

Case Report

Subacute Sclerosing Panencephalitis (SSPE) Associated with Basal Ganglia Involvement - Two Case Reports

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Abstract

Magnetic resonance imaging in subacute sclerosing panencephalitis (SSPE) usually demonstrates changes in periventricular and subcortical white matter. Basal ganglia, Cerebellum, Spinal cord are less commonly involved. In this article, we report two cases of SSPE associated with basal ganglia involvement. The 1st case a 11 years old boy had the severe symptoms of SSPE with bilateral basal ganglia involvement. The 2nd case a 8 years old girl had right sided hemiplegia.

Keywords: Subacute sclerosing panencephalitis; Basal ganglia.

Introduction:

Subacute sclerosing panencephalitis is a slow viral infection caused by the hypermutated measles virus with defective M-protein¹. The diagnosis of SSPE is based on personal history, typical findings and elevated serum and CSF anti measles antibody titres^{2,3}. Magnetic resonance imaging (MRI) in SSPE usually demonstrates changes in white matter signal intensity, cerebral atrophy or in early disorder normal findings. In SSPE most commonly involved areas are periventricular and subcortical white matter. The cerebellum, spinal cord, corpus callosum and basal ganglia are less commonly involved^{3,4}. Prominent basal ganglia involvement may occur very rarely in SSPE. Our two SSPE patients presented with basal ganglia involvement.

Case-1:

A 11- years old male child was in good health 4 months back. Then he developed gradual deterioration of school performance, jerky movement of the body, behavioral changes and slowed speech. Aphasia, gait disorder and generalized seizure started in last 2 months before admission. There was no history of measles in childhood nor was immunized against this disease. He was born after an uneventful pregnancy and had normal motor and mental development. The parents were nonconsanguineous and other siblings were healthy.

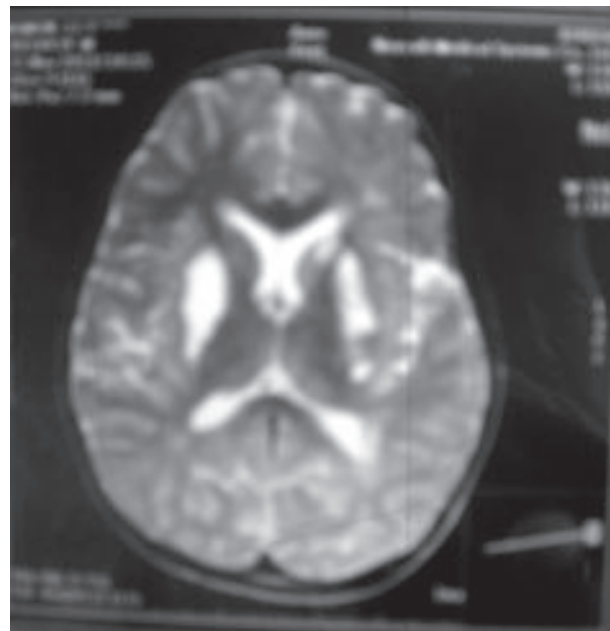


Fig.-1: Case-1: MRI of brain (T2 image) shows bilateral basal ganglia hyperintensity.

Examination reveals patient was well alert, jerky movement present & vital signs were normal. The cranial nerve functions were normal, deep tendon reflexes hyperactive, planter responses were bilaterally extensor. Haemogram, routine blood chemistry & urine analysis were normal. In CSF study, there was elevated titer of measles antibody. Serum measles antibody was also positive. EEG shows epileptiform discharges. MRI of brain T1 & T2 weighted images showed hyperintensity in both lentiform nucleus, and caudate nucleus. These findings were consistent with SSPE. The patient was prescribed anticonvulsant (Na valproate- clonazepam) and antiviral drug amantadine and discharged with counseling.

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Case-2:

A 8- years old girl, 1st child of nonconsanguinous parents, developed normally up to 7 years of her age. Then she developed gradual deterioration of speech and memory for last 1 year. It was associated with periodic myoclonic spasm, progressive weakness of left side of the body for last 6 months.

Her perinatal period was uneventful and family history of illness were non contributory. She did not received measles vaccine and suffered from measles at 18 months of age.

On admission she was drowsy and responded poorly to simple commands and needed help for daily life. Her gait was hemiplegic which was interrupted by periodic myoclonic spasm. Deep tendon reflexes were exaggerated and planter responses were extensor and clonuses also present. Her cranial nerves were intact and sensory examination revealed normal findings. Eye examination showed pale optic disc. Routine blood chemistry and urinalysis were normal. An electroencephalogram revealed generalized epileptiform discharge. The CSF fluid analysis showed normocytosis, slightly raised protein and CSF anti measles antibody IGg was positive. MRI of brain showed T1 hypo intense & T2 hyper intense shadow involving the right lentiform nucleus with hypoxic

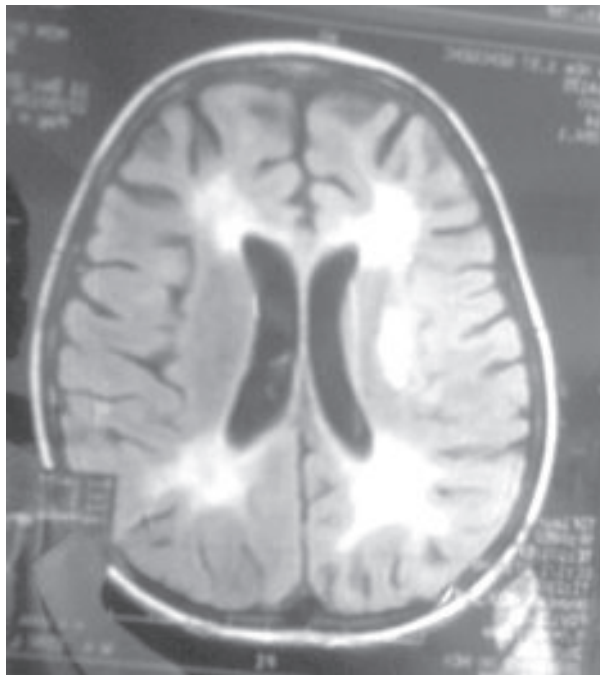


Fig.-2: Case-2: MRI of brain showed Hyperintense lesion in left sided basal ganglia with periventricular white matter lesion.

ischemic encephalopathy and periventricular degeneration. The diagnosis of SSPE was done. This patient was treated by oral sodium valproate and oral amantadine. Patient was discharged after counseling.

Discussion:

SSPE is a chronic and debilitating slow-virus infection and its incidence is around 1/1000000⁵. The diagnosis of SSPE is based on clinical manifestation, typical EEG findings, and laboratory findings^{2,3}. Neuroimaging can be used to show the extent of cerebral disease, and for differential diagnosis, but it has no impact on treatment or prognosis. MRI as the most sensitive modality and findings have been reported in a limited number of studies^{3,4,6,7}. MRI abnormalities in SSPE include focal areas of increased signal intensity on T2 weighted images (hypo or isointense on T1 weighted images) in the cerebral cortex, subcortical and periventricular white matter, corpus callosum, basal ganglia, thalamas and brainstem^{2,4,8}. On the other hand, MRI can be normal in the first few months and early clinical stage of the disease^{3,4,6,7}. Kulczycki, et al⁶. noted that white matter lesions began in the occipital region and progressed to frontal white matter in patients with SSPE. These studies demonstrate that patients with severe disease may have normal MRI findings and patients with mild clinical disease may have intense MRI lesions.

In the early stage of the disease, the CT and MRI may be normal, or may show slight cerebral edema^{3,4,6,7}. In the later stages, however bilateral symmetrical or asymmetrical low density enhancing or no enhancing lesions in the cerebrum involving the gray and white matter may be seen. Cortical atrophy is found in the advanced stages of disease⁷.

Basal ganglia lesions are infrequent but they tend to appear in patient with advanced clinical states specially in those with longer disease duration. Initial involvement in basal ganglia as a first site in SSPE is unusual.

First patient (case -1) of this series was admitted with severe clinical picture and MRI showed involvement of bilateral basal ganglia without any white matter change or any cerebral atrophy. Case-2 of these series presented with myoclonic jerks and Rt sided hemiplegia and MRI showed left basal ganglia and periventricular white matter change.

Although it is reported that MRI in SSPE followed a consistent pattern involving primarily the white matter, the striking finding in these patients was the prominent basal ganglia involvement which is a constant features of mitochondrial disorders (e.g Leighs disease) and Neurodegenerative disorder (e.g Wilson disease).⁹ So these patients demonstrate that SSPE should be considered in the differential diagnosis of basal ganglia lesion observed by MRI. The observations expand the differential diagnosis of focal MRI and of the radiologic presentation of SSPE.

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