Leber hereditary optic neuropathy (LHON) causes visual loss, predominantly in healthy young men. We recently examined a patient who previously had bilateral macular holes and subsequently developed LHON at 74 years of age. Although his central scotomas were initially attributed to the macular holes, his visual acuity declined following an initial improvement after operative closure of the macular holes; thus, other diagnoses, including LHON, were considered. Furthermore, macular optical coherence tomography (OCT) images remained unchanged in this time. A mitochondrial genetic analysis identified a 11778G→A mutation. From this case, we propose that LHON remains in the differential diagnosis even in older patients, as has previously been reported.

Introduction

Leber hereditary optic neuropathy (LHON) is a mitochondrial DNA-mediated disease that causes acute or sub-acute visual loss, predominantly in healthy young men (typically 18–35 years of age). Cases of late-onset LHON have been reported—for instance, 11778G→A mutation in 50-, 58-, 59-, 62-, 63-, 67-, 70-, 72-, 73-, 75-, 76-, 78-, 79-, 81-, 87-, and 88-year-old men, 14484T→C in 59- and 81-year-old men, and 3460G→A mutation in a 62-year-old woman and in 75- and 81-year-old men. Onset in older age ranges increases the likelihood that the patients will previously have had other systemic or eye diseases, which can make the diagnosis of LHON more challenging. We recently examined a patient who developed LHON at the age of 74 years and previously had proliferative diabetic retinopathy and bilateral macular holes. His central scotomas were initially attributed to the macular holes.

Case presentation

In February 2014, a 74-year-old man presented with a central scotoma in his left eye. His family history was unremarkable; as far as he knew, none of his immediate relatives had a visual problem. He was a former smoker (up to 20 cigarettes/day until the age of 54). He had type 2 diabetes mellitus, complicated by proliferative diabetic retinopathy for which he had received bilateral photocoagulation (Figure 1A). In December 2007, he was diagnosed with a macular hole in his right eye (Figure 2A), followed by his left eye in January 2013 (Figure 2B). The macular holes resulted in central scotomas and decreased his visual acuity to 15/100 in the right eye and 20/100 in the left eye. Both macular holes were closed by a combined vitrectomy with phacoemulsification: the right eye in December 25, 2007, and the left eye in February 14, 2013; his visual acuity subsequently improved to 20/40 in his right eye and 20/30 in his left eye on examination in August 2013 (Figure 2C).

The relapse of the central scotoma in his left eye (Figure 3A) was observed a year after the surgery and initially attributed to the macular hole. However, optical coherence tomography (OCT) images showed that the macular hole was still closed. Brain/orbital magnetic resonance imaging (MRI) failed to determine another cause of the scotoma. A 3-day course of intravenous methylprednisolone (1 g/day) followed by oral administration of...
Figure 1. Fundus photographs in February 2014 (A) and fluorescein angiography in April 2014 (B).

Figure 2. Optical coherence tomography images. (A) Stage 2 macular hole (arrow) in the right eye December 2007. (B) Stage 2 macular hole (arrow) in the left eye January 2013. (C) Bilateral macular holes were successfully closed after a combined vitrectomy/phacoemulsification, images taken in February 2013.
prednisolone had no effect except to increase his blood sugar level. Instead, he developed a similar central scotoma in his right eye in April 2014, 2 months after the onset in the left eye (Figure 3B). His visual acuity decreased to 20/500 in his right eye and 20/250 in his left eye. The treated macular holes remained unchanged throughout this time.

Fluorescein angiography, which had until then been withheld due to his renal condition, did not identify a pathogenic change in the fovea or the optic disc (Figure 1B). Neuromyelitis optica immunoglobulin G (NMO-IgG) was negative. Ultimately, we completed a workup for bilateral painless optic neuropathy, including mitochondrial genetic analysis, which revealed a 11778G→A mutation.

Discussion

A macular hole can be closed by vitrectomy, but it may have after-effects that could mask signs of other diseases. Although bilateral macular holes are relatively rare, as described in a recent study of 4507 cases with bilateral presentations in only 2.4% (108 patients), they can potentially present with bilateral central scotomas. In this case, however, his visual acuity declined despite the initial improvement postoperatively and the unchanged macular OCT, all of which suggested an alternative aetiology. From this case, similar to previous reports, we suggest that LHON be considered as a possible diagnosis even in older patients.

An onset of LHON can be not only associated with but also triggered by many factors, including diabetes, optic neuritis, anti-tuberculous medications, antiretroviral therapy for human immunodeficiency virus, carbon monoxide poisoning, solvent exposure, smoking, trauma, and even an increase in the intraocular pressure. For example, a 58-year-old man had an onset of visual loss 1 year after an elevation in his intraocular pressures was first recorded, and a 11778G→A mutation was identified. In our patient, a number of events, including an intraocular pressure fluctuation that was induced by the surgery, intraocular

Figure 3. Goldmann visual field testings. (A) Central scotoma OS and an asymptomatic paracentral relative scotoma OD in February 2014. (B) Central scotomas OU in July 2014.
tamponade, and postoperative posturing, might have induced the late onset of LHON.

**Declaration of interest**

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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