Atypical magnetic resonance imaging features in subacute sclerosing panencephalitis

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Abstract

Objectives: Subacute sclerosing panencephalitis (SSPE) is rare chronic, progressive encephalitis that affects primarily children and young adults, caused by a persistent infection with measles virus. No cure for SSPE exists, but the condition can be managed by medication if treatment is started at an early stage. **Methods and Results:** Heterogeneity of imaging findings in SSPE is not very uncommon. But pial and gyral enhancements are very rarely noticed. Significant asymmetric onset as well as pial-gyral enhancements is not reported. Herein we present a case of 16 years adolescent of SSPE having remarkable asymmetric pial-gyral enhancements, which were misinterpreted as tubercular infection. **Conclusion:** Early diagnosis and treatment is encouraging in SSPE, although it is not curable with current therapy. Clinico-radiological and electrophysiological correlation is very important in diagnosis of SSPE, more gravely in patients having atypical image findings as in our index case.

Key Words

Encephalitis, measles, myoclonic jerks, pial and gyral enhancement, subacute sclerosing panencephalitis (SSPE)

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Clinical Description

A 16-year-boy presented with 2 months history of progressive cognitive decline and myoclonus without other focal neurological deficit. There was history suggestive of measles at 8 months of age. He was started on antitubercular treatment based on magnetic resonance imaging (MRI) findings elsewhere. Examination revealed generalized myoclonic jerks occurring periodically at the interval of 7-8 s, involving dominantly right side of the body. Cognitive examination revealed mild inattention, disorientation to time, acalculia as well as impaired judgment, and insight. In view of clinical picture with classic myoclonic jerks and cognitive decline, a possibility of subacute sclerosing panencephalitis (SSPE) was considered and he was evaluated. Electroencephalography showed periodic slow wave complexes slightly more prominent on the left side along with mild background slowing consistent with SSPE. In gadolinium enhanced MRI brain, left posterior parietal and occipital lobes hyperintensities on T2-weighted and fluid attenuation inversion recovery (FLAIR) sequences [Figure 1c and d], affecting both grey and white matter with pial and gyral contrast enhancement (CE) [Figure 1a and b], were noted. Though MRI brain findings were not classic of SSPE, in view of raised cerebrospinal fluid (CSF) antimeasles antibody titers (1>:625), diagnosis of SSPE was kept. Antitubercular therapy was stopped and valproate was started for myoclonus. At the 6-month follow-up, he has shown further deterioration in cognitive functions, though severity of myoclonic jerks was under control.

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Table 1: Spectrum of abnormal MRI findings in subacute sclerosing panencephalitis (SSPE)

MRI findings in subacute sclerosing panencephalitis (SSPE)	T2 and FLAIR hyperintensities in bilateral occipital and parietal regions involving both gray and white matters are common findings
	Asymmetric white matter abnormalities are common followed by basal ganglionic and cortical gray matter involvement
	Edema, mass effect, and contrast enhancement are unusual
	Pial and gyral enhancement is very rare

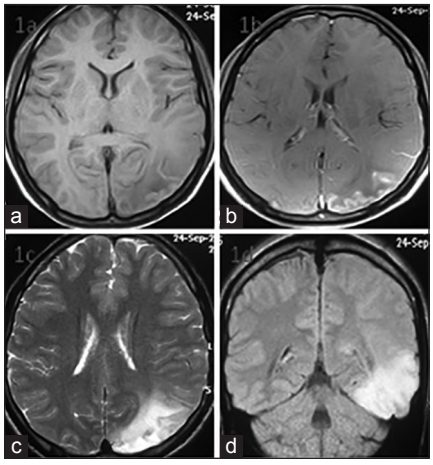

Figure 1: Axial T1-weighted plain (a) and contrast (b) showing left parieto-occipital gyral and meningeal enhancement; T2 (c) and coronal FLAIR (d) images showing hyperintensities involving the same region

Discussion

MRI is quite sensitive in detecting abnormalities in SSPE. However, the MRI abnormalities in SSPE are not well described in literature. The commonest abnormality on MRI includes signal changes (hyperintensities on T2-weighted and FLAIR sequences) in bilateral occipital and parietal regions involving both gray and white matter [Table 1]. Edema, mass effect, and CE are unusual.[1-3] There are two large series that have described imaging abnormalities in SSPE. In one series, CT scan was carried out in 42 patients and no enhancement was observed. In same series, MRI brain (N = 44) revealed white matter abnormalities, basal ganglionic, and cortical gray matter involvement in 75%, 40%, and 25% of patients, respectively.[4] In another series (N = 26), most common abnormalities were seen in parietal and temporal lobes (both gray and white matter), which were asymmetrical in approximately 33%. CE was seen only in two MRIs.[5] In addition to the two cases described earlier, there are isolated case reports of CE on neuroimaging in SPPE.[6-8]

Our case adds further to repertoire of heterogeneity of imaging findings in SSPE as is obvious from above discussion. Only two cases of both pial and gyral enhancement in same patient have been reported so far,[5] while the combination of remarkable asymmetry as well as gyral and pial enhancement as in our case is not reported. The likely explanation for these findings in our patient is intense inflammatory necrosis of brain parenchyma with disruption of blood-brain barrier (BBB) and extension of inflammatory process into the meninges. The necrosis may be caused either by severe viral infection *per se* or due to immune response or due to both the process occurring simultaneously. Myoclonic jerks in our patient were dominantly right-sided that correlated very well with MRI findings, thereby strengthening the previous reports that suggested similar correlation.[5]

Conclusion

To conclude, the spectrum of radiological changes in SSPE is ever expanding, and one needs to keep this not so uncommon entity in mind to avoid unnecessary diagnostic errors. A high index of suspicion especially in the third world countries, knowledge of rare imaging findings plus a proper clinical and electroencephalogram (EEG) correlation will go a long way in diagnosing this rare condition.

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Conflicts of interest
There are no conflicts of interest.

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