ACUTE DISSEMINATED ENCEPHALOMYELITIS OR SUBACUTE SCLEROSING PANENCEPHALITIS? A CASE REPORT

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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 twelve to eighteen 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.[4]

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 bedridden. 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. 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, acute hemiparesis, cerebellar ataxia, brainstem syndromes, optic neuritis, myelitis and coma. Different degree of residual deficits may remain, ranging from mild clumsiness to hemiparesis. The encephalitic illness is more common in children younger than three years of age. 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.\textsuperscript{[9]} Different
sizes of lesions may be seen\textsuperscript{[10]} 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.\textsuperscript{[11]}

The CSF immune tests usually show pleocytosis and/or
an increase in proteins indicating an inflammation in the
cerebrospinal fluid.\textsuperscript{[11, 12]} High serum titers of IgG
specific for myelin oligodendrocyte glycoprotein (MOG)
may be observed.\textsuperscript{[13]} 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.\textsuperscript{[14, 15]} Usually, SSPE
occurs between five and fifteen years of age.\textsuperscript{[16]} The
incidence of SSPE is increased in the area where the
vaccination program against measles is not widespread.\textsuperscript{[17]}

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.\textsuperscript{[18, 19]} The initial presentation
with focal neurologic deficit rarely occurs, and may
make the diagnosis more difficult.\textsuperscript{[20, 21]} In SSPE, the
CSF analysis has normal cytology, glucose and total
protein, but there are elevated levels of gammaglobulin
and anti-measles antibodies.\textsuperscript{[22, 23]} 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 is called “the Rademecker
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\textsuperscript{[24]}, 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.\textsuperscript{[25-27]} No definitive curative
treatment is so far available in SSPE.\textsuperscript{[14]} The available
therapies as immunoglobulins and intrathecal interferon
are mainly supportive treatment. Most patients die within
one-three years of disease manifestation.\textsuperscript{[28]} Fortunately,
the incidence of acute measles infection and SSPE has
decreased markedly due to a widespread administration of
measles vaccine.\textsuperscript{[29]}

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