

Amblyopia Associated with Prominent Cilioretinal Artery

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INTRODUCTION

The cilioretinal artery, which derives from the circle of Zinn, is formed by small branches from the short posterior ciliary arteries. This artery goes through the optic nerve to participate as a parallel source for perfusion and blood circulation to the inner retina. Cilioretinal arteries, which are the most common congenital vascular anomalies of the retina, have been reported in 6–30% of normal eyes.^[1-3] The significance of this artery is preservation of macular perfusion in eyes with occlusive retinal vascular diseases. Herein, we report the spectral domain optical coherence tomography (SD-OCT) features of a patient with aberrant and prominent cilioretinal artery extending to the center of the fovea resulting in macular distortion, visual disturbance and amblyopia.

CASE REPORT

A 7-year-old girl was referred to the ophthalmology clinic for amblyopia. Best corrected visual acuity was 10/10 and 7/10 in the right and left eyes, respectively. Full cyclorefraction showed no significant refractive error in both eyes and no deviation was evident. Anterior segment examination yielded no pathologic finding. Dilated funduscopy revealed an aberrant and branching cilioretinal artery, the tip of which extended to the center of the fovea with concurrent distortion of the foveal reflex. In red reflectance fundus image, an aberrant cilioretinal artery reaching the center of the fovea was detected [Figure 1]. Macular thickness map showed nasal macular thickening in the same

eye [Figure 2] which was also evident on SD-OCT imaging which revealed irregularity of the nerve fiber layer with posterior shadowing in the nasal macular region extending to the center of the fovea as compared to the normal right eye. In spite of nerve fiber layer irregularity, the configuration and regularity of deeper retinal layers seemed to be preserved on SD-OCT images [Figures 3 and 4].

DISCUSSION

A wide variety of retinal vascular anomalies have been reported. Aberrant macular vessels are one such anomalies. These congenital vessels are aberrantly large branches of retinal arteries or veins which typically cross the horizontal raphe to supply or drain the macular region and may be associated with arteriovenous shunts, telangiectatic vessels or abnormal capillary networks.^[4,5] Visual impairment may result if the anomalous vessels pass across the foveola, foveolar cysts develop, or hemorrhage occurs.^[6]

de Crecchio et al^[7] reported 13 cases with anomalous venous macular macrovessels. Visual impairment in these cases was evident when the anomalous vessel crossed the fovea or caused foveolar cysts, or when hemorrhage occurred. In another report, Petropoulos et al presented three cases of anomalous retinal vessels without visual disruption.^[5] Ceylan et al,^[8] reported macular thickening detected on SD-OCT in a case with congenital retinal macrovessel leading to reduced but stable visual acuity. The patient reported herein had

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Figure 1. Red reflectance image shows an aberrant cilioretinal artery reaching the center of the fovea in the left eye.

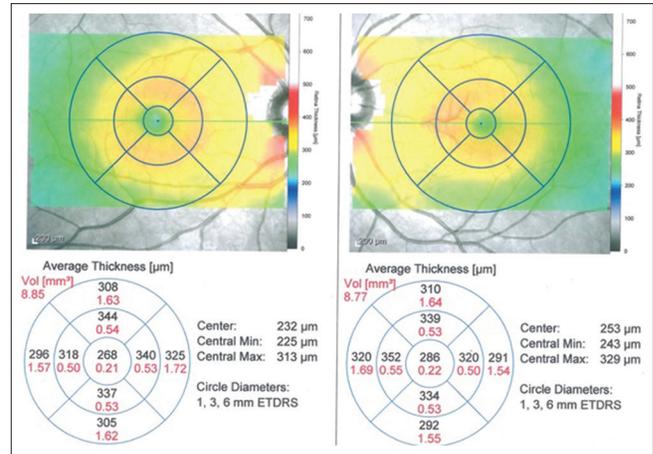


Figure 2. Macular thickness map shows nasal and central foveal thickening in the same eye.

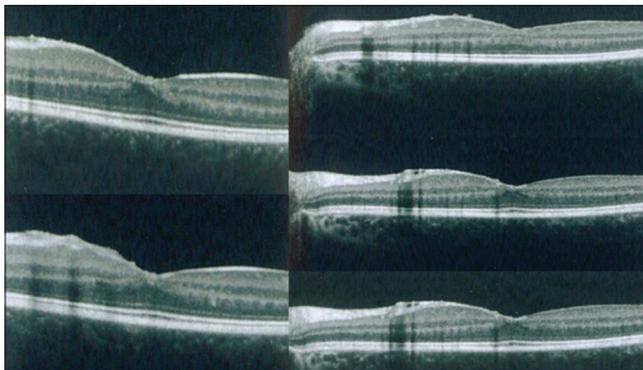


Figure 3. Macular spectral domain optical coherence tomography images of the left eye show posterior shadowing of the cilioretinal artery and nerve fiber layer irregularity in nasal foveal region up to the center of the fovea.

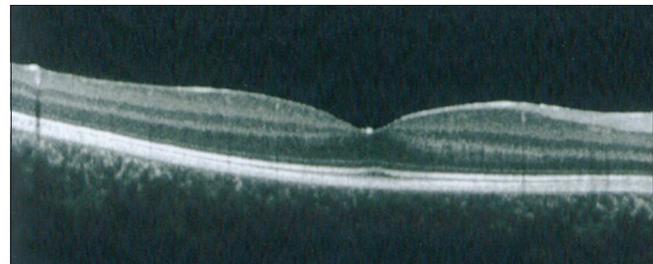


Figure 4. Macular spectral domain optical coherence tomography image of the right eye with no pathologic finding.

reduced visual acuity and mild amblyopia due to an aberrant branching cilioretinal artery with macular thickening. Irregularity of the nerve fiber layer with posterior shadowing in the nasal and central foveal region was detectable on SD-OCT imaging while the configuration of deeper retinal layers was preserved.

The central foveal region lacks the inner nuclear and ganglion cell layers of the retina. Although the outer plexiform layer is thicker in the macula, it becomes almost parallel to the internal limiting membrane toward the center of the fovea. This anatomical configuration allows light to be focused on the central fovea with minimal aberration through retinal layers. Thickening of the nerve fiber layer and an aberrant structure in the central fovea interrupts precise focusing of light on foveal photoreceptors. The current case showed that even fine changes in this structure can result in irreversible visual disturbance.

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