Optical coherence tomographic findings in optic nerve hypoplasia

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We investigated a case of unilateral optic nerve hypoplasia using spectral domain optical coherence tomography (SDOCT). Optical coherence tomography was done on both eyes using 5-line Raster scan for the fovea to analyze the retinal nerve fiber layer thickness, inner retinal layer thickness, outer retinal layer thickness, and optic disc cube scan for the disc. Retinal nerve fiber layer thickness, inner retinal layer thickness, and outer retinal layer thickness were manually measured at 21-points of each five lines, and results were compared between both eyes. Retinal nerve fiber layer thickness and inner retinal layer thickness of optic nerve hypoplasia were significantly thinner than the opposite eye, but there was no significant difference in the thickness of the outer retinal layer between both eyes.

Key words: Inner retinal layer thickness, optic nerve hypoplasia, outer retinal layer thickness, retinal nerve fiber layer thickness, spectral domain optical coherence tomography

Optic nerve hypoplasia (ONH) is a nonprogressive congenital abnormality of one or both optic nerves associated with a diminished number of axons in involved nerves with normal development of supporting tissues and the retinal vascular system.[1] A reduced number of axons can be demonstrated as having a smaller disc size than normal.[1] Furthermore, the retinal nerve fiber layer (RNFL) is diminished and ganglion cells are reduced in number, but the outer retinal layer (ORL) appears normal.[1]

The diagnosis of ONH may be difficult in some cases in which disc shape and size are normal or involves only a segmental part.[1,2] In this case, optical coherence tomography (OCT) may be useful since OCT can provide useful information about the optic disc, RNFL, and retina.

We herein report the quantitative spectral domain OCT (SDOCT, Carl Zeiss Meditec INC, Dublin, CA) analysis in a patient with unilateral ONH of an unknown cause.

Case Report

A 67-year-old woman presented with blurring of the left eye. The patient presented with best-corrected visual acuity of count fingers at 2′ oculus dexter (OD) and 20/40 oculus sinister (OS). A relative afferent pupillary defect was observed in the right eye. There were nuclear sclerosis (Lens Opacities Classification System (LOCS) III grading = 1) in both eyes. A small optic disc and double ring sign were observed in the right eye and left was normal.

In order to evaluate the thickness of each layers of retina, OCT imaging was performed for both eyes. 5-line Raster scan, which makes five line scans with 6 mm width in horizontal and each separated by 250 μm, and optic disc cube scan were generated. We selected the results with minimum signal strength seven and the third row of the five line scans that passed the fovea to compare both eyes.

To compare and analyze each layers of the retina, 21-points of each Raster scans were selected using the caliper tool in the device. Each points were located at 250; 500; 750; 1,000; 1,250; 1,500; 1750; 2,000; 2,250; and 2500 μm apart from the fovea to nasal and to temporal side.

Our OCT device did not provide thickness maps of each layers of retina, which was the main concern in this study. Therefore, for quantitative analysis of retinal layers, segmentations were done to three parts, which were RNFL, inner retinal layer (IRL), and ORL; on the basis of internal limiting membrane, posterior boundary of the RNFL, posterior boundary of the outer plexiform layer, and anterior boundary of the cone outer segment tip line.[3,4] RNFL thickness (RNFLT), IRL thickness (IRLT), and ORL thickness (ORLT) were measured manually using the caliper tool in the device. The Wilcoxon's signed-rank test and *t*-test were used for the comparison of RNFLT, IRLT, and ORLT; between both eyes. Statistical analysis was performed using SPSS®, version 14.0 (SPSS Inc., Chicago, IL). The results were considered significant at $P < 0.05$.

The average of RNFLT of ONH was thinner than the opposite eye [Fig. 1] and the thickness analysis of each five line scans were represented by a graph and fundus photograph map, respectively [Fig. 2]. RNFLT and IRLT of

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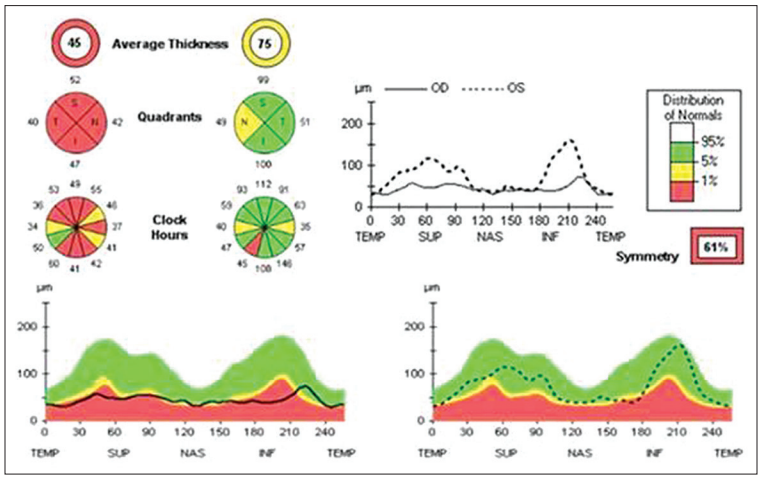

Figure 1: Comparison of average retinal nerve fiber layer (RNFL) of both eyes. The average thickness of RNFL was thinner in optic nerve hypoplasia (ONH) (45 vs 75 µm) and diffuse thinning of RNFL in ONH is apparent by RNFL thickness (RNFLT) analysis program of Cirrus HD-OCT

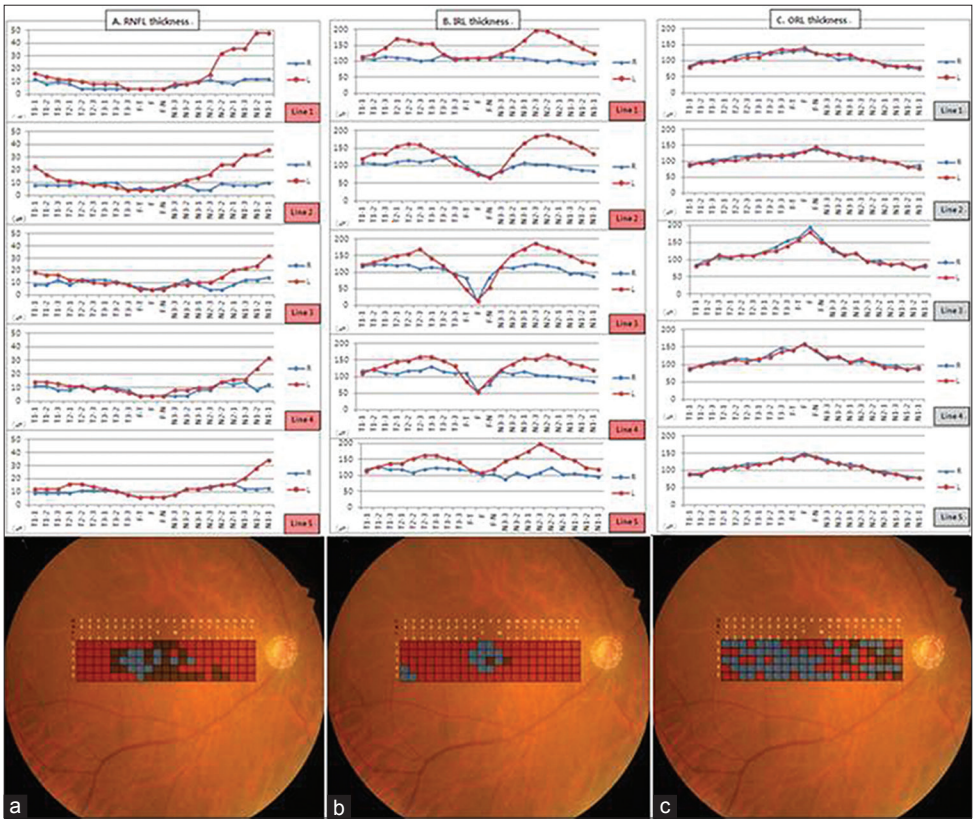

Figure 2: Comparative fundus photograph maps and graphs of each retinal layers of both eyes. Each graphs and fundus photographs with blocks are the comparison of five line scans and point comparison of both eyes, respectively; comparing (a) RNFLT, (b) IRLT, and (c) ORLT. Each points have been represented by the two colors on the thicker side (blue-right, red-left) or gray color, if both sides have the same thickness. RNFLT and IRLT of the left eye is significantly thicker than the right eye, for all five lines ($P < 0.000$; Graph A, B, fundus photograph map A, B). However, ORLT shows no significant difference between both eyes for all five lines ($P > 0.05$, Graph C)

ONH were significantly thinner than the opposite eye, for all five lines ($P < 0.05$). However, ORLT did not show significant difference between both eyes for all five lines ($P > 0.05$). In addition, to compare the thickness of retinal layers of the nasal and temporal areas from the fovea, each 50 points from the fovea to the nasal and temporal sides were divided into two groups. The results were the same as above. RNFLT and IRLT of the right eye were significantly thinner than the left eye on the temporal and nasal side ($P < 0.000$), but ORLT did not show significant difference between both eyes ($P = 0.879$, 0.164). The differences of RNFLT and IRLT between both eyes are greater on the nasal side than the temporal side as shown in Fig. 2 (difference of RNFLT between both eyes on the temporal and nasal side: 2.04 vs 9.02 µm, IRLT: 20.86 vs 45.02 µm).

Discussion

ONH results from congenital deficiency of retinal ganglion cells and axons that lead to disorganization of the retinal ganglion cell layer, thinning of RNFL, and a small optic disc.[4] These results cannot be exactly explained, but they are thought of as a result of any insult to the visual pathway before the completion of development, especially in the early developmental period.[1] The diagnosis of ONH remains primarily on a clinical basis, but it becomes difficult when the degree of hypoplasia is less severe or even segmental in nature,[5] because the thinning of RNFL and disc appearance are variable. Therefore, OCT, which is an *in vivo*, noninvasive, valid imaging method that provides accurate, quantitative, and reproducible cross-sectional measurements of the optic nerve and macula, is useful for diagnosing any optic disc and retinal anomalies.[6] Nevertheless, there have been few reports on detecting ONH using OCT worldwide.

In our patient, the clinical characteristics of the optic nerve head showed typical features of ONH. The outcomes obtained through using SD-OCT and manual segmentation in our report demonstrated the thinning of RNFL and IRL and preserved ORL in the patient with congenital, unilateral ONH; which coincided with a conventional histological report.[1]

Our patient showed that best corrected visual activity (BCVA) of ONH was count fingers at 2'. Visual potential of ONH is related to the degree of papillomacular bundle defect.[1] Our report demonstrated uniformly thin RNFLT and IRLT on both sides of the fovea and showed more significant difference on the nasal side. These results may be related to VA of our patient. Since it can be difficult to assess the visual potential by the appearance of disc alone[1], SDOCT can be helpful for evaluating the visual capacity of ONH.

In conclusion, SDOCT findings of ONH, namely the analysis of the thickness of the retinal layers in our case showed significantly thinner RNFLT and IRLT than the normal eye, but ORLT did not show any difference between ONH and the normal. These results coincided with a conventional histological report.[1]

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