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Case Report of Subacute Sclerosing Panencephalitis

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ABSTRACT
BACKGROUND: Subacute Sclerosing Panencephalitis(SSPE) is a chronic complication of Measles with a delayed onset and an outcome that is nearly always fatal. It appears to result from a persistent infection with an altered Measles virus that is harboured intracellularly in Central Nervous System (CNS) for several years. After 7-10 years the virus apparently regains virulence and attacks the cells in CNS that offered the virus protection. **CASE REPORT:** A 6 year old girl was admitted at our tertiary care hospital with the complaints of inability to speak, to sit and walk without support, decreased appetite and excessive cry since past 5-6 days before admission. Her mother gave past history of Measles infection. Cerebro-Spinal Fluid Measles IgG titre was positive. **DISCUSSION:** Diagnosis of SSPE can be established through documentation of a compatible clinical course and atleast one of the following supportive findings: 1) Measles antibody detected in CSF 2) Characteristic EEG findings 3) Typical histological and/or isolation of virus or viral antigen from brain tissue obtained by biopsy or post-mortem examination. **CONCLUSION:** Clinical Manifestations of SSPE begin insidiously 7-13 years after primary Measles infection. Subtle changes in behavior or school performance appear, including irritability, reduced attention span. and temper outbursts.

KEYWORDS: Subacute Sclerosing Panencephalitis, Measles, Electroencephalogram, Interferon, Antibody titre

I. INTRODUCTION:

Measles is still a common communicable disease, particularly in Africa and Asia. Subacute Sclerosing Panencephaltis (SSPE) is a progressive neurological disorder caused by persistent Measles virus infection; characterized by progressive mental decline, myoclonus and raised anti-measles titre in CSF[1]. Electroencephalography (EEG) in SSPE characteristically reveals generalized periodic complexes or discharges. The latent period between Measles and SSPE is commonly 6-8 years.

II. CASE REPORT:

A female child, 6 years old, was admitted at our tertiary care hospital with the complaints of inability to speak; to sit and walk without support along with the complaints of decreased appetite and excessive cry since 5-6 daybefore admission. The routine investigations sent which were within normal limits. Her mother gave history of fever followed by maculopapular rash at a time well before the patient receiving his scheduled MMR vaccine around 9 months of age. Measles IgM and IgG assays were ordered when Measles IgG was found to be detectable in the blood serum. Cerebrospinal fluid tapping and examination done where CSF microscopy showed exclusive lymphocytes with 5 counts per high power field. Also CSF total protein and glucose were within normal limits. Neurophysician opinion obtained and advised Magnetic Resonance Imaging (MRI) Brain and EEG. MRI Brain SOS Contrast study didnot demonstrate any brain abnormality at that point of time. According to the clinical history the course suggested regression of developmental milestones in this patient. During the later part of her stay, after about 10 days, the patient started having tremor-like movements in her right upper and lower limbs which lasted about 10 sec and appeared intermittently throughout the day. An EEG was done to rule out the possibility of non-convulsive status epilepticus. A possibility of SSPE was considered and hence, repeat CSF tapping done to detect Measles CSF IgG. Upon recieving the result for CSF/Serum quotient reference for Measles, it was found out to be positive suggesting the confirmation of diagnosis. Repeat Neurophysician opinion obtained, relatives were counseled about the condition, danger signs explained and patient has been advised for monthly follow-up.

III. DISCUSSION:

Most of the patients with SSPE have a history of primary Measles infection at an early age. Children infected with Measles under age of one year carry 11 times greater risk of SSPE than those infected at age 5 year or later. Diagnosis can be established through documentation of compatible clinical course and atleast one of the following finding:1) Measles antibody detected in CSF 2) Characteristic EEG findings and 3) Typical histological findings in and/or isolation of virus or viral antigen from brain tissue obtained by biopsy or Post-Mortem examination. Latent period between Measles infection and SSPE is around 6-8 years in most of cases[2], but may range between 3 months -18 years. In this child, latent period of approximately 5 years was noted. The initial phase (stage 1) may at times be missed; fever, headache and other signs of encephalitis are absent. The hallmark of 2nd stage is Myoclonus. Involuntary movements and repetitive jerks begin in single muscle groups. Consciousness is maintained. In the 3rd stage, involuntary movements disappear and are replaced by choreoathetosis, immobility, dystonia and lead pipe rigidity. The sensorium deteriorates into dementia, stupor and then coma. The 4th stage is characterized by loss of critical centres that support breathing, Heart rate and blood pressure[3]. No curative treatment is available for SSPE but therapy with immunomodulator and interferons; and antiviral drugs like Ribavirin may help in halting the progression of disease.

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