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Rapidly progressive subacute sclerosing panencephalitis presenting with acute loss of vision

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Abstract

A 10-year-old male presented with vision loss and behavioral changes. He had midpoint pupils with no reaction to light and normal funduscopic examination. Cranial magnetic resonance imaging revealed bilateral cortical lesions at parieto-occipital lobes. Elevated measles antibody titers in the cerebrospinal fluid confirmed the diagnosis of subacute sclerosing panencephalitis. Despite oral inosiplex and supportive care, patient developed generalized seizures with frequent myoclonic jerks and rapidly progressed into coma. Cortical blindness in subacute sclerosing panencephalitis can be an early indicator for fulminant course.

Key words: Subacute sclerosing panencephalitis; cortical blindness; fulminant course.

Introduction

Subacute sclerosing panencephalitis (SSPE) is a chronic encephalitis occuring after measles infection. The onset of the disease is usually insidious, and behavioral problems are prominent. Gradually, myoclonic seizures led to a final stage of akinetic mutism and death (1). Most cases follow a slowly progressive course, but about 10% of the cases progress more rapidly and the patients die within a few months (2). The patient reported here had a rare rapidly progressive form of SSPE, which is presented with acute loss of vision.

Case

A 10-year-old boy was admitted with acute vision loss and behavioral changes. There were no history of fever, headache, trauma and a recent infection. He had painless loss of vision in right eye followed by left eye in a couple of hours, 5 days prior to admission. He developed irritability and sleepiness after vision loss. He received measles vaccination at the age of 9 months but had measles at 16 months. There was no family history of vision problems and neurological disease. Prior to the event, he had been a student at 5th grade with good school performance.

On examination, he was irritable but cooperative and well oriented to time, place and person. He had midpoint pupils with no reaction to light. Funduscopic examination was normal. Neurological examination was normal except for the brisk tendon reflexes. Cranial MR imaging revealed bilateral cortical lesions at parieto-occipital lobes (Fig. 1 and 2). CSF cytology, glucose and protein levels were all normal. MR venography and blood pressure monitoring were not diagnostic.

Acyclovir was started for suspected herpes encephalitis. Despite the treatment, patient developed generalized seizures with frequent myoclonic jerks and progressed into coma. EEG showed high voltage periodic slow-wave discharges that were often associated with jerks.

Myoclonic jerks of the patient subsided dramatically with carbamazepine. Patient was diagnosed as subacute sclerosing panencephalitis (SSPE) with measles IgG titers by enzyme-linked immunosorbent assay at 1.88 IU/ml and 2.24 IU/ml (0.1-1.1 IU/ml) in CSF and in serum, respectively. CSF IgG index was 1.56 (< 0.7).

Discussion

Ocular findings are reported in nearly half of the SSPE patients. The most characteristic fundus finding is chorioretinitis but correlation between the clinical stage of SSPE and the chorioretinitis is unclear (3). It was hypothesized that inclusion bodies, that occur in the retina of SSPE patients with chorioretinitis are due to actual viral invasion. But

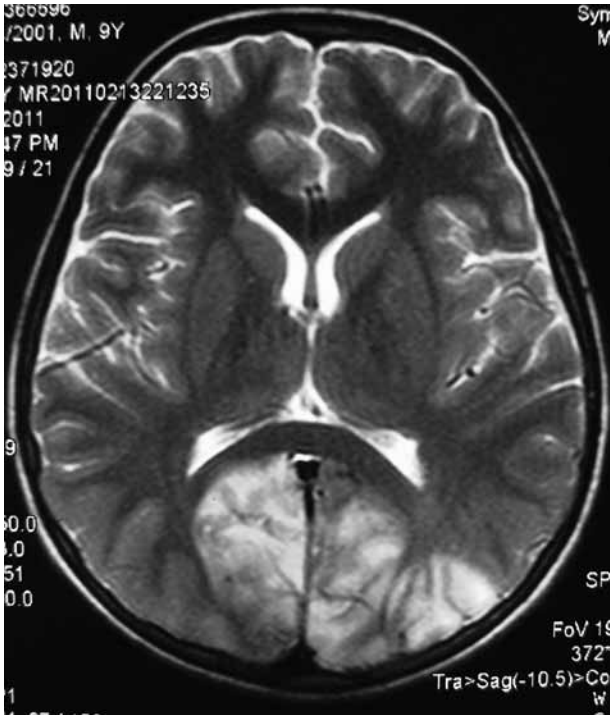

FIG. 1

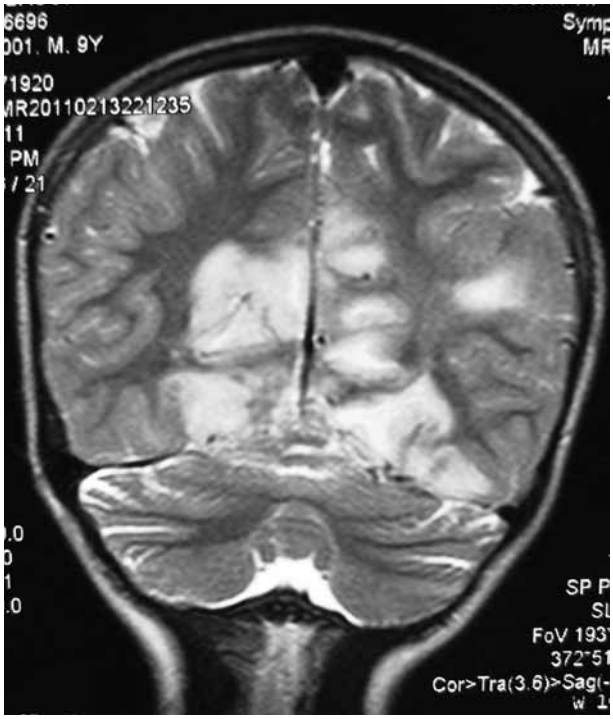

FIG. 2

chorioretinitis can also be the presenting sign of an acute form of SSPE (7).

While adult patients with an interval of several years between visual symptoms and the onset of other SSPE neurological signs were reported (4, 5, 6), cortical blindness presenting before classical neurological symptoms is rare in childhood SSPE. Up to our knowledge, previously four children with cortical blindness preceeding the disease were reported. The first patient was reported by Kabra *et al.* (8) with sudden loss of vision and normal funduscopic examination. Brain computed tomographic (CT) imaging was normal. The patient's central nervous system findings developed 4 months later. There was an interval of 7 months between the visual symptoms and the clinical deterioration in the 4 year old patient reported by Şenbil *et al.* (9). Cranial MR revealed bilaterally symmetric lesions in the optic radiations. Sharma *et al.* (10) reported the other patient similiar to our case with rapidly progressing encephalopathy within 15 days. Brain CT imaging revealed right occipital hypodensity causing visual symptoms as in our case. Another rapidly progressive SSPE patient presenting with blurred vision is reported by Takayama *et al.* (11), with high intensity areas in the lateral geniculate bodies on T2-weighted images, but the occipital region appeared normal. Subacute sclerosing panencephalitis causes visual symptoms due to different lesions along optic pathway.

It is known that a few patients with SSPE have an acute and rapidly fulminating course. Being younger at the onset of disease and measles ınfection prior to the immunisation are proposed as risk factors for a fulminating course (12, 13). It is also remarkable that, three of 5 patients presenting with cortical blindness had a fulminant course, which contrasts with the expected 10% overall in SSPE patients.

In conclusion, subacute sclerosing panencephalitis can cause lesions along to the optic pathways and visual symptoms can be the presenting sign of the disease. Cortical blindness can herald a rapidly progressing course of SSPE.

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