A 4-year-old girl with normal development presented with progressive ocular deviation of 3 years’ duration; past medical and ocular history was unremarkable. Upon presentation, cycloplegic refraction revealed refractive error of +1.00 -0.50×180 and -14.00 in the right and left eyes. Visual acuity was 20/20 and 20/400 in the right and left eyes with optical correction of plano and -14.00, respectively. Relative afferent pupillary defect was negative and cover test revealed 40 prism diopters of constant left esotropia (Fig. 1). Slitlamp examination was normal and dilated fundus examination was unremarkable in the right eye, but revealed extensive Myelinated retinal nerve fibers in the left (Fig. 2). A-scan ultrasound biometry showed axial length of 20.75 and 25.57 mm in the right and left eyes, respectively (Fig. 3). The patient was diagnosed with extensive unilateral myelinated retinal nerve fibers associated with axial myopia, deep anisometropic amblyopia and esotropia in her left eye. She received spectacle correction together with part time patching of the right eye six hours every day. The patient was scheduled for a three month follow-up visit to monitor improvement in visual acuity.
DISCUSSION

Myelinated retinal nerve fibers are found in 0.57 to 1% of the population; they appear as white to gray-white patches on funduscopic examination corresponding to the distribution of retinal nerve fibers. Myelination of retinal ganglion cell axons starts at the lateral geniculate body proceeding through the optic tracts, chiasm and optic nerves during intrauterine life. The process of myelination normally terminates at the level of the lamina cribrosa; however it occasionally continues into the retinal nerve fiber layer. Myelination of retinal nerve fibers seems to be due to anomalous distribution of oligodendroglia within the retina. The lamina cribrosa is considered to be a barrier against myelination, therefore abnormal scleral formation is hypothesized to be responsible for retinal nerve fiber layer myelination. Ellis et al have categorized myelination into three forms: type I affects one temporal arcade; type II affects both temporal arcades, and type III which is not contiguous with the disc.

Ellis et al found that 83% of patients with myelinated retinal nerve fibers had myopia greater than 6 diopters. It remains unknown whether myelination of retinal nerve fibers is the reason for, or the result of myopia. It is possible that myelinated fibers may blur retinal images and induce visual deprivation. Such deprivation at a critical stage of ocular development may contribute to myopia by including axial enlargement. On the other hand it is also possible that axial elongation predisposes to retinal nerve fiber myelination. In axial myopia, lamina cribrosa development is prolonged, therefore myelination may be allowed to continue down into the optic nerve and retina. Straatsma et al found that 10% of patients with myelinated nerve fibers have myopia, amblyopia and strabismus.

The cause of amblyopia in patients with myelinated retinal nerve fibers is a matter of controversy. Some authors believe in an organic etiology and postulate that myelinated nerve fibers result in elevation of the optic disc and disorganization of neural elements leading to amblyopia. In contrast, others believe that myelination per se does not cause organic visual loss.

In our patient, three factors possibly contributed to development of amblyopia, namely myopia, esotropia and myelinated retinal nerve fibers. It remains unclear which one plays the major role and which are complementary. The first step in treatment of amblyopia in these patients is full optical correction based on a cycloplegic refraction. However the patient described herein, seems to have poor prognosis for visual improvement because of considerable unilateral myopia, deep amblyopia, strabismus and extensive myelinated nerve fibers.

REFERENCES