Short Communication

Subacute Sclerosing Panencephalitis with Generalized Seizure as a First Symptom: A Case Report

Deniz Tuncel*, Ayda Ertugrul Ozbek¹, Gülenc Demirpolat² and Hamza Karabiber³

Department of Neurology, ¹Department of Pediatrics and ²Department of Radiology, Kahramanmaras Sütçü Imam University, Faculty of Medicine, Kahramanmaras, Turkey

(Received March 24, 2006. Accepted June 16, 2006)

SUMMARY: Subacute sclerosing panencephalitis (SSPE) can show variations in its clinical course. Typical initial symptoms consist of intellectual decline, deterioration in school performance, and myoclonic jerks. Atypical forms of presentation such as generalized seizures and acute or subacute coma can be observed rarely. This report describes a 5-year-old boy with an atypical feature of SSPE, generalized seizures. For 1 month his only symptom was seizures, followed by personality and behavioral changes and myoclonic jerks. A diagnosis of SSPE was made based on the boy’s history of measles, characteristic electroencephalogram changes and compatible magnetic resonance, and elevated anti-measles antibody titers in the cerebrospinal fluid and serum. The case presented in this article is a good example of SSPE in which, at early stages, some of the signs and symptoms can lead to an erroneous diagnosis.

Subacute sclerosing panencephalitis (SSPE) is chronic encephalitis of uncertain pathogenesis, caused by persistent measles virus infection of the central nervous system. The incidence of SSPE is 1.6 in 100,000 measles cases. Most SSPE patients have a history of natural measles infection before 2 years of age (1). Although there are cases reported as young as 4 months and as old as 52 years, the first clinical symptoms and signs attributable to SSPE are usually observed in children and young adults 6 to 15 years of age after acute measles infection. However, in immunized populations where the age at which measles is contracted increases, SSPE can be expected to start in later adulthood (2,3).

This report describes a boy with atypical initial symptoms consisting of generalized seizures. The presence of atypical early symptoms can lead to an erroneous diagnosis other than SSPE.

A 5-year-old boy was admitted to our department because of behavioral changes, myoclonic jerks of the head, and agnosia for a week. His parents stated that he had experienced generalized seizures without high fever a month earlier and been admitted to a local hospital. The boy was diagnosed with epilepsy and started carbamazepine; he had two more attacks of seizures during antiepileptic therapy. He was diagnosed with epilepsy and started carbamazepine; he had two more attacks of seizures during antiepileptic therapy. He was diagnosed with epilepsy and started carbamazepine; he had two more attacks of seizures during antiepileptic therapy. He was the fourth child of healthy and nonconsanguineous parents. His prenatal and natal history as well as developmental milestones were normal. He had suffered from measles, which consisted of a rash on the body and face, at the age of 2 years and improved gradually over a week without sequelae. He had not been immunized with measles vaccine. On examination he was irritable, and stereotypic movements and myoclonic jerks of the head could be seen. Deep tendon reflexes were hyperactive. Mental status examination was highly abnormal, with lack of visual and emotional contact, confusion, and disinhibition with unmotivated laughter. The patient did not cooperate while undergoing other neurological examinations.

Investigations revealed hemoglobin of 12.7 g/dl, total leukocyte 7,000/mm³ with 68% polymorphs, an erythrocyte sedimentation rate (ESR) of 30 mm/h, and normal urine and stool examination. Blood tests for toxoplasma, cytomegalovirus, rubella, and herpes simplex virus IgG and IgM were negative. Cerebrospinal fluid (CSF) was grossly clear with normal pressure and without cell proliferation protein 28 mg/dl, and sugar 42 mg/dl against blood sugar of 93 mg/dl.

Electroencephalogram (EEG) examination revealed characteristic abnormality consisting of periodic (every 15 to 20 min) bursts of 3- to 5-per-second high-voltage waves. After intravenous (i.v.) diazepam administration to the patient, the periodic bursts on EEG were not suppressed (Figure 1). Magnetic resonance imaging (MRI) showed high signal intensity on fluid-attenuated inversion recovery (FLAIR) images symmetrically in the periventricular white matter and left hippocampus (Figure 2).

In view of the EEG changes, MRI findings, and past history of measles, an estimation of antimeasles antibody titers was ordered. Specific measles antibody titers of serum and CSF were 1:10,240 hemagglutinin (HI) and 1:320 HI, respectively.

Diagnosis of SSPE stage 2a was suggested by the clinical presentation and confirmed by typical CSF and EEG findings. Oral isoprinosine and intraventricular interferon-α treatment were started. We have maintained contact with the family during 2 months of follow-up.

Measles is a highly contagious viral infection that kills more children than any other vaccine-preventable disease. Measles and its complications are severe problems in Turkey, with thousands of measles cases observed in our country every year due to immunization failure. Between 15,000 and 30,000 measles cases have been reported annually since the 1990s (4). According to the data of the Turkish SSPE Registry Center, from 1975 to 1999, the mean age of measles onset fell from 29 months to 20 months and the mean age of SSPE onset also fell from 13 years to 7.6 years (5). In contrast, in developed countries, the total incidences of measles and SSPE have declined significantly since the advent of the measles immunization, with the mean age of SSPE onset increasing from ~10 years to approximately 14 years (2). These find-
ings indicate that immunization against measles has still not reached the desired level (nationally 84%) (4). Measles immunization is performed in the 10th month of age as part of the routine immunization program in our country. The World Health Organization (WHO) and the United Nations Children’s Fund (UNICEF) have recommended that, in addition to achieving high coverage with the first dose of measles vaccine, all children should be offered a second opportunity for the measles vaccine to maximize both individual and population immunity (6).

SSPE is one of the most important complications of measles in children and young adults. The five diagnostic criteria of SSPE include clinical presentation, a characteristic EEG, abnormal CSF findings, high measles antibody titers in serum and CSF, and positive findings on brain biopsy. A diagnosis of SSPE can be reliably established if the patient fulfills three of the five criteria (7). The typical initial symptoms of SSPE usually involve regressive changes in intellect and personality. Myoclonic jerks usually follow the mental disturbances. Atypical forms of presentation such as generalized seizures, acute or subacute coma, onset at a very young age or older age, and visual loss as the initial presentation can be observed rarely (1,8,9). Our patient’s initial clinical course reflected this atypical presentation.

The CSF in SSPE will typically have normal cellular components, glucose, and total protein, but markedly elevated values of gammaglobulin and anti-measles antibodies (1). A high titer is sufficient to establish the diagnosis. Although serum antibodies are also increased, they are not diagnostic in the absence of CSF antibodies (1). Bilateral synchronous periodical complexes on the EEG consisting of bi-, tri-, or polyphasic high-amplitude waves (200-600 μV), sometimes followed by suppression periods, are typically seen in patients with SSPE (1). Although similar EEG findings can be observed in degenerative CNS diseases, severe encephalitis, or encephalopathy, the discharges in SSPE have a distinguishing feature: they are not suppressed by intravenous diazepam given during EEG recording. All of these characteristic immunologic and electrophysiological abnormalities were observed in our patient.

MRI is more helpful than computed tomography in the diagnosis of SSPE. Because MRI came into clinical use after SSPE became rare in industrialized countries as a result of widespread measles vaccination, few reports have been published on MRI findings in SSPE (10,11). A fairly typical order of involvement is observed as SSPE progresses: the cortex, subcortical white matter, and periventricular white matter sequentially show high signal intensity lesions on T2-weighted images (1).

According to the data, combined therapy with inosine pranobex and intraventricular interferon-α appears to be the most effective regimen at present: remission, improvement, or stabilization are reported in 44-55% of cases (12,13), and this therapy is the most efficient in cases of slowly progressing SSPE (1). With the consultation of pediatric neurology our patient has been receiving combined therapy for 2 months, and he is still under supervision.

In conclusion, SSPE is a rare and fatal complication of a common and preventable viral infection. The presented case had an atypical initial symptom of this rare illness, which led to an erroneous diagnosis of epilepsy. Such erroneous diagnoses can occur with atypical initial presentations of the disease. The diagnosis of SSPE, although relatively easy with the use of serological and laboratory techniques, still depends on clinical suspicion, and there is currently no 100% effective treatment for SSPE. The implementation of a two-dose measles vaccination program in the childhood period is the most safe, effective, and inexpensive method to increase population immunity and therefore prevent measles infection and its subsequent complications such as SSPE in developing countries like Turkey where measles is still a severe problem.

REFERENCES