Saudi Journal of Ophthalmology (2015) 29, 85–88

Case Report

Myelinated retinal nerve fibers (MRNF) – Dilemmas related to their influence on visual function

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Abstract

Myelinated nerve fibers (MNF) occur in less than 1% of the population, however, they might be responsible for diagnostic dilemmas in cases with visual loss. The case report of an aged pseudophakic patient with visual deterioration in the right eye and MNF in both eyes is presented. The documentation provided by the patient proved recent several examinations of both fundi, and all of them were described as normal. Physical examination revealed the posterior capsule opacification in the right eye, white lesions on the retina of the right eye around the optic disk, and in the left eye – the peripheral, which could correspond to the myelinated fibers. Although visual field changes and OCTs corresponded to the NMF, it turned out, however, that visual acuity loss was in fact caused by PCO and was reversed by the YAG capsulotomy procedure. This case shows some problems related to MNF diagnosis and evaluation of their influence on visual function.

Keywords: Myelinated nerve fibers, Visual loss, Visual field defects, OCT

http://dx.doi.org/10.1016/j.sjopt.2013.09.002

Introduction

The first description of myelinated retinal nerve fibers (MRNF) was given in 1856 the German pathologist Rudolf Virchow based on eye preparations. He wrote that ''retina was white, very thick and wrinkled. Macula was normal and near the optic disc, though more deeply situated, were thick, opaque, chalk-white spots, which spread around the disc in the shape of a star, so that when I wanted to draw the line between the disc and macula on each side of the two had the same divergence. In the other eye, I found, without much surprise, in the same place, the ring around the disc with a width of 2–2, 5 mm, regressing towards the outside''.[1] Today this developmental anomaly occurs in less than 1% of the population.[2]

MRNF may be congenital or acquired in nature. It is usually asymptomatic. It presents as white or greyish white spots with jagged edges, in conjunction with the optic disk, around it, or not related. It is a rare case of family inheritance.[3]

Case report

The patient, aged 66, was admitted because of the deterioration of vision in the right eye, which appeared in the last 2–3 months. The patient was pseudophakic in both eyes for 3 years. There were no other eye diseases or eye injuries in the history.

The documentation provided by the patient proved recent several examinations of both fundi, and all of them were

Received 24 April 2012; received in revised form 18 August 2013; accepted 8 September 2013; available online 21 September 2013.

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Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

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described as normal. The corrected visual acuity in the right eye was 0.3, in the left eye −0.6.

Physical examination revealed the posterior capsule opacification (PCO) in the right eye, white lesions on the retina of the right eye around the optic disk (Figs. 1 and 2), in the left eye – the peripheral, which could correspond to the myelinated fibers. The patient claimed that she repeatedly went through fundus examination and was never informed of the existence of such lesions. Visual fields showed defects corresponding to the MRNF of the retina in both eyes. (Fig. 3) OCT scans confirmed the typical picture of MRNFs around the optic disk in the right eye (Fig. 4).

The question was if the reason of visual acuity loss was MRNF, PCO or both of them. The evaluation of the role of MRNF was complicated by co-existence of two factors: the lesions were not described in previous examinations (MRNF are usually present from childhood) and the decreased visual acuity (usually MRNF are not accompanied by blurred vision). We decided to conduct YAG laser posterior capsulotomy in the right eye, after which an improvement of vision was noticed to the corrected visual acuity of 0.6. It was found that blurred vision was caused by posterior capsule fibrosis.

Figure 1. MRNF of the right eye: the unusual aspect of the MRNF with borders close to vessels.

Figure 2. MRNF of the left eye.

Discussion

The presented case shows that even in non-typical MRNF cases, including diagnosis in adulthood related to decreased visual acuity, the other accompanying disorders should be firstly excluded. This also confirms that MRNF typically give visual field deficits adequate to its localization (Fig. 3), and can be visualized by OCT (Fig. 4), but only uniquely might be the reason for visual deterioration.

MRNFs are formed because of an error during the process of myelination of fibers of retinal ganglion cells. For this process are the responsibility of oligodendrocytes. Physiologically, it starts already around the 5th month of fetal life from the corpus geniculatum laterale, following along the visual pathway and ending on the lamina cribrosa in childbirth or shortly thereafter.[4,5] Lamina cribrosa is a protective barrier against the penetration of fiber myelin in the area of the retina. And so when a disturbance affects the integrity and underdevelopment, oligodendrocytes penetrate between the fibers and ganglion cells produce myelin, the more, the lower the density of these fibers. And that is why myelinated nerve fibers are so rare in macula and on the nasal retina. Myelin fibers unrelated to the optic disk arise when the lamina cribrosa allows ofigodendrocytes go into space with reduced density of retinal ganglion cell axons. Such fibers usually do not cause problems with vision. Patients have usually a profound physiological cup. Situs inversus of retinal vessels or tilted optic disk sometimes happens.[4] The development of myelinated nerve fibers associated with the optic disk depends on the degree of hypoplasia of the disk itself. And that is why this type of fiber is often accompanied by amblyopia, myopia and different presentations of strabismus.[3,5,6]

In clinical studies, the fibers related with the disk are frequently observed, but according to autopsy studies, peripheral fibers are three times more common.[5] Symptoms of visual impairment, which rarely appear are due to amblyopia, anisometropia[5] and hypoplasia of the disk,[7] found when macula is associated or very large fibers. These can sometimes look like leukocoria and retinoblastoma can even be suspected.[3,5] Patients often report visual field defects. These are relative scotoma and blind spot enlargement, which correspond to the areas covered by the fibers.[5] In angiography, masking of structures behind fibers is visible, but there are no circulation disorders within the retinal and choroid.[5] In areas occupied by the myelinated nerve fibers may appear vascular anomalies, such as neovascularization, bleeding and thrombus and also retinal abnormalities like degeneration, traction, detachment and epiretinal membrane. It happens to find the fibers in patients with diseases such as neurofibromatosis type I or Gorlin syndrome and developmental disorders such as coloboma, polycoria, keratoconus, or various forms of dyscrania.[3]

Myelinated nerve fibers may also progress.[8,9] There are cases of spontaneous appearance of fibers in adults with pre-tests showing no such changes. Some of them can be explained by post-traumatic optic nerve decompression.[9] Described are several cases of disappearance of the fibers

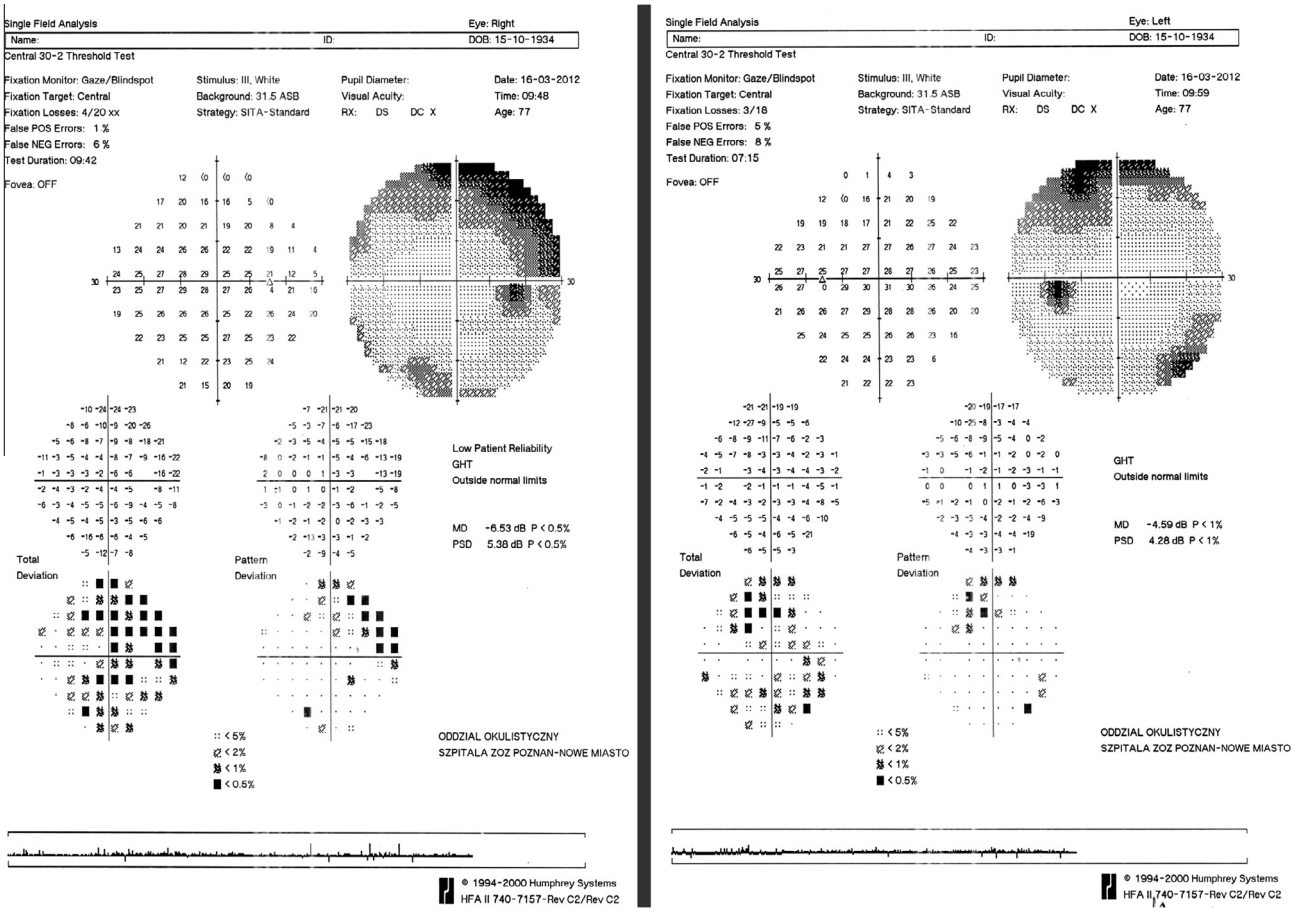

Figure 3. Visual field defects corresponding to MRNF changes in the right and left eye.

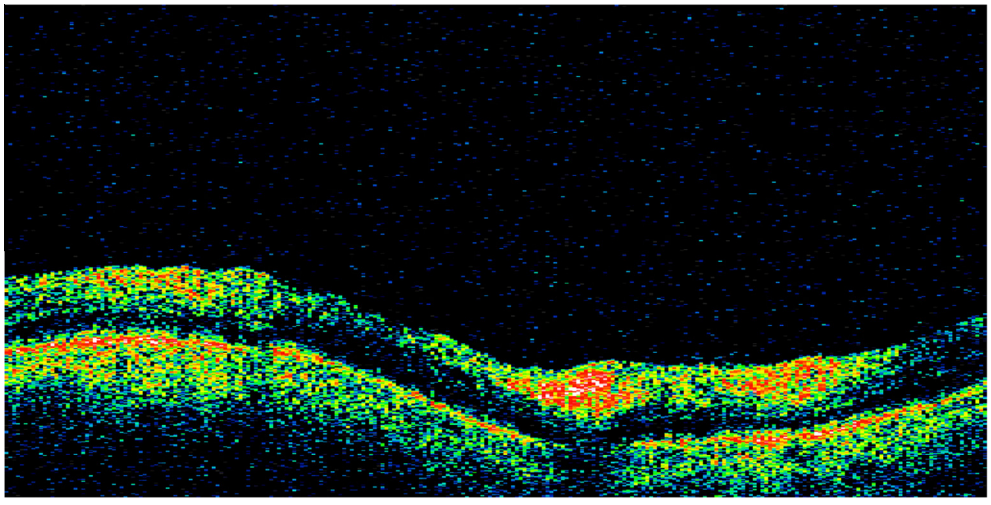

Figure 4. OCT scan of the MRNFs.

after the closure of ischemic retinal vessels, in the course of ischemic neuropathy or Leber neuropathy (LHON), chronic glaucoma, diabetic retinopathy, in patients after laser photocoagulation and surgical resection of pituitary adenoma.[3] Most cases require no treatment. However, for those with decreased visual acuity, amblyopia, myopia, and strabismus there is little possibility of effective therapy and occlusal correction.

In conclusion, we described the case of an aged patient with MRNF complicated by the lack of earlier fundus description of the lesions and by the recent visual deterioration. Although visual field changes corresponded to the MRNF, it turned out, however, that visual acuity loss was in fact caused by PCO and was reversed by the YAG capsulotomy procedure. This case shows some problems related to MRNF diagnosis and evaluation of their influence on visual function.

Conflict of interest

The authors declared that there is no conflict of interest.

Acknowledgment

Authors wish to thank Dieter Schmidt, Emeritus Professor of Ophthalmology, University of Freiburg, Germany for his valuable comments during the work on the manuscript.

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