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Unilateral Visual Disturbance Due to Posterior Ischemic Optic Neuropathy in Eosinophilic Granulomatosis with Polyangiitis: A Case Report

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Dear Editor,

Posterior ischemic optic neuropathy (PION) is an acute optic neuropathy caused by ischemia of the posterior optic nerve, which requires early identification of its etiology to initiate timely treatment and preserve vision. Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare small- and medium-sized vessel vasculitis that leads to widespread damage to peripheral organ systems including the respiratory, neurological, and renal systems with rarely reported ocular manifestations such as PION. We describe the first documented case of PION due to EGPA in Korea. This study was approved by the Institutional Review Board of Gangnam Severance Hospital, Yonsei University College of Medicine, with a waiver of informed consent (No. 2023-0728-002).

A 55-year-old male patient with a fever of unknown origin, presented with an acute visual disturbance in the inferior visual field of his left eye (Fig. 1A). On examination, his visual acuity was 1.0 in both eyes. The left eye showed relative afferent pupillary defect (RAPD) grade 2–3, while no RAPD was observed in the right eye. Ocular movement, anterior segment, and intraocular pressure were normal in both eyes. While no tenderness over the bilateral temporal region was noted, right median nerve palsy and generalized skin rash were observed.

Diffusion-weighted imaging (DWI) findings showed increased signal intensity of the left posterior optic nerve, without any significant mass, inflammation, or lymphadenopathy (Fig. 1B). Fundus imaging showed a normal cup-disc ratio of 0.44 (Fig. 1C). No thinning of the retinal nerve fiber layer and the ganglion cell-inner plexiform layer (GCIPL) were observed (Fig. 1D, 1E). As initial fundus imaging findings did not show disc swelling with posterior optic nerve diffusion restriction on DWI, PION was suspected [1,2].

Laboratory findings showed a white blood cell count of 18,900/μL with 12.9% eosinophilia (normal range, 4,000–10,800/μL with 0%–7%), C-reactive protein of 111.7 mg/L (normal range, 0–8 mg/L) and the erythrocyte sedimentation rate of 120 mm/hr (normal range, 0–15 mm/hr). The patient tested positive for antinuclear antibodies at a titer of 1:80, although both myeloperoxidase-antineutrophil cytoplasmic antibodies (MPO-ANCAs) and proteinase 3-ANCAs (PR3-ANCAs) were negative.

With the suspicion of giant cell arteritis, a superior temporal artery biopsy was performed. The biopsy revealed only focal lymphocytic infiltration in the adventitia without the presence of multinucleated giant cells. Additional

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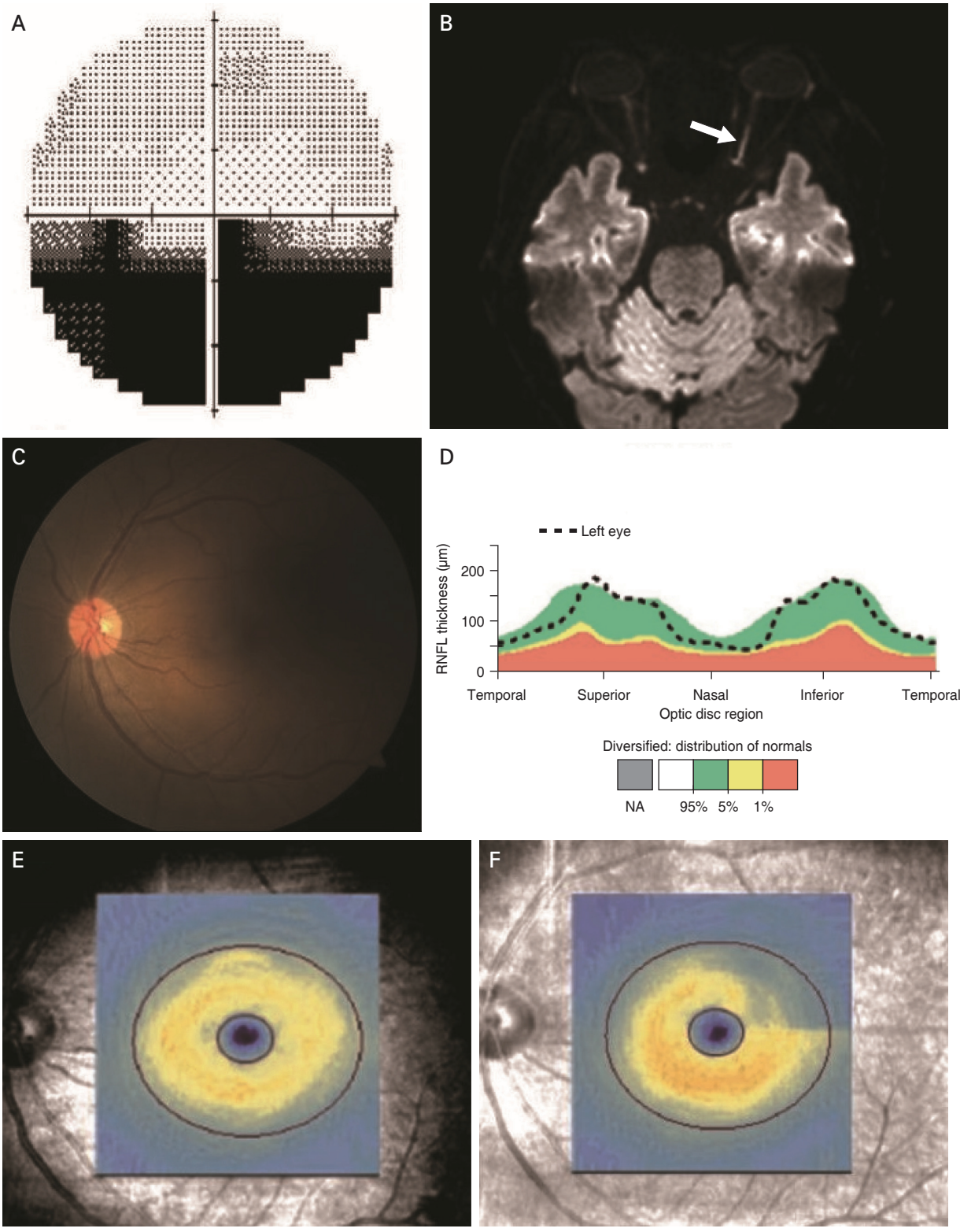

Fig. 1. Clinical and imaging findings. (A) Humphrey visual field test showing inferior visual field defect. (B) Diffusion-weighted imaging showing increased signal intensity of the left optic nerve (arrow). (C) Fundus imaging with no definite optic disc swelling. (D) Retinal nerve fiber layer (RNFL) thickness within normal range on optical coherence tomography (OCT). (E) OCT test showing normal ganglion cell layer. (F) Ganglion cell-inner plexiform layer thinning was observed in the upper left on OCT after 4 months of follow-up. NA = not applicable.

skin biopsy of the patient's trunk showed eosinophil predominant lymphocytic infiltrate suggesting EGPA. As this patient met the criteria for eosinophilia, mononeuropathy, and extravascular eosinophilic predominant inflammation on biopsy from the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for EGPA, the diagnosis of EGPA was made [3]. Patient did not present with other systemic findings and bronchial provocation test for obstructive airway disease was negative.

To prevent further optic nerve damage, the patient was started on corticosteroid treatment with a daily 30 mg dose of prednisolone and an immunosuppressive agent with cyclophosphamide 750 mg. Follow-up after 4 months revealed decreased GCIPL thickness in the superotemporal sector with the GCIPL thickness down from 82 to 71 μm. (Fig. 1E, 1F) Over the following 2 years, no further deterioration in the visual field test or optic nerve was observed. This positive outcome suggests the effectiveness of immunosuppressive therapies in managing PION associated with EGPA.

In conclusion, our case emphasizes the importance of considering EGPA as a possible underlying cause in patients presenting with clinical features of PION. Although exceedingly rare, when the etiology of optic nerve damage remains unidentified, it is essential to conduct a comprehensive evaluation for a systemic cause of the disease. Timely administration of high-dose corticosteroid and immunosuppressive agents is crucial to prevent further visual impairment in such cases. This report adds to the limited literature on EGPA-associated PION, paving the way for future research and enhancing clinical management strategies for patients with a similar presentation.

Conflicts of Interest: None.
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References

1. Robson JC, Grayson PC, Ponte C, et al. 2022 American College of Rheumatology/European Alliance of Associations for rheumatology classification criteria for granulomatosis with polyangiitis. *Ann Rheum Dis* 2022;81:315–20.
2. Abel A, McClelland C, Lee MS. Critical review: typical and atypical optic neuritis. *Surv Ophthalmol* 2019;64:770–9.
3. Kupersmith MJ, Garvin MK, Wang JK, et al. Retinal ganglion cell layer thinning within one month of presentation for optic neuritis. *Mult Scler* 2016;22:641–8.