High-dose intravenous pulse steroid therapy for optic disc swelling and subretinal fluid in non-arteritic anterior ischemic optic neuropathy

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ABSTRACT

Non-arteritic anterior ischemic optic neuropathy (NAION) is a disease with microvascular abnormality that causes acute optic disc swelling (ODS) and, in severe cases, subretinal fluid (SRF) accumulation. ODS causes compartment syndrome and subsequent axonal degeneration and loss of retinal ganglion cells by apoptosis. No treatment modalities have been effective, although some cases improved after the intake of oral systemic steroids. We reported a case of a 72-year-old man who was referred due to a visual defect in the right eye. At first presentation, visual acuity and visual field were disturbed; critical flicker frequency (CFF) was decreased; and optic coherence tomography (OCT) showed ODS and SRF. Microscopic examination revealed parapapillary hemorrhage and fluorescence angiography showed non-filling, temporal-superior choroidal lesion adjacent to the optic disc at an early phase. After high-dose intravenous steroid treatment, SRF and ODS were decreased, and completely resolved after 30 days. Visual acuity and CFF were improved, and visual field was enlarged. High-dose intravenous steroids could possibly resolve SRF and ODS and improve visual function of patients with NAION. Some cases in NAION improved visual acuity and visual function in natural course, more cases were needed to evaluate the efficiency.

Key Words: ION, macular edema, NAION, OCT, systemic steroids

INTRODUCTION

Non-arteritic anterior ischemic optic neuropathy (NAION) is a microvascular disease process affecting the optic nerve head and resulting in optic disc swelling (ODS), vascular attenuation, variable degrees of optic nerve hemorrhage, and neuroretinal rim pallor. The optic nerve ischemia leads to consequent swelling of the optic nerve head, resulting in optic nerve compression and inflammation through a compartment syndrome. Crowding of the optic disc leads to mechanical compression of the optic nerve fibers through the lamina cribrosa, followed by impairment of axonal flow and capillary perfusion of the optic nerve head. Subsequent vasogenic and cytotoxic optic nerve edema probably increase the compartment syndrome and worsen the ischemia through the release of cytotoxic factors, with subsequent axonal degeneration and
loss of retinal ganglion cells through apoptosis. In severe cases, optic nerve edema can cause subretinal fluid (SRF) accumulation in the macula; SRF accumulating in the parapapillary region is thought to be a sign of poor clinical course. These pathological changes in NAION could be followed up by optical coherence tomography (OCT) of the eyes.

Decompression of the optic nerve head in the case of compartment syndrome can prevent optic nerve damage. Many options, including a wide range of agents, have been proposed for empiric treatment of NAION, but none of them have facilitated significant improvement. The rationale for the use of oral steroids for the treatment of NAION was based on the hypothesis that steroids possibly decrease capillary permeability. Faster resolution of ODS reduces the compression of capillaries in the optic nerve head and improves blood flow, which restores function and survival. Here, we reported a case of severe NAION that was treated by high-dose intravenous pulse steroids; excellent improvement of ODS and SRF was obtained and documented by follow-up OCT observation.

CASE PRESENTATION

A 72-year-old man was referred to our department due to a visual defect in the right eye. His right visual acuity and visual field were suddenly impaired upon waking up. Two weeks prior, he had similar, but milder symptoms that spontaneously resolved after 2 h. He had no headache, ocular pain, or diplopia. For 13 years, he had hypertension and hyperlipidemia that

Fig. 1 Color photos, fluorescence angiography, and visual field of the right eye at first visit
Color photo (A), visual field (B), and fluorescence angiography in the early (C) and late (D) phases.
were well controlled by medications. Five years ago, he was diagnosed to have NAION in the left eye and observed with no treatment.

At first presentation, best corrected visual acuity (BCVA) was 20/660 OD and 20/200 OS. Critical flicker frequency (CFF) was 10 Hz OD and 15 Hz OS; intraocular pressure was 13 mmHg OD and 12 mmHg OS. Both direct and indirect light reflexes were absent. Microscopic examination found mild cataract and parapapillary hemorrhage OD (Figure 1A). The remaining visual field in the right eye was only in the shape of an island (Figure 1B). Fluorescence angiography showed non-filling of a temporal-superior choroidal lesion that was adjacent to the optic disc at the early phase and leakage from the disc and pooling of macular SRF at the late phase (Figures 1C and 1D). OCT detected SRF accumulation in the macula extending to the ODS (Figure 2). Central retinal thickness (CRT) was 546 μm. Serum examination showed only a slight increase in C-reactive protein (0.28 mg/dl), without any changes in erythrocyte sedimentation rate and serum fibrinogen. With a diagnosis of NAION with SRF and ODS by papillary inflammation

![Fig. 2 Changes in macular structure on optical coherence tomography](image)

Macular structures of the right eye at days 0, 1, 2, 3, 7, 14, and 30. White arrows indicate SRF
in the right eye, intravenous pulse steroid therapy at 1000 mg methylprednisolone per day for 3 days was administered. Following this treatment, there was an immediate decrease in SRF and ODS. In particular, SRF decreased from 546 μm to 405 μm on day 1, 383 μm on day 2, and 327 μm on day 3 (Figure 2); ODS decreased simultaneously with a decrease in SRF (Figure

Fig. 3  Changes in optic disc swelling on optical coherence tomography  
ODSs of the right eye at days 0, 1, 2, 3, 7, 14, and 30.

Fig. 4  Changes in visual field  
The visual field of the right eye at days 0, 1, 3, 7, 14, and 30.
3). Visual acuity and CFF improved to 20/320 and 18 Hz, respectively; visual field enlarged to include the superior area on day 3 (Figure 4). After the intravenous steroid pulse treatment, we started oral prednisone at a dose of 60 mg daily for three days, followed by gradual tapering by 10 mg every three days before discontinuation. On day 30, there was complete resolution of SRF (Figure 2) and ODS (Figure 3); BCVA and CFF were 20/125 and 20 Hz, respectively. Visual field enlarged gradually, but the inferior visual field defect was not recovered (Figure 4).

**DISCUSSION**

In some patients, NAION has been reported to cause ODS and SRF accumulation\(^2\); in fact, Hedges *et al.* showed that SRF accumulated in 8 out of 76 eyes with NAION.\(^5\) Visual acuity in patients with SRF accumulation is severely disturbed than the visual acuity of patients without SRF. In a previous report, the visual acuities of two patients with CRT of more than 400 μm were 1/200; this value was worse than the visual acuity in patients with SRF accumulation of less than 400 μm. In the current case, the CRT at first presentation was 532 μm and visual acuity was 20/660, which were almost the same as those previously reported for severe cases. After intravenous 1000 mg methylprednisolone, ODS and SRF were decreased and resolved at day 30. In addition, visual acuity recovered to 20/125 (six lines improvement) and CFF to 18 Hz at day 30.

ODS may sometimes contribute to a compartment syndrome that compresses the fine capillary blood supply of the optic nerve head, resulting in ischemia and axonal damage. In eyes with severe NAION, compartment syndrome worsens the ischemia by the release of cytotoxic factors; therefore, the clinical course would be worse.\(^3\) A faster resolution of the compartment syndrome would be important for treating severe NAION. Systemic steroids were used to shorten the duration of ODS and may improve visual outcome in patients with NAION. A previous study reported that oral prednisone 80 mg daily during the acute phase resolved ODE faster, and resulted in a significantly higher probability of improvement in visual acuity and visual field, than observation without treatment. They described that faster resolution of ODS with corticosteroid therapy could decrease compression of the capillaries in the optic nerve head, thus better blood flow in the capillaries and improve circulation in the optic nerve and function of hypoxic axons.\(^12\) In this case, a slight increase in C-reactive protein without any changes in erythrocyte sedimentation rate and serum fibrinogen was observed at first visit, thus this neuritis was not caused by systemic inflammation disease, such as Takayasu disease. After administration of systemic steroid, ODS and SRF were decreased. It could be considered that these mechanisms occurred. Another study showed that for treating optic neuritis, high-dose intravenous steroids significantly improved disc edema, visual acuity, and visual field faster than oral steroids.\(^14\) For orbital compartment syndrome, high-dose intravenous steroids could be used as initial therapy to decrease optic nerve pressure.\(^15, 16\)

However, there are some limited and conflicting evidence. Rebolleda *et al.* used the same dose of prednisone during the acute phase of NAION and found no significant changes in visual acuity and visual field.\(^13\) To date, there is no class I evidence of benefit for the treatment of NAION, and in natural course of NAION, some cases improved visual acuity and visual field after 6 month.\(^17\) This report is one case report, more cases would be needed to evaluate the efficiency.

In conclusion, SRF and ODS caused by severe NAION were resolved by high-dose intravenous steroid medication. Clinical studies on the use of intravenous steroids for NAION would be necessary to validate this efficacy.
COMPETING INTERESTS

No author had any funding support related to the manuscript. The authors declare that they have no competing interests.

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