Case Report

Subacute Sclerosing Panencephalitis Presenting with Isolated Positive Psychotic and Catatonic Symptoms

Arpit Parmar, Rajeev Ranjan[1], Rajesh Sagar

ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a rare slowly progressing neurological illness. Although patients with SSPE initially present with symptoms such as myoclonic jerks, cognitive decline, and personality/behavioral changes usually, rarely pure psychiatric symptoms (e.g., mania, psychosis, and catatonia) have also been reported during the initial course of the illness. We report an unusual case of an adolescent with SSPE presenting with prominent positive psychotic and catatonic symptoms with the absence of classical symptoms of SSPE in initial course of illness and further discussed the relevant literature.

Key words: *Catatonia, psychosis, schizophrenia, subacute sclerosing panencephalitis*

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a progressive illness with an invariably fatal outcome. It is usually seen in children and adolescents affecting the central nervous system caused by defective measles virus. Although psychiatric symptoms have been reported in patients with SSPE, such presentations are rare. Till date, only a few cases have been reported in which psychosis or catatonia was a presenting complaint.[1] We report an unusual case of an adolescent with SSPE who presented with prominent positive psychotic and catatonic symptoms in the initial course of illness which misdiagnosed as a case of schizophrenia.

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CASE REPORT

A 15-year-old boy with no past or family history of psychiatric illness presented to a tertiary care hospital with an illness of 4–5 months duration. His illness started with behavioral changes observed by his parents. He would be restless and would pace around in his home for a long period without any reason. After a few days, his sleep was also disturbed. He would be found standing in a particular posture for 1–2 h without getting tired for which he did not give any reason even after repeatedly being asked by family members. He also started remaining fearful for which he stated the reason that someone was coming to harm him. He

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Department of Psychiatry, All India Institute of Medical Sciences, New Delhi, [1]Department of Psychiatry, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India

Address for correspondence: Dr. Arpit Parmar
Department of Psychiatry, Room No. 4096, Teaching Block, All India Institute of Medical Sciences, New Delhi - 110 029, India.
E-mail: dr.arpitparmar@yahoo.in

would repeatedly check doors and windows and also ask his parents to do so. Despite repeated reassurances given by the parents, he would not get convinced of the fact that anyone was going to harm her. He also reported seeing "Bajrang Bali" (The Hindu God) in his clear consciousness during awaken state multiple times; however, he would not elaborate further. Parents also found him smiling and muttering to self on many occasions while he was alone as if he would be talking to someone. However, he did not give a history of hearing any voice. By now, he would also pass urine in his clothes (he would be aware of the urge but could not reach the bathroom). He became completely mute after 4–5 weeks of onset of illness. He was found doing some motor acts repeatedly in a specific manner (clapping continuously for 10–15 min with his hands kept above head). He used to maintain some unusual postures for a long period. He stopped taking a bath, taking food, and his mother had to assist him in daily activities of life.

He was taken to a private psychiatrist initially and was started on risperidone tablet 1–2 mg which led to some improvement in his psychotic symptoms. However, catatonic symptoms did not improve. After about 1½ months of treatment, he was referred to us for further management. A working diagnosis of schizophrenia – undifferentiated subtype versus organic psychotic disorder (ICD-10) – was kept. The dose of risperidone was optimized, and tablet lorazepam was added. It led to improvement in psychotic symptoms, however, as the catatonic symptoms and bladder dysfunction did not improve; a neurology referral was also sought. The patient underwent multiple investigations including computed tomography (CT) scan, magnetic resonance imaging (MRI) scan, and electroencephalogram (EEG). His CT scan was normal. Brain MRI demonstrated periventricular white matter lesions. EEG reported typical findings suggestive of SSPE (i.e., high-voltage slow-wave complexes interspersed with short rhythmic waves) [Figure 1]. Considering this, antimeasles antibody titers were also performed which were found to be increased (blood - 1:32, cerebrospinal fluid [CSF] - 1:4). Serum creatine phosphokinase level was normal. Based on these findings, a diagnosis of SSPE was considered. In the follow-up course, the patient also developed myoclonic jerks and disorientation, and he was advised to follow-up at neurology outpatient department for long-term management.

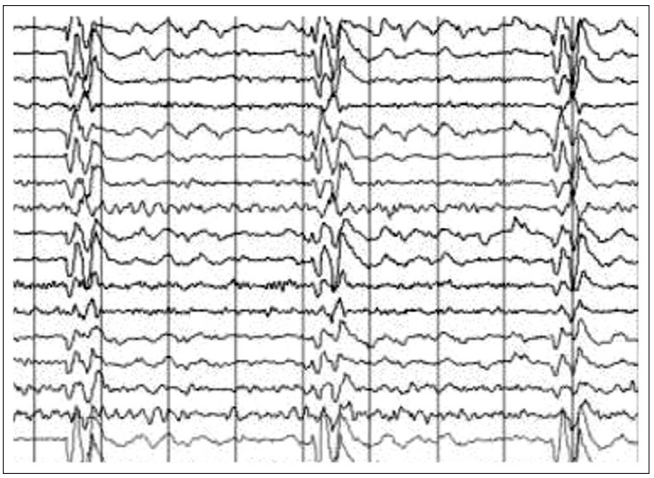

Figure 1: Electroencephalogram characteristic of periodic complexes consisting of synchronous, polyphasic, stereotyped delta waves

DISCUSSION

SSPE is a rare but slowly progressive illness leading to severe neurological deficits and death. It progresses in stages which vary from person to person. During the initial stage, symptoms such as cognitive decline, myoclonus, and behavioral and personality changes are seen in up to 97% of cases. As the illness progresses, hemiplegia, blindness, and seizures may occur. Eventually, the patient loses ability to walk as muscles stiffen. Such progressive deterioration leads to persistent vegetative state eventually.[2,3]

There are cases reported in the literature where SSPE presented with isolated psychiatric complaints, which lead to misdiagnosis.[1] Such presentations include psychosis, mania, and catatonia. Catatonia is relatively rare, especially during the early stages of the illness. Although catatonia is seen in patients with many neurological illnesses as presenting symptom, but akinetic mutism has been seen only in advanced illness in case of SSPE.[4] Till date, only three cases have been reported in the literature in which patients with SSPE presented with catatonic symptoms in the initial course of the illness.[1] Similarly, psychosis has only been reported in four children with SSPE till date in authors' knowledge.[2,5]

The absence of typical symptoms of SSPE with the presence of typical symptoms suggestive of schizophrenia (paranoid delusion, visual hallucinations, and possible auditory hallucination) led to initial misdiagnosis of our case as possible schizophrenia. However, the presence of catatonic symptoms along with sphincter dysfunction led to suspicion of possible organicity in this case. A diagnosis of SSPE was entertained based on typical MRI of the brain, EEG, and serum and CSF antimeasles antibody titers. The previous case suggests that catatonic symptoms in patients with SSPE may not respond to usual management with lorazepam and antipsychotics as also seen in our case.[6] However, no further deterioration of catatonic features was seen in our patient, unlike the previous case. One more interesting finding, in this case, is the absence of the previous history of measles infection during childhood.

This has been reported in many cases previously, even among childhood-onset SSPE.[7]

This case highlights the need to keep a differential of viral encephalitis, especially when a child presents with catatonia along with sphincter dysfunction. The absence of a history of measles does not rule out such a possibility. Treatment with lorazepam and antipsychotics might not benefit such patients, which is an indicator for the clinician to think of this rare illness. The case is also important considering the fact that the prevalence of SSPE has been found to be as high as 21 cases per million people in some settings.[8]

CONCLUSION

This case suggests the need to screen children presenting with psychosis and catatonia for an illness such as SSPE. Psychiatrists need to be aware of this rare but important possibility. Atypical SSPE (presenting with catatonia or psychosis) when presented needs a high index of suspicion for detection by psychiatrists.

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Conflicts of interest
There are no conflicts of interest.

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