International Journal of Sciences & Applied Research

www.ijsar.in

Case Report
Atypical presentation of Subacute Sclerosing Panencephalitis: a case report

Garg V.K.*, Wadhwani P., Sengar G.S., Chahar C.K.

Department of pediatrics, S. P. Medical College & Associated group of hospitals, Bikaner, Rajasthan, India
Correspondence Address: *Dr. Vijendra Kumar Garg, S/o Mr. P. L. Gupta, H. no. 227, Joshi colony, Shahpura road, Neem ka thana, sikar, Rajasthan-332713, India

Abstract
Subacute Sclerosing Panencephalitis (SSPE) is a rare progressive disorder with a prominent childhood onset, caused by persistence of mutant measles virus in the central nervous system. In the classic form of SSPE, slow progression of neurological symptoms goes through four characteristic stages. Atypical form of SSPE occurs in about 10% of patients and a high index of suspicion is needed to detect these atypical cases. We report a Patient of SSPE who presented to us as rapidly progressive fulminant acute encephalitis.

Keywords: SSPE, atypical, acute encephalitis, measles

Introduction
Subacute sclerosing panencephalitis (SSPE) is a rare progressive inflammatory disorder of central nervous system related to persistent and aberrant measles virus infection[1], which affects primarily children and young adults. The annual incidence of SSPE in both adults and children after measles infection dropped from 1/million in the pre-immunization era to 0.06/million after immunization was implemented[2]. Measels at an early age predisposes the development of SSPE, upto 75% cases of SSPE have been reported to have measles before 4 years.

In the classic form of SSPE clinical features begins insidiously. Slow progression of neurological symptoms goes through four characteristic stages[3]. In the initial stage I, the individual's behaviour becomes abnormal and erratic, and often accompanied by memory loss and mental deterioration characterised by intellectual difficulty. Fever, headache, and other signs of encephalitis are usually absent.The hallmark of II stage is involuntary movements and repetitive myoclonic jerks which usually begins in single muscle group but give way to massive spasm and jerks involving both axial and appendicular muscles. Consciousness is maintained. In the III stage, involuntary movements disappear and are replaced by choreoathetosis, immobility, dystonia and lead pipe rigidity. Sensorium deteriorates into dementia, stupor, and then coma.The IV stage is characterized by loss of critical centres that support breathing, heart rate and blood pressure.

However, in about 10% of patients clinical manifestations of SSPE are not typical and that may cause a delay in the diagnosis and treatment of the disease. The diagnosis may be established by the characteristic EEG pattern (suppression – burst episodes), elevated anti-measels IgM and IgG(>1:8) in

CSF, and raised measles antibody titres, presence of measles antigen on immuno-fluorescent studies and nuclear inclusion bodies in brain biopsy. Brain biopsy is no longer routinely indicated for diagnosis of SSPE.

We report a case of 7-year-old boy presenting as acute encephalitis and later diagnosed to have SSPE. The aim of this report is to emphasize the importance of considering a different clinical presentation of SSPE.

Case report

A 7 year old previously healthy boy came with sudden onset of fever, headache, altered sensorium, involuntary movements, difficulty in walking and abnormal speech since 2 days. Prior to the illness he didn't have any complaints except discharge from right ear since last 2 years. He attended the school properly till date of illness and there was no behavioural and intellectual deterioration prior to illness. He had history of fever, cough and redness of eyes with generalised morbiliform exanthema, clinically diagnosed as having measles infection at around 1 year of age. He was not immunised against measles. He recovered uneventfully within a week.

On presentation, his weight was 17 kg, BP 100/50 mmHg, Temperature 98.7^0F, pulse rate 108/min, and respiratory rate 30/min.He had Glasgow coma score of six, bilateral involuntary movements(myoclonic jerks), brisk deep tendon reflexes and extensor plantar reflexes. Fundus examination showed bilateral papilledema. Other systemic findings were found to be normal. Acute encephalitis was suspected and supportive treatment was started.

Cerebrospinal fluid (CSF) examination was found to be normal, protein= 45mg/dl, sugar = 68mg/dl, cell count < 5 mainly lymphocytes. MRI showed normal study except maxillary sinusitis. As the CSF examination was normal and clinical condition of the patient deteriorated rapidly with persistent myoclonic jerks and the patient had history of measles infection in the past patient was suspected to have atypical presentation of SSPE.

EEG revealed periodic generalized complexes consisting of bilaterally symmetrical, high voltage bursts of sharp waves and delta waves which repeated at an interval of 3 to 20 seconds with a slow background (Fig. 1). A sample of CSF and serum was obtained for anti-measles antibody. CSF and serum anti-measles antibody were 19174U/ml and 17709U/ml respectively. Based on typical EEG and presence of anti- measles antibody in CSF diagnosis of SSPE was established.

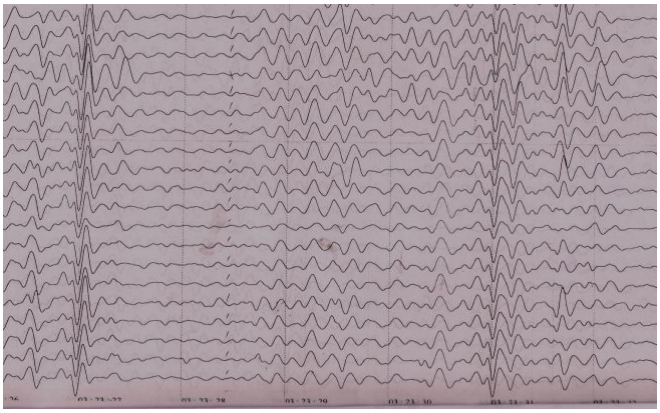

Fig 1: Generalised periodic EEG pattern with a slow background.

During the stay in hospital patient was given supportive treatment. He was given sodium valproate and clonazepam for myoclonic jerks. After 10 days, seizures persisted though the frequency of seizures decreased somewhat, patient's relatives left the hospital against medical advice.

After around two and half month patient was brought to Pediatric emergency room with profound shock and irregular gasping respiration. Prompt resuscitation measures were started and supportive therapy was provided but patient expired on second day of re-admission.

Discussion

SSPE usually runs a relentlessly progressive clinical course, resulting in death in most cases within 1 to 3 years after diagnosis[4]. The disease characteristically progresses insidiously through the stages of cerebral dysfunction in the form of cognitive impairment and behavioural changes, motor and convulsive phenomena especially myoclonic jerks, and deterioration of consciousness (sometimes culminating in coma).

Atypical form of SSPE occurs in about 10% of all patients. Unlike classical SSPE, in atypical form there are no defined stages in clinical presentation due to rapid course[5]. Atypical features also include unusual age of onset, visual loss, seizures and other focal symptoms as initial presentations, a lack of SSPE-specific EEG pattern, and atypical fast progression of disease. A patient could have more than one of these atypical features [6].

This case highlights the atypical nature of SSPE, as the case presented actually without intellectual delay and behaviour deterioration. This case presented to us as acute encephalitis with fever, headache, involuntary movements and deranged sensorium and also had fulminant course as he died within 3 months of diagnosis. High index of suspicion is needed to detect SSPE with atypical presentation, as the disease can mimic acute encephalitis, it is important to include SSPE in the list of differential diagnosis of acute encephalitis, especially if the patient presents with myoclonic jerks and positive history of measles in early childhood.

The cerebrospinal fluid (CSF) in SSPE will typically have normal cellular components, glucose and total protein, but markedly elevated values of gammaglobulin and anti-measles antibody[1, 4].Typically serum anti-measles antibody titers are grossly elevated. The EEG pattern is virtually diagnostic with periodic complexes consisting of bilaterally symmetrical, synchronous, high voltage bursts of polyphasic, stereotyped delta waves which repeat at fairly regular 4-10 second interval and have a 1:1 relationship with myoclonic jerks[7].All these characteristic CSF and EEG findings were seen in our case.

No curative treatment is available for SSPE but therapy with immunomodulators such as isoprinosine and interferons; and antiviral drugs like ribavarin may help in halting the progression of the disease[8].

Measles is still an important medical problem in the developing countries, so SSPE should be considered when a patient with history of measles presents with atypical clinical features like loss of consciousness, acute partial-generalized convulsion, acute encephalitis, visual loss, ataxia, and hemiparesis. EEG of these patients should be evaluated carefully and serum and CSF measles antibodies should be examined.

References

1. Garg RK. 2002. Subacute sclerosing panencephalitis. Postgrad Med J. 78:63-70
2. Honarmand S, Glaser CA, Chow E, et al. 2004. Subacute sclerosing panencephalitis in the differential diagnosis of encephalitis. Neurology. 63:1489-149
3. Jabbour JT, Duenas DA, Modlin J. 1975. Subacute sclerosing panencephalitis: clinical staging, course and frequency. Arch Neurol. 32:493.
4. Dyken PR. 1985. Subacute sclerosing panencephalitis. Neurolclin. 3:179-195.
5. Kravljanac R, Jovic N, Djuric M, Nikolic L. 2010. Epilepsia partialis continua in children with fulminant subacute sclerosing panencephalitis. NeuroloSci. 32:1007-1012.
6. Cruzeria MM, Vale TC, Pires LA, Franco GM. 2007. Atypical subacute sclerosing panencephalitis. ArqNeuropsiquiatr. 65:1030-1033.

7. Dunn RA. 1991. Subacute sclerosing panencephalitis. Pediatr Infect Dis J. 10:68-72.
8. Gascon GG. 2003. Randomized treatment study of inosiplex versus combined inosiplex and intraventricular interferon-alpha in subacute sclerosing panencephalitis (SSPE): international multicenter study. J Child Neurol. 18:819-827.