Taiwan Journal of Ophthalmology 4 (2014) 21-27

Contents lists available at ScienceDirect

Taiwan Journal of Ophthalmology

journal homepage: www.e-tjo.com

Original article Choroidal changes by ocular coherence tomography in white dot syndrome

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ARTICLE INFO

Article history: Received 5 February 2013 Received in revised form 31 May 2013 Accepted 7 June 2013 Available online 16 September 2013

Keywords: choroid indocyanine green angiography inner segment/outer segment ocular coherence tomography white dot syndrome

ABSTRACT

Purpose: To evaluate the findings of optical coherence tomography (OCT) in the acute and convalescent stages in patients with white dot syndrome. Patients were followed up at our clinic for at least 6 months. *Materials and methods:* A consecutive case series of patients with white dot syndrome were enrolled in this study. Only patients with disease onset less than 1 week were included in this study. Slit-lamp biomicroscopy, indirect ophthalmoscopy, fluorescein angiography, indocyanine green angiography (ICGA), OCT, visual field test, and corrected decimal visual acuity test were performed on all patients. *Results:* A total of eight eyes from eight patients were analyzed in this study, including cases with acute zonal occult outer retinopathy (AZOOR), punctate inner choroidopathy and AZOOR, multiple evanescent white dot syndrome, and multifocal choroiditis. In the acute stage, OCT demonstrated diffuse or segmental attenuation/loss of inner segment/outer segment (IS/OS) signal. Choroidal thickening with increased choroidal vascular porosity as compared with the fellow eyes was noted in all eyes. The ICGA showed hypofluorescence patches in the late phase. In the Convalescent stage, complete or partial thickening and choroidal vascular porosity in OCT. The ICGA also demonstrated resolved choroidal hypofluorescence in the convalescent stage.

Conclusion: Choroidal thickening and increased choroidal vascular porosity in addition to disruption of photoreceptor IS/OS were characteristic OCT features of white dot syndrome. Recovery of vision was accompanied with restoration of OCT findings in both retina and choroid.

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1. Introduction

White dot syndrome is composed of a group of disorders including acute zonal occult outer retinopathy (AZOOR), multiple evanescent white dot syndrome (MEWDS), multifocal choroiditis (MFC), punctate inner choroidopathy (PIC), and acute posterior multifocal placoid pigment epitheliopathy, etc. Inflammation located on the outer retinal layer, retinal pigment epithelia, and inner choroid have been proposed as the etiology.¹ Clinical characteristics include acute deterioration of vision, blind spot enlargement, visual field (VF) loss, and photopsia. Loss of inner

segment/outer segment (IS/OS) line and attenuation of outer nuclear layer (ONL) in optical coherence tomography (OCT) have been described in cases with AZOOR, MFC, and MEWDS in previously published reports.^{2–7} To the best of our knowledge, there are still no reports discussing the choroidal changes in OCT. In this study, manifestations of the retina and choroid in Fourier domain OCT and indocyanine green angiography (ICGA) at the acute and convalescent stages are described and discussed.

2. Materials and methods

This prospective study evaluated the consecutive case series of white dot syndrome from June 2010 to December 2011 in Changhua Christian Hospital, Taiwan. Only patients with disease onset less than 1 week were recruited in this study. This study was approved by the Institutional Review Board of Changhua Christian Hospital and was carried out in accordance with the World Medical Association's Declaration of Helsinki. All patients







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Conflicts of interest: The authors have no conflicts of interest relevant to this article.

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underwent comprehensive examinations including corrected decimal visual acuity (VA), slit-lamp biomicroscopy, indirect ophthalmoscopy, spectral domain OCT (Cirrus OCT; Carl Zeiss Meditec Inc., Dublin, CA, USA), and color and autofluorescence fundus photography. Fluorescein angiography (FA), ICGA (HRA-2; Heidelberg Engineering, Heidelberg, Germany), and VF test (Humphrey 30-2, Carl Zeiss Meditec, Germany) were also performed.

2.1. Spectral domain OCT

Cirrus HD-OCT (Carl Zeiss Meditec, Inc) was used to obtain spectral domain OCT images. The scanning rate was 27,000 A-scan/ second with an axial resolution of 5 μ m. Both 6 mm \times 6 mm cubic scan and high-definition one-line raster scan centering on the foveola were performed. Multiple hyper-reflective bands corresponding to the different histological layers of retina were identified and achieved. The subfoveal choroidal thickness was measured according to the horizontal high-definition one-line raster scan for both eyes on every visit. We also performed a comparison of changes in hyper-reflective bands and choroidal thickness between fellow eyes and each visit.

2.2. Fluorescein and ICGA

Simultaneous fluorescein and ICGA were carried out using HRA-2 (Heidelberg Engineering).

3. Results

A total of eight eyes from eight patients (3 males and 5 females) who fulfilled the study criteria were analyzed. All eyes were myopic. The average refractive status was -8.91 ± 3.27 D (range from -5.25 D to -13.5 D). The average age of the participants was 35.1 ± 16.0 years (range: 18-43 years). Based on the subclassification of disease entity, one patient had AZOOR, one patient had PIC

Table 1	l
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Demographic data of patients.

combined with AZOOR, four patients had MEWDS, and two patients had MFC. All patients complained of blurred vision with or without VF defect. Corrected decimal VA ranged from 20/100 to 20/ 20 at the initial visit. The IS/OS boundary attenuation or loss was noted in every patient in the acute phase of the disease. The four patients with MEWDS had complete recovery of IS/OS boundary without treatment. The patient with AZOOR had IS/OS boundary that completely recovered after immunomodulation therapy. The patient with AZOOR combined with PIC and the two patients with MFC had partial recovery of IS/OS boundary after immunomodulation therapy. Increased choroidal thickness compared with the fellow eyes was noted in all but one patient (Case 7), in whom MFC occurred 1 year earlier in the fellow eye. Increased choroidal vascular porosity as compared with the fellow eyes was also noted in all eyes. Decreased choroidal thickening was noted in all eyes during the convalescent phase. Hypofluorescence in late-stage ICGA was noted in all eyes at the acute phase and was later partially or completely resolved in all cases. Patients' demographic data are listed in Table 1.

3.1. AZOOR

3.1.1. Case 1

An 18 year-old myopic male patient complained of loss of vision in the left eye for 1 day. His corrected decimal VA in the left eye was 20/100 and 20/20 in the right eye. Relative afferent papillary defect was noted in the left eye. Indirect ophthalmoscopy and slit-lamp microscopy showed a tessellated myopic fundus without any other abnormalities (Fig. 1A). Humphrey 30-2 VF test showed a defect in the temporal lower, upper, and nasal periphery areas in the left eye. Subtle obliteration at the nasal upper area was also noted in the right eye (Fig. 1B). He also had a decreased electroretinographic amplitude in A wave in the left eye. An FA showed no particular findings, and ICGA showed delayed choroidal filling and multiple hypofluorescent patches at the posterior pole at the late phase (Fig. 1C). Spectral domain OCT

Case/ gender/ age/eye	Refractive status (D) diseased eye/ fellow eye	Diagnosis	IS/OS changes, acute/final stage	Choroidal thickness, initial/ final (µm)	Choroidal thickness in the fellow eye	Late-stage ICGA acute/convalescent stage	Initial/ final VA (decimal VA)	Duration of follow-up (mo)
1/M/18/ OS	12.75/-12.00	AZOOR	Diffuse attenuation, hyper-reflective spots at INL/complete	120/96	68	Hypofluorescence spots/resolved	20/100 20/20	11
2/F/36/OS	-11.50/ -11.50	PIC and AZOOR	Diffuse attenuation/partial recovery	165/108	80	Hypofluorescent spots and patches/partially resolved	20/20 20/20	10
3/M/43/ OS	-5.25/-6.25	MEWDS	Segmental disruption, hyper-reflective spots at ONL/complete recovery	316/284	244	Hypofluorescent spots/resolved	20/40 20/20	9
4/F/35/OD	-8.25/-8.00	MEWDS	Segmental disruption/complete recovery	195/142	167	Hypofluorescent spots/resolved	20/40 20/20	6
5/F/37/OS	13.5/-13.5	MEWDS	Segmental disruption with triangular elevation, hyper-reflective spots at ONL/complete recovery	174/108	104	Hypofluorescent spots/resolved	20/40 20/20	10
6/M/43/ OS	-7.0/-4.0	MEWDS	Segmental disruption with triangular elevation, hyper-reflective spots at ONL/complete recovery	204/201	172	Hypofluorescent spots/resolved	20/30 20/20	6
7/F/31/OS	-7.75/-5.00	MFC	Segmental disruption/partial recovery	285/218	221	Hypofluorescent spots/partially resolved	20/100 20/70	12
8/F/38/OD	-5.25/-5.50	MFC	Segmental disruption/partial recovery	271/221	175	Hypofluorescent spots/partially resolved	20/20 20/20	6

AZOOR = acute zonal occult outer retinopathy; F = female; ICGA = indocyanine green angiography; INL = inner nuclear layer; IS/OS = inner segment/outer segment; M = male; MEWDS = multiple evanescent white dot syndrome; MFC = multifocal choroiditis; mo = months; OD = oculus dexter; ONL = outer nuclear layer; OS = oculus sinister; PIC = punctuate inner choroidopathy; VA = visual acuity.

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