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Case Report

# A Rare Case of Subacute Sclerosing Panencephalitis Presenting as Schizophrenia - A Case Report

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## A B S T R A C T

Subacute sclerosing panencephalitis (SSPE) is a rare, progressive and often fatal late neurological manifestation of measles infection which occurs after a latent period of several years after primary infection in immunocompetent hosts. It is characterised by mental state changes, myoclonus and often ataxia. In this case, an 18-year-old male presented with mood swings, irritability, and cognitive decline, leading to an initial misdiagnosis of schizophrenia. The misdiagnosis stemmed from overlapping psychiatric symptoms, such as behavioural changes, forgetfulness, and decreased attention, as well as the absence of prominent neurological findings at the onset. The diagnosis is based on the detection of intrathecal anti-measles antibodies of the IgG class and the presence of the IgM class of antibodies usually confers a bad prognosis. Pathological examination reveals inflammatory changes in brain tissue, gliosis, demyelination and typical inclusion bodies containing measles viral antigens or RNA.

Electroencephalographic findings in SSPE are highly characteristic showing bilateral generalised high amplitude slow or sharp-slow wave complexes. Therapeutic modalities often prove disappointing, the outcome and prognosis is generally poor and most patients die over a period ranging from several months to a few years. The cause of death is generally respiratory tract infection or involvement of brain stem centres controlling vital functions. Although the incidence of SSPE has dramatically declined after successful immunisation programmes, it still occurs in developing countries where coverage is lower. It is very important to correctly diagnose and manage SSPE and not attribute the symptoms to a mere psychiatric illness. Further research is highly needed for early diagnosis and treatment options for these patients to have a decent quality of life, which at present is difficult.

**Keywords:** Demyelination, Anti-Measles IgG Antibodies, EEG, Schizophrenia, Myoclonus

## Introduction

Subacute sclerosing panencephalitis (SSPE) is a chronic encephalitis occurring after infection with the measles virus.[1] The prevalence of the disease varies depending on the uptake of measles vaccination, with the virus disproportionally affecting regions with low vaccination rates.[2] The disease often begins with behavioural and cognitive changes, which can mimic psychiatric disorders, leading to diagnostic challenges.[3,4] This is illustrated in the case of an 18-year-old male who presented with mood swings, irritability, and memory deficits. These symptoms, along with the absence of clear neurological signs during early evaluation, led to an initial misdiagnosis of schizophrenia.The patient's psychiatric symptoms, including verbal abuse and decreased attention, further supported this misdiagnosis, while a history of measles infection and lack of vaccination were initially overlooked.[4] It was only after the emergence of motor symptoms, such as myoclonic jerks and urinary incontinence, that a neurological cause was suspected, leading to the diagnosis of SSPE.

## Patient Information

An 18-year-old male, a painter by profession, was brought to the psychiatry department of a civil hospital with many complaints. Relatives reported changes in mood and memory of the patient over 2 months. They also reported easy irritability, temper tantrums, verbal abuse, forgetfulness, decreased attention, inability to sleep, and changes in behaviour for 2 months. The symptoms had increased in the past 1 week to the extent that the patient had burnt a finger at work because of lack of attention. He was a chronic tobacco chewer. There was a history of measles in early childhood when he was about 2 years old, and the illness lasted for about 1 week. There was no history of measles vaccination.

On mental state examination, the patient was found to have several abnormalities. The patient had an impaired sense of insight and judgment. Abstract thinking was also impaired. Attention and memory function were also abnormal. After a thorough psychiatric evaluation, the patient was diagnosed with schizophrenia and was started on medications.

Relatives reported no improvement and onset of new symptoms. The patient had developed complaints of involuntary jerky movements of both arms and legs for the past 3–4 days. He also had urinary incontinence for 2–3 days. The movements were initially occurring 4–5 times per day which progressed to 22–24 times per day. Each episode lasted for a few seconds. Patients also reported walking difficulty and a sense of imbalance while walking.

On neurological examination, the patient was conscious and cooperative but somewhat irritable. He was oriented to time, place and person. Sensory examination was normal. On motor examination, the tone in all four limbs was normal. Power was normal. Reflexes of bilateral upper and lower limbs were brisk. Bladder sensation was lost. Bowel sensation was intact. The cranial nerve examination was normal. Cerebellar signs (finger nose test, heel shin test, and dysdiadochokinesia) were not elicitable as the patient was not cooperative. There was no nystagmus. The Romberg test was negative. Other systems examination was unremarkable.

## Discussion

SSPE is a rare neurological manifestation of measles infection in the past. It is the result of natural measles infection especially when it occurs at a young age, particularly less than 2 years.[1] In addition to age, other factors like rural residence, crowded households and adverse socio-economic conditions have been associated with SSPE. [2]It has a long and variable latent period ranging from 1 to 10 years. Incidence in males is slightly higher than that in females. It is usually progressive and fatal.

### Clinical Features

The patients present initially to a psychiatrist because the usual course of illness starts with changes in mental status, behavioural disturbances and memory and attention deficits.[4,5] This is typically followed by motor impairment, usually in the form of myoclonic jerks with or without ataxia. Myoclonic and atonic movements are characteristic. They begin with head dropping, unilateral arm or facial twitching without loss of consciousness. These episodes last only a few seconds and repeat every few minutes to hours. Atypical manifestations are also common. Neurological examination may be normal during the early course of illness, or it may reveal hypertonia, spasticity, tremor, ataxia and apraxia. Less common features are seizures, encephalopathy, raised intracranial tension, hemiparesis and visual loss.[5] Symptoms tend to worsen over weeks to months. They are followed by loss of speech and ambulation in most patients. Clinical staging systems are of limited value due to a lack of standardisation and the absence of typical clinical features in a subset of patients. The staging system adopted by most clinicians for a typical course of illness is as follows:

- **Stage 1:** Behavioural changes, memory deficits
- **Stage 2:** Motor impairment including myoclonic jerks, ataxia
- **Stage 3:** Loss of independent ambulation

In the later stages, myoclonia is diminished or absent and patients may be left with spastic quadriparesis. About 20% of patients show fulminant course of illness.

Diagnosis is made by detection of intrathecal production of anti-measles antibodies of subclass G.[6] Detection of M subclass of anti-measles antibodies usually

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predicts a fulminant course and poor outcome.[6,7] Electroencephalographic findings are characteristic. Typical EEG pattern is bilateral high amplitude slow or sharp-slow wave complexes usually seen from clinical stage 2. MRI of the brain is non-diagnostic and often not needed to diagnose SSPE, but it is done to exclude other differential diagnoses. It may be normal in the early course of the disease. The most common finding is altered T2 signal intensity in periventricular and subcortical white matter.[3]

Treatment options are often limited and extremely disappointing and the disease usually follows a progressive course.[8] Treatment with oral inosiplex and interferons is reported to temporarily stabilise and induce remission in a few patients (30%).[9] It leaves the patient with severe disability and dependence and quality of life is hampered to a great degree. The only effective remedy is prevention by immunisation against the measles virus and maintenance of immunisation coverage to more than 90% of the population.

Conclusion This case highlights the diagnostic challenges posed by Subacute Sclerosing Panencephalitis (SSPE), particularly in its early stages when psychiatric symptoms may dominate or coexist with neurological manifestations. It highlights the importance of maintaining a high index of suspicion for neurological conditions in patients presenting with behavioral and cognitive changes, especially in regions with low vaccination coverage or a history of measles infection.The disease causes severe morbidity and disability and Patients are usually young and belong to economically productive strata of society, which adds on to the burden on family and the Nation as a whole. It further emphasizes the critical role of vaccination programs in preventing measles and its devastating complications, such as SSPE. Continued research into early diagnostic markers and novel therapeutic approaches is the need of the hour to improve outcomes and quality of life for these patients.

## Conflict of Interest: None

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