

Spectral domain optical coherent tomography demonstrates structural retinal changes in isolated cilioretinal artery occlusion

Mansooreh Bagheri, MD; Hosein Ashraf, MD; Mohammad Hosein Nowroozzadeh, MD

Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

This report demonstrates the structural retinal changes observed in-vivo by spectral domain optical coherent tomography (SD-OCT) in a case of isolated cilioretinal artery occlusion. A 32-year-old woman presented one week following acute sudden painless loss of vision. Ophthalmoscopy revealed macular edema and a cherry red spot. Fluorescein angiography one week post-infarction demonstrated a large patch of macular hypoflorescence in the distribution of the cilioretinal artery, with perfusion of the cilioretinal artery. Within this same macular distribution, SD-OCT demonstrated increased reflectivity, with increased reflectivity of the inner nuclear, inner plexiform, and ganglion cell layers. A sharp boundary was present between normal and infarcted macula on SD-OCT imaging. SD-OCT is a useful tool for diagnosing and identifying the extent of retinal vascular occlusion.

Introduction

Isolated cilioretinal artery occlusion is a rare condition, comprising 5% of all retinal arterial occlusions.^{1,2} Mechanisms underlying cilioretinal artery occlusion are either a reduction in perfusion pressure, often from embolus, or a mechanical compression of the artery as a result of an increase in venous pressure.³ The three clinical settings in which cilioretinal artery occlusions are: (1) an isolated

Correspondence: Mansooreh Bagheri, MD Department of Ophthalmology Pootschi Opthalmology Research Center Zand Avenue, Shiraz, Iran Email: mansooreh_bagheri@yahoo.com Phone: 00989173201260



arterial occlusive event, (2) an occlusive event associated with venous occlusion, specifically a central retinal vein occlusion, mechanically compressing the artery, and (3) an occlusive event associated with anterior ischemic optic neuropathy, often arteritic, namely as a result of giant cell arteritis.⁴ The cilioretinal artery and its occlusion may be diagnosed by funduscopy, fluorescence angiography, optical coherent tomography (OCT) imaging, as well as OCT angiography.⁵ This report demonstrates the structural retinal changes observed *in-vivo* by spectral-domain OCT (SD-OCT) in a case of isolated cilioretinal artery occlusion.

Case Report

A 32-year-old woman presented one week following acute sudden painless loss of vision in her left eye. The patient's past medical history and family history were unremarkable. Her best corrected visual acuity (BCVA) was count fingers at half a meter in the left eye and was 20/20 in the right eye. On clinical examination, the anterior segment in each eye was unremarkable and intraocular pressure in each eye was within a healthy range. Fundus examination of the left eye demonstrated macular edema with retinal opacification throughout the central macula in an area approximately 2 to 3 disc diameters in radius around the fovea. There was a central cherry red spot. There

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was no retinal hemorrhage or exudate. The retinal vasculature appeared normal.

Fluorescein angiography (FA) of the left eye revealed a large double branch cilioretinal artery perfusing the main parts of the macular area (Figure 1). Although the cilioretinal arteries were patent on FA, retinal ischemia along the distribution of these arteries was indicative of previous occlusion of the double branch cilioretinal artery. No other pathologic changes were observed on FA of the left eye.

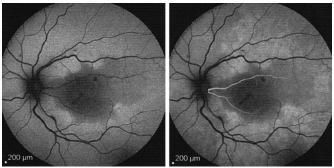


Figure 1

Fluorescein angiography in choroidal flush phase shows a large hypofluorescent area due to macular edema. A double branch cilioretinal artery perfuses the main parts of this area. Retinal ischemia along the distribution of these arteries is indicative of previous occlusion of the cilioretinal artery.

The structural changes of macular area were evaluated by SD-OCT (version 5.4.6 software, Spectralis OCT, Heidelberg engineering, Heidelberg, Germany), which revealed increased reflectivity of the inner nuclear, inner plexiform, and ganglion cell layers in the involved parts of the macula, as well as increased retinal thickness with a normal looking general contour in the parafoveal and perifoveal areas (Figure 2). Noticeably, on SD-OCT imaging, there was a sharp border between the infarcted and noninfarcted portions of retina.

Formal visual field testing (Humphrey Visual Field Analyzer 30-2 SITA-FAST) of the left eye demonstrated poor fixation and confirmed a large central scotoma. Systemic evaluations were unremarkable.

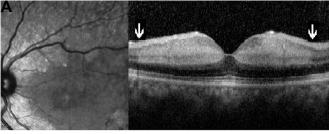


Figure 2

SD-OCT in acute phase demonstrated increased reflectivity and thickness of the inner nuclear, inner plexiform, and ganglion cell layers in the involved parts of macular areas. SD-OCT also demonstrated a sharp border (white arrows) between the infarcted and non-infarcted portions of retina.

Two months after onset, the patient's visual symptoms persisted and examination of the left eye revealed atrophic changes in macular area. SD-OCT demonstrated profound thinning and decreased reflectivity of the parafoveal inner retinal layers with loss of the foveal depression (Figure 3).

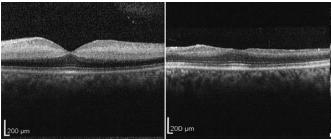


Figure 3

Comparison of SD-OCT of macular area taken 1 week (left image) and 2 months (right image) after cilioretinal artery occlusion revealed profound change over a 2 month perios. SD-OCT imaging at 2 months demonstated decreased reflectivity and thinning of the parafoveal inner retinal layers and loss of the foveal depression.

Discussion

Cilioretinal artery occlusion is a rare condition, which is often diagnosed by funduscopy and fluorescence angiography.¹ Specific anatomical bifurcation, lower perfusion pressure compared to the central retinal artery, and lack of self-regulation of this





perfusion pressure make the cilioretinal artery vulnerable to occlusion.²

SD-OCT is a non-invasive, high-resolution, and highly sensitive optical imaging modality, which offers new insight into retinal structural changes in ocular disease, including retinal vascular occlusive diseases.^{4,6-8} In patients with retinal artery occlusions, SD-OCT reveals increased thickness and reflectivity of the inner retinal layers (specifically the inner nuclear layer, inner plexiform layer and ganglion cell layer) and decreased reflectivity of the outer retinal layers, as the inner retina is edematous; prolonged ischemia results in subsequent atrophy of these layers.⁸⁻¹⁰

In this case report, we present a young patient with an isolated occlusion of a double branch cilioretinal artery, supplying main portions of the macula. Although the cilioretinal artery was patent with no filling delay in the FA, inner retinal edema followed by inner retinal atrophy identified on SD-OCT along the distribution of the cilioretinal artery was indicative of the previous occlusion of the cilioretinal artery. The boundaries of the cilioretinal artery distribution were sharply demarcated on SD-OCT both in terms of the initial edema and the subsequent atrophy.

Conclusions

SD-OCT demonstrates the structural retinal changes that occur in the acute and subsequent phases of cilioretinal artery occlusion. SD-OCT is a valuable imaging modality which assists in the diagnosis of retinal vascular occlusion.

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