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Letter to the Editor

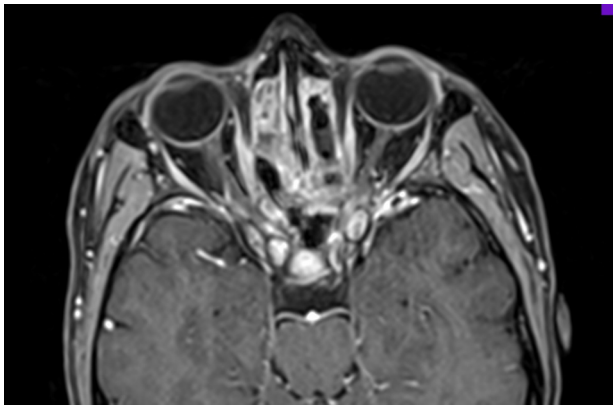

Fig. 1. MRI, axial sections, high-intensity appearance of the left optic nerve in the orbital apex.

Postoperative amaurosis after ethmoidectomy revealing Leber's hereditary optic neuropathy

1. Case report

Mrs. A., 32-year-old woman with no particular medical history, presented with stage 4 nasal polyposis. Bilateral ethmoidectomy was performed, with no intraoperative incident. On recovery, the patient reported loss of visual acuity in the left eye with no oculomotor deficit, rapidly progressing to amaurosis.

Emergency CT scan was performed, revealing no signs of compression or bone splinter, with an intact appearance of the optic nerve. Gadolinium-enhanced T1-weighted MRI (Fig. 1) showed a slightly infiltrated appearance of the optic canal and left orbital apex.

Ten months later, the patient consulted for sudden loss of visual acuity in the contralateral eye. MRI demonstrated left optic nerve atrophy with signal abnormalities extending as far as the optic chiasma. Contrast enhancement was observed in the intracranial portion of the right optic nerve and optic tract (Fig. 2).

The patient was then referred to a geneticist and gene sequencing revealed an *m.11778G>A* mutation (Mitochondrial DNA), indicative of Leber's hereditary optic neuropathy with a heteroplasmy rate of 98%.

2. Discussion

Loss of visual acuity is a very rare complication. In this case, a postoperative complication was the first diagnosis considered, as loss of visual acuity can usually be attributed to retro-orbital haematoma with an incidence of 0.1% [1], or a bone splinter. Imaging excluded these two causes. Complementary investigations then demonstrated optic neuropathy. Because of the two-stage progression, a genetic cause, including LHON, was proposed. Gene sequencing confirmed the presence of a G11778A/ND4 translocation implicated in LHON.

LHON is a mitochondrial disease caused by a *mtDNA* mutation, altering the respiratory chain and leading to selective death of retinal ganglion cells (RGC), resulting in optic nerve degeneration [2]. This genetic disease is known to be leading cause of hereditary optic neuropathy inducing central loss of vision, and is the most frequent genetic mitochondrial disease [3].

LHON generally remains asymptomatic for a long time before inducing subacute, painless, unilateral cecocentral visual loss. Bilateral loss of vision is observed almost systematically, after an average of 6 to 8 weeks [3]. MRI sometimes rapidly shows a nonspecific high-intensity signal of the retrobulbar optic nerve, and more frequently and later, optic nerve atrophy [4].

Incomplete penetrance and the presence of as yet poorly defined environmental factors appear to modify the onset of visual impairment [2]. Several cases of amaurosis associated with LHON have been reported after head injury or eye injury [5]. Similarly, endoscopic endonasal surgery may also induce the release of free radicals by oxidative stress [3] adjacent to a particularly vulnerable anatomical site, inducing death of RGCs, which would explain how simple trauma in this region can induce acute visual decompensation in an optic nerve that is already at risk due to mitochondrial dysfunction.

This case report is the first report of visual decompensation of LHON occurring after endoscopic endonasal surgery.

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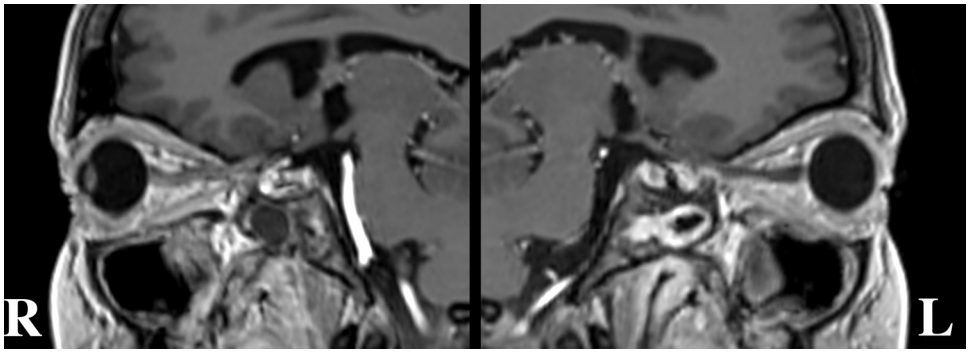

Fig. 2. MRI, sagittal sections, comparison of the contrast enhancement observed in the two optic nerves.

Disclosure of interest

The authors declare that they have no competing interest.

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