Subacute Sclerosing Panencephalitis Presenting with Isolated Positive Psychotic and Catatonic Symptoms

Arpit Parmar, Rajeev Ranjan, 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.

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...
Parmar, et al.: Subacute sclerosing panencephalitis and psychosis

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.

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.[2,3]

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.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Dayal P, Balhara YP. Catatonia as presenting clinical feature of subacute sclerosing panencephalitis. J Pediatr Neurosci 2014;9:57-9.
2. Kayal M, Varghese ST, Balhara YP. Psychiatric manifestation of SSPE. J Neuropsychiatry Clin Neurosci 2006;18:560.
3. Kartal A, Kurt AN, Gürkas E, Aydin K, Serdaroglu A. Subacute sclerosing panencephalitis presenting as schizophrenia with an alpha coma pattern in a child. J Child Neurol 2014;29:NP111-3.
4. Gadoth N. Subacute sclerosing panencephalitis (SSPE) the story of a vanishing disease. Brain Dev 2012;34:705-11.
5. Baran Z, Hanagasi H, Uçok A. An unusual late presentation of subacute sclerosing panencephalitis with psychotic symptoms. J Neuropsychiatry Clin Neurosci 2010;22:123.
6. Aggarwal A, Jain M, Jiloha R. Catatonia as the initial presenting feature of subacute sclerosing panencephalitis. J Neuropsychiatry Clin Neurosci 2011;23:E29-31.
7. Bonthius DJ, Stanek N, Grose C. Subacute sclerosing panencephalitis, a measles complication, in an internationally adopted child. Emerg Infect Dis 2000;6:377-81.
8. Garg RK, Karak B, Sharma AM. Subacute sclerosing panencephalitis. Indian Pediatr 1998;35:337-44.