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SSPE–A Rare Complication of Measles with Short Onset Latency

Dr.Shrikant S.Patil, Dr. Sanjay Joshi,Dr.VilasWadile.
Department of paediatrics, acpm medical college, Dhule. (maharashtra)

Abstract:*SSPE is a rare complication of measles which commonly manifests after about 7-10 years after the initial measles attack.Presenting case which presented to us at 3.5 year with some atypical features of sspe such as non responding focal seizures and rapid progression in about 2 month with short onset latency.*

I. Introduction

SSPE (SubacuteSclerosingPanencephalitis) is the result of a chronic measles infection. SSPE is achronic and slowly progressive inflammatory disease of the central nervous system caused by a persistent measles virus usually affecting the childhood and adolescent age group. Typically there is a primary measles infection at very early age, often before 2 years, followed by 6- to 8 years asymptomatic period.Clinical features at onset are very subtle and nonspecific. The illness evolves in several stages. Thetypical clinical course of SSPE is characterized by intellectual deterioration, personality and behavioralchanges, myoclonic jerks, visual disturbances and sometimes pyramidal and extrapyramidal symptoms.

II. Incidence

SSPE has an annual incidence from under 0.1 cases to 5 or 6 cases per million in nonimmunized populations. In areas of high early-life measles attack rates, SSPE accounts for a proportion of childhood neurodegenerative diseases.

III. Case report

3.5 year old male child a product of normal full term pregnancy and nonconsanguinousmarriage.with history of measles at the age of 9 months of life and prior to which was vaccinated for measles.Presented to us with complaints of frequent falls since 2 months before admission and had rapid progression and signs of neuroregression. Patient also developed convulsionssubsequentlypatient also developed myoclonic jerks with head drop at regular intervals and also loss off bladder and bowel control recently. On clinical examination vitals were stable had mild pallor was bedridden and restless with abnormal movements. Childhad intellectual deterioration , speech and behavioral abnormality and unable to recognize the relatives, power was reduced in all four limbs and DTR were exaggerated and extensor plantar reflex. Cranial nerve examination was normal.

Investigations revealed normal serum electrolytes, CSF examination and routine tests. CSF-IgG for measles showed raised total IgG in CSF. EEG suggests periodic sharp complexes and overlapping on each other.MRI showed periventricular hyperintensities near posterior horn of lateral ventricles.

IV. Discussion

SSPE is a slowly progressive disease characterized by seizures and progressive deterioration of cognitive and motor functions.The diagnosis of SSPE can be reliably established if at least three of the following five criteria are met:(a) Progressive sub-acute mental deterioration with typical signs like myoclonus;

(b) Periodic, stereotyped, high voltage discharges on EEG; (c) CSF globulin levels greater than 20% of total CSF protein; (d) Raised titres of measles antibodies in blood and/or CSF in the absence of other antibodies, including against Herpes simplex virus (HSV) and Varicella zoster virus (VZV); and (e) Typical histopathological findings on brain biopsy or autopsy. This patient meets criteria a,b,c and d and so no need for brain biopsy was felt. The pathogenesis of SSPE is related to defective measles virus maturation in neural cells. Aberrant M (matrix) proteins as well as other envelope proteins interfere with assembly and budding of infectious virus. The virus remains in intracellular form and spreads by cell to cell contact.

V. Conclusion

Hence we diagnosed a case of SSPE with atypical presentation on the basis of history, examination and investigations.