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A RARE CASE OF SUBACUTE SCLEROSING PANENCEPHALITIS IN ADULT

General Medicine

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a rare progressive neurological disorder of early adolescence caused by persistent infection of the measles virus, which remains prevalent worldwide despite an effective vaccine. SSPE is a devastating disease with a characteristic clinical course in subcortical white matter. Typically, measles is self-limiting; however, measles may result in infrequent complications involving the CNS. SSPE is a fatal long-term complication of measles infection, caused by the intracerebral spread of the measles virus leading to neuronal destruction. The initial symptoms of SSPE are subtle and include mild mental deterioration (such as memory loss) and changes in behavior followed by disturbances in motor function including uncontrollable involuntary jerking and slowly progress to spasticity ,ataxia ,blindness and persistent vegetative state . The latency period between acute measles and the first symptoms of SSPE is usually 4 to 10 years, but can range from one month to 27 years .Most patients survive for one to three years after diagnosis, with a mean survival of about 18 months

KEYWORDS

measles, neurodegeneration, subacute sclerosing panencephalitis. Myoclonic jerks Progressive deterioration

INTRODUCTION

Subacute Sclerosing Panencephalitis, a serious disorder of Central Nervous System is a slow viral infection, caused by defective measles virus. It has an incidence of 1:1,00,000 cases, usually affecting children(10-14 year). There is a higher incidence among males than females(M/F:3/1). Most children with SSPE have a history of measles infection at an early age of less than 2 years, followed by latent period of 6-8 years before development of neurological symptoms.[1]

SSPE has been reported from all parts of the world, but in the West it is considered a rare disease with fewer than 10 cases per year reported in the United States. The reported frequency of SSPE in the United States was approximately one per million childhood population from 1960 to 1978 The incidence declined substantially after introduction of an effective measles vaccine. The annual incidence of SSPE is still quite high but variable among developing countries. Saha et al reported an annual incidence of 21 per million population in India, in comparison with 2.4 per million population in the Middle East.[2]

SSPE is progressive neurological disorder of children and young adults that affects central nervous system. It is slow but persistent viral infection caused by defective measels virus.It is single stranded RNA virus of the paramyxoviridae family .[4]

Vaccination implementation has been effective in reducing the number of the measles cases ,thereby reducing the cases of SSPE .

Case presentation

A 31 year old male patient school teacher presented to us with chief complaints of rapidly progressive painless vision loss for 2 months, rapidly progressive dementia for 1 month and occasional muscle twitch in both arms for 15days.

The patient was apparently normal before 2 months ,after which he developed painless insidious onset difficulty in vision, simultaneously starting in both eyes and then followed by complete vision loss in both the eyes After 1 month, relatives noticed rapidly progressive change in his behaviour and memory issues in form of irritability, outburst of angerness, easy distractibility and forgetfulness and followed by relatives noticed involuntary movements in form of muscle twitch in both the arms .

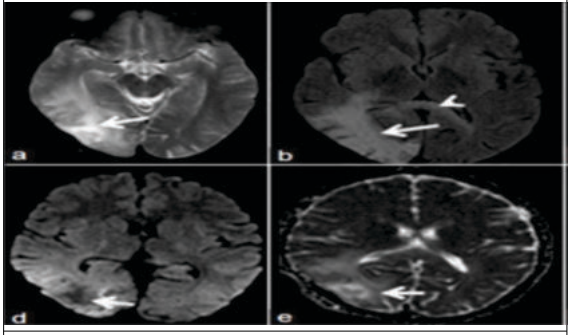
Patient had significant past history of measles at the age of 1 year which was recovered .There was no other significant Personal or Family history. Pt was on mixed diet with normal bowel,bladder and sleep pattern.

On admission pt was vitally stable with MMSE score 9/26,conscious and oriented to time only and rest of the general examination was normal. On systemic neurological evaluation- the Cranial Nerves and sensory system examination were not elicited because of MMSE score of 9/26.Hypertonia in form of spasticity and DTR-symmetrical and exaggerated, extensor planter. Myoclonic jerks were found typically involving upper extremities (deltoid) with spastic gait.Rest of systemic examination was unremarkable.

On admission patient was evaluated for detailed blood and radiological investigations.

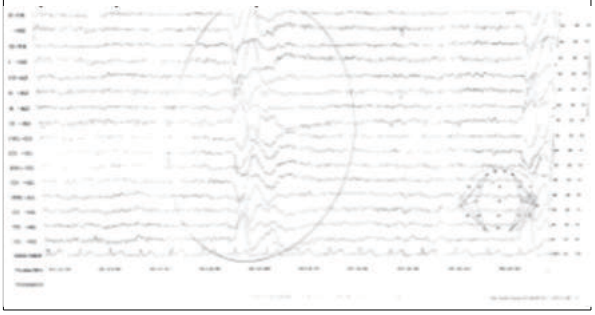
Blood Investigations:

Complete blood count	Comprehensive metabolic panel
White blood cell: 6960/uL (4000-10000)	Sodium: 139 mEq (136-145)
Hemoglobin: 14.9 g/dL (13-16)	Potassium: 4.5 mEq (3.5-5.0)
Platelets: 179000/uL (150000-350000)	BUN: 29.5 mg/dl (8-20)
	Creatinine: 0.8 mg/dl (0.7-1.3)
Differential count:	Calcium: 8.95 mg/dl (8-9.5)
Neutrophils: 67% (42-72)	Alkalaine phosphatase: 106 U/L (36-92)
Lymphocytes: 21% (25-45)	ALT:10.9 U/L (0-35), AST:30.5 U/L (0-35)
Monocytes: 8% (0-10)	T. Bilirubin: 1:2 mg/dL (0.3-1.2)
OTHER SPECIFIC INVESTIGATIONS:	
MRI Brain: Small Focal non enhancing abnormal signal intensity involving Right Occipital and Sub cortical White Matter. Imaging may represent Brain Edema/Focal Encephalitis	



CSF-R/M: Opening Pressure-140mmHg, Cells-05(100% Polymorphs), Protein-45, Sugar-53.
CSF-Measles IgG Index: 62(Normal <9)

EEG: Generalised High Voltage Periodic Long Duration polymorphic Delta waves with superimposed sharp waves s/o SPPE



Patient was treated with Intravenous Immunoglobulin 140gm and started on antivirals (T.Ribavarin and T.Isoprinosine) and other symptomatic treatment were given (anticonvulsants medications). Despite of the aggressive treatment , there is no significant outcome happened in patient and he died within 2 months.

DISCUSSION:

SSPE is a progressive illness leading to severe neurological deficits and death. A high index of suspicion is must for clinicians to diagnose this condition.[3]

SSPE virus differs from wild measles virus. Measles virus genome recovered from the autopsied brain tissues demonstrates clustered mutations in virus genome particularly in the M gene. These mutations destroy the structure and functioning of the encoded proteins. Complete infectious virus particle has rarely been recovered from the brain. Recent in vitro studies suggest that mutations in F protein confer hyperfusogenic properties to measles virus facilitating transneuronal viral spread. The inflammatory response in the brain leads to extensive tissue damage.[4]

During the initial stages, symptoms such as cognitive decline, myoclonus and behavioural abnormalties are seen. These may slowly progress to Spasticicty, Ataxia, Blindness and persistent Vegetative State.[5]

DIAGNOSTIC CRITERIA OF SSPE	
1. Clinical	Progressive, subacute mental deterioration ;with typical signs like myoclonus
2. EEG	Periodic, stereotyped, high-voltage discharges
3. Cerebrospinal fluid	Raised gamma-globulin or oligoclonal pattern
4. Measles antibodies	Raised titer in serum (1:256) and/or cerebrospinal fluid (1:4)
5. Brain biopsy	Suggestive of panencephalitis
Definitive: Criteria 5 with three more criteria; probable: Three of the five criteria	

Stages of SSPE:

Stage 1: Behavioural and Cognitive decline (lethargy, inattention or temper tantrums)
Stage 2: Myoclonic jerks, seizures and dementia
Stage 3: Rigidity, Extrapyramidal Symptoms, Progressive Unresponsiveness
Stage 4: Autonomic instability, akinetic mutism,Vegetative state and Coma.[4]

The classical presentation of SSPE is seen in the age group of 10-14 years. However, it has been documented that adult patient suffering from SSPE develops visual signs and symptoms followed by other typical manifestations.[4] But in our patient who was 31 years old male presented with first symptom of vision loss followed by other classical symptoms as above stage. Based on CSF, MRI and EEG, diagnosis of SSPE STAGE 2 was made and patients relatives were explained about the disease, its course and prognosis.

Prognosis:

SSPE is a progressive disorder and death usually occurs in 1–3 years. Apart from this classical course, a chronic very slowly progressive form, a very fulminant form leading to death in weeks, and a "stuttering" form of disease with remission and relapses, have been observed. Approximately 5% of the patients can have substantial spontaneous long term improvement. Spontaneous remission may occur during any stage of the disease and last for a variable period of time before eventual relapse occurs.The age of onset of SSPE less than 12 years, disappearance of periodic complexes, the tendency for normalisation of the background of follow up EEGs, and a progressive increase in measles antibody titres in cerebrospinal fluid are the factors that appear to be associated with favourable outcome in SSPE.[6]

CONCLUSION:

SSPE is an uncommon and fatal complication of childhood measles. It is a progressive and incurable condition resulting in death typically within one to three years of onset of symptoms. Clinical presentation widely varies ranging from progressive weakness, seizures, pyramidal and extrapyramidal symptoms, and coma. SSPE is still common neurodegenerative disorder despite increase in measles vaccination. Further studies and investigations are warranted to better understand the natural history and appropriate management of this condition.

SSPE is common in low-socioeconomic status. The profile of adult onset did not differ from childhood onset SSPE, except for a longer interval between measles infection and presence of the ophthalmic symptom as presenting feature in adult onset group.

The presence of SSPE in vaccinated patients indicates either previous subclinical infection prior to measles vaccination or poor maintenance of cold chain. SSPE is a devastating disorder that deserves elimination through the immunization of all children worldwide.

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