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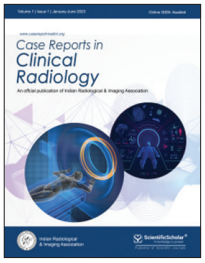

Case Report

Atypical imaging and clinical presentation of fulminant subacute sclerosing panencephalitis

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a late progressive fatal central nervous system sequelae of measles infection. The diagnosis is made based on clinical and electroencephalography (EEG) findings and confirmed by elevated titers of anti-measles antibodies in cerebrospinal fluid. The usual clinical presentation is in the form of progressive behavioral change, cognitive decline, and myoclonic jerks with some cases presenting in an atypical manner in the form of ataxia, epilepsy, and stroke. EEG is quiet characteristic in the form of periodic discharges of slow wave complex. Magnetic resonance imaging (MRI) early in the disease is usually normal with abnormal scans showing focal leukodystrophy, predominantly in the posterior cerebral white matter, and development of cortical atrophy with disease progression. Cord involvement is not common in SSPE. We report a child with SSPE who had initial presentation with cerebellar ataxia, acute progression of encephalitis, and atypical EEG findings. MRI brain on the initial presentation showed very subtle and focal abnormality which later progressed to have disseminated brain lesions and dorsal cord myelitis and further in the disease course showed cerebral venous sinus thrombosis. This case report emphasizes that SSPE can have very atypical presentation with rapid deterioration and can mimic as acute encephalitis or demyelinating disease.

Keywords: Subacute sclerosing panencephalitis, Ataxia, Dorsal cord, Diffusion restriction, Sinus thrombosis

INTRODUCTION

Subacute sclerosing Panencephalitis (SSPE) is a fatal CNS complication of measles infection with a chronic progressive course. Usually there is a history of incomplete immunization or measles infection in childhood, but it can occur in a non-infected, immunized child as well. The diagnosis is based on clinical presentation, EEG findings and elevated anti measles titters in CSF. The typical clinical presentation is in form of behavioral changes and myoclonic jerks, but atypical presentations have been described. We report a case of a child who had no history of measles infection, presented with atypical clinical, imaging and EEG findings and progressed very rapidly to death.

CASE REPORT

A 7-year-old boy presented with complaints of fever and imbalance while walking for 6–7 days. The parents noticed the child swayed to one side while sitting and had abnormal behavior with irrelevant laughter. The child was conscious, oriented, and had normal speech. He was ataxic and his lower limb reflexes were brisk. There was no history of measles infection, and he was immunized with MMR vaccine. During the first 3 days of admission, the child developed excessive sleepiness. Magnetic resonance imaging (MRI) brain, electroencephalography (EEG), and

cerebrospinal fluid (CSF) analysis were normal. A possibility of acute cerebellitis/autoimmune encephalitis was suspected, and pulse methylprednisolone was given for 5 days, followed by oral steroids. The child was discharged as there was significant improvement in sensorium. After 3 days, he was again brought with altered sensorium. He had new-onset left upper limb dystonia. EEG showed generalized slowing (Theta-delta waves) with the right posterior intermittent epileptiform discharges. Repeat MRI brain done after 20 days showed subtle restricted diffusion with T2 hyperintensity in bilateral middle cerebellar peduncles [Figure 1]. A possibility of autoimmune encephalitis or demyelinating disease was considered. Anti MOG Ab test, CSF autoimmune panel (NMDA, LGI1, CASPR2, and GABA), and CSF biofire panel were negative. He developed herpes zoster in the meantime, so steroids were tapered, and injection acyclovir started for a period of 10 days with IVIg at the dose of 2 g/kg. A repeat MRI brain after 27 days showed extensive confluent asymmetric signal abnormality in bilateral cerebral white matter with parieto-occipital predominance, involvement of corpus callosum, bilateral thalami, posterior limb of internal capsule, crus cerebri, pons, and middle cerebellar peduncles [Figure 2a] with patchy restricted diffusion [Figure 2b]. Subtle enhancement was seen along bilateral 3rd, left 6th, and bilateral 7th–8th nerve complexes [Figure 2c]. There was mild cerebral volume loss as compared to the previous MRI scan. Spine images showed non-enhancing abnormal T2 hyperintense signal in the anterior and lateral aspect of the spinal cord from D1 to D6 vertebral levels; suggestive of myelitis. [Figure 2d]. A suspicion of subacute sclerosing panencephalitis (SSPE) was raised with subsequent positive CSF measles antibody from two separate laboratories. The child deteriorated with poor sensorium and neurogenic respiration requiring mechanical ventilation. He was started on anti-measles virus medications (Ribavirin, Amantadine, Inosiplex, and Lamivudine). On the last MRI brain study, there was increase in the extent of white matter signal abnormality with the involvement of frontal lobes as well [Figure 3a]. There was new development of the left transverse, sigmoid, and internal jugular vein thrombosis, for which

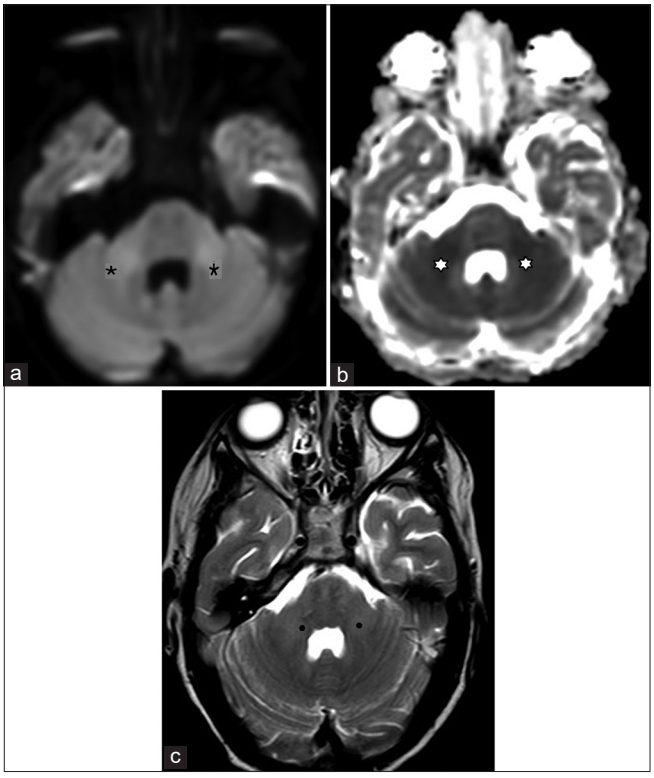

Figure 1: A 7-year-old boy presented with fever and imbalance while walking. First MRI brain shows (a) mild hyperintensity [asterisk] on diffusion-weighted images in bilateral middle cerebellar peduncles. (b) Low signal on ADC images [star]. (c) High signal on T2-weighted images [dots].

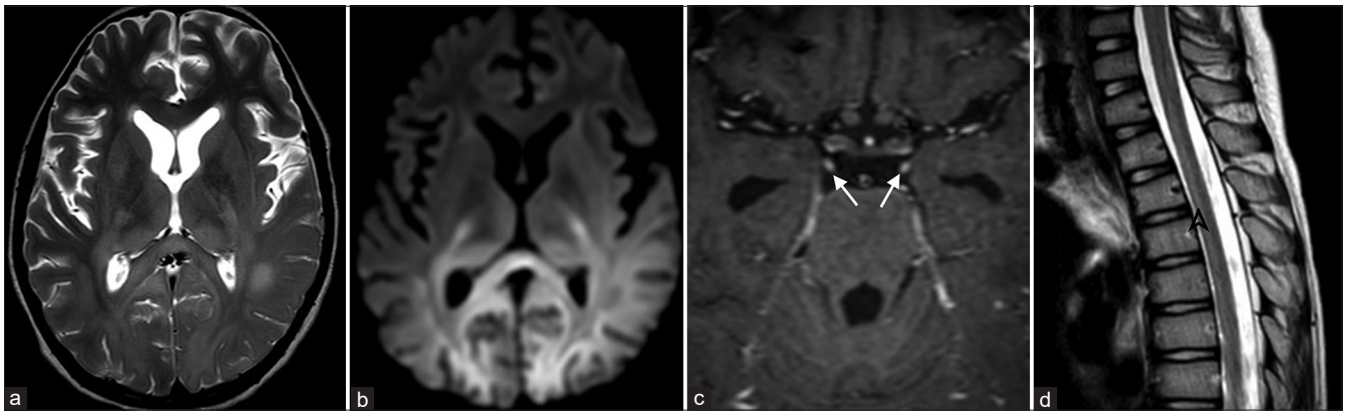

Figure 2: New-onset progressive dystonia and altered sensorium. Follow-up MRI brain shows: (a) Abnormal T2 hyperintense signal in bilateral parieto-occipital white matter, thalami, posterior limb of internal capsule, and corpus callosum, (b) restricted diffusion in the involved brain parenchyma, (c) post contrast 3DT1 images show enhancement along the cisternal segments of 3rd cranial nerves (arrows), and (d) intramedullary T2 hyperintensity in anterior part of dorsal cord [arrow head].

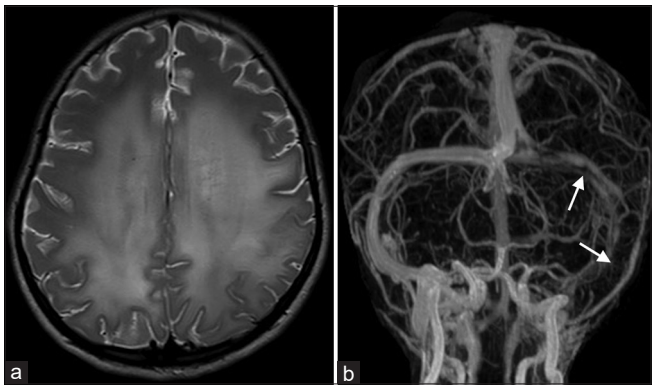

Figure 3: Last MRI brain performed due to further neurological deterioration shows: (a) Increase in the white matter involvement with abnormal frontal white matter. (b) Left side transverse, sigmoid, and jugular venous thrombosis (arrows).

subcutaneous low molecular weight heparin was started [Figure 3b]. The patient developed autonomic instability, continued to worsen, and eventually succumbed to illness.

DISCUSSION

Measles is a paramyxovirus which causes acute febrile exanthematous illness early in childhood. Central nervous system measles infection is a rare occurrence and can present in one of the four forms: Primary measles encephalitis, acute post infectious measles encephalomyelitis, measles inclusion body encephalitis, and subacute sclerosing encephalitis.[1] Patients generally have a history of measles infection in childhood or incomplete immunization, but few cases of SSPE have been described in vaccinated patients with negative history of infection.[2] SSPE is associated with a mortality rate of 95%, the average life span after the initial presentation being about 3.8 years, with a range of 45 days to 12 years.[3] In fulminant cases, the overall survival is few weeks to months, and they are likely to show atypical presentation. Typically, the child usually presents with progressive cognitive, behavioral abnormalities, and myoclonic jerks. In atypical cases, the child may present with stroke, seizures, ataxia, or Parkinsonian features.[4,5]

As described by Cece *et al*., MRI changes in SSPE are non-specific, but most commonly occurs as posterior predominant T2 hyperintensities in periventricular and subcortical white matter with less common and later involvement of corpus callosum, basal ganglia, brainstem, and cerebellum.[6] Involvement of just the brainstem in SSPE has been described,[5,7] but isolated involvement of middle cerebral peduncles initial in the course of illness has not been reported to the best of our knowledge. Brainstem involvement was found to be a part of progressing supratentorial disease in a study by Alkan *et al*.[8] and Anlar *et al*.[9] The brainstem lesions in their patients showed increased ADC values suggesting neuronal loss and gliosis. However, in our case, there was acute restricted diffusion suggesting cytotoxic injury. Such DW restriction has been described to be indicative of fulminant SSPE in a few case reports.[10,11]

Involvement of spinal cord in SSPE is also rare, described only in few case reports[12-13] as cervical cord involvement. In our case, thoracic cord was involved with abnormal signal in the anterior and lateral cord. Furthermore, cerebral venous thrombosis and cranial nerve enhancement have not been described in patients with subacute sclerosing pan encephalitis.

Lack of literature on these findings might be because they represent later manifestations of the disease, and such patients are not always serially imaged once the diagnosis is made.

CONCLUSION

Atypical MRI findings of early involvement of middle cerebellar peduncles with rapid progression, involvement of dorsal cord and cranial nerves should raise the suspicion of SSPE.

MCQs

1. Which of the following MR brain findings can be seen in a patient suspected for SSPE?
   a. Normal Brain MR
   b. Oval lesions with restricted diffusion
   c. Microhemorrhages in basal ganglia
   d. Bilateral symmetrical frontal white matter involvement

Answer Key: a

2. The lesions in SSPE on MR Brain can show all except
   a. DW restriction
   b. Meningeal enhancement
   c. Incomplete ring enhancement
   d. High ADC signal

Answer Key: c

3. The investigation which confirms the diagnosis of SSPE is:
   a. MRI Brain
   b. CSF measles IgG
   c. EEG
   d. It's a diagnosis of exclusion

Answer Key: b

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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