

SHORT COMMUNICATION

Analysis of progressive ophthalmic lesion in a patient with subacute sclerosing panencephalitis

M. ZAKO¹, T. KATAOKA¹, A. OHNO-JINNO¹, Y. INOUE¹, M. KONDO², M. IWAKI¹

¹Department of Ophthalmology, Aichi Medical University, Nagakute

²Department of Ophthalmology, Nagoya University, Nagoya, Aichi - Japan

PURPOSE. To evaluate the progressive lesions affecting the visual system in a patient with subacute sclerosing panencephalitis (SSPE).

METHODS. The authors observed a 15-year-old boy with SSPE. Since the diagnosis was made before the appearance of ocular manifestations, the authors recorded the progressive ocular lesions using various ophthalmic examinations.

RESULTS. The patient showed no ophthalmic abnormalities until he developed a left homonymous hemianopia with sudden bilateral disturbed visual acuity. Severe progressive macular lesions including a pigment epithelial window defect by fluorescein angiography, a marked decrease in foveal thickness by optical coherence tomography, and an extensive disorder mainly specific to cone cells in the central retina by electroretinography were demonstrated. Novel findings such as a transient relative afferent pupillary defect and an anterior uveitis were also observed.

CONCLUSIONS. Analyses over a long period of time showed progressive ophthalmic findings in a patient with SSPE. (*Eur J Ophthalmol* 2008; 18: 155-8)

KEY WORDS. Subacute sclerosing panencephalitis, Uveitis, Optical coherence tomography, Electroretinogram, Window defect, Relative afferent pupillary defect

Accepted: July 30, 2007

INTRODUCTION

Ocular manifestations occur in up to 50% of subacute sclerosing panencephalitis (SSPE) cases (1, 2). A variety of ocular manifestations, such as papilledema, papillitis, retinitis, chorioretinitis, optic nerve pallor, homonymous visual field defects, and cortical blindness, have been described (1, 2). Although these ophthalmic manifestations generally precede the neurologic signs (1-3), we commenced analyses before the appearance of ocular manifestations in our case. Novel findings such as a transient relative afferent pupillary defect and an anterior uveitis were observed, and their relations to SSPE are also discussed.

Case report

A 15-year-old boy presented with a diagnosis of SSPE, and showed no pathologic ocular findings in March 2001. In March 2002, he showed a sudden left relative afferent pupillary defect (RAPD), but his best-corrected visual acuity (VA) in both eyes was 1.0. The ophthalmoscopic appearances (Fig. 1A), a Humphrey field analyzer, and MRI of the brain showed no abnormal results. The RAPD was completely resolved by December 2002.

In February 2003, he presented with sudden blurred vision, and his best-corrected VA in both eyes was 0.1. There was serous detachment in the macular region in both eyes (Fig. 1A). He showed a left homonymous hemi-

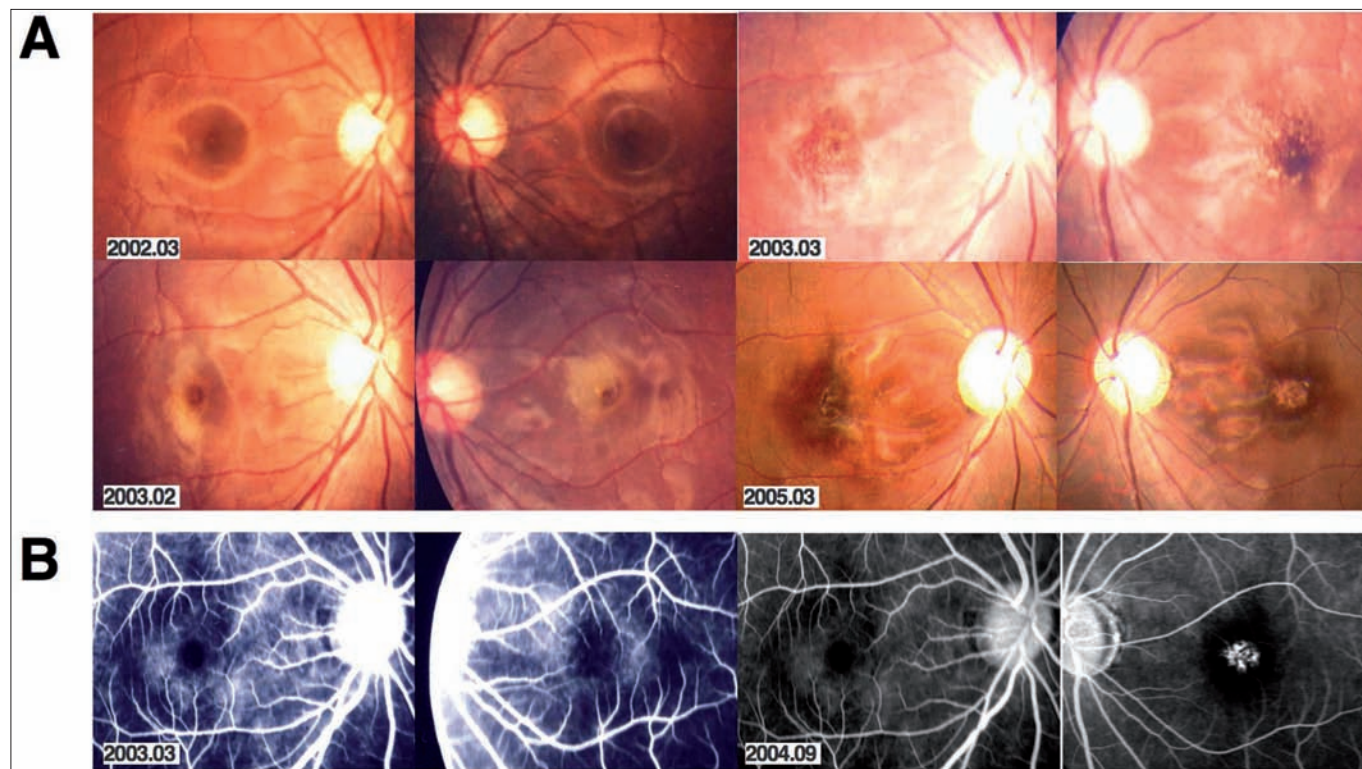


Fig. 1 - (A) Photographs showing the progressive macular lesions in both eyes. **(B)** Fluorescein angiography demonstrating a transient posterior uveitis in both eyes. Finally, a pigment epithelial window defect is present in the left eye.

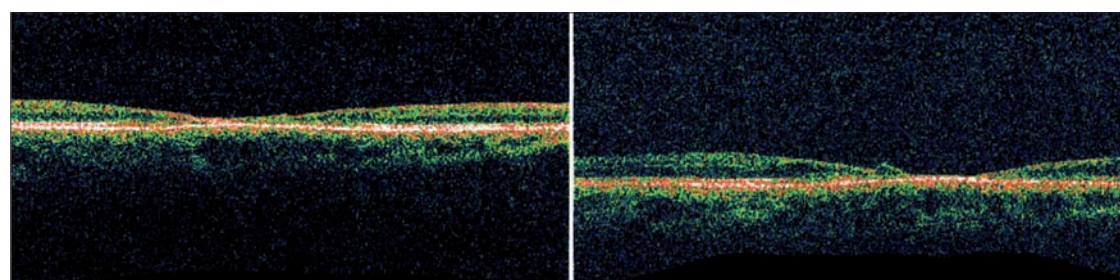


Fig. 2 - Horizontal sections of optical coherence tomography images of the macular lesions. A marked decrease in foveal thickness is present (left, right eye; right, left eye).

anopia, and MRI demonstrated abnormal intensities in the right temporo-occipital area. In March 2003, there was a pigmentation of the fovea circumscribed by irregular whitish lesions with retinal folds in both eyes (Fig. 1A), and fluorescein angiography revealed staining of the lesions (Fig. 1B). Anterior uveitis, including anterior chamber cells, flare, and keratic precipitates, was present, but it disappeared in April 2003. Subsequently, depigmentation of the fovea encircled by pigmented lesions and pallor of the optic discs were observed in both eyes (Fig. 1A). In January 2004, optical coherence tomography revealed marked decreases in the foveal thickness in both eyes

(Fig. 2). The thickness of the foveola was estimated to be 0.04 mm in the right eye and 0.00 mm in the left eye. We detected the myopic changes in our patient by -1.25 D for the right eye and -2.50 D for the left eye by refractometry after cycloplegia treatment. A pigment epithelial window defect in the left eye was revealed (Fig. 1B). Full-field and multifocal electroretinograms (ERGs) showed that although both the rod and cone systems were impaired, the cone was most depressed in the central retina (Fig. 3). ERGs showed the amplitude of rod response was 65% of normal value, and the amplitudes of the a- and b-waves elicited by a bright white stimulus were 55% and 61% of

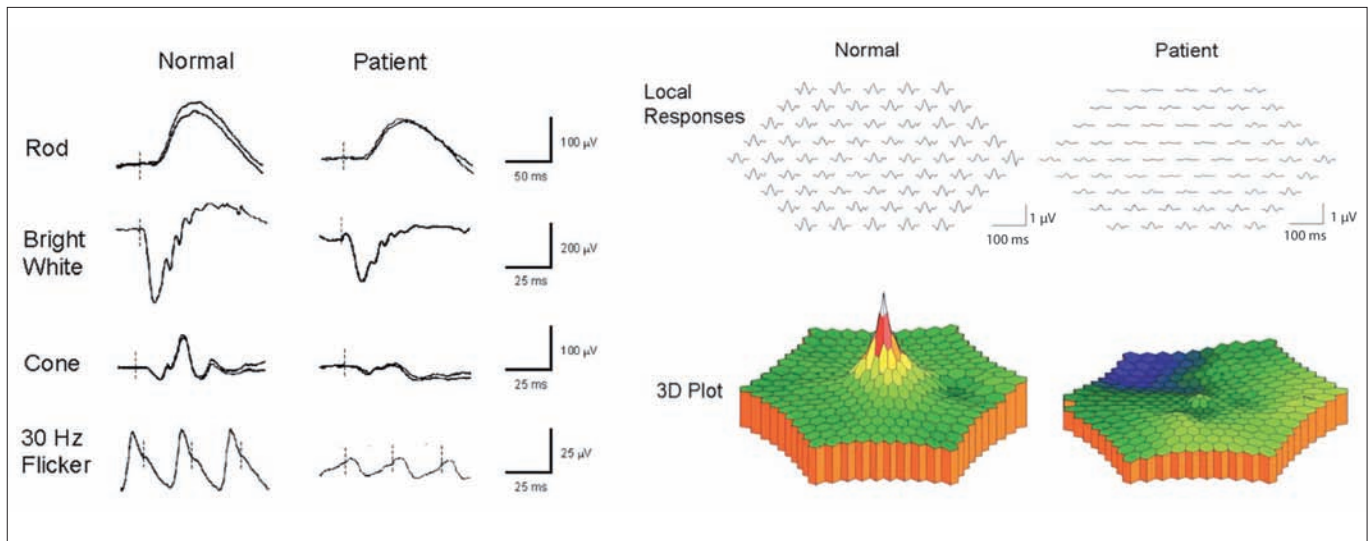


Fig. 3 - Electroretinograms showing the function of the cones is more deteriorated than that of the rods. The dotted lines represent the onset of the stimuli.

normal values, respectively. The single flash cone response and 30 Hz flicker response were reduced to about 25% and 28% of normal values, respectively. The amplitude of multifocal ERGs was relatively preserved in the peripheral retina but nearly undetectable in the central retina. No other significant ophthalmic changes have been recorded afterward.

DISCUSSION

Novel clinical findings, not only transient RAPD and anterior uveitis but also pathologic macular changes analyzed with fluorescein angiography, optical coherence tomography, and ERGs, were demonstrated to characterize the disease.

Previous reports showed RAPD in patients with SSPE (3-5). All these cases demonstrated monocular loss of visual acuity after severe irreversible manifestations, mainly on the disc and/or macula. The etiologic cause in our case was different, since it was transient without any lesions on the disc and/or macula. The RAPD improved just before the onset of visual loss and hemianopia in our case. We observed, however, obvious anterior uveitis just after the onset of visual loss and hemianopia. Although the etiology is unknown, the following hypothesis could be set up. The defective measles virus might have latently but exten-

sively infected central nervous system. Initially, the extent of the infection of his left optic nerve might have been more severe than of his right optic nerve, but finally both optic nerves were equally infected. Then the onset of the infection with an inflammation suddenly appeared on his eyes and brain.

The thinning of the retina and retinal pigment epithelium which resulted in elongation of the axial eye length might have been involved in the myopic change.

Proprietary interest: None.

Reprint requests to:
Masahiro Zako, MD
Department of Ophthalmology
Aichi Medical University
Nagakute
Aichi 480-1195, Japan
zako@aichi-med-u.ac.jp

REFERENCES

1. Hiatt RL, Grizzard HT, McNeer P, Jabbour JT. Ophthalmologic manifestations of subacute sclerosing panencephalitis (Dawson's encephalitis). *Trans Am Acad Ophthalmol Otolaryngol* 1971; 75: 344-50.
2. Green SH, Wirtschafter JD. Ophthalmoscopic findings in subacute sclerosing panencephalitis. *Br J Ophthalmol* 1973; 57: 780-7.
3. Caruso JM, Robbins-Tien D, Brown WD, Antony JH, Gascon GG. Atypical chorioretinitis as an early presentation of subacute sclerosing panencephalitis. *J Pediatr Ophthalmol Strabismus* 2000; 37: 119-22.
4. Salmon JF, Pan EL, Murray AD. Visual loss with dancing extremities and mental disturbances. *Surv Ophthalmol* 1991; 35: 299-306.
5. Park DW, Boldt HC, Massicotte SJ, et al. Subacute sclerosing panencephalitis manifesting as viral retinitis: clinical and histopathologic findings. *Am J Ophthalmol* 1997; 123: 533-42.