A case of IgG4-related disease with unilateral visual field impairment

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Abstract

A 76-year-old man receiving maintenance therapy with oral steroids for IgG4-related disease presented to our hospital with the chief complaint of visual disturbance. After detailed history-taking for IgG4-related disease, an imaging diagnosis of an intracranial lesion was made. Intensified steroid therapy was performed, which led to symptom resolution.

Introduction

IgG4-related disease is an autoimmune disease of unknown etiology in which IgG4-positive plasma cells infiltrate the body organs and cause swelling, nodules, and hypertrophic lesions in target organs. Approximately 20% and 5% of patients have vision loss and visual field defects, respectively¹. Two major pathological conditions can cause optic neuropathy in this disease: compression or encasement of the optic nerve by an enlarged tissue in the orbit and compression of the optic chiasm by the enlarged pituitary gland. In the former case, compression or encasement of the optic nerve by the enlarged external ophthalmic muscle or localized or diffuse tissue proliferation in the orbit has been reported². The latter is a condition that can develop in any disease that causes pituitary enlargement, including the present case; however, few studies have reported such cases³ and there are no case reports of visual impairment alone.

Case presentation

Initial evaluation

A 76-year-old man visited his family physician at the end of April X (year) due to visual disturbance in the left eye and was referred to our department on May 27, X. Ophthalmological history included bilateral cataract surgery, left ptosis (both in X-4 years), intrascleral fixation of left intraocular lens dislocation (in X-1 year), Irvine Gass syndrome, and ocular hypertension. Detailed history-taking revealed that the patient received treatment for IgG4-related disease (retroperitoneal fibrosis; prednisolone 2.5 mg/day) from his primary care physician.

Current medical history

At the initial visit on June 2 X, there was no conjunctival hyperemia in both conjunctivae, and anterior chamber cells were mildly present on the left eye. The left optic nerve papilla was mildly erythematous and swollen. Visual acuities were 1.2 (1.2 × S +1.75D) in the right eye and 0.6 (0.7 × S +2.00D) in the left eye, and intraocular pressures were 18 mmHg in the right eye and 16 mmHg in the left eye. Critical flicker frequency (CFF) was decreased in the left eye (right —38Hz—41Hz, left —36Hz—36Hz), and Humphrey visual field test revealed inferior auriculotemporal one-quarter blindness in the left eye (Figure 1). Brain magnetic resonance imaging (MRI) showed no abnormal signal in the infraorbital optic nerve; however,
marked pituitary/pituitary sclerenchyma enlargement with associated optic chiasm compression was observed (Figure 2).

Intervention description/results

Based on the history and initial evaluation findings, a diagnosis of left optic neuropathy associated with pituitary/pituitary scrofula enlargement caused by IgG4-related disease was made. On June 10, visual acuity and visual field defects improved (Figure 3), and upon examination on July 7, the pituitary gland and pituitary pattern were almost normalized (Figure 4), and the CFF decreased (right —42Hz—43Hz, left —37Hz—43Hz) (39 Hz). The steroid dose was gradually decreased, and maintenance therapy with 2.5-mg prednisolone daily was resumed on August 11.

Discussion

In this case, no optic nerve swelling or signal changes were evident on imaging, but fundus examination revealed mild erythema of the optic nerve papilla. Although this finding could be attributed to iritis, the possibility of optic neuritis caused by an autoimmune mechanism in IgG4-related disease could not be ruled out. However, the visual field findings were not consistent with those commonly observed in optic papillitis. Moreover, brain MRI findings suggested optic chiasm compression due to pituitary enlargement. The patient’s visual acuity and visual field defects improved following pituitary gland enlargement after steroid treatment, suggesting that pituitary gland enlargement and pituitary scrofula were the main pathophysiology of vision loss and visual field defects.

The ophthalmologic clinical hallmark of IgG4-related ocular disease is bilateral lacrimal gland enlargement with three features: suborbital nerve enlargement, exophthalmos, and pressure optic neuropathy. IgG4-related optic neuropathy is commonly caused by compression of the optic nerve by enlargement of the lacrimal glands or other intraorbital tissues. Few studies have reported on optic neuropathy caused by compression of the optic chiasm by an enlarged pituitary gland as in the present case. Although bilateral hemianopsia is generally observed with pituitary enlargement, the present case showed mild visual field defects in not only the auriculolateral lower one-quarter blindness but also the nasal lower one-quarter region. A previous study found visual field defects in only one eye in two of 10 patients with symmetrical pituitary enlargement. Because the present case also showed optic chiasm compression due to pituitary enlargement, it is possible that it caused an atypical visual field defect and associated visual acuity impairment.

Alternatively, as other studies have shown infiltration of IgG4-positive plasma cells around the trigeminal nerve in the same disease, a deep infiltrative or inflammatory mechanism may be involved in this case of optic neuropathy.

In conclusion, it is necessary to consider that the effects of IgG4-related disease may be involved when patients complain of vision loss and visual field impairment.

Declarations:

1. State of Ethics: Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.
2. Conflict of Interest: Not applicable.
3. Funding Sources: No funding was obtained for this study.
4. Author contributions and Data Availability: Shintaro Kohno, Hitoshi Tabuchi and Atsuki Fukushima evaluated the findings of patients and collected data. Shintaro Kohno and Atsuki Fukushima wrote the manuscript. All data generated or analysed during this study are included in this article.

References


Figure Legends

Figure 1. HFA visual field test on June 2. A. right eye. No abnormal findings. B. Left eye. Note inferior auriculotemporal 1/4 blindness in the left eye.

Figure 2. Brain MRI on June 2. A. Coronal Section. B. Sagittal Section. Note that marked pituitary/pituitary sclerenchyma enlargement with associated optic chiasm compression without any abnormal findings in optic nerve.

Figure 3. HFA visual field test on June 2. A. right eye. B. Left eye. Note that abnormal findings in the left eye disappeared.

Figure 4. Brain MRI on June 2, X year. R. A. Coronal Section. B. Sagittal Section. Note that the pituitary gland and pituitary pattern were almost normalized.

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