Optical Coherence Tomography Demonstration of Macular Infarction in Sickle Cell Retinopathy

Sickle cell retinopathy is caused by retinal ischemia secondary to the sickling of red blood cells in retinal arterioles, which supply nutrients to the ganglion cell layer, inner nuclear layer, and Muellerian glia of the retina. Macular infarction due to sickle cell disease has been documented using fluorescein angiography, electroretinography, and histopathologic examination. However, optical coherence tomography (OCT) of sickle cell retinopathy has, to our knowledge, never previously been reported. We report a case of macular atrophy secondary to retinal arteriolar occlusion in a patient with sickle cell disease, documented by standard-resolution OCT and ultra-high-resolution OCT, a new imaging technology capable of 2- to 3-µm resolution in the axial direction compared with 10 µm with standard-resolution OCT.

Report of a Case. A 26-year-old man with sickle cell disease (HbSS) went to the emergency department during an acute sickle cell crisis and was admitted to the medical intensive care unit with myocardial infarction, acute renal failure, and cholecystitis. He was referred to the retina service at the New England Eye Center, Boston, Mass, with a sudden decrease in visual acuity in both eyes and with central distortion in the left eye. On examination, uncorrected visual acuity measured 20/60 OD and counting fingers OS, without improvement on manifest refraction in the left eye. Intraocular pressures were 10 mm Hg OD and 12 mm Hg OS. Anterior segment examination was unremarkable in both eyes. Retinal whitening secondary to occlusion of branch arterioles was present in both eyes, with involvement of the fovea in the left eye (Figure 1A and B). Peripheral retinal examination demonstrated involuted neovascular fronds with evidence of peripheral nonperfusion in both eyes. Visual acuity remained stable 1 month following the patient’s visit, with resolution of the retinal whitening. Residual, fine retinal pigment epithelium changes in the area of arteriolar occlusion were visible (not shown).

Standard-resolution OCT images obtained at the 1-month follow-up visit demonstrated marked thinning of the retina in the temporal macula of both eyes, with greater foveal involvement in the left eye (Figure 2A and B). Ultra-high resolution OCT was performed, which again showed thinning of the temporal macula in both eyes, specifically involving inner retinal layers while sparing the photoreceptor and retinal pigment epithelium layers (Figure 2C).

Comment. The retinal vessels supply blood to the ganglion cell and inner nuclear layers of the retina whereas the choriocapillaris nourishes the photoreceptors and the retinal pigment epithelium. As vessels in the choriocapillaris are of larger caliber, it is rare for them to occlude and cause outer retinal ischemia. However, the inner retinal layers are prone to ischemia, as the retinal vessels are end arterioles and capillaries. Histopathologic studies of sickle cell retinopathy and other vaso-occlusive diseases have previously shown selective atrophy of the inner retinal layers of the macula in several eyes after retinal infarction.

Our patient had clinically visible whitening in circumscribed areas of the macula on his initial visit. Five weeks later, macular thinning was noted in these ischemic areas on standard-resolution OCT, contrasting with regions of normal retinal thickness where vessels were left unoccluded. Ultra-high resolution OCT showed the retinal atrophy to specifically involve the inner retinal layers while sparing the photoreceptors and the retinal pigment epithelium. We would expect to see similar findings in other arteriolar occlusive diseases of the retina.

Figure 1. Fundus photograph of the right eye at the initial visit, showing retinal whitening in the distribution of the retinal arteriolar occlusions in the macula (A), and fundus photograph of the left eye at the initial visit, showing retinal whitening in the distribution of the retinal arteriolar occlusions in the macula, including the fovea (B).
The measurement of retinal thinning with OCT might therefore be useful to document retinal infarction and its repair in patients with known vaso-occlusive disease.

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