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Case Report

Subacute sclerosing panencephalitis presented with visual loss: Proton magnetic resonance spectroscopy and diffusion weighted MR imaging findings

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Abstract. We present brain magnetic resonance imaging, diffusion weighted imaging and proton magnetic resonance spectroscopy findings in a 13-year-old patient who presented with vision loss and behavioral changes after trauma. Elevated measles antibody titers in the plasma and cerebrospinal fluid confirmed the diagnosis of subacute sclerosing panencephalitis.

Keywords: Subacute sclerosing panencephalitis; magnetic resonance spectroscopy; diffusion magnetic resonance imaging; visual loss

1. Case Report

A 13-year-old boy with loss of vision was referred to our hospital. On admission, he had visual loss in his eyes (greater in the left than the right) that started 15 days ago. His family stated that he fell with his bike and had a mild head injury two months ago. They noticed mild behavioral changes after bicycle accident. Medical history revealed that he was born by normal vaginal delivery and had normal mental-motor development. Prior to admission he had no known disease and his vaccines were administered routinely. On his neurological examination, he was conscious but his speech was slow and sometimes meaningless. Pupillary responses to light in both eyes were weak. Bilateral pupils were isochoric. Cranial nerves and deep tendon reflexes were normal. On ophthalmologic examination, there was mild temporal pallor of the right optic disk and bilateral maculopathy with retinal pigment epithelial changes. In laboratory examination, his blood cell count, routine blood biochemistry and urine analyses were unremarkable. The first cerebrospinal fluid (CSF) examination showed no cells, and normal protein and glucose levels. Magnetic resonance (MR) imaging examination was requested;it was performed with a 1.5-T system (Achieva; Philips, The Netherlands). There were bilateral symmetrical hyperintensities in the white matter and cortex of the parietal and occipital lobes on axial T2-weighted images (Fig. 1a). On postcontrast T1-weighted image, minimal leptomeningeal enhancement was observed (Fig. 1b).

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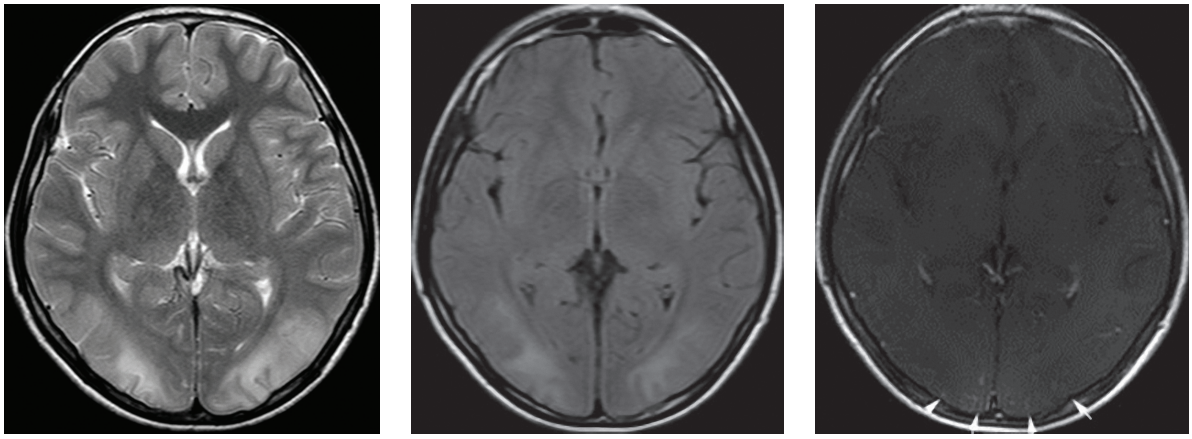
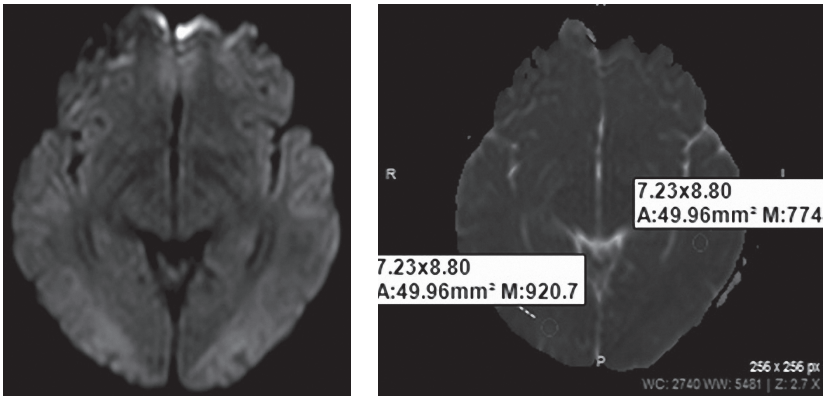


(a) (b) (c)

Fig. 1. T2 TSE (a) and T2 FLAIR (b) images show bilateral symmetric hyperintensities in the white matter and cortex of the parietal and occipital lobes. Contrast enhanced T1 weighted image (c) reveals minimal leptomeningeal enhancement (arrows).



(a) (b)

Fig. 2. Diffusion-weighted image (a) show high signal intensity at the lesion sites and on ADC map (b) the lesions had higher ADC values than normal appearing white matter.

The lesions were hyperintense on diffusion weighted imaging. On the apparent diffusion coefficient (ADC) map, the lesions had higher values (mean ADC value: 0.954×10^{-3} mm^2/sec) than normal appearing brain parenchyma (range between, 0.756×10^{-3} mm^2/sec and 0.824×10^{-3} mm^2/sec) (Fig. 2). Single voxel proton MR spectroscopy (PRESS, TR: 2000, TE: 40 ms) from the lesions revealed decreased N-acetylaspartate (NAA)/ creatine (Cr) and slightly increase myo-inositol (mI)/ Cr ratios. Choline (Cho)/ Cr ratio was normal. There were slight lipid and lactate peaks (Fig. 3). A diagnosis of subacute sclerosing panencephalitis (SSPE) was suspected from the clinical and ophthalmologic findings. Elevated anti-measles antibodies were found in the plasma and second CSF analysis. Three months later; the patient came back with status epilepticus.

2. Discussion

SSPE is a rare, slowly progressive fatal neurodegenerative and inflammatory disorder of the central nervous system that develops after measles infection, following an asymptomatic period of 6 to 8 yr [1]. It usually occurs in childhood and early adolescence. The diagnosis is based on clinical findings, typical EEG (electroencephalogram) results and increased titer of the antibodies in plasma and CSF. Clinically, the disease is characterized by

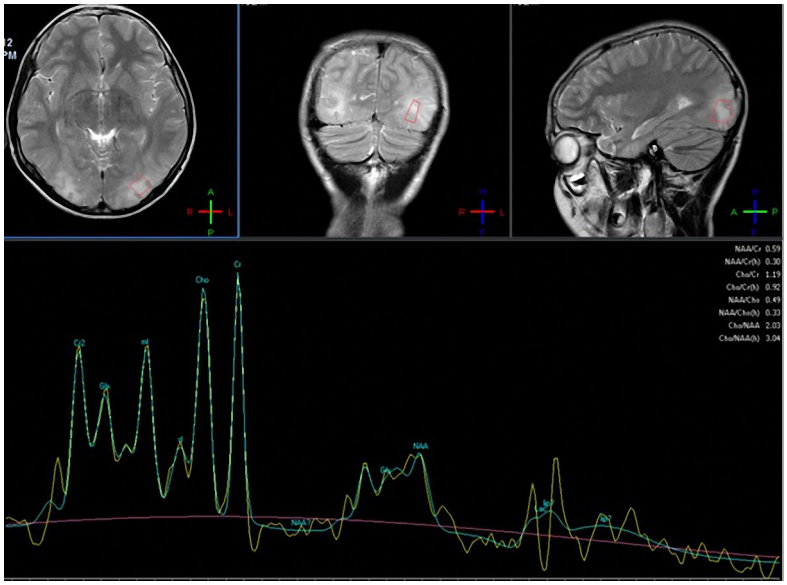


Fig. 3. Proton MR spectroscopy (single voxel, PRESS; 2000/40) shows a decreased NAA/Cr ratio and slightly increased mI/Cr ratios. There are also slight lactate and lipid peaks.

personality changes, mental deterioration, myoclonic seizures, and other focal neurological deficits [2]. Acute loss of vision was reported previously as a predictor of fulminant course [3]. Our patient had gradual visual loss with a more favorable short-term outcome. But this case, and a few other previously reported cases, indicate that visual loss might be the first manifestation of the disease.

Inflammation and edema in the early stages, and demyelination and gliosis in the late stages, are responsible for the imaging findings [4]. The parieto-occipital region is the most frequently involved region. The involvement of subcortical and deep white matter becomes most prominent as the disease progresses [5]. Brainstem and basal ganglia involvement can be observed in SSPE [6,7]. Increased diffusion, on diffusion weighted MR imaging, in the affected regions was reported [8]. Proton MR spectroscopy has proven to be very useful in the early diagnosis of SSPE. The NAA/ Cr ratio might be normal at the beginning of the disease but it becomes decreased in the later stages due to neuronal loss. mI/ Cr ratio was found to be increased in all patients with SSPE and becomes more prominent when the disease progresses. Cho/ Cr ratios might slightly increase in the early stages due to inflammation and demyelination. Lactate and lipid peaks might be seen because of cellular necrosis, and anaerobic metabolism [1,9,10]. Jabbour et al. [11] classified clinical SSPE in four stages (stage I, cerebral signs (mental, behavioral); stage II, convulsive motor signs (myoclonus, incoordination, choreoathetosis, and tremors); stage III, coma, opisthotonus, decerebrate rigidity, and no responsiveness to any stimulus; stage IV, mutism, loss of cerebral cortex function, less frequent myoclonus, and diminished hypertonia) according to clinical findings. Proton MR spectroscopy may have the potential to differentiate the stages of the disease [10].

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