role of iron export in AMD needs further investigation, but this report provides evidence that iron overload can occur in AMD not only in the RPE and Bruch’s membrane but also in the neurosensory retina. Because iron can cause oxidative stress, it may be toxic to both the RPE and photoreceptors, cells in which cell death leads to blindness in AMD. Iron chelation therapy may one day prove useful in the prevention of vision loss due to AMD.

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Optical Coherence Tomographic Findings of Combined Hamartoma of the Retina and Retinal Pigment Epithelium in 11 Patients

Combined hamartoma of the retina and retinal pigment epithelium (RPE) is an uncommon fundus tumor with classic clinical features. In 1984, Schachat et al2 published 60 cases collected from the members of the Macula Society and described the clinical features and natural course of this benign lesion. They noted that...
The general clinical data included patient features at initial examination such as age, sex, and race. The ocular data included visual acuity, symptoms, tumor color, tumor quadrant and anteroposterior location, tumor distance to the optic disc and foveola, and largest tumor basal dimension and thickness. Additional clinical data at the site of the combined hamartoma included the presence of posterior vitreous detachment, preretinal fibrosis, retinal exudation, retinal edema, retinal hemorrhage, retinal atrophy, subretinal fluid, and RPE hyperplasia or atrophy. The OCT data at the site of the combined hamartoma included presence of posterior vitreous detachment, vitreoretinal traction, preretinal membrane, retinal striae, retinal disorganization, retinal edema, subretinal fluid, and RPE irregularity or detachment. Optical reflectivity of the inner and outer portions of the tumor were recorded. Retinal edema was classified as cystoid vs noncystoid based on the presence or absence of intraretinal optically lucent cysts. The macula was defined as the round area within 3 mm of the foveola.

Results. There were 11 consecutive patients who had a combined hamartoma of the retina and RPE imaged with OCT. The clinical features are listed in Table 1. In all cases, the tumor was unilateral and unifocal and appeared to be gray-brown. The initial symptom at presentation was blurred vision (n = 6), strabismus (n = 2), or asymptomatic (n = 3). The meridional location of the tumor was macula (n = 8), inferior (n = 1), and nasal (n = 2). The

<table>
<thead>
<tr>
<th>Patient No./ Age, y/Sex/Race</th>
<th>Tumor Location</th>
<th>Tumor Basal Dimension, mm</th>
<th>Tumor Ultrasound Thickness, mm</th>
<th>Posterior Vitreous Detachment</th>
<th>Preretinal Fibrosis</th>
<th>Retinal Traction</th>
<th>Foveal Ectopia</th>
<th>Intraretinal Hemorrhage</th>
<th>Intraretinal Exudation</th>
<th>Subretinal Fluid</th>
<th>Adjacent RPE Alterations</th>
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<tr>
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<td>14</td>
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<td>13</td>
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Abbreviations: A, Asian Indian; F, female; M, male; RPE, retinal pigment epithelium.
The mean basal dimension of the tumor was 7 mm (median, 7 mm) and the mean thickness of the tumor as measured by ultrasonography was 1.8 mm (median, 2.0 mm). The tumor margin was a mean of 0.2 mm to the optic disc (median, 0 mm; range, 0-2.0 mm) and a mean of 0.4 mm from the foveola (median, 0 mm; range, 0-2.0 mm). Choroidal neovascularization was not present in any patient by clinical examination or OCT.

The OCT findings are listed in Table 2. A preretinal membrane was visualized by OCT in 10 patients. The preretinal membrane was configured in peaks, reflecting vitreoretinal traction in 3 patients (Figure 1). The retina was drawn into folds or striae by preretinal membrane in 9 patients (Figures 2, 3, and 4). Retinal anatomical disorganization with loss of identifiable retinal layers at the site of the mass was noted in all patients (Figures 1-4). The mean retinal thickness at the site of the lesion was 766 µm (median, 790 µm). The adjacent flat retina appeared to be of normal thickness and anatomical configuration.

Ultrasonography was performed on 8 patients and revealed no evidence of posterior vitreous detachment, vitreous traction, or preretinal membrane in any patient. The lesion showed acoustic features of echolucency (n=2), echodensity (n=0), and isodensity (n=6). There was no acoustic shadowing or intraleSIONAL calcification in any patient.

Comment. Epiretinal membrane formation is a prominent clinical feature of combined hamartoma of the retina and RPE. In the collaborative report by Schachat et al, only 3 of 60 patients had surgical removal of the epiretinal membrane; visual improvement was achieved in 1 patient. Histopathologic findings of those membranes showed fibrous astrocytes, fibrocytes, fragments of the internal limiting membrane, new collagen, and vitreous collagen. McDonald et al described 2 patients with combined hamartoma of the retina and RPE in which surgical removal of epiretinal membrane was performed and visual acuity did not improve in either case. It has been speculated that irreversible visual loss was present in these slightly older patients (ages 44 and 26 years) with a presumed congenital tumor. Perhaps earlier intervention would have been warranted. Stallman and Mason and Kleiner each reported a case of visual improvement following surgical removal of epiretinal membrane in younger patients with combined hamartoma. The benefits of surgery in such cases and the overriding influence of amblyopia continues to be debated.

In a discussion, Gass speculated that the membrane might be intricately interwoven within the dysplastic retina. Stallman later commented that the membrane was surgically separable from the tumor so...
it most likely was extrinsic to the retinal tumor. He suggested newer imaging modalities such as OCT to delineate the precise location of the membrane as it could influence the surgeon’s approach to this mass.

In 2002, Ting et al reported the first observations on OCT of combined hamartoma of the retina and RPE in 2 adult patients. They noted important findings such as a thickened retinal mass with hyperreflective surface and deep shadowing and they commented that the adjacent retina appeared to be normal and separate from the mass. Cystoid edema was found in 1 patient. In our series of 11 patients, we contribute several additional observations regarding OCT of this lesion. Eight of our 11 patients were teenagers or children. Optical coherence tomography is fairly well tolerated by children and provides more clinical detail of the retina than is visible by indirect ophthalmoscopy or ultrasonography. We observed a distinct epiretinal membrane with secondary retinal folds and striae in almost all patients. The membrane showed horizontal traction in 9 patients and vertical traction with peaks in 3 patients. The membrane was preretinal with no evidence of intertwining into the tumor. Additional findings included retinal disorganization in all patients. The adjacent retina was normal in architecture and seemed to gradually thicken into the disorganized tissue. One might speculate that the tractional component from the epiretinal membrane was the sole source for the distorted retinal findings. However, others have speculated that the epiretinal component could be secondary to the retinal tumor.

Despite the fact that all but 3 of our patients were children or teenagers and the disease seemed to be
detected early in life, the tractional forces of the epiretinal membrane were associated with severe retinal microarchitecture compromise. Future studies are needed to evaluate the effect of earlier detection and elimination of the epiretinal membrane in prevention of permanent retinal disorganization and visual loss. Optical coherence tomography can provide important information regarding the vitreoretinal interface of this tumor and could influence surgical decisions.

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Dynamic Atypical Optic Nerve Coloboma Associated With Transient Macular Detachment

Though infrequently encountered by most ophthalmologists, optic nerve pits and optic nerve colobomas (typical and atypical) are well known cavitary optic disc anomalies.1-2 Maculopathy characterized by macular schisis, outer layer detachments, and occasionally outer layer holes are frequently associated with cavitary optic disc anomalies.3-4 The etiology of the maculopathy and source of the subretinal fluid are unknown but both systemic and intraocular factors have been proposed.3-9 Fluctuations, and even spontaneous resolution, of the described maculopathy can occur but are uncommon.1,3,5,10 Though other cavitary optic disc anomalies have rarely been reported to fluctuate in appearance,11-13 we are unaware of any previous reports of fluctuating optic pits or atypical colobomas. We present 3 unique cases in which the fluctuating appearance of an atypical optic nerve coloboma is associated with spontaneous resolution of the associated maculopathy.

Report of Cases. Case 1. A 40-year-old white woman sought care because of a 1-week history of decreased vision in her right eye. The family history was negative for colobomas or optic nerve disease but 1 brother had a history of cataract and retinal detachment. Best-corrected acuity was measured at 20/200 in both eyes. The visual acuity of the left eye had slowly deteriorated 4 years prior due to progression of low tension glaucoma. At initial examination, she was using Timoptic-XE (Merck & Co Inc, Whitehouse Station, NJ) and Azopt (Alcon Laboratories Inc, Fort Worth, Tex) in both eyes. Previous neuroimaging results with computed tomography and magnetic resonance imaging were both normal. When first examined, the optic nerve of the right eye was normal in size and color and contained a small slit-like cup (Figure 1A). No optic pit or peripapillary pigment changes were seen. A large macular schisis with a large outer layer detachment was noted. The fovea appeared cystic and no outer layer hole could be identified. A posterior vitreous detachment could not be identified. Late frames of the fluorescein angiogram showed mild disc hyperfluorescence. The left disc was of normal size without peripapillary pigmentary changes and contained a large, uniformly deep cup (cup-disc ratio of 0.95). The macula appeared normal. We could identify the base of the cup and no distinct excavation was seen. Observation was recommended. Over the next 6 months, the acuity of the right eye improved to 20/40. The schisis and outer layer detachment resolved, but macular retinal pigment epithelial changes corresponding to the area of the schisis were still present. A distinct optic nerve cup containing a central excavation became visible (Figure 1B). Funduscopic examination findings of the left eye were unchanged throughout the course of observation. Optical coherence tomography (OCT) was not available. The optic nerve and macular appearance could not be altered by changes in body position or by light stimulation of the eye.

Case 2. A 30-year-old white man was seen for a 1-week history of blurred vision in the left eye. Family history was negative for colobomas or optic nerve disease. Corrected acuity measured 20/20 OD and 20/160 OS. The patient had previously undergone bilateral myopic photorefractive keratectomy and had a postkeratectomy uncorrected acuity of 20/20 in both eyes. The optic nerve of the right eye appeared normal with a cup-disc ratio of 0.3. The floor of the cup was clearly visible and the macula appeared normal. The left optic nerve was normal in size without peripapillary pigment changes. The nerve was pink with slightly blurred margins and contained a thin, centrally located, slit-like cup without a visible excavation (Figure 1C). A large macular schisis cavity with a smaller,