Intraocular melanocytoma is an uncommon tumor. To our knowledge, in none of the cases reported to date, either individually or in case series, has this tumor been associated with bone formation. We report 2 such cases.

Report of Cases. Case 1. A 14-year-old white girl had pain in her left eye of 5 weeks' duration. There was no history of ocular trauma or disease. Her visual acuity was 20/60 OS, and the intraocular pressure was 11 mm Hg. There was no evidence of glaucoma or uveitis. The lens was displaced inferonasally by a suprtemporal ciliary body mass.

Local resection of this mass was performed with deep scleral lamellar dissection. Postoperatively, the visual acuity was 20/60 OS; 6 months later, it had decreased to 20/200 OS because of ocular hypotony, macular edema, and epimacular membrane formation. The hypotony resolved with a short course of systemic steroids. At the last follow-up visit (28 months postoperatively), her visual acuity was 20/40 OS. The gross pathologic specimen consisted of deep sclera (18 × 11 × <1 mm) with an attached, deeply pigmented mass (15 × 11 × 5 mm).

Microscopy revealed a melanocytoma with extensive necrosis and bone formation (Figure 1). There was partial necrosis of the heavily pigmented tumor with extensive cholesterol cleft formation and adjacent fibrosis. The tumor extended into the sclera. Extensive calcium deposition consisted of an admixture of granules, coarse clumps, and calcopherites. Bone was present at the outer, scleral margin of the tumor, where densely pigmented viable tumor cells involved the sclera (Figure 1). The tumor cells had abundant cytoplasm and were large and either polyhedral or spindle shaped (Figure 2). Most nuclei were round or oval with small nucleoli; a proportion of the cells were more variable in nuclear and nucleolar size (Figure 2).
Results of immunohistochemistry showed that the surviving tumor cells were negative for melanoma marker (HMB-45), desmin, smooth muscle actin, glial fibrillary acidic protein, cytokeratins, and neurofilament protein. Staining with paninflammatory cell marker (CD45) and macrophage marker (CD68) confirmed the presence of multiple melanomacrophages, and smooth muscle actin delineated muscle elements in the ciliary body, vessel walls, and fibroblasts with adjacent fibrosis.

Case 2. A 48-year-old white man was referred with a pigmented ciliary body mass with a base of 6 × 7 mm in the left inferotemporal region; this mass extended for 5 mm into the peripheral fundus. There was no inferotemporal ciliary body involvement, and the tumor mass extended for 5 mm into the peripheral fundus. There was no angle involvement. The visual acuity was 20/25 OD and 20/25 OS. The intraocular pressure was 18 mm Hg OU. Ultrasound biomicroscopy showed that the mass arose at the posterior edge of the ciliary body and involved the pars plana. It was 2.6 mm thick.

Clinically, the tumor appeared to be a melanoma of the ciliary body and peripheral choroid. It was not as heavily pigmented as one might expect with a melanocytoma. The patient was given the option of observation, but because of anxiety about ocular tumor growth and spread as well as a history of thyroid cancer, he wanted the tumor removed.

Results of computed tomographic studies of the abdomen were negative. Cryopexy over a band 2 to 3 mm to the base of the tumor was performed to secure the retina. Ten weeks later, scarring was present around the base of the tumor, and the retina was attached. Laser treatment reinforced the posterior boundary of the tumor preoperatively. Excisional biopsy was performed through a scleral lamellar–hinged opening. Vitreous hemorrhage complicated the immediate postoperative period; pars plana vitrectomy was performed. Six months later, the visual acuity was 20/25 OS. The gross pathologic specimen included a portion of the left inferotemporal ciliary body that was circular in outline, 8 mm in diameter, 10 mm at the base, and 5 mm in height (Figure 3). Microscopy revealed a melanocytoma with bone formation. Bleached sections showed uniformity of plump, rounded cells with abundant cytoplasm, small nuclei, and small nucleoli (Figure 4). There was no evidence of mitoses or necrosis. A plaque of mature bone lay within the uvea, superior to the tumor mass and covered by an intact pigment epithelium; there was no evidence of pigment hyperplasia (Figure 5).

Comment. Melanocytoma of the uvea is a rare, benign, heavily pigmented, and slow-growing tumor. To date, 41 cases of melanocytoma of the ciliary body have been reported.1 To our knowledge, neither bone nor calcification has previously been described in association with this tumor.

Melanocytoma may become necrotic with the production of cholesterol clefts. The sclera may also be involved. In case 1, the presence of bone and a few dendritic cells raised the possibility of a necrotic teratoid or choristomatous lesion. Such lesions may become necrotic and calcify.2,3 The lack of muscle, epithelial, or neural...
markers in the surviving tumor cells argued against this diagnosis, and the cell nuclei were compatible with those of a melanocytoma. The clinical features, morphologic characteristics, and lack of cytokeratins argued against a reactive or neoplastic process of the pigment epithelium of the ciliary body. We concluded that the tumor in case 1 was a variant of necrotic melanocytoma in which the calcification and bone formation were secondary degenerative changes. The few cells with atypical nuclei or cytoplasmic morphologic features were probably macrophages or vascular elements.

In case 2, the tumor showed neither necrosis nor malignancy. The microscopic appearance of the characteristic melanocytoma cells was that of type 1 cells with giant uniform melanosomes. The lack of smaller, type 2 spindle-shaped cells may be attributed to tissue sampling but does support a pattern of slow tumor growth.

The plaque of bone that overlay the apex of the tumor was within the uveal tissue. Bone may be present in congenital malformations and as choroidal osteoma. Chan et al described bone in association with a necrotic choroidal melanoma; the plaque of calcified bone was within an area of fibrosis at the site of the ruptured Bruch membrane and was entirely surrounded by malignant melanoma cells. Interestingly, bone has been reported in association with extraocular pigmented and nonpigmented melanoma.

Bone appearing external to the retinal pigment epithelium (RPE) may have originated from uveal mesenchyme. In nonocular tissue, the fibroblast is the source of heterotopic bone formation. The RPE is considered responsible for the bony metaplasia that occurs in end-stage eye disease.

Neither case 1 nor case 2 showed evidence of RPE hyperplasia. In case 2, the RPE internal to the tumor appeared intact. The bone was confined to the uveal tissue, and there was no evidence of neovascularization due to cryotherapy; thus, bone formation in a neovascular membrane could not have occurred. In case 1, the bone lay adjacent to the sclera, remote from the RPE, and may have been part of the degenerative process in this necrotic tumor. In contrast, the tumor cells in case 2 were viable. Because a melanocytoma is a form of nevus and a nevus is a hamartoma, in this case the association with bone...
may be considered part of the hamartomatous process.

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Atypical Manifestation of Multiple Evanescent White Dot Syndrome With Large Peripapillary Lesion

Multiple evanescent white dot syndrome (MEWDS) is on the spectrum of diseases that includes acute zonal occult outer retinopathy, acute idiopathic blind spot enlargement syndrome, punctate inner choroidopathy, multifocal choroiditis and panuvitis, and acute macular neuroretinopathy.1 We present an atypical case of MEWDS in which a strikingly large white peripapillary outer retinal lesion evolved into a lesion with a more classical MEWDS appearance.

Figure 1. Fundus photograph obtained at the patient’s initial examination demonstrated a peripapillary chorioretinal scar in the right eye (A) and a peripapillary lesion 7 disc areas in size in the outer retina or retinal pigment epithelium layer in the left eye (B). Digital fluorescein angiography in the left eye demonstrated patchy peripapillary choroidal filling and disc hyperfluorescence at 13 seconds (C) and staining of the disc and peripapillary vessels at 15 minutes (D).