Case Report

Atypical clinical course subacute sclerosing panencephalitis presenting as acute Encephalitis

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ABSTRACT

We report a 14-year-old boy who presented with loss of consciousness and gait instability. The electroencephalogram (EEG) showed generalized slowing with irregular activity and cerebral magnetic imaging revealed asymmetrical nonspecific signals on basal ganglia. His second electroencephalogram revealed periodical generalized high-voltage slow wave complexes which did not disappear with diazepam induction. Subacute sclerosing panencephalitis (SSPE) was considered and the diagnosis was confirmed with the identification of measles antibodies in cerebrospinal fluid. Our findings show that SSPE should be in mind in the differential diagnosis of meningoencephalitis and acute disseminated encephalomyelitis and highlight the significance of EEG in the diagnosis of unidentified cases.

Key words: Atypical clinical, child, encephalitis, SSPE

Introduction

Subacute sclerosing panencephalitis (SSPE) is a progressive inflammatory and degenerative disease of central nervous system caused by persistent infection of immune-resistant measles virus. The average period between exposure and onset of SSPE ranges between 7 and 12 years.[1,2] The typical clinical course of SSPE is characterized with intellectual deterioration, personality and behavioral changes, myoclonic jerks, and sometimes pyramidal and extrapyramidal symptoms. However, in about 10% of patients clinical manifestations of SSPE are not typical and that may cause a delay in the diagnosis and treatment of the disease.[2,3]

Here, we report a case of SSPE presenting with atypical clinical signs as loss of consciousness and gait instability. We report this patient in order to emphasize the importance of serial electroencephalogram (EEG) in SSPE diagnosis and to remind SSPE with atypical course.

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Case Report

A 14-year-old boy was referred to us with loss of consciousness and gait instability. There was fatigue and decreased consciousness for 2 days and loss of consciousness in the last 24 h. There was no fever and vomiting. He had measles at age 3 years. His developmental milestones were normal. His family history was noncontributory. Physical examination in the emergency room revealed normal vital signs. He was confused and responding to verbal stimulus with nonspecific sounds and withdrawing in response to pain. There was neck stiffness with positive Kernig's and Brudzinski's signs. Deep tendon reflexes were brisk and bilateral plantar reflexes were extensor.

Complete blood count and serum chemistries including lactate and ammonia were normal. Serologies for Epstein–Barr virus, cytomegalovirus, Herpes simplex virus, and mycoplasma were negative. The EEG showed irregular activity with generalized slow waves and cerebral magnetic resonance imaging (MRI) showed asymmetrical nonspecific

signals on basal ganglia [Figure 1]. Cerebrospinal fluid (CSF) examination showed no cells, and normal protein (35 mg/dL) and glucose (68 mg/dL).

Meningoencephalitis and acute disseminated encephalomyelitis (ADEM) were suspected with present clinical and laboratory findings and he was treated with intravenous acyclovir initially. Also, intravenous immunoglobulin (IVIG) 400 mg/kg/d for 5 days were administered because of the cerebral MRI findings which might be compatible with ADEM. Clinical findings partially recovered with IVIG therapy and his awareness has improved. Gait difficulty was ongoing, but there was no vision problem. His second EEG revealed periodical generalized high-voltage slow wave complexes discharges that did not disappear with diazepam induction so that SSPE was considered. The diagnosis of SSPE was suspected and CSF total and measles IgG index, and CSF oligoclonal bands were detected. Increased titers of measles antibody in serum (1/256) and CSF (1/8) (normal: 1/4 or lower) were found by complement fixation test. These results confirmed the diagnosis of SSPE, and treatment with isoprinosine (100 mg/kg/day) was administered. Our patient achieved remission with IVIG and isoprinosine treatments. He is in follow-up for 6 months without any abnormal clinical findings except minimal personality changes.

Discussion

SSPE is one of the most important complications of measles in children and adolescents. The characteristic initial clinical signs of SSPE are personality and behavioral changes and myoclonic jerks.[1] SSPE can also present with unusual features like generalized convulsions, loss of consciousness, acute-subacute coma, visual loss, ataxia, and hemiparesis. To date, in the literature many cases of SSPE have been reported with atypical features.[3-5] The patient described here manifested atypical clinical findings and initially the diagnosis of SSPE was not considered. Unlike previously reported cases with fulminant presentation, we could achieve remission in our patient with IVIG and isoprinosine.

Periodical high-amplitude slow wave complexes which cannot be suppressed with diazepam is a characteristic EEG sign of SSPE. Although periodical complexes are usually generalized and may be detected at the onset of the disease, they may rarely be focal and unilateral.[5] Ozyurek *et al*. reported that a 14-month-old girl was admitted with hemiparesis and focal convulsion, and they confirmed the SSPE diagnosis only after a long-term follow- up and periodical high-amplitude slow wave complexes detected on serial EEGs.[5] Similarly, our patient was admitted with atypical clinical presentation and EEG findings were normal at the onset but there were periodical generalized high-voltage slow wave complexes that cannot be suppressed with diazepam on 7th day EEG. So we suspected SSPE, and high serum and CSF measles antibody titers confirmed the diagnosis. That is why when a patient with a primary measles infection in history admitted with atypical clinical signs did not respond to treatment, and EEG should be evaluated carefully. Serial EEGs and lumbar puncture for CSF measles antibodies should be considered to rule out SSPE.

MRI is the best radiological imaging method for SSPE. Generally, MRI is normal at the onset of the disease. Cerebral atrophy and ventricular enlargement may also be seen on MRI, but none of these findings are specific for SSPE.[6,7] Brismar *et al*. reported that in some of the patients despite severe clinical course, MRI are normal. So that they concluded that there is no correlation between MRI and clinical course. The reported patients were managed as ADEM because of the periventricular white matter involvement in cerebral MRI.[6,7] Akdal *et al*. reported that basal ganglia involvement is not infrequent in SSPE.[8] On admission, cerebral MR T2A and FLAIR images of our patient showed nonspecific hyperintense asymmetrical lesions on basal ganglia.

There is no cure for SSPE. However, studies have shown that 30–35% of individuals benefit from various therapies. Isoprinosine is the first drug that was shown to be effective against SSPE. IVIG, alfa interferon, ribavirin, steroids, and plasmapheresis have also been reported to be effective in SSPE treatment.[1] These drugs cannot supply cure, they only slow the disease progression or prolong survival. Prashanth and Gurer reported that temporary remission could be rarely seen with isoprinosine and IVIG in patients with SSPE.[9,10] Our patient showed improvement with IVIG therapy and has significantly benefited from isoprinosine. He is in follow-up for 6 months without any abnormal clinical signs and symptoms

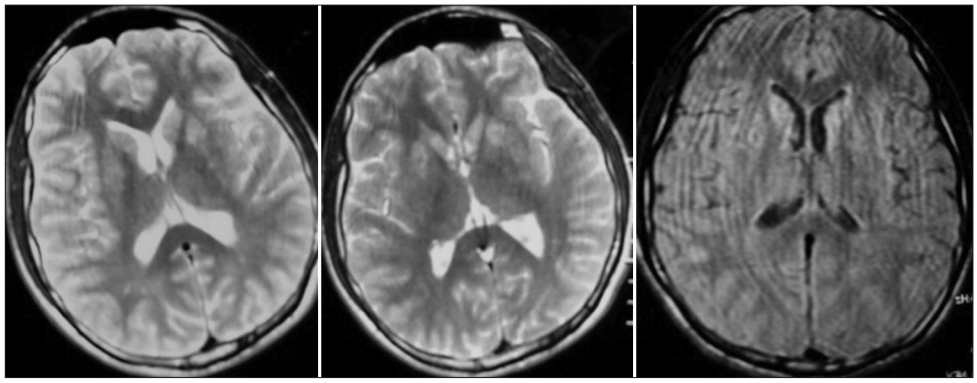

Figure 1: Axial T2-weighted and FLAIR MRI showed asymmetrical nonspecific signals on basal ganglia

except minimal personality changes. Also, we think that our patient is in temporary remission.

As a result, SSPE is still an important medical problem in the developing countries. So SSPE should be considered when a patient with measles history is admitted because of atypical clinical features like loss of consciousness, acute partial-generalized convulsion, acute-subacute coma, visual loss, ataxia, and hemiparesis. EEG of these patients should be evaluated carefully and serial EEGs with diazepam should be performed in SSPE-suspected cases. Despite these surveys if SSPE cannot be ruled out, serum and CSF measles antibodies should be examined.

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