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Atypic Subacute Sclerosing Panensefalitis in a Six Years Old Male

Serap Bilge*, Neslihan Özcan, Gülen Gül Mert, Şakir Altunbaşak, M Özlem Hergüner

Cukurova University Faculty of Medicine, Department of Pediatric Neurology, Adana /Turkey

ABSTRACT

Background: Subacute sclerosing panencephalitis is a rare, slow, and insidious neurodegenerative disease caused by measle. This disease mostly has a classic course. However, sometimes it can be presented with atypical manifestations. In this paper,
Case Presentaion: A six years old male patient that was hospitalized due to seizures and ataxia. Cerebral magnetic resonance imaging was normal on the first day of admission, but within few days the patient started to be apathetic. On the seventh day, magnetic resonance imaging showed hyperintense lesions in the thalamic, brainstem, and periventricular area. Periodic epileptiform discharges were detected in the repeated electroencephalogram. Investigations from the cerebrospinal fluid showed markedly elevated measles virus IgG at 230U/ml consistent with the diagnosis of SSPE
Conclusion: Subacute sclerosing panencephalitis should always be ruled out when a patient comes in with uncontrollable seizures, ataxia, and apathy.

Keywords: Subacute sclerosing panencephalitis, myoclonic seizure, atypical course, ataxia.

Abbreviations: SSPE: Subacute sclerosing panencephalitis; CSF: Cerebrospinal Fluid; MRI: Magnetic Resonance İmaging; NCSE: Nonconvulsive Status Epilepticus.

Background

Subacute sclerosing panencephalitis (SSPE) is a rare, slow, and insidious neurodegenerative disease caused by measles. The disease can develop due to reactivation of the measles virus or an improper immune response to this virus. Behavioral problems, personality changes, seizures, cognitive deterioration, pyramidal, or extrapyramidal findings are the main features of the clinic. However, SSPE can also follow a fulminant course [1]. Dyken's criteria are based on the diagnosis which includes two major and four minor criteria. Raised anti-measles antibody titers in cerebrospinal fluid (CSF) greater than or equal to 1:4 or ratio greater than or equal to 1:256 in serum, typical or atypical clinical history are major criteria. Minor criteria include characteristic electroencephalographic findings that include periodic, generalized, bilaterally synchronous, and symmetrical high-amplitude slow waves that recur at regular intervals of 5–15 seconds called periodic slow-wave complexes, CSF globulin levels greater than 20% of the total CSF protein, characteristic histopathological findings on brain biopsy including inflammatory changes and specialized molecular diagnostic test to identify wild-type measles virus mutated genome. Usually, two major criteria plus one minor criterion are required [1]. Here we reported a case of SSPE that showed interesting clinical courses and radiologic features.

*Correspondence:
Serap Bilge, Cukurova University Faculty of Medicine, Department of Pediatric Neurology, Adana /Turkey

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Case Presentation

A six years old male patient was hospitalized due to frequent uncontrollable seizures. and progressive ataxic walk. The patient was born for consanguineous parents, antenatal and perinatal follow-up reported no insults. There was no family history of epilepsy or neurological diseases. At the age of one, seizures started and they were taken under control with levetiracetam Metabolic screenings and cerebral Magnetic resonance imaging (MRI) performed at that time were normal. He was admitted to our clinic due to frequent seizures and ataxia. On physical examination, the patient was conscious, cranial nerve and motor examination were normal but ataxia of the trunk and lower extremities were noticed. The epileptiform anomaly with multifocal myoclonic features was detected on EEG. Antiepileptic treatment was adjusted. Metabolic screenings, viral, autoimmune encephalitis panel were normal. Only high blood measles IgG (+) was detected. On the first day of hospitalization, cerebral MRI didn't show any abnormalities, the patient started to be apathetic within few days, on the 7th day, MRI showed hyperintense lesions in the T2A sequences in the area of the brainstem, thalamic region, periventricular, and caudate nucleus (Figure 1). Periodic epileptiform discharges were detected in the repeated EEG (Figure 2). Investigations from the cerebrospinal fluid (CSF) showed markedly elevated measles virus IgG at 230 U / ml (N: 0-25) consistent with the diagnosis of SSPE. On the 35th day of hospitalization, the seizures were taken under control using clobazam and carbamazepine and the patient was discharged with a vegetative state.

Discussion

SSPE is a progressive neurological disease that usually occurs during childhood and early adolescence. SSPE has 4 stages; (First stage) Behavioral disorders and intellectual regression, (Second stage) Involuntary movements, (Third stage) Severe pyramidal-extrapyramidal findings, (Fouth stage) Chronic vegetative period, and death. Generally, all phases last between months and years [1]. Atypical features such as acute encephalitis [2], dysarthria, ataxia [3,4] , abrupt vision loss [5] Epilepsia partialis continua [6], focal deficits, and asymmetric myoclonus [7] have been reported. SSPE can be manifested as myoclonic-astatic epilepsy. An Italian girl that died within four months after the onset of the disease was reported. Unfortunately, delay in the diagnosis and treatment of SSPE can occur mostly in patients with an atypical course [8]. Seizure disorders poorly controlled with medication could be the only manifestation of atypical cases. Catching the measles virus before the age of two, increased viral virulence, and coinfection with other viruses are risk factors for a more fulminant, atypical course [9]. Another case of SSPE that presented with ataxia and right hemiplegia with no visual symptoms seizures or slow generalized myoclonus have been reported. The asymmetrical slowing on the initial EEG only suggested a left hemispheric insult [4]. Our patient who was diagnosed with epilepsy earlier had uncontrollable seizures on admission and developed progressive ataxia. The first EEG showed multifocal discharges which urged us to think of the diseases that cause progressive myoclonic seizures. Our patient became apathetic on the third day the possibility of nonconvulsive status epilepticus (NCSE) was suspected. Therefore there was a need for a second EEG. Continuous EEG was recorded on the third day which showed periodic slow-wave discharges that were consistent with the diagnosis of SSPE. In this article, we present an atypical case to highlight that SSPE is still a cause of encephalopathy that should be always kept in mind when investigating a patient with neurological symptoms.

Neuroimaging can be beneficial and supportive but it is not characteristic and it can be even normal in the early stages of SSPE. MRI of the brain may show decreased gray matter volume, especially within the frontotemporal cortex, amygdala, and cingulate gyrus during the early stages, Hyperintensities on T2-weighted images in the periventricular white matter of cerebral cortex, brainstem, and basal ganglia, may develop. As the disease progresses. MRI could also reveal diffuse cortical atrophy [10]. Our patient's MRI was normal on admission but within 7 days it revealed hyperintensities in the thalamic, brainstem, and periventricular area. Antiviral drugs and immunomodulators are used in the treatment, although there is no standard treatment for SSPE. The most commonly used drugs in the clinical practice are Inosine pranobex, interferon alfa, ribavirin, and lamivudine. Steroids and intravenous immunoglobulin are no longer recommended. Antiepileptics and the ketogenic diet are supportive in the treatment of SSPE.

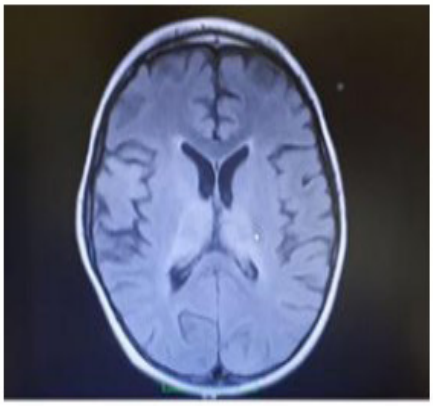

Figure 1: *Hyperintensities on T2-weighted images in the periventricular white matter cerebral cortex and basal ganglia, thalamus in a six years old boy with SSPE.*

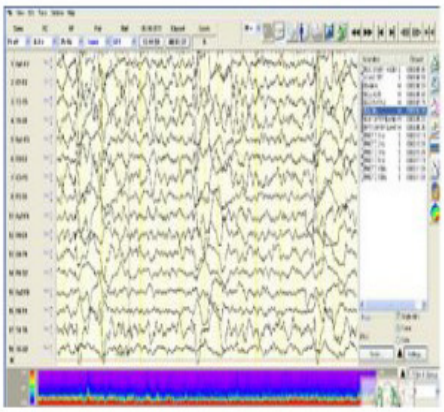

Figure 2: *EEG shows the periodic discharges in six years old patients with SSPE.*

Conclusion

Atypical presentations of SSPE should be always kept in mind and ruled out in every patient comes with uncontrollable seizures, ataxia, and apathy to prevent further delays of the diagnosis

Informed consent statement

The parents or the child's legal guardians provided informed written consent before study enrollment.

Conflict-of-interest statement

Authors declare no conflict of interest for this article.

Data sharing statement

No additional data are available.

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Strobe statement

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Reference

1. Jabbour J, Duenas D, Modlin J (1975) SSPE-clinical staging, course, and frequency. Arch Neurol. 32(7): 493–494.
2. Komur M, Arslankoylu AE, Okuyaz C, Kuyucu N (2012) Atypical clinical course subacute sclerosing panencephalitis presenting as Acute Encephalitis. J Pediatr Neurosci 7: 120-122.
3. Hergüner MO, Altunbaşak S, Baytok V (2007) Patients with acute, fulminant form of SSPE. Turk J Pediatr 49: 422-425.
4. Rukmini Mridula Kandadai, a Praveen Yada (2014) Fulminant subacute sclerosing panencephalitis presenting with acute ataxia and hemiparesis in a 15-year-old boy, J Clin Neurol 10(4): 354-357.
5. Ekici B, Calişkan M, Tatli B, Aydinli N, Ozmen M (2011) Rapidly progressive subacute sclerosing panencephalitis presenting with acute loss of vision. Acta Neurol Belg 111: 325-327.
6. Kravljanac R, Jovic N, Djuric M, Nikolic L (2011) Epilepsia partialis continua in children with fulminant subacute sclerosing panencephalitis. Neurol Sci 32:1007-1012.
7. Zaitoon M. Shivji, Ibrahim S. Al-Zahrani, Yousef A. et al. (2003)Subacute Sclerosing Panencephalitis Presenting with Unilateral Periodic Myoclonic Jerks. Can J Neurol Sci 30: 384-387.
8. Magurano F, Marella GL, Marchi A (2017) A case of fulminant subacute sclerosing panencephalitis presenting with acute myoclonic-astatic epilepsy, Ann Ist Super Sanità 53(2): 167-169.
9. 9.Jafri SK, Kumar R, Ibrahim SH (2018) Subacute sclerosing panencephalitis - current perspectives. Pediatric health, medicine, and therapeutics.
10. Akdal G, Baklan B, Çakmakçi H, Kovanlikaya A (2001) MRI follow-up of basal ganglia involvement in subacute sclerosing panencephalitis. Pediatr Neurol 24(5):393–395.