A rare presentation of subacute sclerosing panencephalitis with acute fulminant course and atypical radiological features

Sir,

Subacute sclerosing panencephalitis (SSPE) is progressive, fatal encephalitis of children and adolescents caused by a persistent infection of immune resistant measles virus.[1] SSPE usually begins insidiously and follows a subacute course with relentless but slow progression to death. However, SSPE can present with atypical symptoms and more acute fulminant course due to various undetermined reasons. Magnetic resonance imaging (MRI) brain in SSPE shows white matter affection typically involving temporal and parietal lobes.[2]

A 14-year-old boy presented with acute onset rapidly progressive cognitive decline and myoclonic jerks of 15 days duration. There was no history of measles vaccination. On examination, patient was in a state of akinetic-mutism with rigidity of all the four limbs. Characteristic myoclonic jerks were observed having rapid onset followed by slow relaxation. Often myoclonus was elicited by loud sound (startle response). His MRI brain showed hypointensities on T1W image with corresponding hyperintensities on T2W image and T2 flair [Figure 1] in bilateral frontal regions, predominantly involving white matter including corpus collosum as well as parieto-occipital area to some extent.

His electroencephalogram revealed classical quasiperiodic complexes with stereotypy and burst suppression pattern [Figure 2]. Cerebrospinal fluid (CSF) showed reactive lymphocytosis. Serum and CSF measles antibody titers were strongly positive, confirming the diagnosis of SSPE.

SSPE is a progressive inflammatory disease of the central nervous system caused by a persistent measles virus.[3] Acute fulminant form of SSPE is extremely rare[4] and it is diagnosed when patient develops at least 66% neurologic disability in the first 3 months or dies within 6 months.[5] Several factors such as exposure to measles at an early age, viral virulence, concurrent infections with other viruses, derangement of T-cell subsets, all have been postulated for fulminant course of SSPE.[2,6] In the early stages, MRI of the brain shows patchy asymmetric white matter affection, typically involving temporal and parietal lobes.[2]

Our patient apart from having atypical fulminant clinical course also had atypical radiological findings involving predominantly and extensively, bilateral frontal regions. Gökçil *et al*.,[7] reported a case of fulminant SSPE in which the course was rapidly progressive, leading to death in 2 months, and MRI showed hyperintensities in the occipital poles. Hergüner (Special character missing) *et al*.,[2] reported three cases of fulminant SSPE but all of them had normal MRI. Alexander *et al*.,[8] reported a case of fulminant SSPE whose MRI revealed ill-defined signal intensities predominantly involving the parietal lobes.

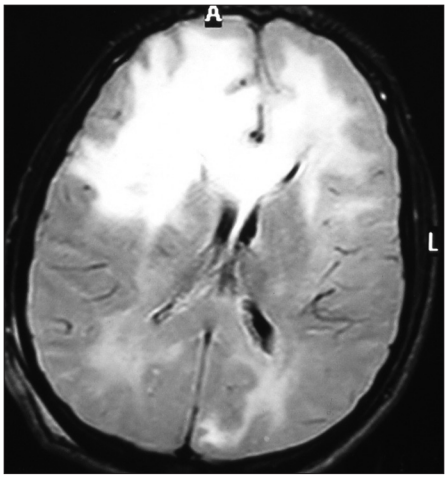

Figure 1: Magnetic resonance imaging of the brain showing hyperintensities on T2 flair image in bilateral frontal regions, predominantly involving white matter including corpus collosum as well as parietooccipital area to some extent

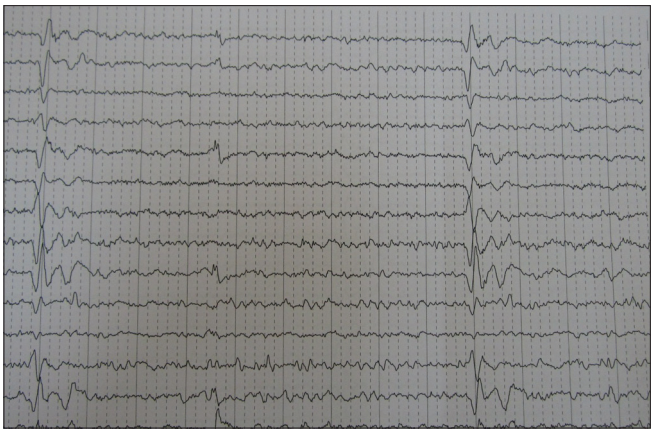

Figure 2: Electroencephalogram showing quasiperiodic complexes with stereotypy and burst suppression pattern

Mahadevan *et al*.,[4] reported a case of fulminant SSPE with MRI showing demyelinaton in frontal and parieto-occipital areas.

To conclude, fulminant SSPE is an uncommon entity which can present with atypical features, without passing through various stages sequentially. Our patient of fulminant SSPE had predominant frontal region involvement on MRI which further adds to its rarity.

Rajendra Singh Jain, Raghavendra Bakki Sannegowda, Trilochan Srivastava, Rahul Jain, Tarun Mathur,

Pankaj Gandhi

Department of Neurology, Sawai ManSingh Medical College Hospital, Jaipur, Rajasthan, India

For correspondence:
Dr. Rajendra Singh Jain, 126-Janakpuri-II, Imli Phatak, Jaipur, Rajasthan, India.
E-mail: drrsjain@yahoo.com

References

1. Miller DH, Robb SA, Ormerod IE, Pohl KR, MacManus DG, Kendall BE, *et al.* Magnetic resonance imaging of inflammatory and demyelinating white-matter diseases of childhood. Dev Med Child Neurol Suppl 1990;32:97-107.
2. Hergüner MÖ, Altunbaşak Ş, Baytok V. Patients with acute, fulminant form of SSPE. Turk J Pediatr 2007;49:422-5.
3. Dyken PR. Subacute sclerosing panencephalitis. Current status. Neurol Clin 1985;3:179-96.
4. Mahadevan A, Vaidya SR, Wairagkar NS, Khedekar D, Kovoor JM, Santosh V, *et al.* Case of fulminant-SSPE associated with measles genotype D7 from India: An autopsy study. Neuropathology 2008;28:621-6.
5. PeBenito R, Naqvi SH, Arca MM, Schubert R. Fulminant subacute sclerosing panencephalitis: Case report and literature review. Clin Pediatr (Phila) 1997;36:149-54.
6. Garg RK. Subacute sclerosing panencephalitis. Postgrad Med J 2002;78:63-70.
7. Gökçil Z, Odabaşi Z, Aksu A, Vural O, Yardim M. Acute fulminant SSPE: Clinical and EEG features. Clin Electroencephalogr 1998;29:43-8.
8. Alexander M, Singh S, Gnanamuthu C, Chandi S, Korah IP. Subacute sclerosing panencephalitis: CT and MRI imaging in a rapidly progressive case. Neurol India 1999;47:304-7.

Access this article online	
Quick Response Code:	**Website:** www.annalsofian.org
	DOI: 10.4103/0972-2327.120437