



## Correspondence

Korean J Ophthalmol 2021;35(2):168-170  
<https://doi.org/10.3341/kjo.2020.0100>

### Antiphospholipid Syndrome of Bilateral Nonarteritic Anterior Ischemic Optic Neuropathy Patient Deteriorating Over Time

Dear Editor,

We reported the case of a patient with antiphospholipid syndrome (APS) in bilateral nonarteritic anterior ischemic optic neuropathy (NAION). APS is an autoimmune disease affecting various organs, associated with hypercoagulability and vascular thrombosis [1]. APS rarely accompany NAION, and the invasion of both eyes is unknown as far as authors know.

A 70-year-old male reported painless reduced vision in left eye suddenly occurred 2 days ago. There were no underlying diseases, and corrected visual acuity was 1.0 in right and 0.1 in left eye. Relative afferent pupillary defect was observed in left eye. During fundusoscopic examination, mild inferior focal disc swelling in right eye and overall disc swelling with inferior flame shape hemorrhage in left eye were observed. During visual field examination, increased superior blind spot through sensitivity degradation in right eye and blind spot along horizontal line were observed. Focal disc leakage and nonperfusion in right eye and overall leakage in left eye were observed from fluorescent angiography (FAG) (Fig. 1A-1G). Magnetic resonance imaging revealed no clear high signal in optic nerve region. Erythrocyte sedimentation rate and C-reactive protein were normal. One gram of methylprednisolone was administered daily for treating suspected severe NAION and optic neuritis in left eye. During the treatment, laboratory

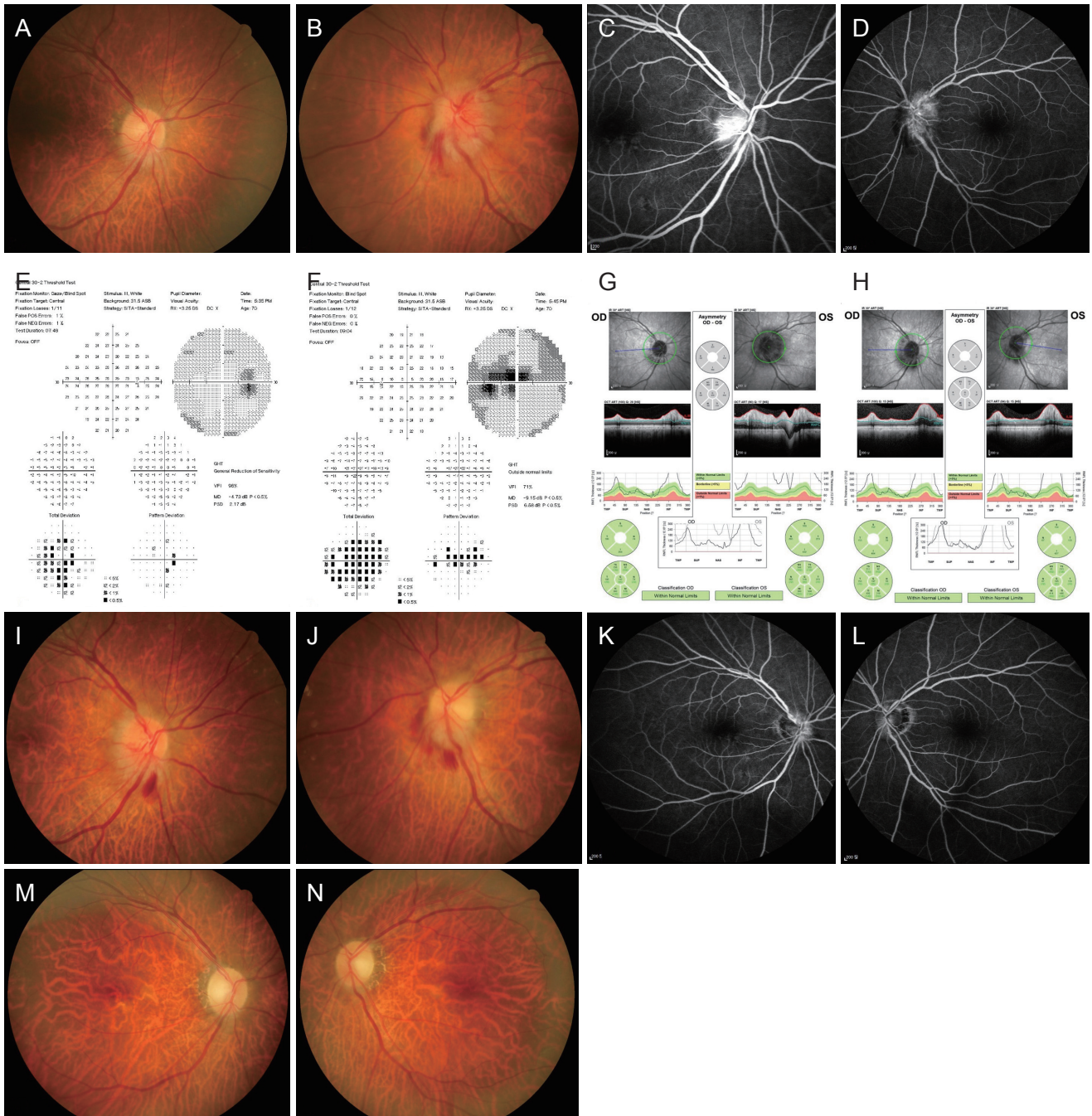
tests for blood coagulation factor were prothrombin time (PT) 16.6 (11.9–14.3 seconds), PT 64 (86–127%), PT 1.35 (0.8–1.2 international normalized ratio), and activated partial thromboplastin time 44.1 (29.1–43.5 seconds). Fluorescent antinuclear antibody showed positive findings. An additional antibody test was performed after rheumatologist's consultation. Rheumatologic evaluations resulted in no positive results. Anticardiolipin test showed positive IgM with high titer. IgM positivity in the anticardiolipin test was observed again 12 weeks after the previous test, which confirmed antiphospholipid antibody syndrome. Also, bone marrow study for differential diagnosis of cancer and other diseases resulted in normal. As hematologic abnormalities were observed during treatment, warfarin (Kufarin, Hana, Seoul, Korea) 3 mg was used. Left eye disc swelling and hemorrhage improved, but visual acuity did not recover. Corrected visual acuity remained at 1.0 on right eye, and was 0.06 on left eye.

During the observation of the outpatient course after steroid tapering, sudden loss of vision occurred in right eye 1 month after the steroid treatment, and corrected visual acuity was 0.1 in right eye and 0.06 in left eye. Upon examination, exacerbated focal disc swelling with flame shape hemorrhage was observed in right eye (Fig. 1H-1J). After consultation with rheumatologist, 60 mg of enoxaparin (Clexane, SA, Seoul, Korea) was administered intravenously twice a day, and tapering was conducted after re-administering 1 g of methylprednisolone for 3 days. One month after the treatment, right eye hemorrhage and disc swelling improved, but there was no improvement in vision. After 1 year of observation, optic nerve became pale, and nonperfusion areas were observed in both eyes from FAG (Fig. 1K-1N). Corrected visual acuity was 0.1 in right eye and 0.06 in left eye, showing no improvement in vision.

Received: July 31, 2020 Final revision: November 24, 2020  
Accepted: November 25, 2020

© 2021 The Korean Ophthalmological Society

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.



**Fig. 1.** Fundus photographs, visual field, optical coherence tomography spectralis in axon mode and fluorescein angiography of the patient. (A) Right eye showing mild inferior disc swelling at the first visit. (B) Left eye showing total disc swelling and inferior flame shape hemorrhage at the first visit. (C) Right eye showed focal disc leaking and nonperfusion at the first visit (captured at 2 minutes 45 seconds). (D) Left eye showed overall disc leakage at the first visit (captured at 1 minutes 3 seconds). (E) Right eye showing increased superior blind spot at the first visit. (F) Left eye showing blind spot along the horizontal line at the first visit. (G) Right eye showing mild inferior focal disc swelling and overall disc swelling in left eye at the first visit. (H) Right eye showing exacerbation superiotemporal and inferior disc swelling and improved disc swelling in the left eye 1 month after the treatment. (I) Right eye showing exacerbation disc swelling with flame shape hemorrhage 1 month after the treatment. (J) Left eye showing improved disc swelling and flame shape hemorrhage 1 month after the treatment. (K,L) Both eyes showed overall disc nonperfusion area one year later (captured at 1 minute). (M,N) Both eyes showed pale disc 1 year later.

NAION is discovered with a painless loss of vision accompanied by optic disc edema. It needs to be distinguished from optic neuritis. The authors believed that visual acuity did not respond to sudden vision loss treatment. In addition, splint hemorrhage and focal swelling on disc with focal perfusion decrease from FAG were observed. Also, no magnetic resonance imaging contrast enhancement showed symptoms appropriate for the diagnosis of ischemic optic neuropathy [2,3].

Ophthalmologically, APS can make various invasions such as vitiritis, hemorrhage, leakage, and edema from anterior to posterior [1]. However there are few reports in which it appears as NAION. In APS cases reported by Serrador-Garcia et al. [4] and Tugcu et al. [5], NAION occurred unilaterally. However, no study reported bilateral NAION caused by APS.

Standard treatment strategy of APS is reducing thrombotic risk [1]. There are reports about use of warfarin, antiplatelets, anti-inflammatory drug and hydroxychloroquine, after APS diagnosis [5]. Authors also started using warfarin after confirming blood abnormality, and intensively used antithrombotic agents intravenously at the second episode, but the prognosis was poor. Intravenous steroid treatment in NAION is controversial, but authors used this for optic neuritis treatment. If NAION occurs in both eyes of old age and does not respond to treatment, APS should be suspected and related systemic examination might be necessary. Also, the prognosis of visual acuity can be poor despite active treatment.

Yongwun Cho, Byoung Seon Kim

*Department of Ophthalmology, Gyeongsang National University Hospital, Gyeongsang National University College of Medicine, Jinju, Korea*

Woong-Sun Yoo, Seong Wook Seo, In Young Chung

*Department of Ophthalmology, Gyeongsang National University Hospital, Gyeongsang National University College of Medicine, Jinju; Institute of Health Science, Gyeongsang National University, Jinju, Korea*  
*E-mail (In Young Chung): in0chung@gnu.ac.kr*

## Conflict of Interest

No potential conflict of interest relevant to this article was reported.

## References

1. Utz VM, Tang J. Ocular manifestations of the antiphospholipid syndrome. *Br J Ophthalmol* 2011;95:454-9.
2. Abel A, McClelland C, Lee MS. Critical review: typical and atypical optic neuritis. *Surv Ophthalmol* 2019;64:770-9.
3. Peeler C, Cestari DM. Non-Arteritic Anterior Ischemic Optic Neuropathy (NAION): a review and update on animal models. *Semin Ophthalmol* 2016;31:99-106.
4. Serrador-Garcia M, Santos-Bueso E, Saenz-Frances F, et al. Non-arteritic anterior ischemic optic neuropathy as first manifestation of antiphospholipid syndrome in a young patient. *Arch Soc Esp Oftalmol* 2014;89:368-72.
5. Tugcu B, Acar N, Coskun CT, et al. Nonarteritic anterior ischemic optic neuropathy as the presenting manifestation of primary antiphospholipid syndrome. *Indian J Ophthalmol* 2014;62:642-4.