CASE REPORT

A case of subacute sclerosing panencephalitis presenting as depression


Summary: Subacute sclerosing panencephalitis (SSPE) is rare in adult patients. Clinical presentation in initial phases of SSPE may be non-specific leading to diagnostic delay. We present a 24-year-old patient with depressive syndrome of five months’ duration prior to the onset of typical features of SSPE, which is a rare presentation. This patient had responded partially to Sertraline, for a brief period, before he was diagnosed to have SSPE. This case illustrates affective symptoms can be the presenting features of SSPE in adults.

Subacute sclerosing panencephalitis (SSPE) is very rare in developed countries but is still common in developing countries. Saha et al. (1) reported an annual incidence of 21 per million population in India. SSPE usually presents in childhood and adolescence. The initial symptoms are subtle and include mild intellectual deterioration and behavioural changes without any apparent neurological signs or findings. Parents and teachers may notice progressive deterioration in scholastic performance.

It is uncommon after 18 years of age, and the disease has a more aggressive course in adults. Patients with adult onset SSPE present at a mean age of 25.4 years (range 20–35 years). Clinical features at presentation usually include cognitive decline, behavioural changes and myoclonic jerks. Features later in the disease include decerebrate and decorticate posturing, dysphagia, dysarthria and coma (2).

The diagnosis is often made at a later stage and may be missed in the initial stages as the signs and symptoms are subtle or atypical (3). The diagnosis of SSPE is suspected clinically and confirmed with the characteristic electroencephalogram (EEG) findings and cerebrospinal fluid (CSF) measles antibody titres (4).

Here, we present a case of an adult onset SSPE that initially presented with a depressive syndrome for 5 months till the onset of typical myoclonic jerks, which is a rare presentation.

A 24-year-old man was referred to the department of psychiatry from the internal medicine department with the complaints of a 5-month history of feeling sad, loss of interest in pleasurable activities, increased fatigue, lethargy, somatic complaints and insomnia. In addition to the above depressive syndrome, he had two episodes of transient loss of awareness of his surroundings. Both these episodes occurred in the presence of others and inside his house. There were no episodes during sleep nor were there any injuries sustained during these episodes. There was no history suggestive of myoclonic jerks, urinary incontinence, lip-smacking movements or any other seizure equivalents. Detailed neurological examination did not reveal any focal neurological signs. Other systemic examinations were also within normal limits. A mental state examination revealed a reduced attention span. The patient’s speech was slow and laconic. Thought content revealed depressive cognitions. Affect was depressed and nonreactive. After an outpatient evaluation in the department of psychiatry, he was diagnosed to be...
suffering from moderate depression with somatic symptoms according to ICD-10 classification.

Organic causes for his mood symptoms were suspected. An EEG (Fig. 1) was done to rule out temporal lobe epilepsy. The initial EEG was normal, and the transient falls were attributed to dissociative phenomenon as a part of the depressive syndrome.

The patient was started on Tab Sertraline 50 mg per day with which his depressive symptoms improved to an extent. He was followed-up as an outpatient on a regular basis once in 2 weeks. After 2 months, he presented with repetitive intermittent jerky movements of the upper limbs. The jerks were asymmetrical and affected the right side of his body more than the left. Because of the persistence and periodic nature of the jerks and history of periods of altered sensorium, the patient was referred to the neurology department. Besides higher mental function, an examination revealed that although he was oriented to time, place and person, he had difficulty in obeying complex commands. Recent memory and new learning ability were impaired. Cranial nerve examination was notable for hypometric saccades. Motor system examination showed mild cogwheel rigidity of the upper limbs bilaterally. Gait showed diminished arm-swing. The posture was remarkable for intermittent jerky movements of the upper limbs, more so of the right side.

A repeat EEG revealed bursts of high-amplitude (upto 250 microvolts) sharp and slow wave discharges bilaterally and synchronously, occurring at regular intervals of 15 s. This activity was associated with jerking of the right upper and lower limbs. These periodic complexes were suggestive of fully evolved SSPE. Computerized tomography of brain was done, which was normal. Magnetic resonance imaging was not done because of financial constraints for the patient.

Measles antibody titres in the CSF examined were strongly positive. A ratio of 1/64 and more between paired serum and CSF antibodies was considered diagnostic. Immunofluorescence was used to detect measles antibodies in the CSF. Patient was started on supportive treatment but he died within 2 months.

SSPE is common in developing countries. Although it usually affects children and adolescents, it can present in adults rarely. The diagnosis may not be suspected as the initial phase of
behavioural changes, and cognitive decline is often labelled as functional. Our case had a long period of predominantly depressive symptoms for 5 months with no other features suggestive of cognitive decline or personality change.

The presence of normal EEG in the early stage makes clinicians even more complacent about suspecting a diagnosis of SSPE. The typical EEG findings of SSPE appear in stage II of the disease. EEG in a typical case in stage II is characterized by high amplitude 300–1500 microvolts repetitive polyphasic sharp and slow wave complexes of 0.5–2 s duration, recurring every 4–15 s synchronously with myoclonic jerks (4). EEG in the end stages of the disease again becomes atypical, as the background and sleep activities are disorganized, and there is a gradual reduction in the amplitude till it finally becomes isoelectric.

Our patient had a rapid deterioration and died 2 months after diagnosis. The natural history of the temporal profile of SSPE is highly variable and can be of three types – subacute onset with duration of illness 1 to 3 years (seen in 80% of cases), rapid course with death in 3 months after diagnosis (seen in 10% of cases) and a slowly progressive form with evolution over 4–8 years (5).

The importance of recognizing the spectrum of potential presentations of SSPE and providing an early diagnosis will increase as more effective treatments become available. This report is basically to highlight another atypical presentation of adult-onset SSPE presenting with a depressive syndrome which is a rare presentation in the initial stages. Another atypical feature in this case was the rather long duration of depressive symptoms before the appearance of myoclonic jerks. Therefore, clinicians need to have a high index of suspicion in patients presenting with these symptoms from areas with high prevalence of SSPE for early diagnosis.

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References
1. SAHA V, JOHN TJ, MUKUNDAN P. High incidence of subacute sclerosing panencephalitis in South India. Epidemiol Infect 1990;104:151–156.