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Subacute Sclerosing Panencephalitis- A Case Report

KEYWORDS: Measles Demyelination Myoclonic jerks

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ABSTRACT *Subacute Sclerosing Panencephalitis (SSPE) is a rare chronic, progressive demyelinating disease of the CNS associated with a chronic nonpermissive infection of brain tissue with measles virus.*

Most patients give a history of primary measles infection at an early age which is followed after a latent interval of 6–8 years by the development of a progressive neurologic disorder.

Initial manifestations include poor school performance and mood and personality changes. Typical signs of a CNS viral infection do not occur. As the disease progresses, patients develop progressive intellectual deterioration, focal and/or generalized seizures, myoclonus, ataxia, and visual disturbances. In the late stage of the illness, patients are unresponsive, quadriplegic and spastic

We report a 17 years old male admitted with history of childhood measles infection and recurrent seizures with no other comorbid condition

INTRODUCTION

SSPE is a rare chronic, progressive demyelinating disease of the CNS. The frequency has been estimated at 1 in 100,000–500,000 measles cases. The incidence has declined dramatically since the introduction of a measles vaccine.

Subacute sclerosing panencephalitis is caused by the accumulation of incomplete measles virus that cannot be cleared by B or T cell mechanism. It begins in the cortical grey matter progresses to subcortcal grey matter and white matter. Brain biopsy performed in the early stages shows mild inflammation of the meninges and brain parenchyma.

CASE REPORT

Master. Sudalaimuthu 17y/m ITI student presented with c/o abnormal posturing of jaw, upper limbs & lower limbs past 9 months with difficulty in speaking, swallowing and cognitive decline past 9 months Patient also has history of jerky movements of upper & lower limb for past 5 months. He had h/o exanthematousfever in childhood at 2 years of age, fever with maculo-papular rashes preceded by fever lasting for 3 days, it was uneventful with no post viral immediate complications

He cried immediately after birth, No h/o delayed mile stones. Immunized for age, vaccination details not known.

Examination of central nervous system -Higher mental function: could not be assessed Intelligence: moderate IQ.Speech: slurred, fluency- reduced, volume- reduced , comprehension- normal

Cranial nerve examination: normal. Spino-motor system: normal

Sensory system, cerebellum: normal, bladder& bowel: normal Spine & cranium: normal

Recurrent Myoclonic Jerks +++ - sudden, brief , shock-like movements of upper and lower limb associated with jaw opening, triggered by pain stimulus , sometimes spontaneously.

INVESTIGATIONS showed

CBC, RFT, LFT: WNL CXR , ECG: WNL CPK : 15 U/L HIV : Non Reactive. HBSAg& ANTI-HCV: negative

CSF Analysis: Glucose 62mg% Protein 30 mg Globulin – positive Acellular smear, Serum Ceruloplasmin- 16mg/dl(18-35mg/dl) Serum Copper – 95 microgram/dl(75 -150microgm/dl), 24 hr urine copper- 23.5 microgram/day(20-50mg/d)

TFT – normal. Opthal evaluation: NO KF Ring, no papilledema

Anti measles antibody IgG +ve

Scalp EEG: Generalised Epileptiform abnormalities

CT Brain –NAD MRI Brain – Normal study (figure 1)

Patient was diagnosed as a case of subacute sclerosing panencephalitis and was treated with antibiotics and antiepileptics.

DISCUSSION

Measles is a highly contagious viral disease that is characterized by a prodromal illness of fever, cough, coryza, and conjunctivitis followed by the appearance of a generalized maculopapular rash.

Rare but serious complications of measles involve the central nervous system (CNS). Postmeasles encephalomyelitis complicates 1 in 1000 cases, The finding of periventricular demyelination, the induction of immune responses to myelin basic protein, and the absence of measles virus in the brain suggest that postmeasles encephalomyelitis is an autoimmune disorder triggered by measles virus infection.

MRI is often normal early, although areas of increased T2 signal develop in the white matter of the brain and brain-

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stem as disease progresses. The EEG may initially show only nonspecific slowing, but with disease progression, patients develop a characteristic periodic pattern with bursts of high-voltage, sharp, slow waves every 3–8 s, followed by periods of attenuated ("flat") background.

The CSF is acellular with a normal or mildly elevated protein concentration and a markedly elevated gamma globulin level (>20% of total CSF protein). CSF antimeasles antibody levels are invariably elevated, and oligoclonal antimeasles antibodies are often present

Measles virus can be cultured from brain tissue using special cocultivation techniques. Viral antigen can be identified immunocytochemically, and viral genome can be detected by in situ hybridization or PCR amplification.

Staging of SSPE is done according to modified jabbour classification as follows.

Stage 1- mental and behavioural changes, forgetfulness, irritability and lethargy
Stage 2- myoclonic jerks, dyskinesia, choreoathetosis, ataxia
Stage 3- decerebrate rigidity and decorticate rigidity
Stage 4- severe loss of all cortical function, flexion posturing of limbs and mutism

CONCLUSSION

SSPE is a neurodegenerative disease caused by persistant infection of the brain by an altered form of measles virus. No definitive therapy for SSPE is available.

Treatment with isoprinosine (Inosiplex, 100 mg/kg per day), alone or in combination with intrathecal or intraventricular alpha interferon, has been reported to prolong survival and produce clinical improvement in some patients but has never been subjected to a controlled clinical trial.

One of the most important limitation in treatment of SSPE is the inability to detect early manifestations of the disease. There is a need for more awareness of the disease by primary care physician and pediatrician. Homecare system for these handicapped children is very important and meaningful to increase quality of life in them.

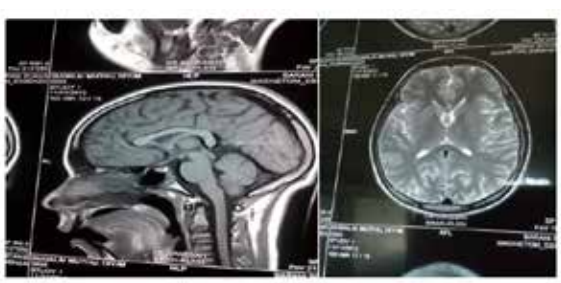

Figure 1

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