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Pediatrics

A RARE CASE OF PROGRESSIVE MYOCLONIC EPILEPSY DUE TO SSPE

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ABSTRACT SSPE is a chronic complication of measles with a delayed onset and nearly fatal outcome. This "slow virus infection" results from a persistent infection with an altered measles virus, that is harboured intracellularly in the CNS. Patients usually presents with cognitive decline and myoclonus but atypical presentations are also quite common. Our case is of a 9 year old male child , previously neurodevelopmentally normal, presented with 4 months history of frequent fall due to jerky movements of the left upper and lower limbs which progresses to involve left half of the body, reduced attention span, cognitive decline and loss of speech. During hospitalisation, despite treatment with antibiotics , steroids , and antiepileptics ,disease progressed with myoclonic jerks , altered sensorium and finally child became comatose. CSF study is normal but MRI and EEG shows abnormalities. Based on clinical features and history of exanthematous illness at 6 months of age, measles igG antibody titres in CSF were tested , which came to be highly elevated confirming SSPE.

KEYWORDS : Measles , SSPE , Anti measles ig G , CSF study , Myoclonus , Neurodegeneration , vaccination.

INTRODUCTION

SSPE is a progressive neurological disorder caused by persistent measles virus infection[1]. The usual age of onset is between 5-15 years[2]. SSPE was first clinically described in 1993 as 'Subacute inclusion encephalitis' due to the presence of inclusion bodies seen in brain biopsies[3]. Usual presentation is with intellectual decline or behavioural issues followed by myoclonic jerks, which becomes generalised involving axial body parts[2]. It has a progressive and downhill course that results in death within 5 years of onset[4]. Overall 4-11 cases of SSPE are expected for every 100,000 cases of measles but the incidence is higher among children aged <5 years [18/ 100,000][5].

we are presenting a case of 9 year old male child with myoclonic jerks involving left lower limb which progresses to involve left half of the body with reduced attention span for 3-4 months. The illness progresses with loss of speech with altered sensorium and inability to do activities of daily living. The child developed repeated myoclonic jerks involving the Right angle of mouth. CSF Study and MRI were normal and EEG shows abnormal findings. As the child had a history of exanthematous illness at 6 months of life, a provisional diagnosis of SSPE was made and antimeasles IgG antibodies were sent, which was highly Positive.

CASE STUDY

9 year old male child from Ganjam, Odisha, product of non-consanguineous marriage, neuro-developmentally normal, fully immunised according to NIS, presented with chief complaints of difficulty while walking due to jerky movements of the left upper and lower limbs for last 4 months. Sudden jerky movements of the left half of the body since last 3 months; reduced attention for last 3 months, loss of speech since last 2 weeks and altered sensorium with vomiting since last 7 days. There is no history of significant illness in the past except a history of an exanthematous fever at around 6 months of age, most probably measles. No significant illness in the family. At the time of admission, child was conscious, afebrile, with stable vitals. CNS examination revealed a conscious and oriented child with reduced speech. Decreased tone on left lower limbs with grade 4 power. Reflexes were normal except Bilateral plantar extensor. There were no meningeal signs. During the course of hospitalisation child becomes unconscious, comatose with repetitive myoclonic jerks involving right angle of mouth. CBC, LFT, RFT, Serum electrolytes were normal. Bidaily RBS recordings were normal. CSF study and fundoscopic evaluation were normal. MRI and EEG done on day 4 of hospitalisation. MRI reveals Periventricular Leucomalacia and white matter paucity in bilateral posterior parieto -occipital, frontal, periventricular and left parietal cortex. EEG Shows background slow 4-5 delta, with poorly marked, sleep waves interspersed with frequent large amplitude spike and polyspike and slow waves lasting for 2-3 Seconds seen throughout the recordings(FIG 3). In view of above findings and typical clinical findings, a normal CSF Study and a history of exanthematous fever at 6 months of age, a provisional diagnosis of progressive myoclonic epilepsy due to SSPE was made after consulting with the department of Neurology and CSF IgG measles antibody were sent. The measles IgG antibody was 294Au/ml (Highly elevated). Sodium valproate and carbamazepine were started and based on request from parents, patient referred to AIIMS Bhubaneshwar for further management.

Test Name	Results	Units	Bio. Ref. Interval
MEASLES (RUBEOLA) ANTIBODY, IgG, CSF @ (CLIA)	274.00	AU/mL	

Fig 1.

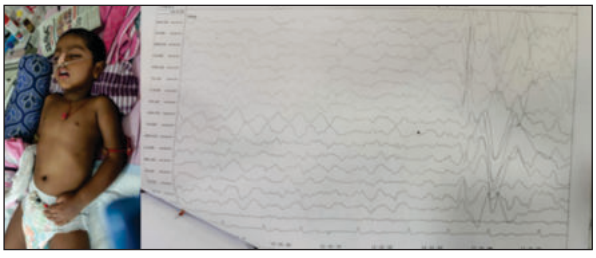

Fig 2.

Fig 3. EEG Showing Abnormal Pattern

DISCUSSION

SSPE is a progressive neurological disorder caused by persistent measles virus infection. Its typical course is a child presenting with intellectual decline or behavioural issue followed by myoclonic jerks, which becomes generalised involving axial body parts. Acute fulminant SSPE is characterised by a downhill course and patient eventually succumbs[6,7,8,9]. Measles at an early age favours the development of SSPE. males are affected twice as often as females. More cases are reported from rural areas. There are 4 clinical stages of SSPE.

Our case is a male child who had given history of measles at 6 months of age. He is from a rural area in Odisha. He had a typical clinical course of reduced attention (stage 1), myoclonic Jerks with consciousness maintained (Stage 2), Progressing to immobility (Stage 3).This downhill clinical Course despite treatment with antibiotics, antiepileptics, steroids and other supportive measures with highly positive measles IgG antibody in CSF (FIG 1) clinch the diagnosis.

Differential diagnosis include autoimmune encephalitis and ADEM. Pediatric autoimmune encephalitis is characterised by rapidly progressive encephalopathy with memory loss, altered mental status or psychiatric manifestations[10]. Other features are new onset seizures ,

focal neurological deficits and inflammatory changes in CSF. In our case the CSF examination reveals normal findings.

ADEM is another DD, inflammatory demyelinating disease of CNS that often follows a viral illness or vaccination[2]. It is clinically characterised by the acute or subacute onset of multifocal neurological disturbance that typically follows a monophasic course with MRI evidence of widespread demyelination. Our case had no history of preceding viral illness or vaccination and MRI findings are not typical.
Long survival and apparent remission are also reported in SSPE[11]. one such Case was reported by WA cobb etal. 14 year old boy deteriorated over a period of 9 months, improved and remained stable for over 7 years before relapse. The final deterioration to death extended over 6 years.

CONCLUSION

Management of SSPE is primarily Supportive. Clinical trials using isoprenaline with or without interferon suggest Significant benefit. Carbamazepine is of benefit in controlling myoclonic jerks. Prevention of SSPE is by prevention of primary measles infection by vaccination. All patients will eventually Succumbs to death.

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