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Adult onset Fulminant form of Subacute Sclerosing Panencephalitis :a case report

KEYWORDS: Subacute Sclerosing Panencephalitis (SSPE) , Myoclonic jerks, Rademecker complexes, Dyken's criteria

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ABSTRACT *Subacute sclerosing panencephalitis (SSPE) is a progressive fatal neurological disorder caused by a persistent measles virus infection. It usually occurs in childhood and adolescence and may not be readily recognized when presenting later in life. The incidence of adult onset SSPE is reported to be between 1-1.75% and 2.6%1,2 . we report a case of 19 year old female patient with complaints of altered behaviour for 30 days and myoclonic jerks for 5 days. She had a past history of measles at the age of 5 years. Her MRI brain showed hyperintensities in bilateral parieto-occipital lobes and frontal lobe in subcortical location. Electroencephalogram showed rademecker complexes. Measles IgG antibodies were detected in CSF and serum. She was diagnosed with SSPE stage II. She deteriorated rapidly and succumbed to death in 40 days. This was an atypical presentation of SSPE with regards to age, rapid progression and normal fundus.*

CASE REPORT : A 19 year old female patient presented to the emergency department with the complaint of altered behaviour for 30 days in the form of decreased attention span, irritability, repeated unprovoked smiling, repeated unprovoked weeping, poor personal hygiene, passing urine in the clothes. She had history of involuntary movements since 5 days from presentation in the form of sudden jerky movements of all the four limbs more of upper limbs occurring at a rate 8 to 10 times per minute not associated with loss of consciousness and frequency diminished during sleep. She had a history of complicated exanthematous viral illness at the age of 5 years which was associated with high grade fever, running nose, redness of eyes, watering of eyes, and rash all over the body. This illness lasted for around 9 - 10 days. Following that she developed productive cough, dyspnoea and fever for which she was hospitalized for around 10 days. It was presumed to be measles. Her body weight is 39kgs, height 148cm, pulse 86/min, blood pressure 100/50 mmHg, axillary temperature 36.6^0 C, respiratory rate 22/min. On examination she had hypotonia, myoclonic jerks and jaunty or apraxic gait. Other systemic examination findings were found to be normal. Her fundus examination was normal. Complete blood picture, erythrocyte sedimentation rate, serum electrolytes, liver function tests and renal function tests were normal. MRI brain showed evidence of T2 weighted and FLAIR hyperintensities in bilateral parieto-occipital lobes and frontal lobe in subcortical location which was suggestive of subacute sclerosing panencephalitis. [Figure 1 here]

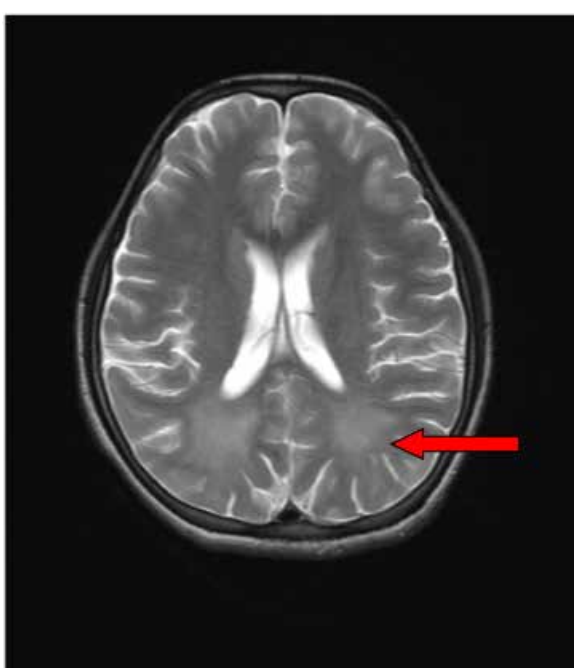

Figure 1 MRI Brain showing hyperintensities in bilateral parieto-occipital regions

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EEG showed RADEMECKER COMPLEXES which consists of high voltage 300 to 1500V repetitive polyphasic and slow waves lasting 0.5 to 2 seconds and occurring regularly at every 4 to 15 seconds. Cerebrospinal fluid [CSF] analysis was done. CSF was clear, there were 4 cells with 100% lymphocytes. CSF protein was 38mg% and sugars 80mg%. Oligoclonal bands were detected in the cerebrospinal fluid which is suggestive of intrathecal IgG synthesis. CSF and serum were tested for measles IgG antibodies by Enzyme Immunoassay. CSF measles IgG was 13978U/ML, serum measles IgG levels were 2672U/ml. total CSF IgG levels were 7.60mg/dl and total serum IgG levels were 1900mg/dl. CSF/serum quotient reference was 6.4 [normal <1.3].

TEST DESCRIPTION	OBSEERVED VALUE	REFERENCE VALUE
Serum IgG measles	2672U/l	-
CSF IgG measles	13978U/l	-
Serum total IgG	1900mg/dl	700 – 1600mg/dl
CSF total IgG	7.60mg/dl	0 – 3.4mg/dl
CSF/serum quotient reference	6.4	Normal < 1.3 Equivocal 1.3 – 1.5 Positive > 1.5

Table 1 : CSF analysis (for measles IgG antibodies by EIA)

Thus she was diagnosed with subacute sclerosing panencephalitis. She was in stage II according to Risk and Haddad staging. The treatment for SSPE is intrathecal interferon alfa and oral isoprenosine[3]. But due to non-availability of drug she was treated with anticonvulsants and supportive care. She deteriorated rapidly and succumbed to death in the next 40 days.

DISCUSSION : Subacute sclerosing panencephalitis is a rare neurodegenerative disorder caused by a persistent mutated measles virus infection. The latent period between measles infection and SSPE is commonly 6 - 8 years. SSPE is a disease of childhood and adolescence commonly occurring between 5 - 15 years of age. It has a gradually progressive course leading to death in 1 - 3 years. It has a slight male preponderance.

Measles is caused by an RNA virus belonging to the genus Morbilivirus and the family paramyxoviridae. SSPE results when extensive point mutations occur in the viral genome resulting in persistent viral infection. Changes in the Hemagglutinin [H] , fusion [F] and matrix [M] are also associated with persistent infection. These proteins are required for viral budding from infected cells and the putative fusion with uninfected cells. Measles virus reaches the brain during acute exanthem phase by infection of cerebral endothelial cells.

The initial manifestations include mild intellectual deterioration and behavioural changes. Later patients have periodic stereotyped myoclonic jerks involving head and subsequently trunk and limbs. Myoclonic jerks do not affect the level of consciousness, exaggerated by excitement and may disappear during sleep. Few patients may develop pyramidal and extrapyramidal signs. Visual disturbances may occur in the form of cortical blindness, chorioretinits and optic atrophy. Patients gradually deteriorate to a comatose state and may develop decerebrate or decorticate rigidity and ultimately death ensues.

The diagnosis of SSPE can be reliably established if the patient fulfills three of the five criteria given by Dyken[4] . (Table 2 here)

Clinical	Progressive subacute behavioural deterioration with typical signs like myoclonic jerks
Electroencephalogram	Periodic, stereotyped, high voltage discharges
Cerebrospinal fluid	Raised gammaglobulins or oligoclonal bands
Measles antibodies	Raised titre in serum and/or CSF
Brain biopsy	Suggestive of panencephalitis

TABLE 2 : Dyken's Diagnostic Criteria for SSPE

Currently there is no adequate therapy for SSPE. According to some non-randomized trials certain antiviral drugs and immunomodulators can prolong life if longterm treatment is given. Combination of intraventricular interferon alfa and oral isoprenasine[3] is currently the treatment given for halting the progression of the disease.

This case was an adult-onset fulminant form of SSPE with delayed onset , rapid progression and normal fundus which is a rare presentation.

REFERENCE 1. Subacute sclerosing panencephalitis in the Middle East: report of 99 cases. Haddad FS, Risk WS, Jabbour JT Ann Neurol. 1977 Mar; 1(3):211-7. 2. . Yalaz K, Anlar B, Renda Y. et al Subacute sclerosing panencephalitis in Turkey: epidemiologic features. 1st meeting of the Association of Mediterranean Child Neurology. Crete, Greece: June 8–11, 1987 3. Gokcil Z, Odabasi Z, Demirkaya S, et al. α-Interferon and isoprinosine in adult-onset subacute sclerosing panencephalitis. J Neurol Sci1999;162:62–4. 4. Dyken PR. Subacute sclerosing panencephalitis. Neurol Clin1985;3:179–95.