Homonymous Hemianopia as the Initial Sign of Posterior Cortical Atrophy

Dear Editor,

Posterior cortical atrophy (PCA), also recognized as a visual variant of Alzheimer disease (AD), is a neurodegenerative syndrome characterized by a progressive decline of higher-order visual function and asymmetric atrophy in the temporal, parietal and occipital cortex [1]. Memory impairment is absent or mild in the early phase of PCA and patients often initially present to ophthalmologists due to diverse visual symptoms such as visual neglect, simultanagnosia or visual field defect [2].

A 57-year-old woman presented with a history of progressive visual disturbance over one year. The patient did not have a history of systemic or ophthalmic disease, brain trauma, or medication use. She reported persistent difficulty in perceiving objects, especially on the left side. General ophthalmologic examination demonstrated no abnormalities in visual acuity, intraocular pressure or on fundoscopy except intraocular lens in the right eye and mild cataract in the left eye. Ocular movement was normal with no evidence of ocular apraxia, optic ataxia or nystagmus. Humphrey automated visual testing revealed left homonymous hemianopia, which was more prominent in lower quadrant area (Fig. 1A). Confrontational visual field test revealed relatively preserved visual perception on the left side to a single visual stimulus; however, significant left-sided visual extinction to double simultaneous stimuli was identified. In neglect exams, rightward deviation was found on line bisection test, but the letter cancellation test showed no definite left-sided neglect. Simultanagnosia was suspected from the Boston cookie theft picture and Navon hierarchical letters. The Seoul Neuropsychological Screening Battery revealed a significant impairment in visuospatial function. Brain magnetic resonance imaging demonstrated asymmetric atrophy in the right temporo-parietal and occipital cortex compared to the left (Fig. 1B). Brain ¹⁸fluoro-

deoxy glucose-positron emission tomography demonstrated hypometabolism in the same cortical area (Fig. 1C). The final diagnosis of PCA was made based on the clinical and radiologic findings. Fourteen months after the diagnosis, neuropsychological examination was repeated. Obvious left-sided neglect was noted on the letter cancellation test and the Seoul Neuropsychological Screening Battery revealed significant cognitive dysfunction, including memory impairment, acalculia and agraphia.

In this report, we described a case of homonymous hemianopia as the initial sign of PCA, with subsequent progression of cognitive impairment. Brain magnetic resonance imaging showed more significant structural atrophy in the right occipito-parietal cortex and ¹⁸fluorodeoxy glucose-positron emission tomography also demonstrated a functional decline in the same cortical area.

AD is the most common etiology of PCA, and 5% of AD cases are categorized as PCA [1]. However, PCA can be related to non-AD pathology in rare cases, such as Parkinson disease, Lewy body dementia, among other neurological conditions [3]. Thus, many synonyms exist for PCA and recent definition has been extended to include progressive syndromes characterized by selective decline in visual processing skills and other functions served by parietal, occipital and occipito-temporal regions [3]. PCA usually affects people at an earlier age compared to typical AD and initial symptoms often include visual discomfort such as blurry vision or difficulty completing daily activities. As the disease progresses, all patients eventually develop higher-order visual dysfunction, ocular motor problems, and cognitive impairment.

PCA mostly originates in the parieto-occipital cortex and involvement of the retrochiasal visual pathway can lead to homonymous hemianopia. In addition, the right hemisphere is more severely affected than the left hemisphere in most patients with PCA [4], as demonstrated in our case. The occurrence of a homonymous visual field defect in PCA is relatively common and varies widely from 47.5% to 78% in the literature; meanwhile, homonymous hemianopic visual field defects in patients with PCA usually appears with higher-order visual impairment, such as visual neglect, hallucination, simultanagnosia, or achromapsia [4].

Fig. 1. Visual testing and radiologic findings of the patient. (A) Humphrey visual field showing a homonymous hemianopia. (B) Brain magnetic resonance imaging demonstrating asymmetric atrophy in the right temporo-parieto-occipital cortex. (C) Brain 18 F-fluorodeoxy glucose-positron emission tomography showing a relatively decreased metabolism (arrows) in the right temporo-parieto-occipital cortex.

In conclusion, as demonstrated in our case, visual symptoms can be the first and the dominant clinical manifestation of PCA [5] and ophthalmologists are often the first to assess these patients. Accordingly, ophthalmologists should be aware of the possibility of neurodegenerative disease in patients presenting with visual symptoms.

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Conflict of Interest

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

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