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Case Report
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A Rare Case of Adult-Onset Subacute Sclerosing Panencephalitis with Atypical EEG Finding

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Abstract

Subacute Sclerosing Panencephalitis (SSPE) is the chronic degenerative inflammation of the brain as a result of persistent measles infection. The onset of SSPE usually occurs in children less than one year of age and is comparatively rare in adults. The disease is fatal accounting for a mortality rate of more than 95%. It has a variety of clinical presentations ranging from behavioral change to coma, which makes the diagnosis challenging. A combination of investigational procedures like MRI, EEG, demonstration of elevated measles antibody titers in Cerebrospinal Fluid (CSF) and serum plays the key role to segregate from its other differentials. It is more common in developing nations due to scarce vaccination, owing to the lack of awareness and low socioeconomic status. Herein, we report a case of newly diagnosed SSPE in a 32-year-old female who presented with fever, altered sensorium, and recurrent seizure episodes. Adult-onset SSPE is very rare and there are very few case reports on it. Though the neuroimaging findings were normal, the EEG gave us a clue towards the diagnosis. It was later confirmed by measles antibody titers in CSF and serum. The patient was conscious and oriented on discharge. She was advised with interferon-2b and regular follow-up visits were scheduled for close monitoring of the symptoms.

Keywords: Subacute sclerosing panencephalitis; EEG; Adult-onset; Seizures

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Introduction

Subacute Sclerosing Panencephalitis (SSPE) is a slow, irreversible neurodegenerative sequela of a hypermutated wild measles virus infection [1]. Children under the age of 1 year have a higher risk of getting the disease when compared to adults. The risk of acquiring SSPE increases if the onset of measles is preferably at a younger age, in lower socio-economic class, poor immunity status, and with overcrowding [2]. The clinical characteristic of the disease is marked by behavioral change, focal or generalized seizure, ataxia which might progress to a persistent vegetative state. The incidence of SSPE in developed countries like the USA is less than 10 cases per 100,000 measles infections per year while it is as high as 210 cases per year in India [3]. The mortality rate of SSPE is 95% and most patients survive only 1 to 3 years after diagnosis [4]. The global prevalence of SSPE has diminished to less than 70 cases per 100,000 due to robust immunization coverage. Cases are underreported in many areas due to a lack of resources and awareness [5]. In the past few years, there has been a re-emergence of the measles virus in developed countries. This is significantly attributed to reduced vaccination due to vaccine hesitancy [6].

Case Presentation

A 32-year-old female with low socio-economic status presented to the emergency department with high-grade fever, chills, and altered sensorium. Her relatives gave a history of severe bilateral proximal myoclonic jerks followed by generalized tonic-clonic seizures on the day of admission. She has had a history of recurrent seizure episodes for the past three years with a progressive decline in cognition. She had no signs of cranial nerve involvement and her visions, as well as eye movements, were normal. The patient had a history of measles in childhood with doubtful measles vaccination status and unremarkable family history.

Clinical examination revealed a disoriented patient with severe disability (MRS=5) and a GCS score of 6/15. The neurological examination depicted an increased tone of bilateral upper and lower limbs. The motor reflexes were normal.

Table 1: Stages of SSPE as described by Jabbour et al. [7].

Stage I	Irritability, dementia, social withdrawal, lethargy, regression of speech.
Stage II	Dyskinesia, dystonia, myoclonus
Stage III	Decerebrate posturing, spasticity and extrapyramidal symptoms
Stage IV	Loss of cerebral function, autonomic failure, signs of vegetative state, akinetic mutism

Table 2: Difference between typical and atypical EEG findings in SSPE.

Typical EEG pattern in SSPE:
Bilateral symmetric high voltage slow wave complexes, synchronous with myoclonic jerks, recurring at 3.5 Hz to 20 Hz intervals. These complexes are remarkably stereotyped and identical in all leads, and almost pathognomonic of SSPE, found in 65%-83% of individuals.
Atypical EEG patterns in SSPE:
1) Rhythmic delta activity in intervals between periodic complexes
2) Electrodecremental pattern
3) Diffuse sharp waves discharge (as in our case)
4) Focal abnormality, like sharp wave discharges

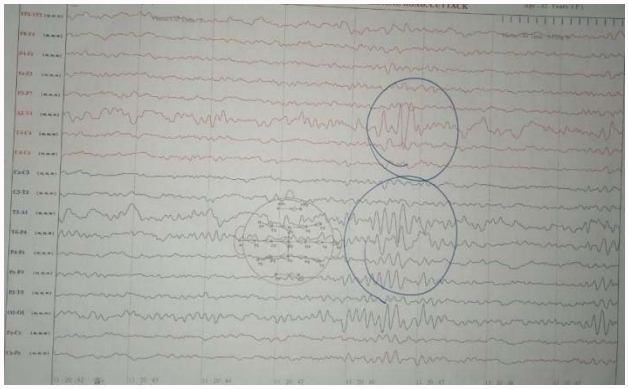

Figure 1: EEG showing generalised sharp wave complexes.

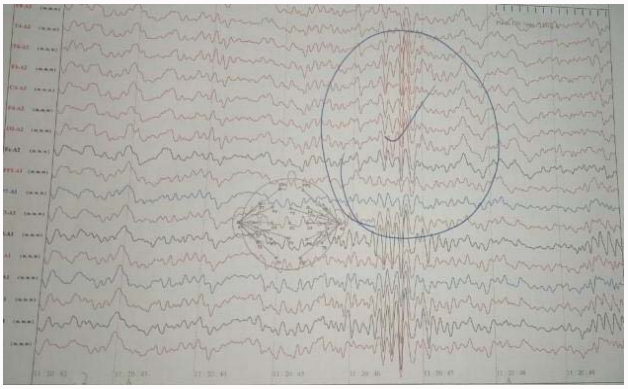

Figure 2: EEG showing synchronous sharp wave complexes of 150 uV to 200 uV with an interval of 0.2 sec to 0.4 sec. (Setting: Amplitude =15 uV/min, paper speed =20 mm/sec).

Complete blood count, serum electrolytes, and urine drug screening were within normal limits except raised total leukocyte count (14,000 cells/cu.mm) with lymphocytic leukocytosis. Blood culture was negative in both aerobic and anaerobic mediums. The MRI sequences had normal signal intensity with no evidence of abnormal enhancement in any region of the brain. Cerebrospinal Fluid (CSF) study depicted normal levels of glucose, protein and white blood cells. The CSF measles IgG antibody (19656.4 IU/ml) & serum measles IgG titer (14929.4 IU/ml) was elevated but negative for all possible autoantibodies including ANA, anti-NMDAR, lupus, etc. The CSF globulin was raised (25.8 mg/dl) and more than 20% of the total CSF protein (34 mg/dl). Electroencephalogram (EEG) revealed bilaterally synchronous sharp wave complexes of 150 μV to 200 μV with a short interval of 0.2 sec to 0.4 sec on a slow background- an atypical EEG finding (Figure 1, 2). Therefore, a diagnosis of Subacute Sclerosing Panencephalitis (SSPE) was made. The patient was managed with weekly doses of intrathecal Interferon 2b (6 million IU) and Isoprinosine 20 mg/kg in three divided doses daily. Antiepileptic drugs like Brivaracetam (4 mg/kg) and sodium valproate (20 mg/kg) in two divided doses were ordered to control ongoing seizures and prevent future episodes.

Following discharge, a few weeks later, she presented with shortness of breath, high-grade fever, and confusion. On further evaluation, she was diagnosed with aspiration pneumonia that progressed into septic shock. The patient was immediately admitted to ICU and critical care management with continuous monitoring was established. She was intubated and resuscitation measures with IV fluids, continuous infusion noradrenaline (4μ mg/min) as well as broad-spectrum antibiotics like piperacillin/tazobactam and clindamycin were started immediately. The patient was conscious, stable, and afebrile on discharge. She was advised to complete the course of weekly Interferon 2b by scheduling follow-up visits and her symptoms were monitored closely.

Discussion

SSPE is a long-term neurological complication of measles that points to the existence of incomplete measles vaccine coverage and illustrates potential challenges such as incomplete coverage in hard-to-reach populations and vaccine hesitancy among parents of children. Long-term neurological sequela of measles arranged in chronological order of presentation include Acute Measles encephalomyelitis which occurs approximately a week after onset of rash, Measles inclusion body encephalitis typically presents 3 weeks to 6 months after initial infection and SSPE occurs 1 to 15 years after primary infection [6].

The clinical features of SSPE have been categorized into 4 stages by Jabbour et al. [7] (Table 1). Atypical presentations have been described including isolated psychiatric manifestations, poorly controlled seizures, and isolated extrapyramidal symptoms, such as dystonia, chorea, and hemiparkinsonism.

In our patient, the presence of high titers of measles IgG in the

CSF (major) along with the presence of chronic progressive history with severe proximal myoclonic jerks and generalized tonic-clonic seizures (major) & CSF globulin greater than 20% of CSF protein (minor) points towards a diagnosis of SSPE by the Dyken criteria [8]. However, there were some atypical findings in the EEG. In our patient, EEG revealed bilaterally synchronous sharp wave complexes of 150 μV to 200 μV with a short interval of 0.2 sec to 0.4 sec on a slow background. This EEG finding is atypical and has not been described before in the context of SSPE (Table 2). Imaging studies, including MRI and CT scans, were also normal.

SSPE is typically a disease of childhood, presenting between 8 to 11 years with subsequent clinical worsening leading to death usually within a few years of diagnosis [9]. Previous studies show mean survival in children to be around 1 year 9 months to 3 years after diagnosis. In adults, 20% survived beyond 4 years after diagnosis [10]. Our patient had a long latent period and was diagnosed late, at the age of 32 years. Fewer than 100 cases of adult-onset SSPE have been reported. It has often been attributed to diagnostic delay due to consideration of alternate diagnoses due to the atypical nature of the presentation and low index of suspicion. Previous studies have shown that adult-onset SSPE has a more atypical presentation with ophthalmological and behavioral disturbance being common. Although eventually fatal, adult-onset SSPE has better rates of remission and longer survival after diagnosis compared to classical juvenile-onset SSPE [10].

SSPE is often diagnosed late, with a previous study stating that the mean duration to establish a diagnosis was 6.2 ± 11.3 months. The study attributed the reasons for such a delay to low geographical prevalence, atypical features, low index of suspicion in adult patients, level of experience of healthcare personnel, acute onset as opposed to the usual indolent course, and fear of diagnosing a disease, which is routinely fatal [11]. Hence, it is important to consider a list of mimicking conditions in which one should keep SSPE as a possible differential: acute Encephalitis, ADEM, Rasmussen encephalitis, Myoclonic epilepsy, Lafora's disease, MERRF, metabolic white matter diseases should be considered [9].

This study adds to the list of reported cases of adult-onset SSPE. It suggests the need to evaluate for SSPE in adult patients with suggestive clinical and radiological features, enabling quicker diagnosis and earlier initiation of therapy.

Human/Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 1964 Declaration of Helsinki and its subsequent amendment.

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