

CASE REPORT

CASE REPORT OF ATYPICAL PRESENTATION OF SSPE WITH ADEM

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ABSTRACT

Sub-acute sclerosing panencephalitis (SSPE) is chronic encephalitis of childhood and young adolescents due to persistent measles virus infection of the central nervous system. In majority of cases, onset occurs from 5-15 yrs of age. In a non-immunised population, the average onset is 8 yrs. SSPE generally occurs 5-10 years after measles infection. In the early stage of the disease, behaviour and personality changes is followed by myoclonic jerks and convulsion. In late stages dementia, stupor and coma develops. Diagnosis is achieved by typical clinical finding, increase measles antibody titre in cerebrospinal fluid and serum, high amplitude slow sharp waves in EEG. Prognosis is poor and death ensues in about 3 yrs. after the diagnosis. 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 ADEM. Effective immunisation against measles is the only solution presently available to the problem of this dreaded disease.

CASE REPORT

A 10 year old boy admitted in our hospital with complaints of involuntary jerky movements of right upper limb, with drop attacks while walking, convulsion, followed by stupor. In past, patient was admitted in private hospital in 2009 for fever & convulsion, diagnosed as febrile convulsion & no anticonvulsants were started. Thereafter, in 2014, patient was having involuntary movement of right upper limb starting from thumb gradually involving the right upper limb lasted for a month and had convulsion two episodes in 15 days with altered consciousness.

CT Brain was done at first which showed S/o enhancement of leptomeningitis. Then after 1 month, MRI Brain was done which showed S/o acute demyelinated encephalomyelitis and patient was advised pulse therapy of steroid but they didn't take the treatment. This time on admission in our hospital, we had a 30kg boy, 140cm in height, vitals normal. CNS examination revealed Babinski positive, with hypertonia in right half of the body with all reflexes absent. CBC, electrolytes, renal function test, Liver function test were within normal range. Chest X Ray was also normal. MRI was done at our hospital showed s/o ADEM. Hence pulse therapy with methyl prednisolone was given but there was no improvement in condition. Also, anticonvulsants like sodium valproate and

phenytoin were given but no improvement in jerks, so, Tab Clobazam and Tab Oxcarbazepine were started.

We also gave immunoglobulin for 5 days, but there was no improvement in condition of the patient. 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.

In CSF examination total cells were 05, all lymphocytes, sugar 122, protein 5, culture showed no growth. CSF anti-measles antibody IgG and serum IgG level showed s/o sub-acute sclerosing panencephalomyelitis.

In CSF, Serum IgG Measles- 9227.5 U/ml; CSF IgG Measles- 15151.6 U/ml; Serum total IgG – 2460 mg/dl; CSF total IgG – 8.49 mg/dl; CSF/Serum quotient reference – POSITIVE (2.35)

From this report the diagnosis of SSPE was confirmed.

In EEG, the wave pattern 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) s/o SSPE.

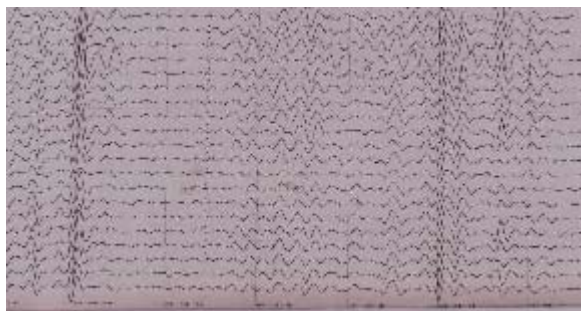


Fig1: EEG pattern of the patient

DISCUSSION

SSPE usually runs a relentlessly progressive clinical course, resulting in death in most cases within 1 to 3 years after diagnosis⁴. 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 course⁵. 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. As rare complication of measles, SSPE is difficult to diagnose & stamp. In our case, patient presented with convulsion, abnormal involuntary myoclonic jerky movements, confusion, stupor, with MRI brain s/o changes of ADEM.

There was a past history of measles infection and patient was unimmunised. Initially pt didn't respond to all treatment given as for ADEM. After the CSF antimeasles antibody report, we can make out the diagnosis of SSPE as atypical rare presentation with ADEM.

As it is the sequel of measles infection, carries poor prognosis. Measles continues to be a major cause of childhood morbidity and mortality in India. Recent studies estimated that 80,000 Indian children die each year due to measles and its complication, accounting to 4% of under 5 death. Hence, SSPE continues to be one of the commonest cause of progressive myoclonia with cognitive decline in

developing countries with incomplete measles immunization coverage. 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. Sedar carmet, Ayca vitrinel, Huzim Alper Gursu, Neslihan Cicek Deniz and Yerseamin Amin Dr. Lutki Kirdch Kartal Education and Research Hospital, Istanbul, Turkey Subacute sclerosing panencephalitis presenting as acute demyelinating encephalomyelitis Indian J Pediatr. 2006 Dec; 73(12):1119-21
2. Garg V.K.*, Wadhvani P., Sengar G.S., Chahar C.K. Department of pediatrics, S. P. Medical College. A Case Report Atypical presentation of Subacute Sclerosing Panencephalitis: IJSAR, 1(2), 2014; 41-44
3. Subacute sclerosing panencephalitis R K Garg Postgrad Med J. 2002 Feb; 78(916): 63-70. doi: 10.1136/pmj.78.916.63 accessed on 26th sep 2015.
4. Kliegman, R.M., Behrman, R.E., Jenson, H. B., Santon, B.F. Nelson (2011). Text book of Pediatrics. 19th edition. Philadelphia: Saunders. Page no. 1072-1073
5. Dyken PR. Subacute sclerosing panencephalitis: current status. Neurol Clin 1985; 3:179-196.
6. Honarmand S, Glaser CA, Chow E, et al. Subacute sclerosing panencephalitis in the differential diagnosis of encephalitis. Neurology 2004; 63:1489-1493.
7. Garg RK, Karak B, Sharma AM. Subacute sclerosing panencephalitis. Indian Pediatr 1998; 35: 337-344.
8. Campbell C, Levin S, Humphreys P, Walop W, Brannan R. Subacute sclerosing panencephalitis: results of the Canadian Paediatric Surveillance Program and review of the literature. BMC Pediatrics 2005; 5: 47.
9. Salehi HR, Delgado E, Wolf SM, McGoldrick PE, Moshé SL. Subacute sclerosing panencephalitis with atypical features. Pediatr Neurol 2005; 33: 280-282.
10. Gascon G, Yamani S, Crowell J, et al. Combined oral isoprinosine-intraventricular α -interferon therapy for subacute sclerosing panencephalitis. Brain Dev 1993; 15: 346-355.
11. P. Singhi, A. G. Saini, and J. K. Sahu. Nonconvulsive status epilepticus on electro encephalography: an atypical presentation of subacute sclerosing panencephalitis in two children. Case Reports in Pediatrics, vol. 2012, Article ID 374232, 3 pages, 2012.