Three-dimensional optical coherence tomography of the optic nerve head with myelinated nerve fibers

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Introduction

Intraocular myelination of the retinal nerve fiber has been reported in 0.3–0.6 % of population by ophthalmoscopy and in 1 % by postmortem examination [1]. Myelinated nerve fibers may be familial, in which case it is inherited as an autosomal dominant trait [2]. Myelination of the anterior visual system begins at the lateral geniculate body at 5 months of gestation and proceeds distally. It reaches the chiasm by 6–7 months, the retrobulbar optic nerve by 8 months of gestation, and the lamina cribrosa at term, although in some cases the myelination may continue for a short period after birth. Normally myelination stops at the lamina cribrosa and does not extend intraocularly.

Several theories have evolved to explain the occurrence of intraocular myelination. Oligodendrocytes, which are responsible for myelination of the central nervous system, are not normally present in the human retina. However, histological sections confirm their presence in some areas of myelinated nerve fibers [3]. Oligodendrocytes may gain access to the retina and produce myelin through a defect in the lamina cribrosa. Presence of fewer axons relative to the size of the scleral canal may also allow access of oligodendrocytes to the retina where they produce myelination [4].

Three-dimensional optical coherence tomography of the optic nerve head in a case of myelinated nerve fibers is reported for the first time.

Material and methods

The authors confirm adherence to the tenets of the Declaration of Helsinki. After taking a written informed consent, spectral domain optical coherence tomography [Cirrus high definition OCT (Carl Zeiss Meditec Inc.), CA, USA] was performed in a case with myelinated nerve fibers.

Case report

A 15-year-old female presented in the outpatient department of our tertiary care center for refraction. Best corrected visual acuity of the patient was 20/20 in both the eyes. There was no systemic abnormality detected. Fundus examination of the left eye revealed a large gray patch obscuring the optic disk margins and the retinal vessels with fan-shaped appearance distally (Fig. 1). The right eye showed no abnormality. On spectral domain optical coherence tomography (SD-OCT), the retinal nerve fiber layer (RNFL) thickness map showed increased thickness of the RNFL of the left eye (212 μm) compared to the right eye (94 μm). In the left eye, the RNFL thickness in the superior, inferior, and nasal quadrants was greater than the temporal quadrant, which corresponded to the presence of myelinated nerve fibers. The RNFL map of the two eyes showed only 9 % symmetry. The extracted RNFL tomogram of the left eye showed an abrupt termination of the structures posterior to the nerve fiber layer (Fig. 2). The five-line raster of the optic disk showed increased reflectivity of the RNFL. The inner

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Fig. 1 Scanning laser ophthalmoscopic picture of the fundus of the left eye showing a large gray patch obscuring the optic disk margins and the retinal vessels with fan-shaped appearance distally

Fig. 2 Retinal thickness analysis map of both eyes showing only 9% symmetry between the two eyes. The average RNFL thickness of the right eye is 212 μm and that of the left eye is 94 μm . The extracted RNFL tomogram of the two left eye shows an abrupt termination of the structures posterior to the nerve fiber layer

segment–outer segment junction and the retinal pigment epithelium (RPE) showed an abrupt end at the beginning of the hyperreflective RNFL (Fig. 3). The 3D-OCT showed an elevation around the disk most marked superonasally (Fig. 4, video 1). The C-scan at the level of internal limiting membrane (ILM) showed areas of hyperreflectivity corresponding to the area of the peripapillary myelinated nerve fibers. A non-reflective area was seen in the center of the hyperreflective region (Fig. 5). The C-scan at the level of RPE corresponded to that at the level of ILM. However, the optic nerve head appeared as a hyporeflective region on this section. The non-reflective area corresponding to that at the level of ILM was seen to be increased in size (Fig. 6). The macular cube showed no abnormality.

Fig. 3 The five-line raster of the optic disk on OCT showing increased reflectivity of the RNFL. The inner segment–outer segment junction and the retinal pigment epithelium show an abrupt end at the beginning of the hyperreflective RNFL

Discussion

Presence of myelinated nerve fiber has been associated with several ocular and systemic disorders. Extensive unilateral myelination of nerve fibers can be associated with anisometropic high myopia and dense amblyopia which is refractory to treatment by occlusion therapy [5, 6]. They may also be associated with occult microvascular abnormalities [7] and Gorlin syndrome [8].

Myelinated nerve fibers do not usually reduce visual acuity. However, it may produce relative scotomas depending upon their location and the number of fibers present. The scotomas are smaller than what the size of the myelinated fiber would suggest.

The time domain and SD-OCT features of myelinated nerve fiber has been described by Salvatore et al. [9]. Increased reflectivity and thickness of the RNFL in the area of the myelinated fibers was reported. Furthermore this

characteristic was accompanied by a posterior cone of shadowing as described by them.

In the present study, diffuse myelination was observed. Increased RNFL thickness in the left eye especially in the superior, inferior, and nasal quadrants corresponded to the presence of myelinated nerve fibers. This resulted in marked asymmetry of the RNFL map between the two eyes. The extracted RNFL tomogram and OCT of the optic disk of the left eye showed an abrupt termination of the structures posterior to the hyperreflective RNFL due to the back shadowing produced by the thickened nerve fiber layer. The steeply elevated area of myelinated nerve fiber as observed on 3D-OCT acted as an optical barrier creating an area of non-reflectivity seen amidst the hyperreflective area as seen on the C-scan.

Three-dimensional OCT documented areas of distribution of myelinated nerve fibers very well and highlighted its topographic distribution. The C-scan also precisely

Fig. 4 The 3D-OCT showing an elevation around the disk most marked superonasally due to myelinated nerve fibers

highlighted the areas of myelinated nerve fibers as areas of hyperreflectivity. Three-dimensional OCT of the optic nerve

Fig. 5 C-scan at the level of ILM showing areas of hyperreflectivity corresponding to the area of the myelinated fibers around the optic nerve head. A non-reflective area is seen in the center of the hyper-reflective region

Fig. 6 C-scan at the level of RPE showing areas of hyperreflectivity corresponding to the area of the myelinated fibers around the optic nerve head. The optic nerve head is seen as hyporeflective area. The retinal vessels and the most elevated part of the myelinated nerve fibers are seen as non-reflective areas

head is a useful tool in documenting the anatomy of the disease.

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