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Case Report

Subacute Sclerosing Panencephalitis

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Abstract:

Subacute sclerosing panencephalitis (SSPE) is a late complication of measles virus infection.It presents with psychiatric manifestations, progressive dementia,myoclonic jerks and other focal neurological signs. The diagnosis is based characteristic clinical manifestations, presence of characteristic periodic EEG discharge and demonstration of raised antibody titer against measles in plasma and CSF. In our case, patient presented with myoclonic jerk, characteristic EEG and raised level of antibody in CSF.

Introduction

Subacute sclerosing panencephalitis (SSPE) is a progressive neurological disorder caused by persistent measles virus infection.[1] The latency period between acute measles and first symptoms of SSPE is usually 4 to 10 years but ranges from 1 month to 27 years.[2] SSPE is characterized by progressive mentaldecline, myoclonus and raised anti-measles antibody titer inthe cerebrospinal fluid. Electroencephalography (EEG) in SSPE characteristically reveals generalized periodic complexesor discharges. A periodic EEG record typically consists of generalized and synchronous bursts of sharp-slowwave complexes. A typical discharge is polyphasic, withduration varying from 0.5 to 2 seconds; high voltage (300-1,500 mV); and repetitive (occurring every 4-15 seconds). We are reporting a child with rapidly evolving encephalopathy and the child's cerebrospinal fluid demonstrates a high titer of anti-measles antibodies.[3,4,5]

Case Report

A 16 yrs old boy from Jurain, Dhaka got admitted in SSMCMH with the complaints of jerky movements of whole body for last 1 year and was unable to speak and stand for last 1 month. From his childhood, he had some behavioral abnormalities and slowness in mental status, so his parents took attention from traditional healer. 7 months back, he suddenly developed convulsion which starts at night, occurs several occasions in a single day, more at night and patient fears to sleep. Subsequently he was maltreated by traditional healer for 8 months. Then he was admitted in BSMMU and got treatment as a case of focal seizure. His symptoms were temporarily relieved for 2 months after starting treatment but subsequently he developed same type of problem in more severe form.

After admission, we found he was unable to speak and stand and myoclonic jerks. His vital sign is normal, not anemic or icteric. His muscle power is reduced in both limbs but bulk is normal with no clonus or fasciculation. His jerks of both upper and lower limbs are exaggerated and planter response is equivocal. His sensory function, cranial nerves are intact. His bowel and bladder habit are normal. On fundoscopic examination, there is no KF ring or sunflower cataract.

On query of his mother, she mentioned that his growth and development was normal except delayed speech. He got vaccination as per EPI schedule except measles vaccine. He had no history of fever with rash in his early childhood.

Investigation report reveals CBC, serum electrolytes, S. creatinine, CXR P/A view is normal. His brain imaging like MRI is normal. CSF study shows total cell count is 20/ mm,^{3} 90% neutrophils and 10% lymphocyte among them. Gram stain and Z. N. stain shows no bacilli. Glucose is 5.6 mmol/L (normal range is 2.2-3.8 mmol/L), protein is 0.86 g/L (normal range 0.15-0.45). Plasma lactic acid level is increased 6.44 mmol/L (normal range 0.5-2.2). His 24hrs urinary copper level is 28.4 microgram/L and serum ceruloplasmin level is 110 mg/L (reference range 200-600).

His EEG demonstrates presence of frequent periodic generalized burst composed of polymorphic slow waves of 1-3 seconds followed by stretches of attenuation at an irregular interval of 5-14 seconds which may be consistent with subacute sclerosing panencephalitis (SSPE). His IgG Antibody against Measles is positive and titer is 352.9 mIU/ml (>200 mIU/ml positive, <150 mIU/ml negative).

Discussion

In this patient, because of the presence of anti-measles antibody in high titer in cerebrospinal fluid, a reasonable diagnosis of SSPE can be considered. There are certain important points that need to be highlighted, initial progressive subacute mental deterioration of the boy was misdiagnosed as psychiatric abnormality by the parents and took attention from traditional healers. His myoclonus was initially diagnosed as partial seizure in a tertiary care hospital and that was reason for delayed diagnosis of the case.

At least three of the following five criteria should be met for SSPE diagnosis:[6,7]

a) a typical clinical picture of progressive subacute mental deterioration with typical signs like myoclonus and neurodiagnostic features consistent with SSPE;

b) characteristic EEG changes;

c) CSF globulin levels greater than 20% of total CSF protein;

d) raised titers of measles antibodies in blood and CSF in the absence of other antibodies, including against HSV and VZV and

e) typical histopathological findings on brain biopsy or autopsy.

According to the WHO definition, acute encephalitis syndrome is characterized with fever and change in the mental status (such as confusion, disorientation, coma, or mutism) with or without seizures. Acute encephalitis syndrome is highly endemic to certain parts of India. In India, Japanese encephalitis is the leading cause of acute encephalitis syndrome. Dengue, West Nile virus, herpes simplex, mumps, Epstein–Barr, and influenza are other viruses that can clinically present with acute encephalitis syndrome. In measles-endemic countries, fulminant SSPE should also be included in the list of causes of acute encephalitis syndrome. [8,9]

Our case also highlights the importance of universal coverage of measles vaccination. The situation remains grim, and approximately 50% of the global measles-associated deaths occur in India.10 Our patient missed measle vaccination in EPI schedule.

Conclusion

SSPE is an uncommon and fatal complication of childhood measles. It is a progressive and incurable condition resulting in death typically within one to three years of onset of symptoms. Clinical presentation widely varies ranging from progressive weakness, seizures, pyramidal and extrapyramidal symptoms and coma. Although SSPE is a very rare disease, still on the background of characteristic clinical manifestation and classical EEG findings, diagnosis is easy.

As available treatments are very costly and are available only at a few centers in the world, so effective measles vaccination seems to be the only solution to problem of this dreadful neurological disorder.

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