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Case report

Interesting MRI finding in SSPE – a case report

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Abstract

Introduction: 10% of the subacute sclerosing panencephalitis (SSPE) presents with atypical features.

Aim: SSPE is rare chronic encephalitis caused by persistent infection with defective measles virus. Characteristic MRI findings include signal changes (T2W and FLAIR hyperintensities) in bilateral occipital and parietal regions involving both gray and white matter. Early involvement of cerebellum and brainstem is not common.

Case study: A 17-year-old male presented with complaints of recurrent seizures, slow walking, and behavioral abnormality. Neurological examination revealed cogwheel rigidity in all four limbs. MRI of the brain revealed asymmetrical cortical and subcortical altered signal intensities (T2W and FLAIR hyperintensity and T1W hypointensity) involving temporal and occipital lobes bilaterally (left more than right) with diffuse atrophy of cerebrum and cerebellum.

Results and discussion: Early onset extrapyramidal features, seizures without myoclonus with MRI finding of posterior predominant asymmetrical cortical and subcortical signal abnormality is uncommon in SSPE.

Conclusions: A high index of clinical suspicion for SSPE has to be maintained in patients haling from endemic areas, unvaccinated individuals presenting with seizures, behavioral abnormality, and extrapyramidal features.

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1. INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a slow virus infection, a rare chronic encephalitis caused by persistent defective measles virus infection of the central nervous system.[1] The disease develops several years after measles infection and presents with altered behaviour, myoclonus, mental deterioration, seizures, extrapyramidal dysfunction and abnormal vision.[2] Saha et al.[2] in 2011 reported an annual incidence of 21 cases per 1 000 000 population in India. Atypical presentation of the disease seen in 10% of the cases,[2–4] presents a stiff challenge to diagnose and a high index of suspicion is required in the endemic areas. There is no definite cure for SSPE. The combination of intraventricular interferon plus oral isoprinosine is effective in halting the progression of the disease.[5]

2. AIM

In this case report we are presenting the case of a young boy who presented with atypical clinical features and MR imaging demonstrating the early involvement of cerebellum and basal ganglia, which are a late feature of the disease.

3. CASE REPORT

A 17-year-old right-handed male born to non-consanguineously married couple presented to us with a 2-year history of recurrent seizures, slow walking, and prominent behavioural abnormality. Patient also complained of difficulty in focusing on objects, visual hallucination and palpitation. Parents reported decreased scholastic performance, difficulty in reading speaking, and decreased memory. Patient was being treated with levetiracetam 1000 mg/day since the last 6 months. Immunisation status against measles was unknown. There was no prior history of measles infection in patient or in the family.

Examination revealed a young male who was cheerful and oriented. On formal mini–mental state examination (MMSE) he scored 17/30 indicating cognitive impairment. Visual field and visual acuity were normal and there were no cranial nerve deficits. Cogwheel rigidity, dystonia was present in all four lower limbs. Patient also had alexia without agraphia. Fundoscopy was normal in both eyes. Routine investigations such as hemogram, erythrocyte sedimentation rate (ESR), urine routine, renal function test, liver function test, ECG, chest X-ray were normal. Serum IgG measles antibody levels was found to be 1679 U/mL, grossly elevated. Serum total IgG level was 1230 mg/dL (normal: 700–1600 mg/dL). Cerebrospinal fluid (CSF) analysis showed no cells with protein 52 mg/dL, sugar 58 mg/dL. CSF IgG measles antibody levels was elevated (12537.6 U/mL). CSF total IgG was 4.09 mg/dL (normal: 0–3.4 mg/dL). CSF/serum quotient reference was 11.1 (normal: >1.5 is positive).

Electroencephalogram (EEG) showed background activity in the α range of 9 Hz to 10 Hz and generalised periodic complexes one in every 3–6 s along with spike-waves and sharp transients.

MRI brain screening study (Figure) was suggestive of asymmetrical cortical and subcortical altered signal intensities (T2W and FLAIR hyperintensity and T1W hypointensity) involving temporal and occipital lobes bilaterally (left more than right) with diffuse atrophy of cerebrum and cerebellum.

At presentation patient's behavioural abnormality was attributed due to levetiracetam, hence it was tapered off. Simultaneously lamotrigine was started after which patient showed some improvement in terms of decreased irritability and stable mood. With EEG and positive measles serology, diagnosis of SSPE was confirmed. Extrapyramidal features subsided with introduction of DOPA and carbidopa quite readily. Patient was started on isoprinosine 2500 mg/day. Treatment with interferon alpha was not considered due to financial constraints currently, the patient is under follow up.

4. RESULTS AND DISCUSSION

Diagnostic criteria for SSPE was described by Dyken.[6] The diagnosis of SSPE requires the fulfilment of at least three of the five criteria:

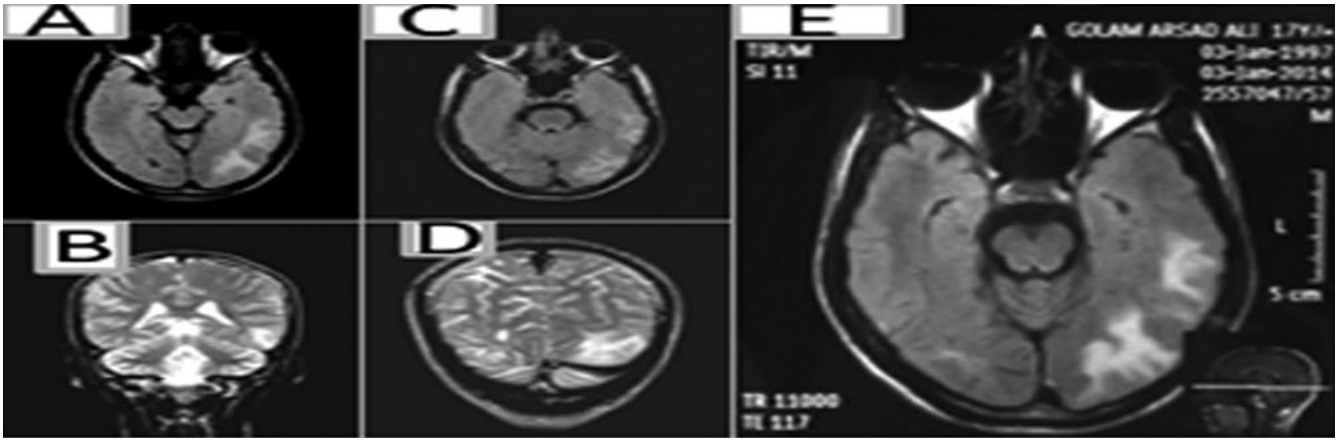

Figure 1. MRI brain screening study: T1W (A and C); T2W (B and D); FLAIR (E).

(1) a typical clinical picture: Personality and behavioural changes, worsening school performance, followed by myoclonic seizures, paresis, dyspraxias, memory impairment, language difficulties, blindness, obtundation, stupor, and coma;
(2) characteristic EEG changes;
(3) elevated CSF globulin levels greater than 20% of total CSF protein;
(4) raised titres of measles antibodies in blood and CSF;
(5) typical histopathological finding in brain biopsy or autopsy.

About 10% of the patients present with atypical features and imaging features.[3,4] Usual course of SSPE involves insidious onset development of personality changes, intellectual disability in the form of difficulties at school. This stage can last from weeks to years. Eventually patient develops myoclonus, dementia along with long tract involvement. Finally patient develops severe neurological dysfunction in the form of decorticate rigidity, autonomic dysfunction. Disease progresses over period of 1–3 years culminating in death of the patient.[1] Combination of intraventricular interferon plus oral isoprinosine is effective in halting the progression of the disease.[5]

Our patient had progressive symptoms, typical EEG findings and raised measles antibody titre both in serum and in CSF which are highly suggestive of SSPE.

Patient presented with seizures, cogwheel rigidity, and inability to walk which are usually seen in the later stages of the disease.[1] Characteristic findings on MRI includes signal changes (T2W and FLAIR hyperintensities) mainly in bilateral occipital and parietal regions involving both gray and white matter.[7] Asymmetric white matter abnormalities are common followed by basal ganglionic and cortical gray matter involvement.[7] Presentation with basal ganglia and cerebellar involvement is rare. In our patient MRI (Figure) showed asymmetrical cortical and subcortical altered signal intensities (T2W and FLAIR hyper intensity and hypointensity) involving temporal and occipital lobes bilaterally (left more than right) with diffuse atrophy of cerebrum and cerebellum. Involvement of predominantly posterior cortical and subcortical signal abnormality is uncommon finding in SSPE.

5. CONCLUSIONS

Early onset extrapyramidal features and seizures without myoclonus with MRI finding of posterior predominant asymmetrical cortical and subcortical signal abnormality is uncommon in SSPE.

Conflict of interest

We hereby declare that there is no conflict of interest.

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