OLGU SUNUMU

A rare case of rapidly progressive subacute sclerosing panencephalitis with atypical radiological involvement

Atipik radyolojik tutulumla giden hızlı ilerleyişli nadir bir subakut sklerozan panensefalit olgusu

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ÖZET

Subakut sklerozan panensefalit (SSPE), kızamık infeksiyonundan yıllar sonra ortaya çıkan, mental kötüleşme, davranış değişiklikleri, miyoklonus ve nörolojik yıkımla seyreden bir hastalıktır. Biz akut konfüzyon ile başlayıp sıradışı radyolojik özellikler gösteren atipik bir SSPE olgusunu sunmayı amaçladık. Öncesinde sağlıklı olan 10 yaşında bir kız çocuğu bir haftadır devam eden baş ağrısı ve sürekli uyku hali ile başvurdu. Dokuz aylıkken kızamık enfeksiyonu öyküsü mevcuttu. Letarjik olup plantar refleks yanıtları bilateral ekstansördü. Sistemik muayene ve rutin biyokimyasal değerlendirmesi normaldi. Beyin manyetik rezonans görüntülemesinde (MRG) beyin sapından serebellar beyaz cevhere uzanan hiperintens lezyonlar görüldü. Rutin beyin omurilik sıvı incelemesi normal olup oligoklonal bantı pozitif, IgG indeksi> 0,7 idi. BOS kızamık antikor titreleri kuvvetli pozitif idi. Elektroensefalografisinde organizasyon bozukluğu ve jeneralize yavaş dalgaları görüldü. Intravenöz (IV) metilprednizolon 5 gün süre ile verildi. Anlamlı bir klinik yanıt alınamadığından IV immunoglobulin tedavisine geçildi. 2.haftada çekilen beyin MRG kontrolünde önceki lezyonlarında artış gözlendi. 4. haftada hasta ani kardiyak arrest nedeniyle kaybedildi. Sonuç: Akut fulminan seyir, beyin sapı ve serebellum tutuluşu SSPE için nadirdir. Baş ağrısı ve akut bilinç değişikliği ile başvuran, kranial görüntülemelerinde atipik bulgular saptanan olgularda subakut sklerozan panensefalit akılda tutulmalıdır.

Anahtar kelimeler:subakut sklerozan panensefalit, fulminan gidiş, akut disemine ensefalomiyelit

ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a progressive disease characterized by mental-neurological deterioration and myoclonus, occurring after years of measles infection. Herein, we report an atypical SSPE case presented with acute confusion and headache which showed atypical radiological features. Case: A 10-year-old previously healthy girl was admitted with headache and constant sleepiness for a week. She had a history of measles infection at the age of nine months. She was lethargic and plantar reflexes were bilaterally indifferent. Systemic examination and routine biochemical evaluation were in normal limits. Brain magnetic resonance imaging (MRI) showed hyperintense lesions extending from brain stem to cerebellar white matter. Routine cerebrospinal fluid (CSF) examination was in normal limits with positive oligoclonal band and IgG index>0,7. CSF measles antibody titers were strongly positive. Her electroencephalography revealed disorganized back ground and generalized slow waves. Intravenous (IV) metilprednizolone was given for 5 days. Because of no significant clinical response, the treatment was switched to IV immunoglobuline. At the second week, following MRI showed extention of previous lesions. On the 4th week of presentation, myoclonus began, the patient developed sudden cardiac arrest and died. Conclusion: Acute fulminant course and involvement of brainstem and cerebellum is rare in SSPE. Differential diagnosis may be difficult from other acute confusional states. This case report seeks to draw attention to the neccessity of keeping the subacute sclerosing panencephalitis in mind while considering the differential diagnosis in patients with headache and acute altered mental state with atypical findings on the brain imaging

Keywords: subacute sclerosing panencephalitis, fulminant course, acute disseminated encephalomyelitis

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is an inflammatory neurodegenerative disease occurring years after measles infection characterized by mental deterioration, behavioral changes, myoclonus and severe neurological destruction. The disease is a fatal slow virus infection of the central nervous system and a mutated wild measles virus is responsible for its ethiopathogenesis (1). SSPE is a rare complication of measles infection and the incidence is about 4-11/105. The risk for developing SSPE significantly increases in children encountering measles virus before the age of one (2). The mean latent period between measles infection and SSPE disease is about 6-8 years. Many patients die within 1-3 years from the onset of symptoms (3,4). There is no definitive treatment of the disease is present yet.

The disease begins clinically with mental impairment and personality changes However, the disease is known to occur with or atypical symptoms or findings (5,6). Patients with SSPE may present with different clinical pictures such as confusion, loss of vision, seizures, hemiparesis, focal neurologic deficits (7).

The diagnosis of the disease is established with characteristic clinical features, abnormal electroencephalography findings and the presence of high titer of measles antibody in cerebrospinal fluid (CSF). Typical electroencephalography (EEG) findings for SSPE include high-amplitude, biphasic, periodic and synchronized wave discharges. The definitive diagnosis is made by the presence of high titer measles IgG in CSF. Cranial MR scans are usually normal in the early period, however, in the later stages of the disease on T2-weighted images, hyperintense signal increases can be seen in the cerebral cortex, periventricular white matter, basal ganglia and brain stem (8).

The clinical course varies in SSPE cases. While 49% of the patients show subacute form, the other forms are: 22.5% chronic, 17.5% acute, 10% fulminant course, respectively (9). Cases with acute and fulminant course die in the first six months. SSPE cases with fulminant course have been reported more frequently in recent years (10,11,12).

We present our SSPE case which started acutely, and showed atypical cranial MR findings and had fulminant course which was different from the well-known features.

CASE REPORT

10-year-old girl was brought to our emergency department with a history of headache, and constant sleepiness for one week. She had no health problems before presentation. Her prenatal and birth history was normal. We learned that she had had an illness characterized by fever, wide-spread rashes across her body at the age of nine months which was diagnosed as measles.

No definite information as to whether the routine vaccines were performed could not be obtained. On examination, the general condition of the the patient was poor; she was lethargic and she opened her eyes with painful stimulus; pupillas were bilaterally isochoric and reactive to light, plantar reflexes were bilaterally indifferent. Systemic examination was not remarkable and the fundus was normal. Routine blood tests were also normal. On the cranial MR, hyperintense lesions were noted in two-sided cerebellar peduncles, brain stem and to a lesser extent in cerebellar white matter and sections in the supratentorial region in T2 and FLAIR sections (Figure 1A-B). There was no contrast uptake of the lesions.

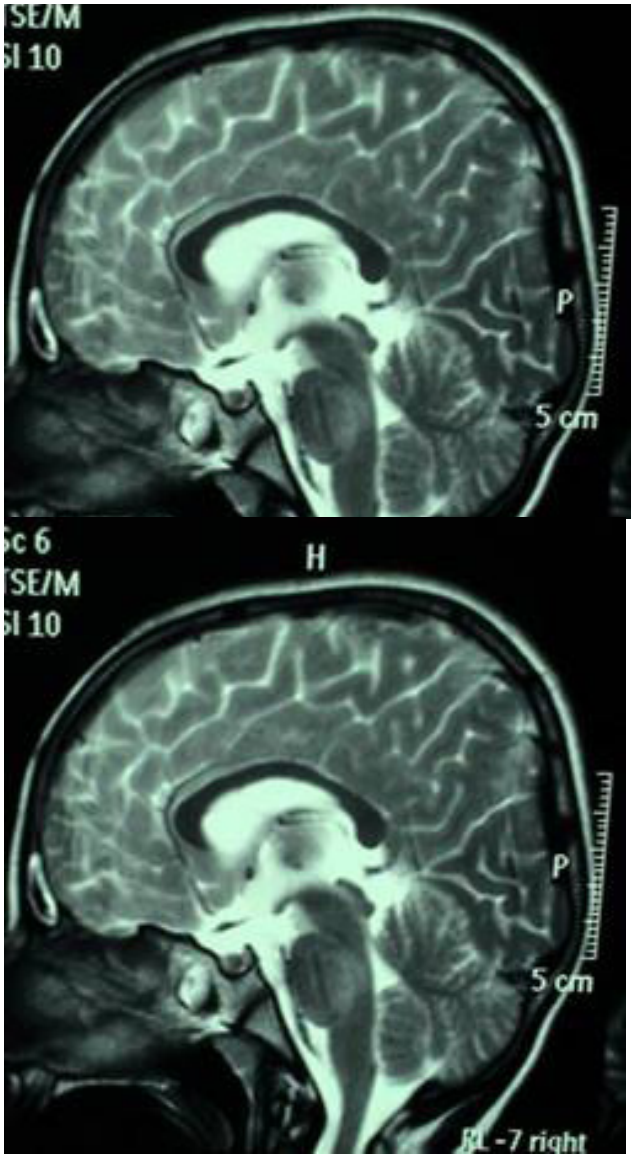

Figure 1. A. Hyperintensity in the pons and B. cerebellar peduncle on T2 sagittal section of the first MRI.

In the differential diagnosis, of encephalitis and acute disseminated encephalomyelitis (ADEM) were considered. CSF was clear in appearance with normal pressure, no cells were present. In CSF, there was 41mg/dl of protein, 57mg/dl of sugar (simultaneous blood sugar was 107mg/dl), positive oligoclonal bands, IgG index > 0.7. EEG showed disorganization and generalized slow waves (Figure-2).

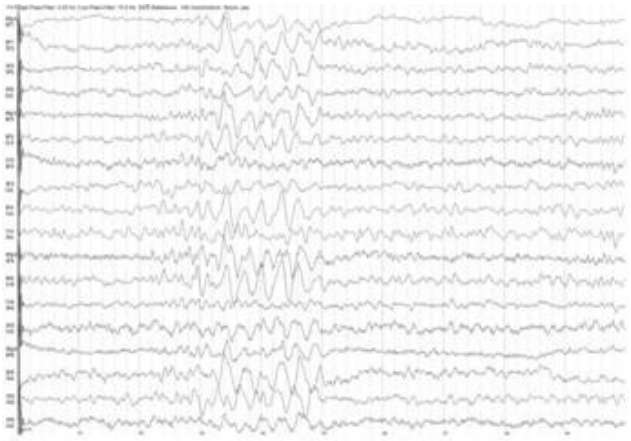

Figure 2. Disorganization and generalised slow wave paroxysm in the EEG.

Upon detecting that measles IgG was positive in the CSF (160 U / ml), the diagnosis of SSPE was established. High dose intravenous corticosteroids was initially given because of the severity of the clinical picture and the findings of brain stem involvement on cranial MRI, and then intravenous immunoglobulin was given; however, no response was received. During hospitalization, she was drowsy and agitated. On the twelfth hospital day, the main activity was found to be slower on repeated EEG which was taken due to increased tendency to drowsiness. A repeat cranial MRI scan after 2 weeks showed the lesions were enlarged in cerebellar peduncles and cerebellar hemispheres while the pons was completely involved As in first MRI, there was no contrast up-

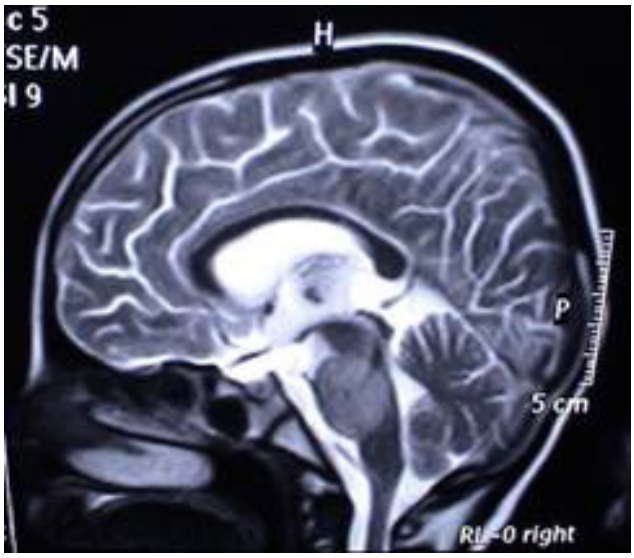

Figure 3.A. Proggression of hyperintensity in the pons and B. the cerebellum T2 sagittal section of MRI On the twenty-second day, myoclonus began and was partially taken under control with anticonvulsant therapy. On the twenty-sixth hospital day plasmapheresis was planned, however, the patient developed sudden cardiac and respiratory arrest and died in spite of active resuscitation.

DISCUSSION

SSPE is an insidious, progressive disease and results in coma and then death 1-3 years after onset of neurological symptoms. In recent years, acute and fulminant course of SSPE cases are more frequently reported and difficulties in diagnosis may lead to delays in management (6,10). As progression of the disease is very rapid in patients with fulminant SSPE, the classical four phases of the disease can not be observed.

Although the diagnostic criteria of the disease do not include the cranial MR findings, abnormal MRI findings are commonly encountered in patients with SSPE (13). Cranial MR imaging is usually normal in the early period; however, in the late stages of the disease, periventricular and subcortical white matter lesions are frequently seen (13,14). Involvement of the cerebellum and pons that we detected in our case is extremely rare in SSPE (14,15). While there is no correlation between clinical staging and cranial MR findings in SSPE, brain stem involvement shortens life expectancy (14,15,16). Our patient with SSPE with involvement of the pons and cerebellum died within a very short period like a month from the beginning of the disease.

Although neuropathological studies show early cortical involvement, conventional MR imaging taken in the early period of the disease are normal. In the study by Aydın et al. bilateral decrease was shown in the volume of gray matter of the fronto-temporal cortex, involving the amygdala and cingulate cortex on the cranial MRI using 'voxel-based, morphometry method in patients with SSPE stage 1 and 2 (8).

Considering the acute onset, clinical and

cranial MRI findings of our case we first assumed the differential diagnosis to be acute disseminated encephalomyelitis (ADEM); however, when IgG antibody of measles was determined in CSF this diagnosis was ruled out (17).

The incidence of SSPE has been reported to be approximately 4-11 per 100.000 cases of measles by World Health Organization's report in 2006 (2). This incidence is as high as 20-100/100.000 in developing countries (18).

The risk for SSPE is still higher in those contracting measles infection before the age of one; the only way of protection from this disease that has no definitive treatment is vaccination (19). The history of measles infection of our case at the age of nine months emphasizes the importance of vaccination.

10% of SSPE cases have a fulminant course (9). The fulminant form of SSPE has to be kept in mind while esablishing differential diagnosis of acute encephalopathies of unknown ethiologies (1).

When the presentation of SSPE is different from classic signs and symptoms, the diagnosis can be late and difficult. This case report seeks to draw attention to the neccessity of keeping the subacute sclerosing panencephalitis in mind while considering the differential diagnosis in patients with headache and acute altered mental state with atypical findings on the cranial imaging.

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