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Isolated Choroidal Macrovascular: A Track-like Choroidal Lesion

Justis P. Ehlers MD¹, Hani Rayess BS^{1,2}, and Richard F. Spaide³

¹Ophthalmic Imaging Center, Cole Eye Institute, Cleveland Clinic, Cleveland OH

²Case Western Reserve School of Medicine, Cleveland OH

³Vitreous Retina Macula Consultants of New York, New York, NY

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Introduction

Enhanced depth imaging optical coherence tomography (EDI-OCT) has provided significant new insights into the choroidal involvement in numerous vitreoretinal diseases, including age-related choroidal atrophy and central serous.^{1,2} In this report, a patient presented with a lesion suggesting a variety of diagnoses including a potential intraocular parasite, but non-invasive visualization of ocular structures, particularly the choroid, established the diagnosis of a rare ocular lesion: a choroidal macrovessel possibly secondary to an aberrant long posterior ciliary artery.

Case

A 76 year-old asymptomatic female was referred for evaluation of a possible “worm” in the right eye. Visual acuity was 20/30. Examination revealed macular drusen with a prominent temporal curvilinear red-orange lesion extending from the temporal fovea to the periphery. The lesion appeared deep, but caused elevation of the overlying retina. Associated pigment rarefaction and isolated pigmentary clumping with a bulbous termination in the macular region were noted. Fundus autofluorescence demonstrated variable hyperautofluorescence over the lesion. Fluorescein angiography showed a subtle transmission defect. Indocyanine green angiography showed a large vascular structure with overlying blocking caused by the focal areas of hyperpigmentation. The vessel filled early suggesting it to be arterial. The intensity of the fluorescence paralleled the fluorescence seen in the surrounding, presumably normal choroidal vasculature (Figure 2). EDI-OCT demonstrated a large hyporeflective tubular structure in the choroidal space consistent with a choroidal macrovessel. Above the

Corresponding Author: Justis P. Ehlers, MD; Cole Eye Institute, 9500 Euclid Ave/i32, Cleveland OH. Phone: 216 318-7129. ehlersj@ccf.org.

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retinal pigment epithelium (RPE), there was debris in the subretinal space (Figure 3A). The areas of focal hyperpigmentation appeared as homogenous mounds in the subretinal space or as focal thickening of the RPE band (Figure 3B and 3C). The lesion remained asymptomatic and unchanged for over a year of follow-up. The left eye was normal except for macular drusen.

Discussion

In this report, we describe a rare clinical entity of a choroidal macrovessel that was characterized using multimodal imaging, particularly indocyanine green angiography and EDI-OCT. The differential diagnosis for the observed lesion includes trauma, neoplasm, intraocular parasites, inflammatory conditions, and anomalous posterior ciliary vessels or nerves. Visualization of a large hyporeflective tubular structure in the choroid with filling characteristics similar to the surrounding choroidal vessels established the diagnosis. Additional data obtained from the multimodal imaging allows us to surmise information about the concurrent physiologic changes that could be present. The OCT images revealed debris in the subretinal space with corresponding hyperautofluorescence originating from the same topographic location. The wavelengths employed for the autofluorescence principally detect retinoids, suggesting there was accumulation of shed outer segments in the subretinal space.

Though originally referred for a possible subretinal “worm,” the patient had no travel outside the United States or other at-risk behaviors. Nematode infestations that may produce subretinal tracts include *Oedemagena tarand*, *Alaria americanus*, and DUSN.³⁻⁵ Although the lesion in the case was “worm-like” in appearance, there was no evidence of inflammation or chorioretinal lesions consistent with nematode infection.

One prior case report was identified that described a choroidal macrovessel.⁶ In that report, a temporal macular orange-red lesion was present that demonstrated vascular perfusion. That patient had unexplained chorioretinal “spots” in the affected eye that suggests the possibility of inflammatory changes in the reported case, which were not present in our patient. Past reported cases of vortex varices involved the ampulla and therefore were venous. In this case, the lesion filled rapidly on ICG angiography suggesting this lesion was an artery.

The overall pathogenesis of choroidal macrovessels remains unclear, including whether it is congenital or acquired and whether there are potential systemic associations. This report demonstrates a rare clinical entity and the importance of a multimodal approach to imaging to facilitate diagnosis.

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Figure 1.

A. Standard fundus photograph of the right eye (OD) revealing reddish-orange serpiginoid temporal macular lesion with overlying RPE changes. B. Ultra-widefield color fundus photo of the right eye showing peripheral extension of the lesion. C. Ultra-widefield fundus autofluorescence with hyperfluorescence along the length of the lesion.

Figure 2.

A. Early phase ultra-widefield fluorescein angiography with faint hyperfluorescence (white arrow) in vicinity of the lesion corresponding to choroidal filling. B. Montage indocyanine green angiography revealing filling of the lesion. C. Ultra-wide field isolated red channel fundus photograph showing similar appearance of the lesion and other choroidal vessel.

Figure 3.

A. Vertical 5-line raster optical coherence tomography (OCT) B-scan through the foveal showing increased reflectivity at the RPE with elevation of the RPE in the area of the lesion with tubular hyporeflectivity of the underlying choroidal lesion. B. EDI-OCT along lesion in posterior pole revealing prominent choroidal hyporeflective tubular structures (white arrow) with overlying increased hyperreflective material at the level of the retinal pigment epithelium. C. Mid-peripheral EDI-OCT along lesion showing similar findings as B.