



Subacute Sclerosing Panencephalitis Presenting with Hemidystonia

Hemidistoni ile Prezente Olan Subakut Sklerozan Panensefalit

Hepsen Mine Serin, Sevcan Bilen*, Ali Cansu**

Firat University Faculty of Medicine, Department of Pediatric Neurology, Elazığ, Turkey

*Karadeniz Technical University Faculty of Medicine, Department of Pediatric, Trabzon, Turkey

**Karadeniz Technical University Faculty of Medicine, Department of Pediatric Neurology, Trabzon, Turkey

Abstract

In this paper, we present a case of subacute sclerosing panencephalitis (SSPE) in an 11-year-old boy who presented with hemidystonia. Electroencephalogram (EEG) revealed periodic epileptiform discharges which did not disappear with diazepam induction. His cranial magnetic resonance imaging was normal. SSPE diagnosis was considered and it was confirmed with the identification of measles antibodies in cerebrospinal fluid. SSPE is a progressive disease. Hemidystonia is not an expected presentation of SSPE. We aimed to emphasize that SSPE may present with different clinical findings such as hemidystonia. (*The Medical Bulletin of Haseki 2014; 52: 137-9*)

Key Words: Subacute sclerosing panencephalitis, hemidystonia, measles

Özet

On bir yaşında hemidistoni ile prezente olan erkek olguyu sunduk. Elektroensefalografisinde diazepam ile kaybolmayan periyodik epileptiform deşarjlar izlendi. Hastanın kranial magnetik görüntülemesi normal olarak değerlendirildi. Hastada subakut sklerozan panensefalit (SSPE) düşünöldü ve tanı beyin omurilik sıvısında kızamık antikorü saptanması ile doğrulandı. Subakut sklerozan panensefalit tipik klinik sunumu ile ilerleyici bir hastalıktır. Hemidistoni SSPE için beklenen bir prezentasyon deęildir ve biz SSPE'nin hemidistoni gibi farklı klinik bulgularla karřımıza çıkabileceęini vurgulamak istedik. (*Haseki Tıp Bülteni 2014; 52: 137-9*)

Anahtar Sözcükler: Subakut sklerozan panensefalit, hemidistoni, kızamık

Introduction

Subacute sclerosing panencephalitis (SSPE) is a slow virus infection, a consequence of persistent measles virus infection of brain cells (1). This progressive and degenerative disease is associated with mutations affecting the measles virus with matrix protein and occurs 2-10 years later than the primary infection (2-4). The average age of onset is 5-15 years and boys are affected two times more frequently than girls (5).

Dystonia is a complex neurological syndrome which causes abnormal posture, iterative movements and distortion-curling with prolonged muscle contractions (6). Secondary dystonia may be a consequence of infectious or non-infectious encephalitis (6).

Typical clinical findings of SSPE include behavior abnormalities, cognitive regression, myoclonic jerks and seizures. Beside the clinical findings, SSPE might present with visual loss, increased intracranial pressure, epileptic seizures and focal neurological deficits (5).

With this particular case, we aimed to draw attention to uncommon clinical manifestations of SSPE such as hemidystonia.

Case

An 11-year-old boy has attended to our hospital with the complaints of bending of the right side of the body and curling of the right leg, once occasionally for a week, then frequently in the past two days. In addition, the patient has been treated with clonazepam in another

Address for Correspondence/Yazışma Adresi: Hepsen Mine Serin
Firat University Faculty of Medicine, Department of Pediatric Neurology, Elazığ, Turkey
Phone: +90 424 233 35 55 E-mail: hepsenmine@yahoo.com

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clinic for shaking in the right hand and limping in the right leg for two months, and after that, these complaints were dissolved.

A thorough history and complete physical and neurological examination were performed. Mental status examination was normal; he was alert, fully-oriented and cooperated.

The vital signs, ophthalmologic examination and pupillary light responses were normal. Cranial nerve examination was normal, deep tendon reflexes were bilaterally brisk, muscle power examination indicated 3/5 for the right upper and lower extremities, and 5/5 for left. The patient was hospitalized with the initial diagnosis of dystonia.

Periodic epileptiform discharges in his electroencephalogram (EEG) indicated SSPE (Figure 1, 2). The second EEG revealed periodical generalized high-voltage slow wave complexes which did not disappear with diazepam induction. The diagnosis was verified by increased plasma measles immunoglobulin G (IgG) levels

(5.13 T.V.) (>0.7 positive) whereas immunoglobulin M (IgM) was negative; increased cerebrospinal fluid (CSF) IgG levels (>100 U/ml) (<25 negative) whilst IgM was negative; and IgG index: 5.58 (0.2-0.6) and oligoclonal IgG positivity. Cranial and vertebral MR images were normal.

After verification of the diagnosis of SSPE, isoprinosine and β -interferon treatments were started and hemidystonia regressed within two days of the treatment. Nine months after beginning of the treatment, the patient was admitted to the hospital with speech impairment and gait instability. EEG revealed periodical generalized epileptiform discharges. Cranial MRI showed increased T2 signal intensity bilaterally in the basal ganglia and parieto-occipital region which was especially predominant in the left one. These findings were compatible with SSPE.

Discussion

SSPE is a fatal and rare disease observed in 1/100.000 of patients with measles infection. Although usual age of onset is between 5 and 15 years, it might present itself from 6 months to 30 years of age (7).

Measles infection in children younger than 1 year of age poses a risk for SSPE 16 times greater than in those aged 5 years or older (8). SSPE can occur an average 7 years post-measles infection (7). However, although rarely, it might occur without a previous measles infection. Our patient also did not have a history of measles infection.

The diagnosis of SSPE is based on typical clinical findings, EEG findings, an elevated anti-measles antibody (IgG) in the serum and CSF (9). There are four stages of the disease: firstly, mild intellectual deterioration and behavioral changes, secondly, different kinds of involuntary movements, thirdly, hypertonia in pyramidal and extrapyramidal muscles and lastly, chronic vegetative state and death (1).

Electroencephalogram may be normal in the early stages of the disease or nonspecific generalized or focal slowing can be observed (5). Typical EEG findings of SSPE include periodic complexes consisting of synchronized, high voltage biphasic waves (5). Our patient had also periodic generalized epileptiform discharges.

Neuroimaging studies are not necessary for the diagnosis of SSPE but it might be helpful. In fact, the neuroimaging studies are generally normal in the early stages of the disease. Neuroimaging is not necessary for diagnosis and do not provide prognostic information. However, it may demonstrate extent of the disease and provide differential diagnosis (10-12). In MRI findings of SSPE, there may be symmetrical, focal areas with increased signal intensity on T2 and hypo-isointense



Figure 1 and 2. Patient's EEG showed periodic generalized epileptiform discharges

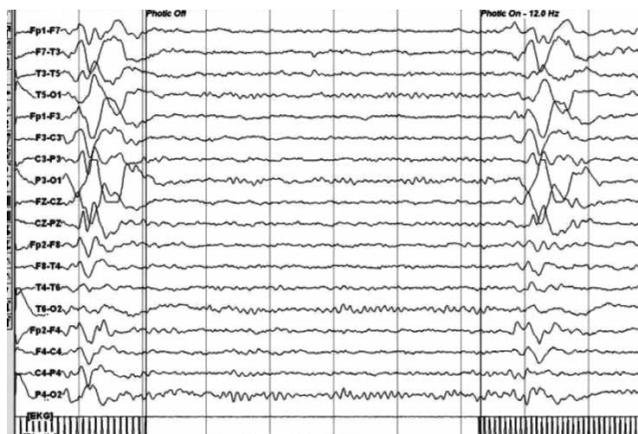


Figure 1 and 2. Patient's EEG showed periodic generalized epileptiform discharges

regions on T1-weighted images. These changes are usually located in the cerebral cortex, subcortical and periventricular white matter and rarely in the corpus callosum, basal ganglia, thalamus, and more rarely, in the brain stem (10-12). In the late phase, cortical atrophy might be seen (12). MRI of our patient was firstly normal and in the follow-up period, MRI T2-weighted images showed increased signal intensity in the bilateral parieto-occipital region and basal ganglia.

The beginning of the disease is usually indistinct. Firstly, behavioral changes and a decrease in scholastic performance occur and myoclonic jerks and seizures might develop (5). There have been several reports of atypical presentation of SSPE. Demir et al. have reported patients with SSPE presented with ataxia, encephalopathy and hemiplegia (13). Ayçiçek et al. have reported five patients with SSPE presenting with pseudotumor cerebri (14). Similarly, Ondo and Verma have identified a 26-year-old young adult patient with SSPE presented with paroxysmal dystonia (15). A previous case report have included a 14-year-old boy with unconsciousness and gait instability who had also encephalitis and was diagnosed with SSPE by serial EEGs (16). The importance of EEG was underlined in this case report. Özyürek et al. have reported a 14-month-old girl with hemiparesis and focal seizures who has been diagnosed with SSPE during long-term follow-up (17). Goraya et al. have presented another case of SSPE in a 9-year-old girl presented with poor balance and ataxia following upper respiratory system infection (18). These case reports have showed that presentation of SSPE might be completely different than expected. In this paper, we reported an atypical initial presentation of SSPE.

We aimed to point out that SSPE may present with different clinical manifestations including hemidystonia.

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