Optical coherence tomography (OCT) plays an important role in the diagnosis of retinal diseases with minimal ophthalmoscopic changes. For example, in eyes with acute zonal occult outer retinopathy (AZOOR), an abnormality of the photoreceptor inner segment–outer segment (IS/OS) junction found by OCT was spatially correlated with the region of visual field defect. Recent high-resolution spectral-domain OCT images have shown a thin line between the IS/OS junction and the retinal pigment epithelium, where photoreceptor OS disc membranes are continuously shed for renewal. Thus, the appearance of the COST line may reflect the normal function of the photoreceptor OSs more closely than the IS/OS junction. In both cases, the fundus appeared normal and the COST line was not present or appeared indistinct in the region of visual field defect. Our findings suggest that the COST line may be an early indicator of cone photoreceptor dysfunction in eyes with minimal ophthalmoscopic abnormalities.

Report of Cases. Patient 1 (a 24-year-old woman) and patient 2 (a 28-year-old woman) both had sudden unilateral visual disturbances following photopsia. The visual acuities were 0.02 OD and 1.5 OS in patient 1 and 0.15 OD and 1.5 OS in patient 2. Goldmann kinetic perimetry revealed a blind spot enlargement and central scotoma in the right eye of both patients (Figure 1 and Figure 2). The anterior segment and fundus were normal; however, fluorescein angiography showed a slightly motled hyperfluorescence around the macula in the affected eye of both patients. The full-field scotopic ERGs were normal, but there were phase delays in the photopic 30-Hz ERGs in the affected eyes: 5.7 milliseconds in patient 1 and 8.0 milliseconds in patient 2. In addition, the amplitudes of the photopic b-waves were reduced in both patients. The focal macular ERGs (ER80; Kowa Co, Tokyo, Japan) in the central 15° were almost flat in the affected eye in both patients. Neither patient had systemic disorders such as viral infections or autoimmune diseases.

Spectral-domain OCT (Carl Zeiss Meditec, Dublin, California) showed the IS/OS junction clearly, even in the region of the scotoma. However, the COST line was not detected in patient 1 and appeared indistinct in patient 2. Moreover, the bulgelike structure of the IS/OS junction at the fovea (with the foveal bulge indicating a domelike appearance of the IS/OS junction due to an elongated cone OS at the fovea) could not be observed in the affected eyes. The visual disturbances of these patients did not recover, and these abnormalities in the OCT images were observed at all examinations for 50 months in patient 1 and 18 months in patient 2 after the onset.

Selective Abnormality of Cone Outer Segment Tip Line in Acute Zonal Occult Outer Retinopathy as Observed by Spectral-Domain Optical Coherence Tomography


Comment. To our knowledge, this is the first report of AZOOR where the boundary of the IS/OS junction in the OCT images was well preserved but the COST line was absent or indistinct from the initial examination through the entire follow-up period. Earlier studies demonstrated that a loss or irregularity of the IS/OS junction observed by OCT corresponded well with the visual field defects even at the early stages of AZOOR, and the abnormality in the IS/OS junction can improve following recovery of the scotoma. These findings have led to the hypothesis that photoreceptor OS dysfunction is the primary lesion in AZOOR. The COST line corresponds to the junction between the photoreceptor tips and the apical processes of the retinal pigment epithelium, where photoreceptor OS disc membranes are continuously shed for renewal. Thus, the appearance of the COST line may reflect the normal function of the photoreceptor OSs more closely than the IS/OS junction. In fact, in all of the AZOOR cases we have recently examined, the COST line was always absent in the region of IS/OS abnormalities, suggesting that the abnormality of the COST line may precede that of the IS/OS junction. In our 2 cases, the fundus appeared normal and the IS/OS junction was clearly observed in the region of the COST line abnormality for 50 and 18 months after the onset. The focal macular ERGs, however, were markedly reduced in the affected areas. In the OCT images, the cone photoreceptor dysfunction corresponding to the region of scotoma could be detected only by the abnormality of the COST line.
Our findings suggest that the dysfunction of the cone photoreceptor OS could be initially reflected by an absence or indistinctness of the COST line and the absence of the foveal bulge. These changes may be followed by the development of abnormalities in the IS/OS junction in the more advanced stages. However, in our cases, the IS/OS junction remained the same during the entire follow-up period. This may suggest another possibility that our 2 cases constitute a subtype of AZOOR.

However, in another case of AZOOR with a blind spot enlargement and relative central scotoma (a 21-year-old woman, data not shown), both the IS/OS and COST lines disappeared in the peripapillary region where visual field disturbance was severe, whereas only the COST line disappeared and the IS/OS line remained normal in the foveal region where the visual field disturbance was milder. These findings support the idea that the visibility of the COST line is more easily affected than that of the IS/OS line.
the IS/OS line at an earlier stage by the pathological changes in a typical case of AZOOR. We should note that care should be taken in evaluation of the COST line because its visibility is dependent on the intensity and direction of the laser light that reaches the photoreceptor layer. However, in patients with AZOOR, the COST line and the foveal bulge observed by OCT could help as indicators of early cone photoreceptor dysfunction in cases with minimal ophthalmoscopic and angiographic abnormalities.

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Adult Ovarian Retinoblastoma Genomic Profile Distinct From Prior Childhood Eye Tumor

We report the first case of a woman, previously cured of childhood intraocular retinoblastoma, who developed tumor in the ovary with histological and genomic characteristics suggesting an independent retinoblastoma, not a metastasis.