Subacute sclerosing panencephalitis with disorganized behavior: a case report from psychiatry ward

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Summary
Subacute sclerosing panencephalitis (SSPE) is a late onset, rare and slowly progressive viral encephalitis with a mandatory fatal outcome. The disease initially presents with myoclonic jerks and seizures, ocular manifestations, cognitive decline and behavioral disturbances. However, few rare cases of SSPE had been reported with the initial presentation of sole psychiatric symptoms creating a diagnostic dilemma. We hereby reported a case of an 18-years-old male presented with a history of disorganized behavior who was further evaluated as a case of SSPE. The importance of a holistic approach to rule out organic causes before diagnosing a psychiatric disorder had been highlighted in this report.

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Introduction
Subacute sclerosing panencephalitis (SSPE) is a rare, slowly progressive, fatal neurodegenerative disorder secondary to measles virus infection. 1,2 The late-onset chronic encephalitis occurs mostly after 10 years since the measles infection but the latency can vary from 1-27 years. 3 To date, the incidence was 1 per million in developed countries whereas in developing countries it was 21 per million. 4 Male were seen to be more affected yet no gender susceptibility was evidenced. 5 The affected age group was mostly children and adolescents but very few adult onsets SSPE had been reported and became an area of interest over years.

Also, the adult-onset SSPE tended to be more aggressive with very fast fatality. 6 The initial presentation of SSPE was mostly neurological therefore, cases with behavioral disturbances as initial complaints were very rare. 1 We hereby reported a case of an 18-years-old male who came to the Psychiatry unit with complaints of disorganized behavior and eventually been evaluated as a case of SSPE.

Case summary
An 18-years-old male was admitted to our psychiatric inpatient department of Bangabandhu Sheikh Mujib Medical University (BSMMU) on 6th January 2022 with the history of disorganized behavior like repeated self-injurious and wandering behavior, hitting others without any provocation, public nudity, sudden jumping into the pond and often eating mud. On query, his parents admitted that 9 months back he had a sudden onset of recurrent jerky movements of left side of body which was shock like in nature, but was absent during sleep. He had multiple history of fall and being injured due to this jerky movements but loss of consciousness, bowel or bladder incontinence during such events were absent. Also, there was no history of fever or unconsciousness. He was then taken to multiple physicians and underwent many investigations. But all of which came out inconclusive. Then after a month, he started having difficulty in performing some basic life skills like putting on his clothes, eating on his own hands and also gradual deterioration in his intellect as he could not understand the concept of calculation and money management. His memory impairment was also significant in terms of immediate and recent memory loss. These symptoms were followed by the disorganized behaviors for which he received treatment from a psychiatrist and had improvement after taking anti psychotics, but his intellectual impairment was getting worse day by day. Then 15 days before hospital admission, he started having tremor and stiffness over all four limbs along with slurring of speech. No history of measles was reported by the parents and they confirmed about immunization against measles virus. With all of these symptoms he first went to the neurology outpatient department, BSMMU and from there he was referred to psychiatry outpatient and further admitted in psychiatry ward. After admission, the patient underwent detailed physical and mental state examination with special emphasis on neurological evaluation. On general physical examination, vitals
were normal. Systemic examinations other than nervous system examination revealed no abnormality. On nervous system examination, motor examination revealed visible left upper and lower limb myoclonus, tremor in all four limbs in both resting and postural state along with grade 3 rigidity in all four limbs. Left knee and ankle jerks were brisk and all other jerks were normal along with planter flexor. Left ankle clonus was present. Gait was slightly high stepping. No abnormality of behavior, no signs of meningeal irritation was found. On mental state examination, his facial appearance was restricted with occasional undue smiling, mood was euthymic, speech was slurred and no other abnormality was found in thought and perception. On assessment of memory; immediate and recent memory found to be impaired and in Wechsler Abbreviated Scale for Intelligence-TM (WASI TM) the patient scored 50, suggestive of moderate level of IQ. Complete blood count, Erythrocyte Sedimentation Rate (ESR), serum glutamate pyruvate transaminase (SGPT), random blood sugar, thyroid profile, urine for copper and serum ceruloplasmin were within the normal ranges of laboratory standards. Slit lamp examination for Kayser Fleischer (KF) ring was not positive. Electroencephalogram (EEG) showed symmetrical high voltage, repetitive, polysphasic sharp and slow wave complexes of 1 to 1.5 second duration recurring every 2-5 seconds. On T2 weighted magnetic resonance imaging (MRI) chronic infarct was found in the right basal ganglia. Results of cerebrospinal fluid (CSF) studies were as follows: total cell count of white blood cell (WBC)=04 cells/cu.mm, neutrophils=50%, lymphocytes=50%, glucose=4.1 mmol/L, CSF-protein=58 mg/dL. Anti-measles Antibody showed IgM<5.0 U/ml and IgG=260.3 mIU/ml where the reference value for IgG was negative below 150mIU/ml and positive above 200mIU/ml. Then after evaluating all these laboratory investigations, he was finally diagnosed as a case of subacute sclerosing panencephalitis (Jabbar stage-II). Initially his treatment started with levetiracetam 500mg/day, trihexyphenidyl 2mg/day and clonazepam 0.5mg/day for the symptomatic control of myoclonic seizure and muscle rigidity. Later the dose of levetiracetam was increased to 1000mg/day and the patient was discharged with these medications on 17\textsuperscript{th} February, 2022 after almost 1.5 months of hospital stay. One month after on 28\textsuperscript{th} March, 2022 the patient came for follow-up and at that time his condition was worse than before. The frequency of myoclonic jerks increased along with loss of speech and profound intellectual impairment. The dose of levetiracetam was increased to 1500mg/day and we advised for his next follow up again after one month. But as he didn’t come for further visit, we called his father on 22\textsuperscript{nd} May, 2022 and found that his condition almost progressed to the vegetative state where he became completely bed ridden with complete loss of bowel and bladder control.

**Discussion**

SSPE is a slow but persistent viral encephalitis with mandatory fatal outcome. The diagnosis can be established by at least three out of five Dyken’s\textsuperscript{7} criteria (Table 1) all of which were fulfilled by our patient- atypical clinical picture, characteristic EEG changes and raised titers of measles antibody in CSF .

The initial stage of SSPE starts with mild impairment of intellect along with behavioral disturbances without any obvious neurological presentations.\textsuperscript{6} These symptoms often remain unnoticed until they cause significant impairment in functional areas of life.\textsuperscript{6} Gradually symptoms progresses to myoclonic jerks, seizures, ocular manifestations and eventually vegetative state.\textsuperscript{5} But confusion regarding diagnosis arises when SSPE presents with isolated primary psychiatric manifestations.

Till date, many case reports had been published where the onset of SSPE came off with sole psychiatric symptoms. Among them three cases were presented as mania two of which presented with classic manic symptoms\textsuperscript{1,8} and few days after the initiation of treatment for mania, neurological symptoms started to appear. In 2018, Bhat et al.\textsuperscript{9} reported a case of SSPE presenting as antidepressant induced childhood mania. In this report, a boy of 8 years initially presented with depressive symptoms and after the starting of antidepressant medications he developed symptoms of mania. However, his symptoms eventually progressed to the typical neurological symptoms of SSPE. Apart from mania, two cases had been reported with sole depressive symptoms.\textsuperscript{10,4} Interestingly, both of the cases had almost similar symptoms of major depressive disorder along with history of two times sudden loss of awareness\textsuperscript{10} and falling off.\textsuperscript{4} Both of these transient episodes were diagnosed as dissociative

<table>
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<tr>
<th>Criteria</th>
<th>Description</th>
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<tr>
<td>1. Clinical</td>
<td>Progressive, subacute mental deterioration with typical signs like myoclonus</td>
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<tr>
<td>2. EEG</td>
<td>Periodic, stereotyped, high voltage discharges</td>
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<td>3. Cerebrospinal fluid</td>
<td>Raised gamamoglobulin or oligoclonal pattern</td>
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<td>4. Measles antibodies</td>
<td>Raised titer in serum (≥1: 256) and/or cerebrospinal fluid (≥1:4)</td>
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<tr>
<td>5. Brain biopsy</td>
<td>Suggestive of panencephalitis</td>
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Definitive: criteria 5 with three more criteria; probable: three of the five criteria.
phenomena as EEG report and other findings from history came out incompatible. Then the psychiatrists started the treatment of major depressive disorder and few months later with little or no improvement of depressive symptoms, further neurological symptoms suggestive of SSPE began to manifest. Rest of the cases of SSPE with primary psychiatric manifestations had been reported as non-specific behavioral disturbances or psychosis with/without catatonia.

In 2017, Parmar et al.\textsuperscript{11} reported a case of a 17-years-old boy whose clinical presentation started with marked positive psychotic symptoms including visual and auditory hallucinations, paranoid delusions and disorganized behavior along with prominent catatonic symptoms by way of mutism, stereotypy and posturing. He was prescribed tablet lorazepam to decrease the catatonic symptoms but when no improvement was seen, psychiatrists became doubtful of non-organic catatonia and sought out multiple relevant investigations including computed tomography (CT) scan, MRI, EEG, anti-measles antibody and based on the findings, the diagnosis of SSPE was confirmed.

Our patient came to us with the complaints of pure psychotic like symptoms as disorganized behavior along with gross loss of intellect and no complaints of typical neurological features seen in SSPE. Also, all the investigations done prior to the hospital admission were inconclusive. Later, the detailed longitudinal history and incompatible neurological signs made us doubtful about the organic basis of his psychiatric symptoms and lead to further investigations and imaging. Literature said that SSPE with atypical presentation was mostly seen in the second decade of life and presentation was most likely psychotic.\textsuperscript{4} Another notable finding, in this case, was the absence of the past history of measles infection and also there was history of immunization against measles virus. Such history had been reported in many other case reports earlier including childhood-onset SSPE.\textsuperscript{11} Our case highlighted that psychiatrists needed to be very meticulous in terms of history taking and clinical examination to pick up any anomaly that might be a subtle clue of an underlying organic disease. Also, vaccination against measles virus and absence of history of measles did not rule out the possibility of SSPE.

### Conclusion

Clinicians especially psychiatrists should have a high index of suspicion for SSPE whenever any child or adult patient presents with sudden behavioral changes or any other primary psychiatric phenomena in composition with atypical history and physical examination, irrespective of the history of previous measles infection. Also, carrying out every relevant and cost efficient neurological investigations including EEG and neuroimaging should be on mind before diagnosing any psychiatric disorder. Hence, an enthusiastic collaborative approach between psychiatrists and neurologists in such cases are not only necessary but also time demanding.

### References