Retinal and Choroidal Findings in Oxalate Retinopathy Using EDI-OCT

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ABSTRACT
A 55-year-old woman with extensive retinal crystal-line deposition secondary to primary hyperoxaluria presented with bilateral loss of vision secondary to oxalate retinopathy. Enhanced depth imaging optical coherence tomography revealed intraretinal, subretinal, and intraretinal and subretinal pigment epithelium, and choroidal focal hyperreflective structures consistent with both neurosensory and uveal deposition of oxalate crystals. Serial optical coherence tomography revealed continued crystalline deposition with progressive retinal atrophy. [Ophthalmic Surg Lasers Imaging 2012;43:S142-S144.]

INTRODUCTION
The retinal deposition of calcium-oxalate crystals in primary hyperoxaluria, an autosomal recessive disorder caused by a deficiency of the liver enzyme alanine-glyoxylate aminotransferase, has been well characterized from a clinical standpoint.1,2 Traditional fundus findings are variable, but can include subretinal black ringlets/hyperpigmentation, optic disc pallor, intravascular crystals, or extravascular crystals.1,2 Only preliminary studies have described the optical coherence tomography (OCT) findings of retinal oxalosis, which have included small localized dome-shaped lesions at the retinal pigment epithelium (RPE) and variable reporting of intraretinal deposits.3,4 Although traditional OCT technology obtains high-resolution cross-sectional images of the retina, it has typically lacked sufficient choroidal visualization for assessment of choroidal details. Enhanced depth imaging OCT (EDI-OCT) provides excellent choroidal visualization, allowing improved understanding of the choroidal involvement of various disease processes.5-7 In this study, we describe the retinal and choroidal architectural features of retinal oxalosis as imaged with EDI-OCT and the architectural changes noted over time.

CASE REPORT
A 55-year-old woman diagnosed as having primary hyperoxaluria presented with bilateral vision loss secondary to proliferative retinopathy secondary to retinal oxalosis. Visual acuity was 20/200 in the right eye and counting fingers in the left eye. Due to bilateral recurrent vitreous hemorrhage, bilateral vitrectomies with endolaser photocoagulation were performed. One year after surgical intervention, visual acuity remained poor at hand motions in the right eye and 20/400 in the left eye with continued crystalline deposition and macular pigmented changes (Figure 1). At presentation, EDI-OCT revealed small hyperreflective deposits throughout the inner and outer retina, consistent with crystalline deposition. Hyperreflective deposits at the level of the RPE were noted, consistent with sub-RPE and/or intra-RPE deposition. In addition to the previously described...
hyperreflective RPE deposits, focal areas of ovoid hyperreflective deposits were also noted (Figures 2A-2B). The disorganization of the RPE line limits definitive identification of deposit location. Additionally, areas of focal hyperreflectivity were noted within the choroid, suggestive of choroidal deposition (Figures 2A-2B). One year later, repeat EDI-OCT demonstrated increased size and extent of the deposits, evidenced by the increased shadowing effect and increased size of hyperreflective areas throughout the retina, RPE, and choroid. Alterations of retinal architecture were also noted and indicative of progressive outer retinal atrophy (Figures 2C-2D).

DISCUSSION

In primary hyperoxaluria, systemic accumulation of oxalate leads to deposition of oxalate crystals in multiple tissues throughout the body, including the retina. In this study, we describe the retinal and choroidal architectural changes in primary hyperoxaluria visualized with EDI-OCT. In our patient, extensive focal areas of hyperreflectivity were noted throughout the retinal and subretinal tissues. Near total retinal arterial obliteration was seen with extensive intraarterial crystalline deposition. Using EDI-OCT, visualization of the choroid was achieved and confirmed the presence of hyperreflective material within the choroid, suggesting choroidal deposition of oxalate crystals. Consecutive OCT imaging over 1 year revealed the dynamic nature of the crystalline deposition with increased amounts and density of the hyperreflective lesions. In our review of the literature, this study represents the first description of EDI-OCT in oxalate retinopathy and the subsequent finding of potential choroidal involvement of crystalline deposition.

Two previous studies have provided insight on the preliminary OCT findings in oxalate retinopathy. One study focused on a 19-year-old man and described...
tiny hyperreflective lesions localized within the areas of dome-shaped RPE associated with retinal oxalate deposits, but intraretinal hyperreflective deposits were not described. The other study described the OCT features in a boy with primary hyperoxaluria, including massive retinal edema and subretinal fluid, a thickened and irregular RPE, and both intraretinal and subretinal hyperreflective deposits. OCT imaging of other crystalline retinopathies has also been described. Bietti crystalline retinopathy has been shown to have outer retinal structures, with a similar pattern of a hyporeflective core and hyperreflective outer area, although larger in size. Comparatively, tamoxifen maculopathy crystals present far less dramatically on OCT, but pseudocysts can appear prominently. West African crystalline retinopathy presents with crystalline maculopathy crystals as small highly reflective bodies.

In this study, we describe the retinal and choroidal OCT features of advanced retinal oxalosis in a woman with continued disease progression over time. Although focused in the intravascular and sub-RPE space, crystalline deposition appears to be a panretinal and choroidal process. Progressive atrophy and retinal architecture loss likely occur due to significant ongoing ischemia and potential direct effects of crystalline deposition. Further research is needed to better understand the role of the choroidal deposition of crystals and disease progression.

REFERENCES