ultrasound biomicroscopy showed intralesional cavities. The inner one-fourth of the sclera adjacent to the base of the tumor was hypechoic compared with the outer three-fourths of the sclera (Figure 2A). The thickness of the sclera over the lesion was deemed to be normal.

Diagnosis of a medium-sized, malignant, ciliochoroidal melanoma was made, and enucleation was performed. Gross examination showed a pigmented superotemporal ciliary body mass measuring 6 × 9 × 4 mm. The ciliary body melanoma was composed of spindle B and epithelioid melanoma cells, with large cavities containing eosinophilic exudate. Tumor cells were noted in the anterior chamber angle and iris root (Figure 2B) and extended to the inner sclera only. The inner sclera adjacent to the tumor showed a pale staining area relative to...