Subacute sclerosing panencephalitis presenting with hypersexual behavior

Sir,

Subacute sclerosing panencephalitis (SSPE) is a rare progressive demyelinating disease of the central nervous system associated with a chronic infection of brain tissue with measles virus. The disease is commonly seen in children and young adults. It is characterized by progressive intellectual deterioration, focal and/or generalized seizures, myoclonus, ataxia, and visual disturbances. The presentation varies according to the age of the patient, and the diagnosis is often delayed in adults due to the atypical presentation. The hypersexual disorder is classically defined in persons aged above 18 years and is characterized by the recurrent and intense sexual urge irrespective of the situation persisting beyond 6 months. The disease is usually associated with significant psychiatric ailments and also with central nervous disorders such as stroke, brain injury, and medications. SSPE presenting with hypersexual behavior is uncommon and is rarely reported in the literature.

We retrospectively reviewed the data pertaining to four patients of SSPE treated at our hospital between 2012 and 2014. The diagnosis of SSPE is based on the Dyken's criteria. SSPE is diagnosed, if the patient meets any three of the following five criteria: (a) Progressive intellectual deterioration with signs of myoclonus; (b) characteristic electroencephalographic (EEG) pattern; (c) elevated cerebrospinal fluid (CSF) globulin levels; (d) elevated CSF measles antibody titers; and (e) brain biopsy suggestive of measles. The demographic profile and investigation findings of all the patients are summarized in the Table 1.

Briefly, all our patients are young adults of both sexes and hail from poor socioeconomic strata of the society. All the patients presented with classical behavioral abnormalities, scholastic regression, and a couple of them presented with the hypersexual syndrome. All the patients showed a characteristic EEG pattern of periodic lateralized epileptiform discharge sequences neuroimaging revealed nonspecific white matter hyperintensities in few patients. The paired serum and CSF samples for measles ELISA IgG were positive in all the patients confirming the diagnosis of SSPE.

The patients were treated with intrathecal interferon, Isoprinosine, and antipsychotic drugs including risperidone. Sodium valproate was used to control the myoclonus and seizure activity. One patient had a persistent worsening illness with recurrent seizures and succumbed to his illness at the end of 1-year of observation. Behavioral abnormalities improved in the remaining three patients, and they are under regular follow-up.

In India, SSPE is still a common neurodegenerative disorder despite an increase in the measles vaccination. The diagnosis and treatment are usually delayed due to the varying presentation of the disease. Our data revealed that three patients were receiving psychiatric treatment for more than a year prior to the proper diagnosis. Poor scholastic performance and cognitive regression are important clues to the underlying diagnosis of SSPE and EEG is often diagnostic in these cases. The clinical presentation of adult onset differs from childhood onset as, personality changes and ophthalmic manifestations are common presenting features and survival with treatment have been better in certain series. Our data had patients from the second and third decades of life, and none of the patients had ophthalmic manifestations. Hypersexual behavior leads to social stigma and is often underreported by the patients. The same has been linked with a variety of neuropsychiatric disorders including obsessive-compulsive disorder, bipolar disorder, drugs such as dopaminergic agonists and other disorders including stroke and brain injury.

The prevalence of SSPE is common among lower
Table 1: Clinical details of all the four patients

| Feature                                           | Case 1      | Case 2      | Case 3      | Case 4      |
|---------------------------------------------------|-------------|-------------|-------------|-------------|
| Age (years)                                       | 12          | 23          | 18          | 27          |
| Sex                                               | Male        | Female      | Female      | Male        |
| Measles vaccination                               | No          | No          | No          | No          |
| History of childhood measles                      | Yes         | Yes         | Yes         | Yes         |
| Socioeconomic status                              | Middle class| Poor        | Middle class| Poor        |
| Gap between measles and the symptoms (years)      | 9           | 14          | 14          | 17          |
| First symptom                                     | Poor scholastic performance | Behavioral abnormalities | Poor scholastic performance | Behavioral abnormalities |
| Myoclonus                                         | +           | ++          | ++          | +           |
| Scholastic regression                             | +           | +           | +           | ++          |
| Jabbour’s stage                                   | 2           | 3           | 3           | 1           |
| Behavior                                          | Normal      | Hypersexual | Normal      | Hypersexual |
| EEG                                               | PLEDs +     | PLEDs +     | PLEDs +     | PLEDs +     |
| MRI findings                                      | White matter hyperintensities | White matter hyperintensities | White matter hyperintensities | Normal |
| Follow-up duration (years)                        | 3           | 1           | 3           | 2           |
| Course                                            | Static, recognizes relatives | Expired at the end of 1-year | Static, poor communication | Ambulant with support |
| Current status                                     | Alive       | Dead        | Alive       | Alive       |

PLEDS – Periodic lateralized epileptiform discharge sequences; EEG – Electroencephalographic; MRI – Magnetic resonance imaging

socioeconomic strata, large family size, and rural areas due to overcrowding. The measles virus is transmitted by respiratory secretions via aerosol exposure and also through direct contact with larger droplets.[1] A variety of metabolic disorders leads to dementia in young including Wilson’s disease, hypothyroidism, B12 deficiency, etc. Our patients were investigated for these conditions and the negative screen resulted in the psychiatric diagnosis. It is essential for all the practitioners to look for myoclonus and also investigate for the rare causes of dementia such as SSPE and prion diseases.

The therapy of SSPE involves the use of isoprinosine and an immunomodulator like interferon. The response to therapy depends on the clinical stage of SSPE at which the treatment was started. SSPE staging is done as per the modified Jabbour’s classification and patients with stage 1 disease respond remarkably to the therapy whereas, stage 2 disease progresses invariably resulting in fatal outcomes in 2–5 years. To conclude, SSPE is a devastating disorder of public health importance in the developing countries. A high index of suspicion for SSPE is required during the evaluation of young adults with myoclonus, behavioral changes, and scholastic regression.

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Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. Tatli B, Ekici B, Ozmen M. Current therapies and future perspectives in subacute sclerosing panencephalitis. Expert Rev Neurother 2012;12:485-92.
2. Borgermans L, Vrijhoef B, Vandevoorde J, De Maeseneer J, Vansintejan J, Devroey D. Relevance of hypersexual disorder to family medicine and primary care as a complex multidimensional chronic disease construct. Int J Family Med 2013;2013:519265.
3. Dyken PR. Subacute sclerosing panencephalitis. Current status. Neurol Clin 1985;3:179-96.
4. Praveen-kumar S, Sinha S, Taly AB, Jayasree S, Ravi V, Vijayan J, et al. Electroencephalographic and imaging profile in a subacute sclerosing panencephalitis (SSPE) cohort: A correlative study. Clin Neurophysiol 2007;118:1947-54.
5. Prashanth LK, Taly AB, Ravi V, Sinha S, Arunodaya GR. Adult onset subacute sclerosing panencephalitis: Clinical profile of 39 patients from a tertiary care centre. J Neurol Neurosurg Psychiatry 2006;77:830-3.

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