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. 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.

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 colosum 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. Acute fulminant form of SSPE is extremely rare and it is diagnosed when patient develops at least 66% neurologic disability in the first 3 months or dies within 6 months. 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. In the early stages, MRI of the brain shows patchy asymmetric white matter affection, typically involving temporal and parietal lobes.

Our patient apart from having atypical fulminant clinical course also had atypical radiological findings involving predominantly and extensively, bilateral frontal regions. Gökçil et al. 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. reported three cases of fulminant SSPE but all of them had normal MRI. Alexander et al. reported a case of fulminant SSPE whose MRI revealed ill-defined signal intensities predominantly involving the parietal lobes.

Mahadevan et al. reported a case of fulminant SSPE with MRI showing demyelination 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.

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Dear Sir,
Moyamoya disease (MMD) presenting with movement disorders in the form of chorea, dystonia, and diskinesias has been reported before; however, MMD with exaggerated startle response has not been reported previously.

Recently, we came across a case of MMD featuring exaggerated startle response.

A 7-years-old boy, presented with the sudden jerky movements of body in the form of blink, flexion of neck and trunk, and abduction of arm especially in response to loud noise since the age of 6 years [Video 1]. It often resulted in fall and injuries. There was no history of loss of consciousness with these jerks. He had past history of recurrent strokes at the age of 3 years. No other family members or siblings were affected with similar illness. At the time of presentation he was conscious, alert, and was able to understand commands. His speech was limited to speaking bi-syllables only. Residual right hemiparesis with spasticity was present. Involuntary movement in the form of exaggerated startle response to sudden, loud noise was present. No spontaneous involuntary movements were observed. Computed tomography (CT) scan of head showed bilateral cerebral hemisphere infarcts in middle cerebral artery territory. He was diagnosed to have MMD on the basis of angiographic findings which showed occlusion of bilateral supraclinoid internal carotid arteries and proximal middle cerebral arteries with bilateral basal collaterals as well as right posterior cerebral artery (P1) stenosis [Figure 1].

Exaggerated startle syndromes (hyperekplexia) consist of an excessive motor response or jump, to unexpected auditory, somesthetic, and visual stimuli. An exaggerated startle syndrome may be due to local brainstem pathology (anoxia, inflammatory lesions, and hemorrhage) and also occurs as an inherited condition transmitted as an autosomal-dominant trait.

Our patient presented with recurrent ischemic strokes at the age of 3 years and exaggerated startle response since the age 6 years [Video 1]. Development of exaggerated startle response in this patient can be explained by the ischemia of brainstem with the progression of moyamoya...