Subacute sclerosing panencephalitis presenting as mania

Ashish Aggarwal, Ashish Khandelwal[1], Manish Jain[2], Jiloha R.C.[1]

Department of Psychiatry, Indira Gandhi Medical College, Shimla, Himachal Pradesh, [1]G. B. Pant Hospital, [2]Dr RML Hospital, New Delhi, India

Abstract

Subacute sclerosing panencephalitis (SSPE) is a rare, invariably fatal degenerative disease of the central nervous system developing after measles infection. Besides neurological symptoms as initial presenting symptoms, rare reports of its presentation with pure psychiatric symptoms have been reported. We here report a case of 14 year old male who initially presented with manic symptoms and then subsequently diagnosed to be suffering from SSPE. Improtance of ruling our organic conditions is emphasized.

Key Words

Manic episode, subacute sclerosing panencephalitis, periodic complex

For correspondence:

Dr. Ashish Aggarwal, Department of Psychiatry, Indira Gandhi Medical College, Shimla - 171 001, Himachal Pradesh, India.
E-mail: drashish1980@gmail.com

Ann Indian Acad Neurol 2011;14:120-1

Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare, invariably fatal degenerative disease of the central nervous system that afflicts mainly children and adolescents. Although for some time now it has been known to develop after measles infection, the exact mechanism of its causation is still not fully understood. Initial presenting symptoms have commonly been reported to be neurological like myoclonic jerks, falling attacks, changing gait, abnormal movements, speech impairment, inability to walk or stand, seizures, dementia, visual disturbances, pyramidal and extra-pyramidal signs. Though behavioral disturbances have been reported to be the initial presentation in some cases, reports of cases with pure psychiatric symptoms as the initial complaints are rare.[1] We hereby report a case of a 14-year-old male who was initially diagnosed as a case of manic episode, and then subsequently developed myoclonic jerks. EEG and CSF findings confirmed a diagnosis of SSPE.

Case Report

A 14-year-old boy, was brought to us by his mother for complaints of behavioral change for the past one month. She reported that he has not been working properly. He would demand for money and would go out frequently to watch movies. He would remain outside house for most time and talked a lot. He would look more cheerful then before and would not obey family members, and easily became irritable and angry. His hygiene and grooming also was increased and he would take bath 3-4 times a day. At home, he would keep pacing here and there. His sleep was markedly decreased from 7-8 h before to 3-4 h and the patient would not report any fatigability. There was no past or family history of any psychiatric illness. Mental status examination revealed a restless boy, with increased psychomotor activity, with eye to eye contact established but not maintained. His speech was increased in pitch, tone, and volume. His affect was elated. Flow of thought was increased and revealed ideas of grandiosity. He had no insight into the illness. His higher mental functions were, however, normal. Detailed neurological examination including fundus examination was normal. A diagnosis of first episode mania was made. Routine hematological investigations including chest X ray and ECG were normal. He was started on olanzapine 5 mg per day increased to 10 mg after 5 days. The patient followed up after about 10 days and her mother reported that he was not able to tolerate the medication. He was unduly sedated and would at times pass urine in clothes. Also, now she reported that the patient would have spontaneous jerky movements for a very short period of time repeatedly. On examination at this time, the patient had myoclonic jerks and his gait was unsteady. His affect was inappropriate. Olanzapine was stopped. He was further investigated. Magnetic resonance imaging of the brain revealed multiple areas of hyper intensities in bilateral brain parenchyma on T2W image which were more in posterior part of cortex. Patient was subsequently referred to neurologist. Blood tests for toxoplasma, cytomegalovirus, and herpes Simplex virus IgG and IgM were negative. Venereal

Access this article online	
Quick Response Code:	Website: www.annalsofian.org
	DOI: 10.4103/0972-2327.82801

disease research laboratories (VDRL) slide test was negative. EEG revealed typical picture of generalized periodic spike and wave formation with periodicity at every 7 seconds. Cerebrospinal fluid (CSF) was grossly clear with normal opening pressure and showed 5 lymphocytes/cumm, protein 20 mg/dl and sugar 70 mg/dl against a blood sugar of 100 mg/dl. There were elevated anti measles IgG and IgM anti bodies. A diagnosis of SSPE was made. His condition deteriorated with increase in frequency of myoclonic jerks with urinary and fecal incontinence over next 4 weeks. He could not take care of himself and had to be assisted by family members for his daily routine. His gait became progressively unsteady with frequent falls as well. He was started on sodium valporate 400 mg per day along with clonazepam 1 mg per day. Subsequently, the patient was lost on follow-up.

Discussion

Our patient fulfilled the criteria for SSPE, which include fulfillment of at least three of the following five criteria:[2]

1. A typical clinical picture: Personality and behavioral changes, worsening school performance, followed by myoclonic seizures, paresis, dyspraxias, memory impairment, language difficulties, blindness, and eventually obtundation, stupor, and coma;
2. Characteristic EEG changes
3. Elevated CSF globulin levels greater than 20% of total CSF protein
4. Raised titers of measles antibodies in blood and CSF
5. Typical histopathological finding in brain biopsy or autopsy.

Our patient had myoclonic jerks, though at a later point of time along with typical EEG findings and CSF findings.

Presentations with psychiatric symptoms have been described in the literature. Cases of schizophreniform psychosis, paranoid psychosis, schizophrenia catatonia and depression are well described.[3-7] However, to the best of our knowledge, case presenting with typical features of mania has not been reported, though aimless wandering, hyper religiosity, irritability, and adamant behavior have been described as presenting feature in few cases.[6]

Since the disease is a slowly progressive disease occurring years after measles infection, it is possible that patient's may initially present with only psychiatric symptoms and may develop full neurological manifestations only after some period of time.

Our case highlights the importance of ruling out organic conditions especially in adolescents without any known risk factors for mania. Also, it highlights the importance of EEG in patients with psychiatric symptoms, which can be a cheap mode of investigation.

In conclusion, SSPE can present initially with pure psychiatric symptoms and pediatricians, neurologists as well as psychiatrists, should be aware of this rare possibility. A high index of suspicion is needed to detect SSPE in its atypical and rare forms. When faced with a young patient with psychiatric manifestations, with no other significant history, organic conditions must be ruled out and SSPE should also be considered in the differential diagnosis.

References

1. Prashanth LK, Taly AB, Sinha S, Ravi V. Subacute sclerosing panencephalitis (SSPE): An insight into the diagnostic errors from a tertiary care university hospital. J Child Neurol 2007;22:683-8.
2. Dyken PR. Subacute sclerosing panencephalitis. Neurol Clin 1985;3:179-85.
3. Forrest G, Stores G. Subacute sclerosing panencephalitis presenting with psychosis and possible sexual abuse. Eur Child Adolesc Psychiatry 1996;5:110-3.
4. Duncalf CM, Kent JN, Harbord M, Hicks EP. Subacute sclerosing panencephalitis presenting as schizophreniform psychosis. Br J Psychiatry 1989;155:557-9.
5. Jahnel M. Paranoid-hallucinatory psychosis as primary manifestation of subactue sclerosing panencephalitis (SSPE) in a 19-year-old man. Psychiatr Prax 2003;30:S70-2.
6. Khadilkar SV, Patil SG, Kulkarni KS. A study of SSPE: Early clincial features. J pediatric neurology 2004;2:73-7.
7. Khwaja GA, Gupta M, Sharma DK. Subacute sclerosing panencephalitis. J Assoc Physicians India 1991;39:928-33.

Received: 12-02-10, **Revised:** 15-03-10, **Accepted:** 30-07-10

Source of Support: Nil, **Conflict of Interest:** Nil