

## **Atypical Subacute Sclerosing Panencephalitis (SSPE): All Postpartum Altered Behavior Isn't CVT!**

Dear Editor,

Subacute sclerosing panencephalitis (SSPE) is a rare progressive neurological disorder caused by persistent defective measles virus infection.<sup>[1]</sup> It is usually seen in children and adolescents, who present with insidious onset behavioral changes, cognitive decline, pyramidal and extrapyramidal

features, myoclonus and visual disturbances.<sup>[2]</sup> The disease is relentlessly progressive and fatal within 3–5 years of onset, although rarely, treatment leads to disease stabilization. We present a 22-year-old woman who developed acute onset altered behavior in the post-partum state and was initially misdiagnosed to have cortical vein thrombosis. We seek to

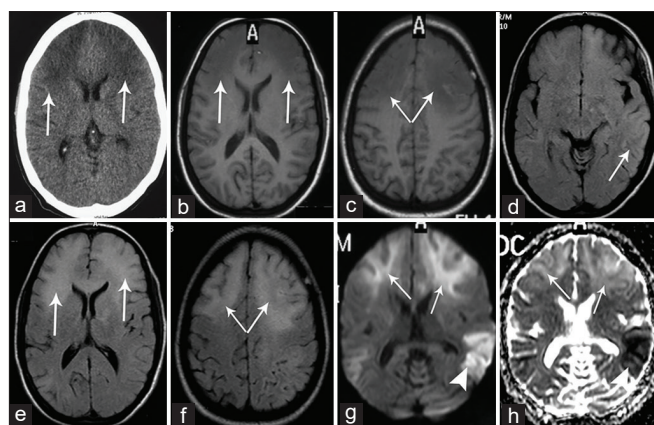
highlight the factors that favor a diagnosis of SSPE even in atypicality.

A 22-year-old primigravida, without any comorbidities, delivered a healthy child at full term by normal vaginal delivery at home assisted by a “dai”. However, immediately postpartum she developed altered behavior and was not able to attend to the needs of the baby. She became mute and was noticed to have intermittent, sudden, brief, jerky movement of all four limbs a day later. These were not stimulus sensitive and disappeared during sleep. She was admitted to a nearby hospital, where a magnetic resonance imaging (MRI) brain (day 3 of ictus) revealed T2-FLAIR hyperintensities in bilateral frontal and left temporal region with diffusion restriction in the latter [Figure 1] without any loss of vascular flow-void in the major venous sinuses. No contrast-MRI studies or Magnetic resonance venography (MRV) was done. Routine blood investigations were found to be within normal limits barring anemia and she was given one unit of packed red blood cells for the same. Even though there was no imaging evidence of sinus thrombosis, she was thought to have cortical vein thrombosis by the evaluating team and was started on anticoagulants. However, she did not improve with treatment and was re-evaluated elsewhere after two weeks of therapy, where a repeat MRI brain and MRV was done. The MRI showed similar findings and the MRV was unremarkable. Considering a possibility of Acute Disseminated Encephalomyelitis (ADEM) she was pulsed with steroids and discharged. However, the frequency of jerks progressively increased and she remained bed bound and incontinent, following which she was referred to our institute 2 months later. She was found to be mute, unresponsive with intermittent, multifocal myoclonic jerks. Spasticity

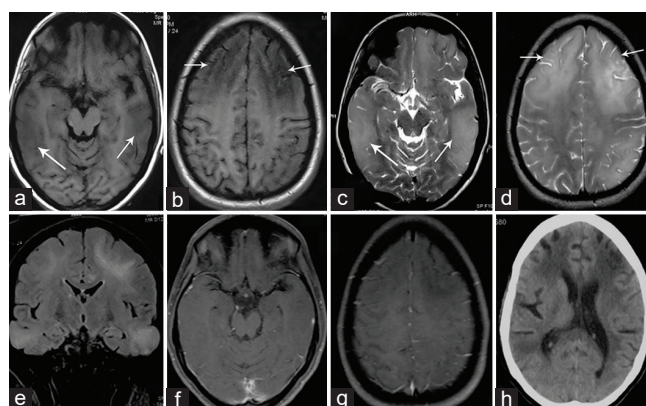
was present in all four limbs with brisk reflexes and bilateral extensor plantars. Sensory and cerebellar system could not be assessed.

Electroencephalogram (EEG) revealed periodic sharp wave complexes synchronized with the jerks [Figure 3]. Cerebrospinal fluid (CSF) analysis revealed nil cells and normal biochemistry but was positive for anti-measles antibody (CSF: Serum ratio = 11.8; normal <1.3). Serum autoimmune panel was negative. Repeat MRI brain showed persistent T2-FLAIR hyperintensities, but no contrast enhancement and the loss of the previous diffusion restriction [Figure 2]. Her parents gave history of incomplete immunization during her childhood. In view of presence of anti-measles antibody in serum, she was diagnosed as SSPE and treated with intrathecal interferon, without significant benefit.

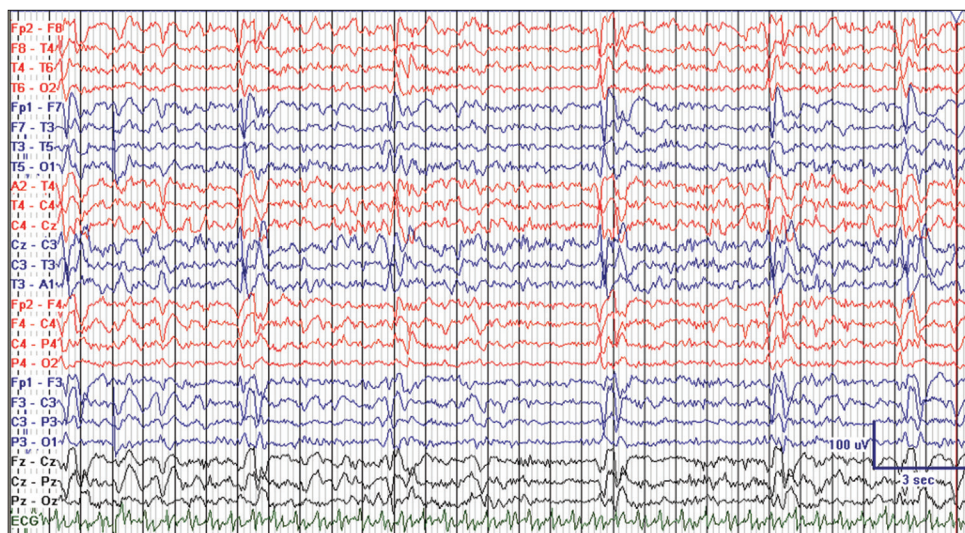
Although SSPE predominantly occurs in children and young adults, it can rarely occur in pregnant women,<sup>[3]</sup> similar to our case. Her clinical presentation, EEG findings along with elevated anti-measles antibody titers are consistent with the diagnosis of probable SSPE according to Dyken’s criteria.<sup>[2]</sup> Her radiological features barring diffusion restriction are consistent with findings of SSPE. ADEM can have similar radiological findings. However, SSPE too can rarely have diffusion restriction.<sup>[4,5]</sup> Our case was peculiar in that diffusion restriction was limited to only the left temporal lobe lesion, which has not been described. Diffusion restriction is indicative of cytotoxic edema in these cases and probably points towards more fulminant disease.<sup>[6]</sup> The initial presence and later resolution of this diffusion restriction on imaging along with the clinical presentation of our patient is consistent with a fulminant disease process. Though SSPE is typically a chronic encephalitic illness, an acute fulminant form with necrotizing encephalitis-like presentation can



**Figure 1:** (a) NCCT head (A) shows hypodensity in bilateral anterior frontal region (arrows in a) with effacement of overlying cortical sulci. Bilateral frontal regions (arrows in b, c) and left temporal region (arrow in d) show hypointense signal involving both grey and white matter with sulcal effacement in T1-WI (b, c) and hyperintense signal on FLAIR Images (d-f). On diffusion maps (g) and ADC maps (h), bilateral frontal regions appear bright suggesting facilitated diffusion (arrows in g and h), while left posterior temporal region appears bright on diffusion-trace images (g) and dark on ADC maps (h) suggesting diffusion restriction (arrowhead in g and h)



**Figure 2:** Axial T1-WIs (a, b) shows hypo-intensity involving both grey and white matter in bilateral temporal region with sulcal effacement (arrows in a). Bilateral frontal lesions seen in previous scans show persistent hypo-intensity with sulcal dilation suggesting chronicity of lesions (arrows in b). These temporal and frontal lesions are hyperintense in FLAIR Images (c-e). No diffusion restriction was seen (images not shown). Following gadolinium administration, no enhancement was seen in T1-WIs (f, g). Follow-up NCCT (h) shows dilation of cortical sulci in bilateral frontal and temporal regions with persistent hypodensity suggesting gliosis



**Figure 3:** Longitudinal bipolar montage EEG showing generalised, long interval periodic discharges with background attenuation

occur.<sup>[4]</sup> Repeat imaging revealed dilatation of cortical sulci in the previously involved regions suggestive of gliosis further strengthening this hypothesis.

SSPE should be considered as a differential in all young individuals presenting with altered behavior along with myoclonic jerks, irrespective of the rapidity of symptom onset since acute presentations can rarely occur. We would also like to add to the existing body of literature on SSPE that diffusion restriction can occur as an atypical manifestation, especially in fulminant presentations.

### LEARNING POINTS

- SSPE should be considered as a differential in all young individuals presenting with altered behavior along with myoclonic jerks, irrespective of the rapidity of symptom onset
- Diffusion restriction can occur as an atypical manifestation, especially in fulminant presentations.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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