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Sub-Acute Sclerosing Panencephalitis Manifesting as Psychiatric Illness: A Case Report

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Abstract **Case Report**

Subacute sclerosing panencephalitis (SSPE) is a chronic progressive inflammatory central nervous system disorder manifesting as a rare complication of measles viral infection in childhood resulting in fatality in all cases. It is likely to have possibility of subclinical or undiagnosed measles in early childhood in absence of prior overt measles infection. Measles vaccine has a protective effect against SSPE and high vaccination gap is associated with higher incidence of SSPE. Children residing in areas with poor vaccination coverage are at increased risk of developing the disease. It is characterised by progressive intellectual deterioration, seizures, myoclonus, ataxia and visual disturbances. The onset of SSPE is insidious and can manifest as significant psychiatric ailments. A 17 years old girl with history of progressive scholastic deterioration for a year presented with focal seizures, myoclonic jerks and psychiatric manifestations like emotional outburst followed by progressive unresponsiveness, altered sensorium and memory lapses since two weeks. A diagnosis of Subacute sclerosing panencephalitis (SSPE) was made on the basis of clinical symptoms, typical EEG changes and presence of anti-measles antibody in cerebrospinal fluid. Patient received treatment for myoclonic jerks, spasticity along with supportive care. SSPE is often a chronic inflammatory and life threatening disease of central nervous system with no cure despite development of antiviral and immunomodulator drugs. There is urgent need to advocate policies to enhance vaccination coverage and for extensive research in field for potential therapies for treatment of such patients to improve clinical outcomes.

Keywords: Subacute sclerosing panencephalitis (SSPE); Measles; EEG.

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a chronic progressive, catastrophic and inflammatory disease of the whole brain caused by a persistent mutant measles virus infection [1]. The disease is characterised by its subacute onset, sclerotic lesions and whole brain involvement [2]. The latency period for onset of SSPE is approximately 9.5 years (2.5-34 years) following the measles infection and fatality can occur within 1-3 years of diagnosis [3]. Higher risk of SSPE was found in males as compared to females [5].

Early clinical course of disease is characterised by memory loss, behavioural changes, irritability followed by myoclonic seizures, mental changes, whereas in late stages dementia, stupor, hypothermia, abnormalities in vital signs or vegetative state can occur [2]. The occurrence of SSPE is found to be more in developing countries due to poor vaccine coverage, suboptimal cold chain maintenance and spread of atypical measles virus strain [4]. In India, incidence rate of SSPE is around 21 cases/million population [4].

Virus causing measles is a paramyxovirus with 22 genotypes and most of them are associated with endemic and epidemic outbreaks [6]. Virus usually enters into the brain through olfactory bulb, viral replication within capillary endothelial cells and infected peripheral blood monocytes [5]. Autoimmune mediated demyelination of white matter can occur due to cross reactivity of reactivated mutant virus with myelin antigens.

CASE REPORT

We present the case of a 17 years female adolescent admitted from Emergency room in a serious condition with complaints of intermittent vomiting and fever since one month. She belonged to a Muslim family with poor socioeconomic background born out of consanguinity as eldest of seven siblings. She had a history of abnormal jerky and dystonic movements of

both upper limbs, which was gradually increasing in frequency with prolonged duration for 2 weeks prior to hospitalization. She developed ataxia, frequent falls, followed by difficulty in walking and sitting along with stiffness of body. She had history of developing an injury on face after falling from bed following a seizure. She had progressive decline in cognition with waxing and waning sensorium.

There was no history of rash, jaundice, joint pain or insect bite noticed by parents but she had decrease in oral intake and had bowel and bladder incontinence. She was an unimmunised girl with normal development prior to illness and uneventful perinatal background except for poor scholastic performance. There was history of uncle death due to tuberculosis one year back in the family and there was no family history of seizure or mental subnormality. There was history of gradual deterioration in her behavior, social withdrawal and school dropout since one year. She was reported to have irrelevant talk, laughter episodes from last 2 weeks for which she was shown to a psychiatrist outside and was started on oral risperidone. In view of progressive uncontrolled dystonic movements, motor weakness and slurring of speech she was taken to various doctors but she showed on response.

On examination she was poorly nourished with BMI less than 3rdpercentile with no pallor, icterus, cyanosis or lymphadenopathy. She was drowsy with Glasgow Coma Scale score 13/15 and had no eye contact and was not oriented to time, place and person. She had numerous myoclonic jerks associated with momentary disturbances in consciousness. Examination of fundi was normal and pupils were bilaterally equal and reacting to light. Neurological examination showed neck rigidity, generalized hypertonia, clasp knife rigidity, brisk deep tendon reflexes with bilateral plantar flexor response. Her sensory examination was normal while motor examination revealed a power of 1/5 in lower limbs and 2/5 in upper limbs. There were no cranial nerve deficits and she had normal gag reflex. There was no tremors, choreoathetosis, tics or fasciculation .Systemic examination was normal. On the basis of history and clinical findings, a provisional diagnosis of tubercular meningoencephalitis, autoimmune encephalitis, viral encephalitis, SSPE (although patient had no history of measles in childhood), neurometabolic encephalopathies and neurosyphilis was considered. Initial investigations like complete blood count, blood glucose levels, electrolyte levels, hepatic function test, and renal function tests were normal. CSF examination showed normal findings with normal cell counts. The results of widal, malarial antigen test, leptospira rapid test, CSF-Acid Fast Bacillus, CSF CBNAAT, markers for viral infection (Hepatitis B, Hepatitis C, HIV) were negative. Thus diseases like typhoid, malaria, tuberculosis, and leptospirosis were ruled out. MRI neuroimaging was normal Test for autoimmune panel came out to be negative (ANA 17 profile: all negative, C3: 163mg/dl, C4: 56mg/dl, ACE: 131U/L). Her initial EEG examination was normal.

She was started on IV antibiotics (Inj Ceftriaxone, Inj Vancomycin), IV fluids and other supportive care. Phenytoin was administered parentrally for the treatment of myoclonic jerks. During hospital stay she had a waxing and waning course and remained delirious with intermittent myoclonic jerks in view of which EEG was again repeated, that showed findings suggestive of two differential diagnoses: SSPE and myoclonic status epilepticus. The findings observed were periodic sharp and slow wave discharges in left frontal field at a frequency of 1-2/second. EEG also showed bilateral frontal low amplitude poly spikes lasting only for one second (time locked myoclonic jerks). Injection valproate 2 g/day and clonazepam 0.5mg three times a day were also added in view of persistent jerky movements. To confirm the diagnosis, CSF examination was repeated. CSF and serum samples were sent for measles antibodies. CSF-Measles-IgG antibodies detection was based on the principle that various CNS infections are characterized by humoral immune response with IgG predominance produced by memory B cells. A CSF/Serum Quotient reference of >1.5 was indicative of measles-specific antibody production in the CNS (Table no. 1). Thus, based on history, clinical examination and laboratory investigations, a final diagnosis of Subacute sclerosing panencephalitis (SSPE) was made.

With treatment, her general condition showed not much improvement. She had improvement in spasticity but had persistent bizarre behaviour with myoclonic jerks and weakness of limbs. Her condition deteriorated, could not recognise her parents, became completely akinetic and mute but parents took her home against medical advice and were not willing for any advanced treatment.

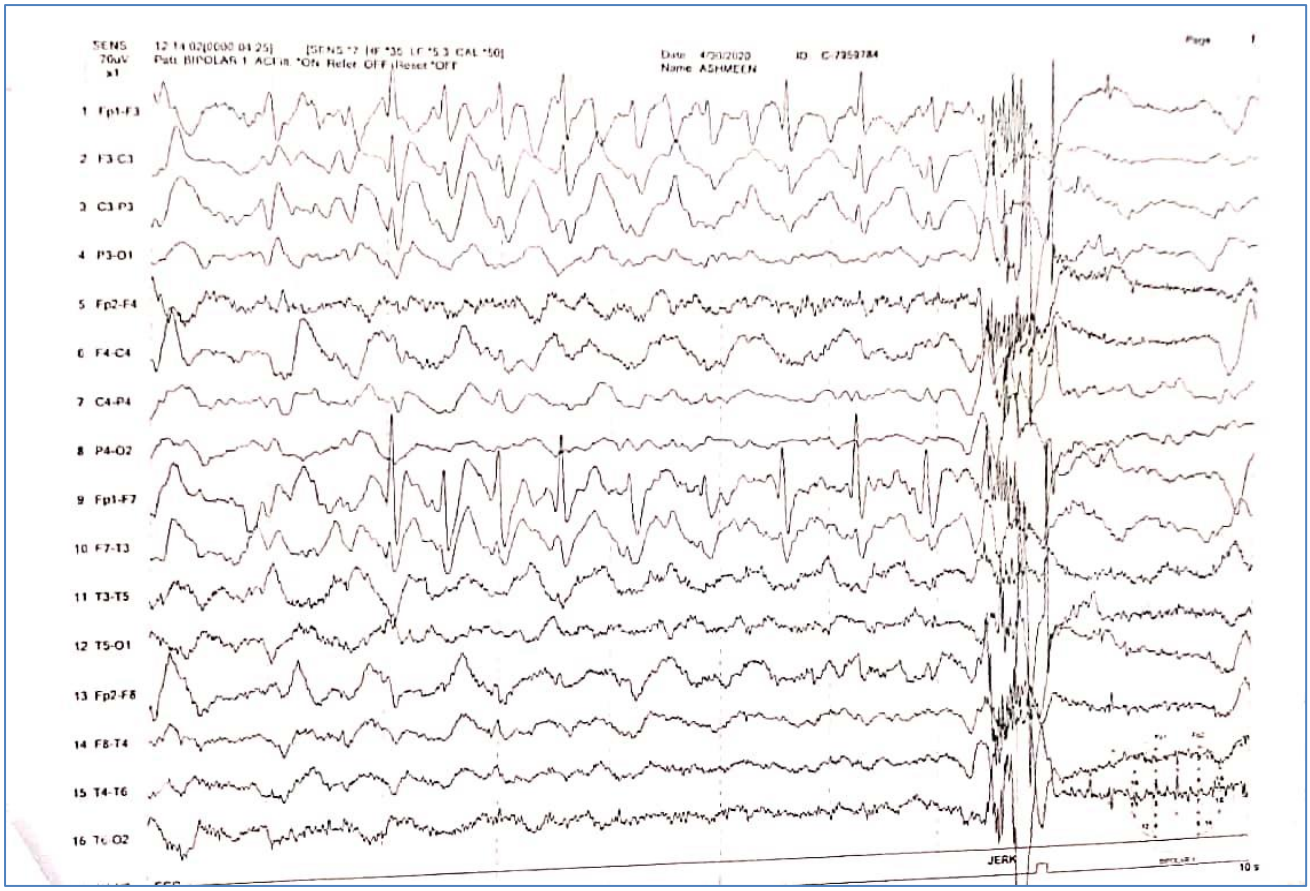

Fig-1: Figure showing findings in EEG of patient

Table-1: Serum and CSF - Measles (Rubeola) - IgG Antibodies by ELISA

	Serum IgG Measles U/ml	CSF IgG Measles U/ml	Serum Total IgG mg/dl	CSF Total IgG mg/dl	Relative CSF/ Serum Quotient
Results	6486	11845.6	1220	5.51	7.77

DISCUSSION

SSPE is a chronic CNS complication of rubeola infection and it is usually diagnosed in age group of 3-35 years [3]. We report a case of 17 years old female belonging to a low socioeconomic family, who because of emotional outburst was receiving psychiatric treatment from last two weeks. Because of initial atypical and nonspecific manifestations of SSPE like behavioural problems, cognitive decline, mood changes and absence of overt measles infection in the past, SSPE is misdiagnosed and often not suspected clinically [3].

The characteristic ocular finding of SSPE is choreoretinitis involving macular region which has been reported but was absent in our patient [5]. For diagnosing SSPE,according to Dyken'scriteria, it requires presence of at least three of the five criteria: a) progressive subacute mental deterioration with signs of myoclonus, b) characteristic periodic high voltage EEG discharges, c) elevated CSF globulin levels greater than 20% of total CSF protein, d) raised titres of measles immunoglobulins in blood (>1:256) and CSF (>1:4) or oligoclonal bands and e) typical histopathological findings in brain biopsy (shows panencephalitis) [7]. We also diagnosed the case as SSPE, based on the above criteria with the presence of positive clinical history (mental and behavioural changes, myoclonic jerks); characteristic changes in EEG; and raised measles IgG levels in serum and CSF. Jabbour *et al.* have divided the clinical manifestations into four stages depending on the progression of neurological symptoms in patient. Stage I is characterized by behavioural as well as intellectual disorientation (irritability, dementia, social withdrawal, lethargy, and regression of speech); Stage II shows various types of movement disorders such as dyskinesia, dystonia, and myoclonus; Stage III is consistent with extrapyramidal symptoms, decerebrate posturing, and spasticity; and Stage IV is characterized by chronic vegetative state and autonomic changes [8]. She came to our hospital in progressive Stage II. Although MRI brain is more sensitive in detecting grey and white matter abnormalities and to exclude other demyelinating lesions but it was normal in our patient [9]. Treatment of SSPE is still under research phase. Supportive care is the mainstay of treatment which includes management of seizures and other complications. Antiviral agents such as

amantadine and ribavirin or immunomodulators such as Isoprinosine, interferon, and immunoglobulin has been tried [5]. We also subjected our patient to supportive treatment with administration of anticonvulsant, benzodiazepines, antibiotics along with IV fluids and multivitamins as she was already in stage II. There was rapid deterioration in clinical stages but parents took child home against medical advice. Thus a high index of suspicion is needed to detect SSPE with atypical presentation.

CONCLUSION

The present case revealed a case report of patient suffering with SSPE stage II who presented with emotional outburst, scholastic as well as personality deterioration and was started on antipsychotics from outside. A careful insight is required to diagnose such cases and progressive nature of illness has to be considered. The diagnosis is mostly based on clinical suspicion, supported by findings of electroencephalography (periodic complexes), brain imaging (demyelination) and immunological tests for diagnosis of measles infection. Management of SSPE includes effective treatment for seizure control and prevention of further complications associated with the progressive disability. Specific therapy like trials of treatment with immunomodulators like Isoprinosine, antivirals such as ribavirin and using different methodologies have reported marginal effects with no definite cure. Thus universal vaccination against measles is the only proven way to prevent SSPE.

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