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Case report

Vasculitis with superior ophthalmic vein thrombosis compatible with neuro-neutrophilic disease

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ABSTRACT

Purpose: To present a unique case of neuro-neutrophilic disease with inflammation and thrombosis of the superior ophthalmic vein (SOV).

Observations: A 43-year-old Japanese man with past histories of oculomotor paralysis, auditory disorder, ischemic enteritis, and recurrent oral ulceration was referred to our hospital because of blurred vision in his right eye. Ophthalmic examination revealed decreased best corrected visual acuity and central scotoma in his right eye. Orbit magnetic resonance imaging (MRI) revealed an enlarged SOV in the right eye, with Gadolinium (Gd) enhancement in the wall of the vein but not inside the vein, indicating thrombosis. Multiple Gd-enhanced hyperintense lesions were also observed in the juxtacortical area of the brain. We diagnosed the patient with vasculitis in the right SOV that was adversely affecting the optic nerve. We ruled out systemic thrombophilia, infections, and malignancy by systemic examinations. The human leukocyte antigen (HLA) typing was Cw1-, B54-, B61-, A2-, A24-, and DR4-positive and B51-negative. We treated the patient with systemic steroid and anticoagulant therapy. After three courses of steroid pulse therapy, his symptoms and the MRI findings of the right SOV and brain improved; therefore, we decided to discontinue the anticoagulant therapy. One month after anticoagulant cessation, MRI revealed recurrence of the thrombus and enlargement of the right SOV despite the lack of vision worsening. We restarted the anticoagulant therapy while continuing the oral prednisolone treatment. At the final visit, 14 months after the onset of the disease, the patient was still receiving oral anticoagulation with warfarin potassium and prednisolone (5 mg/day). His symptoms and the right eye's visual function remained normal with a mildly enlarged SOV; there was less Gd enhancement and no brain lesions on MRI.

Conclusions and importance: We treated a unique case of possible neuro-neutrophilic disease that presented visual disturbances due to right SOV inflammation and thrombosis. Anticoagulation and systemic steroid therapies were required to reduce the inflammation and to prevent the recurrence of thrombosis.

1. Introduction

Neuro-neutrophilic diseases include neuro-Behçet disease (NBD) and neuro-Sweet disease (NSD) that are characterized by aseptic inflammatory lesions of the central nervous system (CNS).[1] Here, we present a unique case of neuro-neutrophilic disease with inflammation and thrombosis of the superior ophthalmic vein (SOV).

2. Case report

A 43-year-old Japanese man was referred to our hospital with the complaint of blurred vision in his right eye. He had past histories of oculomotor paralysis and auditory disorder at the age of 15 and 37 years, respectively, both of which recovered with systemic corticosteroid therapy. He had suffered from ischemic enteritis at the ages of 33 and 42 years. He also had episodes of recurrent oral ulcerations.

Ophthalmic examination revealed best corrected visual acuities (BCVAs) of 0.5 and 1.2 and intraocular pressures of 18 mmHg and 17 mmHg in the patients' right and left eyes, respectively. His pupils were isocoric and the light reflex was normal in both eyes with no afferent pupillary defects. No abnormal findings were observed in the lids, anterior segments, and fundi (Fig. 1A) of either eye. Fluorescein

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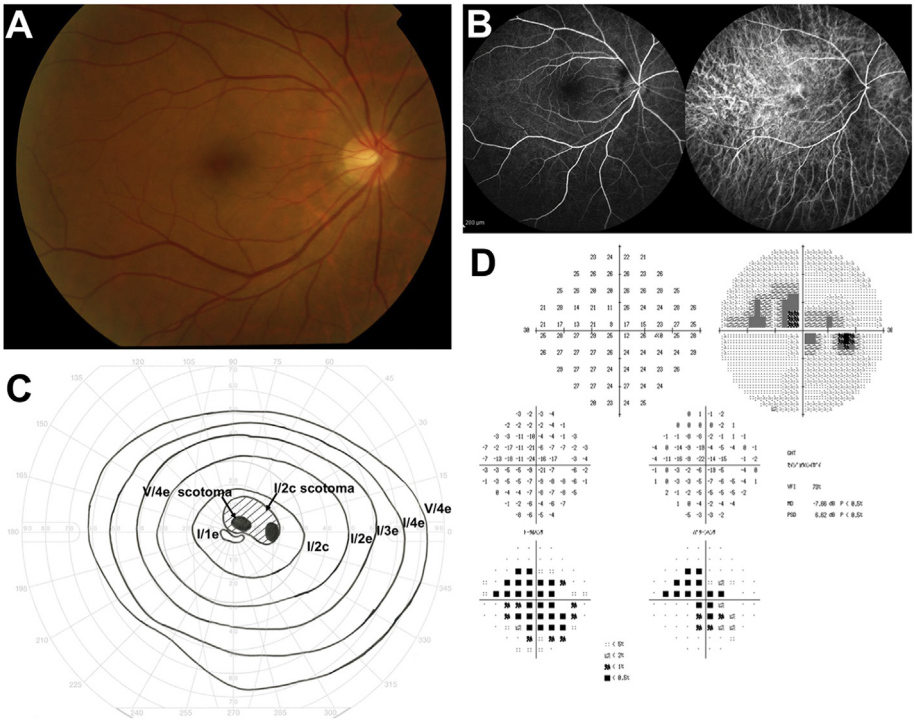

Fig. 1. Ophthalmic tests of the patient's right eye on the first visit.
Funduscopy (A) and fundus angiography (B) show no abnormality. Goldmann kinetic visual field test (C) and Humphrey visual field test (D) showing central scotoma.

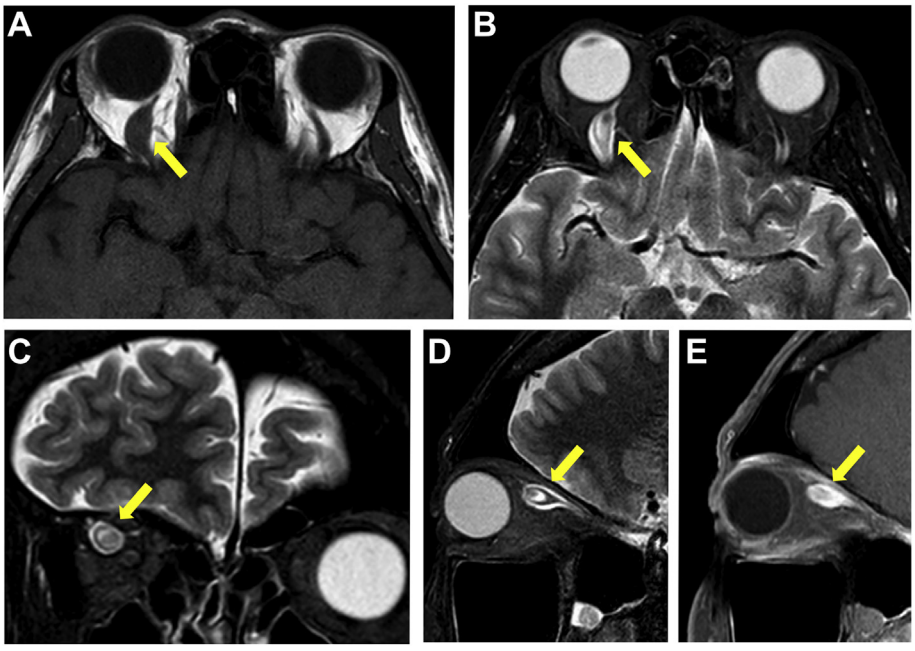

Fig. 2. Magnetic resonance imaging of the right orbit before treatment.
T1-weighted orbit magnetic resonance imaging MRI (A) revealed an enlarged superior ophthalmic vein (SOV) (arrow) in the right eye. Short-T1 inversion recovery (STIR) of the cross-sectional image of SOV showed a thickened wall with a high signal intensity, an intermediate zone with a low signal intensity, and a central spot with a high signal intensity (B–D) (arrow). SOV showed Gadolinium (Gd) enhancement in the wall of the vein but not inside the vein corresponding to the central spot in the STIR image (E) (arrow), and indicating thrombosis in the inflamed SOV.

and indocyanine green angiography (Fig. 1B), optical coherent tomography, and multi-focal electroretinogram findings were also normal for both his eyes. Visual field (VF) tests showed central scotoma in the right eye (Fig. 1C and D). BCVA in the right eye further declined to 0.1 3 weeks after the referral. T1-weighted orbit magnetic resonance imaging (MRI) revealed an enlarged SOV in the right eye (Fig. 2A). Short-T1 inversion recovery (STIR) showed an owl's eye-appearance of the cross-sectional image of SOV, i.e., a thickened wall with a high signal intensity, an intermediate zone with a low signal intensity, and a central spot with a high signal intensity (Fig. 2B–D). The right eye SOV also presented Gadolinium (Gd) enhancement in the wall of the vein but not inside the vein corresponding to the central spot in the STIR image (Fig. 2E), indicating thrombosis in the inflamed SOV. We found no abnormal MRI findings in the optic nerves. However, juxtacortical hyper-intensity lesions with Gd enhancement were also observed in the left temporal and insular lobes (Fig. 3A). Magnetic resonance angiography findings were unremarkable (data not shown); however, magnetic resonance venography revealed stenosis of the left transverse sinus with collateral vascular flow, indicating a chronic disturbance of the venous return (Fig. 4A and B). We diagnosed the patient as having vasculitis with a thrombus in the right SOV that was affecting the optic nerve and resulted in visual disturbance; we admitted him to our