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Case report

Primary antiphospholipid syndrome presenting with homonymous quadrantanopsia

Hee Kyung Yang[a], Ki Won Moon[b], Min Jung Ji[c], Sang Beom Han[c,*], Jeong-Min Hwang[a]

[a] Department of Ophthalmology, Seoul National University Bundang Hospital, Seoul National University College of Medicine, Seongnam, South Korea
[b] Department of Internal Medicine, Kangwon National University Hospital, Kangwon National University Graduate School of Medicine, Chuncheon, South Korea
[c] Department of Ophthalmology, Kangwon National University Hospital, Kangwon National University Graduate School of Medicine, Chuncheon, South Korea

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ABSTRACT

Purpose: To report a case of primary antiphospholipid syndrome presenting with isolated homonymous superior quadrantanopsia.

Observations: A 50-year-old Korean man presented with subjective visual disturbance for 1 month. Visual field testing showed a right homonymous superior quadrantanopsia. Brain magnetic resonance imaging (MRI) revealed an old infarct in his left occipital lobe and multiple lesions in other areas of the brain. Laboratory tests showed a marked increase in serum anti-β2 glycoprotein I antibody, which remained elevated after 12 weeks. He was diagnosed with primary antiphospholipid syndrome and started anticoagulation therapy.

Conclusions and importance: This is the first case report of primary antiphospholipid syndrome presenting with isolated homonymous quadrantanopsia. Antiphospholipid syndrome should be considered as a differential diagnosis in patients with homonymous visual field defects accompanying multiple cerebral infarcts.

1. Introduction

Antiphospholipid syndrome is an autoimmune disease characterized by hypercoagulability, increasing the risk of arterial and/or venous thromboses. It is caused by antiphospholipid antibodies, such as, lupus anticoagulant, anticardiolipin antibody, or β-2 glycoprotein-I IgG or IgM isotype.[1] Ophthalmologic manifestations are reported in 8–88% of the patients with antiphospholipid syndrome, and can be the presenting sign of the disease.[2] Neuroophthalmologic manifestations, such as cranial nerve palsies, ischemic optic neuropathy, transient visual loss, and infarct of the visual pathway are associated with antiphospholipid syndrome.[2,3] However, through a comprehensive search of the MEDLINE database, antiphospholipid syndrome initially presenting with isolated homonymous quadrantanopsia has not been reported. Herein, we report a case of homonymous quadrantanopsia as the only presenting feature of primary antiphospholipid syndrome.

1.1. Case report

A 50-year-old Korean man visited the ophthalmology clinic presenting with vague visual disturbance for the past month. He also complained of mild dizziness. His past medical history was unremarkable. At presentation, his best-corrected visual acuities were 20/20 in both eyes. Color vision were normal and relative afferent pupillary defect was absent in both eyes. Fundoscopic examination showed normal optic discs and macula in both eyes. Humphrey 24–2 (Carl Zeiss Meditec, Inc., Dublin, CA) visual field testing showed a right congruous superior quadrantanopsia, respecting the vertical midline (Fig. 1). Brain magnetic resonance imaging (MRI) revealed an old infarct in the left occipital lobe and subacute infarcts with cortical laminar necrosis in the right occipital lobe (Fig. 2). Old lesions were found in the cerebellum, left parietal lobe, both frontal lobes, basal ganglia, and subcortical white matter of both hemispheres. Moreover, multiple hyperintense foci were present in the subcortical white matter of both hemispheres, suggesting an acute infarct. Magnetic Resonance Angiography (MRA) demonstrated subtle diffuse luminal irregularities at the vertebrobasilar artery and both distal internal carotid arteries, and mild segmental stenosis at both proximal internal carotid arteries. Laboratory tests showed a marked increase in plasma anti- β-2 glycoprotein-I IgG (84.8 CU; normal range ≤ 20). Lupus anticoagulant, anticardiolipin antibody, antinuclear antibody, and rheumatoid factor were within normal range. Other tests—complete blood cell count, liver panel, renal and thyroid function tests, coagulation panel, and blood chemistry tests—were normal. He was finally diagnosed with primary antiphospholipid syndrome.

He received anticoagulant treatment with aspirin 100mg once a

* Corresponding author. Department of Ophthalmology, Kangwon National University Hospital, 156 Baengnyeong-ro, Chuncheon, Kangwon 200-722, South Korea.
E-mail address: m.sangbeom.han@gmail.com (S.B. Han).

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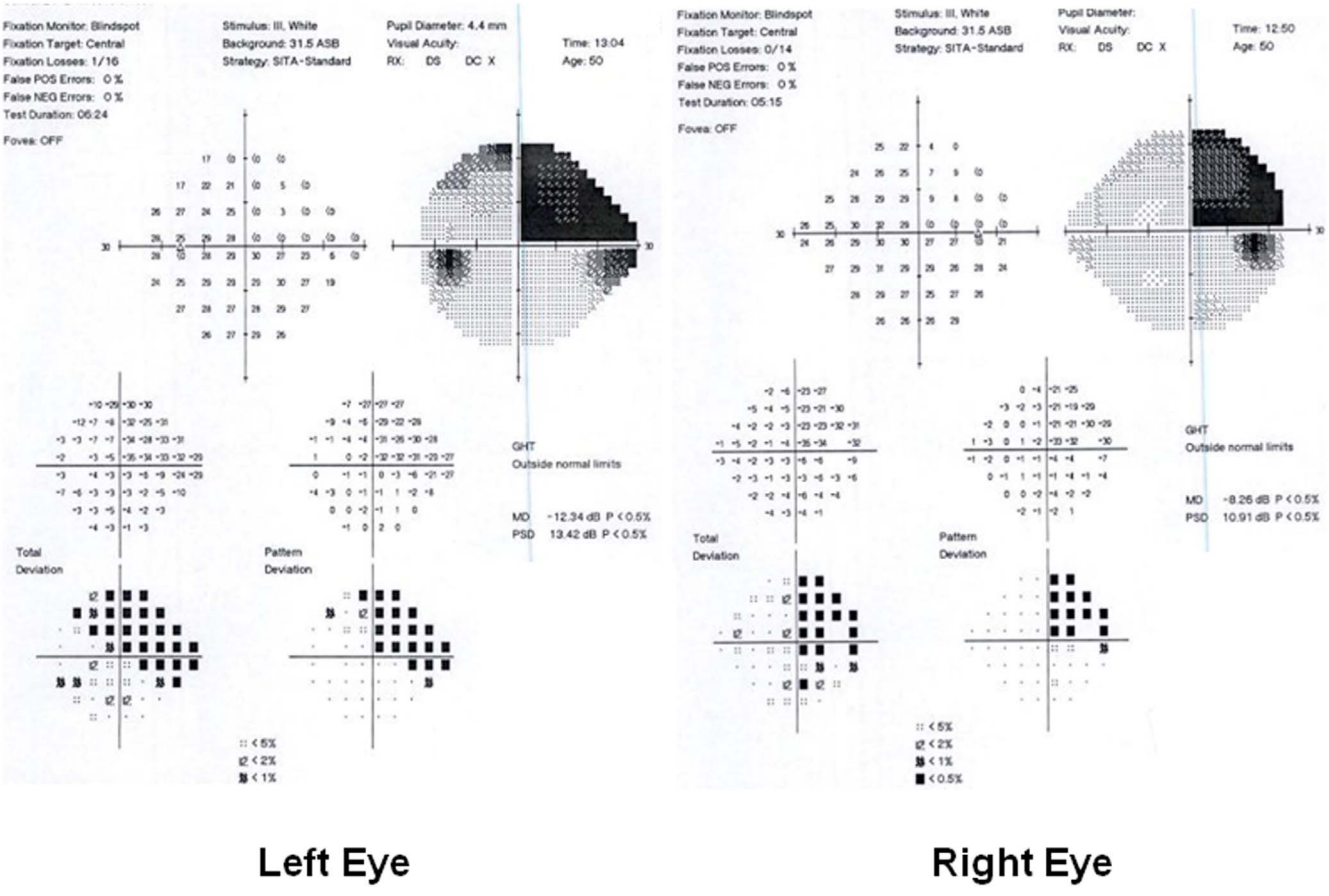

Fig. 1. Humphrey 24–2 (Carl Zeiss Meditec, Inc., Dublin, CA) visual field testing reveals right superior quadrantanopsia respecting the vertical meridian in both eyes.

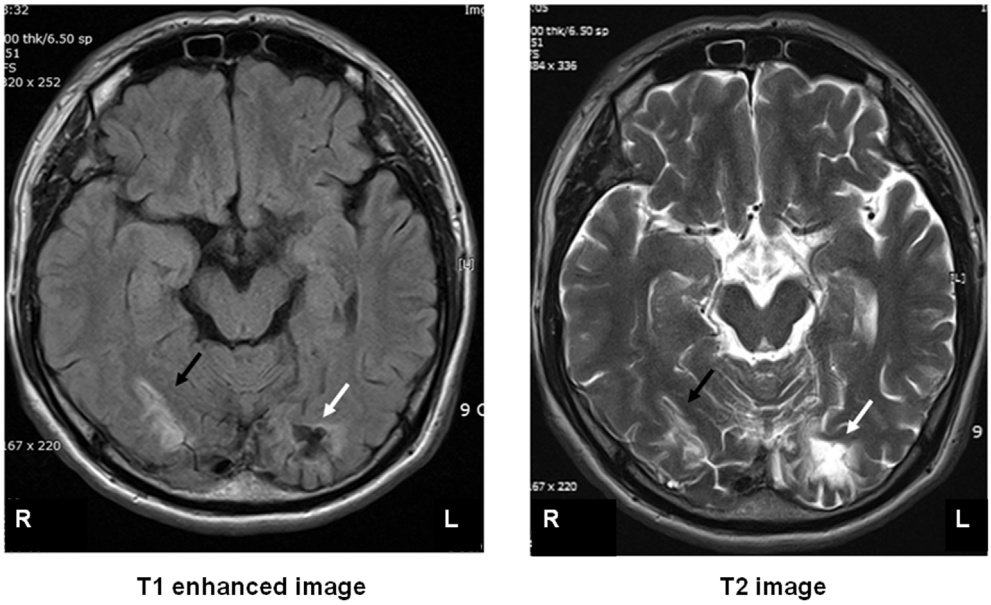

Fig. 2. Brain magnetic resonance imaging (MRI) revealed an old infarct in the left occipital lobe (white arrow) and subacute infarcts with cortical laminar necrosis in the right occipital lobe (black arrow) (Left: T1 enhanced image, Right: T2 image, R: right, L: left).

day, followed by subcutaneous enoxaparin sodium (low molecular weight heparin; LMWH) 60mg twice a day for five days. He was then switched to warfarin 6mg once a day to maintain a target international normalized ratio of 2.5–3.0. After 12 weeks of treatment, plasma anti-β-2 glycoprotein-I IgG remained elevated.

2. Discussion

In this case, antiphospholipid syndrome was diagnosed in a patient with homonymous visual field defects accompanied by multiple cerebral infarcts, in the absence of other cerebrovascular risk factors. Testing for antiphospholipid syndrome is recommended in patients

with autoimmune diseases, neuropsychiatric manifestations, ischemic cerebral events or multiple hyperintensity lesions on brain MRI without other risk factors, especially in those under 40 years of age.[1,2]

Homonymous hemianopsia respecting the vertical meridian indicates that the lesion involves the visual pathway posterior to the chiasm.[4] Our patient featured a right congruous superior quadrantanopsia in both eyes, suggesting a left temporal or occipital lesion isolated to the lower lingual gyrus. While stroke is the most common cause of homonymous hemianopsia in adults,[4] an extensive work-up of autoimmune diseases should be performed in relatively young patients with no cardiovascular risk factors.

Antiphospholipid syndrome can occur either as a primary condition or associated with an underlying systemic autoimmune disease, such as systemic lupus erythematosus. Stroke is the most common arterial thrombotic event in antiphospholipid syndrome, accounting for 22.9% of the initial manifestations of the disease.[5,6] Infarcts of various sizes and focal hyperintense lesions of white matter are the most common findings on brain MRI,[7] as in this case. Optic neuropathy, chronic headache, dementia, cognitive dysfunction, psychosis, transverse myelopathy, chorea and seizures have also been reported.[8]

The main treatment for antiphospholipid syndrome includes antithrombotic medications to prevent secondary thrombotic events.[9] Low molecular weight heparin (LMWH) is the first line treatment for acute thrombotic events, particularly in early pregnancy when warfarin is contraindicated.[9] The duration of treatment is uncertain, and most patients usually require lifelong therapy due to increased risk of recurrence.[10] However, for patients with a first thrombotic event, a known transient precipitating factor, and a low antibody titer, a treatment duration of 3–6 months is recommended.[10] In our patient, plasma anti-β-2 glycoprotein-I IgG levels remained high after 12 weeks of treatment, confirming the diagnosis of antiphospholipid syndrome and excluding other possible causes of transient, nonspecific increase in antiphospholipid antibodies. Unfortunately, lifelong therapy with warfarin is required to prevent secondary recurrences in this case.

This case illustrates the need for visual field testing in patients presenting with vague visual symptoms. The possibility of cerebral infarction may easily be overlooked if a patient presents with normal visual acuity and no other neurologic abnormality, as in our case. Notably, visual field testing revealed a homonymous superior quadrantanopsia respecting the vertical meridian, suggesting a lesion involving the left visual pathway. Prompt visual field testing provided reasonable suspicion of a neurologic brain lesion, which led to the diagnosis of antiphospholipid syndrome after brain imaging and appropriate laboratory tests. Although visual field defects associated with cerebral infarction in antiphospholipid syndrome may not be uncommon, the incidence of ocular signs and patterns of visual field defects have not been reported in detail thus far.[2,3] Other vaso-occlusive manifestations, such as amaurosis fugax and unilateral or bilateral optic neuropathy, have also been reported in antiphospholipid syndrome, which are some of the major causes of blindness in these patients.[2,3]

3. Conclusions

Primary antiphospholipid syndrome may present with isolated homonymous visual field defects. Visual field testing should always be performed in patients who complain of nonspecific visual disturbances. Antiphospholipid syndrome should be considered in the differential diagnosis of homonymous visual field defects associated with multiple brain infarcts. Prompt administration of anticoagulation therapy is important in such cases to prevent secondary thrombotic events.[10]

Patient consent

Written informed consent to publish the report was obtained from the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

None of the authors have financial disclosures.

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