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Case Report

Case of Subacute Sclerosing Pan-Encephalitis (SSPE) presenting as catatonic schizophrenia

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Abstract

Index case presented with insidious onset psychotic symptoms of 6 months duration characterised by excitement, smiling and muttering to self, catatonic posturing, negativism, and periods of mutism, irritability, episodes of aggression and poor self-care which were gradually progressive in nature. For the last 2 months preceding his presentation to the clinic he developed myoclonic jerks predominantly involving the trunks and upper limbs and apraxia. On examination, there was generalised cognitive impairment, subcortical myoclonus occurring every 2-3 minutes, paratonia and echolalia. Non contrast Magnetic Resonance Imaging (MRI) Brain study was unremarkable. Electroencephalogram (EEG) showed slow background and periodic generalised slow waves. His cerebrospinal fluid (CSF) and serum anti-measles antibody as measured by EIA was positive in high titres (15586 U/ml and 8457 U/ml respectively). A diagnosis of SSPE stage 2 was kept. This case demonstrates that SSPE can masquerade as catatonic schizophrenia in early stages.

Key words: SSPE, schizophrenia, catatonia

Introduction

The subacute sclerosing panencephalitis (SSPE) is a rare complication of measles virus seen in children and adolescents caused by latent mutant strain, which affects the central nervous system

[1]. SSPE was previously known as Dawson's inclusion body encephalitis as Dawson in 1933 and 1934 reported cellular inclusions in the cerebral lesions of patients with SSPE [2]. In general, it is unusual to find measles virus affecting brain, and with widespread routine vaccination, the incidence has fallen to approximately 10 cases/year in the US and Western world. However, in India, the prevalence is slightly higher affecting approximately 20 cases/million [3]. The symptoms can range from altered behaviour, alteration in personality, myoclonic jerks, cognitive decline, gradual behavioural changes, and unsteady gait, ataxia, photosensitivity, and even coma [4]. There may be damage to optic nerve and retina (chorioretinitis). The diagnosis of SSPE is based on the Dyken's criteria, which include two major and four minor criteria [5]. The cases are uniformly fatal. The average latent period of SSPE is 7–10 years (range: 1 month to 27 years) after measles virus infection, and death usually occurs within 1–3 years after the onset of symptoms [1].

Case details

Master S.M., a 15-year-old boy presented with insidious onset psychotic symptoms of 6 months duration characterised by smiling and muttering to self, catatonic posturing, negativism, periods of mutism, irritability, episodes of aggression, attempts to run out of home, and poor self- care which were gradually progressive in nature. Two months before presenting to our clinic, he developed myoclonic jerks predominantly involving the trunk and upper limbs and apraxia. The myoclonic jerks were infrequent in the beginning but over the next 3-4 weeks it started occurring at every 2-3 minutes interval. There was marked decline in scholastic performance over a relatively shorter period of time. On examination, there was generalised cognitive impairment, subcortical myoclonus occurring every 2-3 minutes, paratonia and echolalia. Non- contrast magnetic resonance imaging (MRI) brain study was unremarkable. Electroencephalogram (EEG) showed slow background and periodic generalised slow waves. His haemoglobin, blood counts, liver

function tests, thyroid function tests, serum urea and creatinine, lipid profile, fasting blood sugar, serum ceruloplasmin, electrocardiogram, echocardiogram were within normal limit. History of drug addiction was non-contributory. Patient had an uneventful infancy with developmental milestones being achieved at normal age. There was no history of measles infection, febrile seizure or idiopathic epileptic fits prior to this event. There was no family history of any psychiatric disorder and symptom constellation suggestive of any genetic disorder which could give rise to similar set of symptoms.

Initially, the diagnosis of catatonic schizophrenia was entertained and patient was started on amisulpride (up to 300 mg/day) and oral lorazepam (up to 3 mg/day). Patient developed acute extrapyramidal side effects in the form of cervical dystonia, sialorrhoea, static tremor and cogwheel rigidity within one week of starting the treatment. Amisulpride was stopped and switched over to olanzapine (up to 10 mg/day). Trihexyphenidyl (2mg/day) was given for 20 days with which the extrapyramidal symptoms disappeared. Patient was sleeping well and his psychotic symptoms showed sign of improvement over next 2 months. But his self-care and academic performance remained the same and he became completely homebound. He could not even take his bath on his own and had to be fed by his mother.

With the appearance of myoclonic jerks and EEG finding as mentioned above a neurological consultation was sought after 4 months of his presentation to the clinic. His cerebrospinal fluid (CSF) and serum anti-measles antibody as measured by Enzyme Immuno-Assay (EIA) was positive in high titres (15586 U/ml and 8457 U/ml, respectively). A diagnosis of SSPE stage- 2 was kept and patient was started on sodium valproate (1500 mg per day in three divided doses), clonazepam (1.5 mg per day in three divided doses), and oral isoprinosine (500 mg four times a day). Detailed cognitive assessment was attempted with PGI Battery for Brain Dysfunction (PGI-

BBD) but patient either could not complete most of the test domains or scored very poorly. A repeat EEG was done after 4 months of initiation of therapy which showed an interictal record of generalised seizure disorder. There were few breakthrough seizures particularly on missing the dose of the medicine which was managed by proper counselling of parents regarding the disease and emphasis on drug compliance. The patient has been regularly following up for last three and half years. Myoclonic jerks have been absent since past 8 months. A repeat EEG which was done 2 months back came to be within normal limit. But still there is significant cognitive impairment, echolalia, paratonia and bladder-bowel dysfunction.

Discussion

Most of the patients with SSPE have a history of primary measles infection at an early age. Children infected with measles under the age of one year carry a 16 times greater risk of SSPE than those infected at age five year or later [6]. In our case, no history of prior measles infection was forthcoming despite our best attempt. This is quite unusual and separates the index case from other cases which have already been reported in literature.

Catatonia is not a usual clinical presentation of subacute sclerosing panencephalitis (SSPE), especially in the initial stages of 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. Till date, only five cases have been reported in the literature in which patients with SSPE presented with catatonic symptoms in the initial course of the illness [7]. The absence of typical symptoms of SSPE with the presence of mutism, posturing, negativism and excitement led to initial misdiagnosis of our case as possible catatonic schizophrenia. However, as has been described, antipsychotics and lorazepam were not of great help in this case. Non response to psychotropics known to treat catatonia effectively compelled us to investigate the case further.

A diagnosis of SSPE was entertained based on emergence of myoclonus, paratonia, EEG changes, and serum and CSF anti-measles antibody titres.

Following were key points in index case: no history of measles in childhood, initial presentation with catatonic symptoms, hallucinatory behaviour, episodes of aggression and poor self-care, partial response to psychotropic medications which are known to be effective in catatonia, subsequent emergence of myoclonus, paratonia, EEG changes, and serum and CSF anti-measles antibody titres.

There have been very few case reports from India where catatonic symptoms, schizophrenic symptoms, depressive symptoms and mood lability have been reported in the initial phase of the illness [7-12]. This case highlights the need to keep a differential of viral encephalitis, especially when a child presents with catatonia along with myoclonus. The absence of a history of measles does not negate such possibility. Treatment with lorazepam and antipsychotics might not benefit such patients, which should compel the clinician to think of this rare disease.

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