ACUTE DISSEMINATED ENCEPHALOMYELITIS OR SUBACUTE SCLEROSING PANENCEPHALITIS? A CASE REPORT

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DOI: 10.20959/wjpmr20172-544

ABSTRACT

Subacute Sclerosing Panencephalitis (SSPE) is a delayed and fatal complication of measles. It is a slowly progressive encephalitis that usually occurs 6 to 10 years after measles infection and progresses over 12 to 18 months. The early stages of disease consist of an abrupt development of neurologic symptoms such as personality changes, sluggishness, delayed in school performance followed by myoclonic jerk and convulsions. Then, flaccidity or decorticate rigidity and symptoms and signs of autonomic dysfunction appear. In a late stage, dementia, stupor and coma develop. In this article we report a 12 years old boy that developed acute ataxia, aphasia, dysarthria and right hemiplegia.


INTRODUCTION

Subacute Sclerosing Panencephalitis (SSPE) is a delayed and fatal complication of measles.\(^1\) It is slowly progressive encephalitis that usually occurs 6 to 10 years after measles infection and progresses over 12 to 18 months.\(^2\) The early stages of disease consist of an abrupt development of neurologic symptoms such as personality changes, sluggishness, delayed in school performance followed by myoclonic jerk and convulsions. Then, flaccidity or decorticate rigidity and symptoms and signs of autonomic dysfunction appear. In a late stage, dementia, stupor and coma develop.\(^3\) The diagnosis of SSPE can be based on a typical clinical course and positive measles antibody titers in the cerebral spinal fluid (CSF) together with characteristic electroencephalographic findings of high amplitude slow and sharp wave’s complexes, and/or typical histologic findings obtained by biopsy or autopsy.\(^1\)

In this article we present a twelve years old boy who developed acute ataxia, aphasia, dysarthria and right hemiplegia. Due to suggestive clinical and radiological findings, acute disseminated encephalomyelitis (ADEM) was suspected and the patient was started on steroids. His condition didn’t improve; an electroencephalogram (EEG) was done and was specific for SSPE; antibody titers for measles were performed in the CSF and turned out to be positive. The diagnosis of SSPE was then confirmed.

CASE REPORT

A twelve years old previously healthy boy presented to our hospital for a two months history of progressive neurologic deterioration with ataxia, dysarthria, diffuse spasticity and right sided body myoclonic seizures with right hemiparesis. A regression in school performance with was also noted. One week prior to presentation, the patient developed a low-grade fever with several episodes of vomiting. There was no history of a head trauma or a decrease in the level of consciousness. Upon admission, he was afebrile and his vital signs were stable. On physical examination he had a slow, dysarthric and a dysphasic speech, a diffuse spasticity and a right body hemiparesis. The deep tendon reflexes were increased in the lower extremities and the Babinski reflexes were positive bilaterally. Most importantly, right generalized body myoclonic jerks occurred every two-three seconds. Other systemic findings were found to be normal. The patient had an MRI of brain done prior to presentation. It showed diffuse multiple small demyelinating foci in the white matter (figure 1).
His hospital work up showed normal complete blood count and culture, electrolytes, liver and kidney function tests as well as CRP. The blood ammonia, lactate, ANA, ANCA and ESR were all normal. A lumbar puncture was performed and the cerebrospinal fluid was clear without any type of cell in microscopic examination. The glucose and the protein content were normal. The Oligoclonal bands were positive. In view of the clinical picture of this patient and the MRI findings, the diagnosis of acute disseminated encephalomyelitis was sustained and treatment with IV Methylprednisolone with a dose of 30 mg/kg/day was started.

Unfortunately, the patient started to deteriorate quickly and the right-sided myoclonic jerks became more frequent with a frequency of almost two-three/sec. His speech worsened and he became bed ridden. Steroids were therefore stopped after three days of treatment. An EEG was done (figure 2) and showed high-voltage (around 300 μV), generalized, periodic complexes of polyphasic sharp and slow waves lasting half to two seconds and occurring regularly every three-four seconds, with a slow background of low-amplitude activity. The findings were suggestive of Rademecker complex seen in SSPE. Titers of IgG antimeasles antibodies were detected in his cerebrospinal fluid and in his blood and were both extremely elevated. SSPE diagnosis was then confirmed. Due to the poor prognosis of his condition, the family decided to withdraw medical treatment on the patient.

DISCUSSION

ADEM is an acute demyelinating disease of the central nervous system resulting from an immune-mediated inflammatory disorder triggered by a viral infection mainly after upper respiratory tract infection and infectious like varicella, herpes zoster, rubella and mumps or after vaccination. The demyelination mainly involves the white matter of the brain and spinal cord and presents as a monophasic disorder associated with multifocal neurologic symptoms and encephalopathy.[4-6] The clinical presentation is polysymptomatic and can fluctuate in severity from altered mental status, lethargy, delirium, seizures (which can be focal or generalized) to pyramidal dysfunction [7], acute hemiparesis, cerebellar ataxia, brainstem syndromes, optic neuritis, myelitis and coma. Different degree of residual deficits may remain, ranging from mild clumsiness to hemiparesis.[8] The encephalitic illness is more common in children younger than three years of age.[8] The MRI of the brain shows...
typically widespread multifocal lesions, demyelination in the white matter, the basal ganglia and the brain stem with increase in signal intensity in T2 and fluid attenuated inversion recovery sequences. Different sizes of lesions may be seen in the same patient. The electroencephalogram (EEG) is not diagnostic. It may describe a disturbance of normal sleep rhythms and may show focal or generalized slowing of the background activity seen usually in encephalopathic states.

The CSF immune tests usually show pleocytosis and/or an increase in proteins indicating an inflammation in the cerebrospinal fluid. High serum titers of IgG specific for myelin oligodendrocyte glycoprotein (MOG) may be observed. 10% of patients with ADEM have oligoclonal bands in the CSF. Some patients respond well to steroid or immunoglobulin therapy.

The presence of progressive neurologic deterioration and diffuse multiple lesions with signal changes on MRI with oligoclonal bands in CSF in our patient initially suggested the diagnosis of ADEM and treatment was started with steroids. However due to the continuous clinical deterioration and ongoing focal body myoclonus other etiologies were evaluated, SSPE being the most relevant. This was confirmed by the typical findings on EEG and by the detection of anti-measles antibody in CSF that were extremely elevated. SSPE is a rare, fatal and delayed form of encephalitis, causing widespread demyelination and neuronal degeneration in the central nervous system. It occurs in children who are infected with measles virus at a young age. Usually, SSPE occurs between five and fifteen years of age. The incidence of SSPE is increased in the area where the vaccination program against measles is not widespread.

The typical clinical presentations of SSPE begin with a progressive intellectual deterioration, decreased school performance, behavior change, myoclonic jerks, seizures and/or visual symptoms. Atypical presentations of SSPE can occur in 10% of cases. The initial presentation with focal neurologic deficit rarely occurs, and may make the diagnosis more difficult. In SSPE, the CSF analysis has normal cytology, glucose and total protein, but there are elevated levels of gammaglobulin and anti-measles antibodies. The anti-measles antibodies in blood are markedly elevated. Typically, the EEG reveals high-voltage generalized, periodic complexes of polyphasic sharp and slow waves lasting half to two seconds and occurring regularly every three-four seconds, or was called “the Rademeyer complex”. The bursts of abnormal sharp and slow waves appear on a normal background EEG activity in the first stage, but as the disease progresses, this background activity deteriorates to diffuse slow waves, forming a “burst suppression” pattern. The MRI is sensitive in the diagnosis of SSPE. The most common finding on MRI is hyperintensities on T2-weighted and FLAIR sequences involving bilaterally the gray and white matters in the occipital and parietal regions. No definitive curative treatment is so far available in SSPE. The available therapies as immunoglobulins and intrathecal interferon are mainly supportive treatment. Most patients die within one-three years of disease manifestation. Fortunately, the incidence of acute measles infection and SSPE has decreased markedly due to a widespread administration of measles vaccine.

CONCLUSION

SSPE presentation can mimic ADEM. The presence of CSF measles and EEG findings could help differentiating both entities. Though SSPE is nowadays a rare disease, we do encourage vaccine campaigns all over the world hoping to eradicate the measles virus, as prevention is the most essential step in facing the disease.

REFERENCES