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Case Study

Subacute sclerosing panencephalitis presenting as acute cerebellar ataxia and brain stem hyperintensities

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ABSTRACT

Background: Subacute sclerosing panencephalitis is a devastating neurodegenerative disease with a characteristic clinical course. Atypical presentations may be seen in 10% of the cases.

Aims: To describe the atypical clinical and radiological features of SSPE in a child form endemic country.

Methods: A 5-year-old boy presented with acute-onset cerebellar ataxia without associated encephalopathy, focal motor deficits, seizures or cognitive decline. He had varicella-like illness with vesicular, itchy truncal rash erupting one month prior to the onset of these symptoms. He underwent detailed neurological assessment, relevant laboratory and radiological investigations.

Results: Neuroimaging revealed peculiar brain stem lesions involving the pons and cerebellum suggestive of demyelination. With a presumptive diagnosis of clinically isolated syndrome of demyelination, he was administered pulse methylprednisolone (30 mg/kg/day for 5 days). Four weeks later he developed myoclonic jerks. Electroencephalogram showed characteristic periodic complexes time-locked with myoclonus. CSF and serum anti-measles antibody titres were elevated (1:625).

Conclusion: Our report highlights that subacute sclerosing panencephalitis can present atypically as isolated acute cerebellar ataxia and peculiar involvement of longitudinal and sparing of transverse pontine fibres. The predominant brainstem abnormalities in the clinical setting may mimick acute demyelinating syndrome. Hence, it is important to recognize these features of subacute sclerosing panencephalitis in children, especially in the endemic countries.

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1. Introduction

Subacute sclerosing panencephalitis (SSPE) is a devastating neurodegenerative disease secondary to persistent, aberrant wild measles-virus infection in the central nervous system. The characteristic clinical manifestations of SSPE commonly begin as progressive intellectual deterioration, behaviour problems, periodic myoclonic jerks, seizures and/or vision abnormalities.[1] Atypical presentations of SSPE may occur in 10% of cases; the spectrum of atypical clinical and radiological manifestations in children is still expanding and needs high suspicion for accurate diagnosis.[1,2]

Though gait instability commonly accompanies the drop-attacks and encephalopathy in SSPE, initial presentation as acute cerebellar ataxia in the absence of any myoclonic jerks, vision changes or cognitive decline is most unusual. We describe a 5-year-old boy in whom an acute-onset ataxia preceding the characteristic myoclonic jerks by 4 weeks was the initial manifestation of SSPE. This case further highlights the atypical clinical and radiological features of SSPE in children that should always be borne in mind, especially in endemic countries.

2. Case report

A 5-year old, previously well and developmentally normal boy presented with acute onset, gait instability and slurring of speech for the past two weeks. He also developed shaking of hands while reaching out, looseness of the limbs and needed support for walking. The symptoms were preceded by a minor upper respiratory tract infection. There was no history of seizures, behavioural abnormalities, vision impairment, involuntary movements, altered sensorium or history suggestive of cranial nerve palsies. In the recent past, he had an illness with varicella-like vesicular, itchy truncal rash erupting 1 month prior to onset of symptoms. There was no history of drug/toxin exposure, trauma or recent vaccination. He was immunized for measles at the age of 9 months. Perinatal and family history was not contributory.

On examination, he was alert and oriented to time, place and person, memory and intellect were intact and speech was non-fluent and slurred but was comprehensible and coherent. He had truncal ataxia, mild lower limb hypotonia, past-pointing, intentional tremors, impaired finger-nose and knee-heel manoeuvres. His stance was broad-based and gait was ataxic with frequent swaying and occasional falls. Rest of

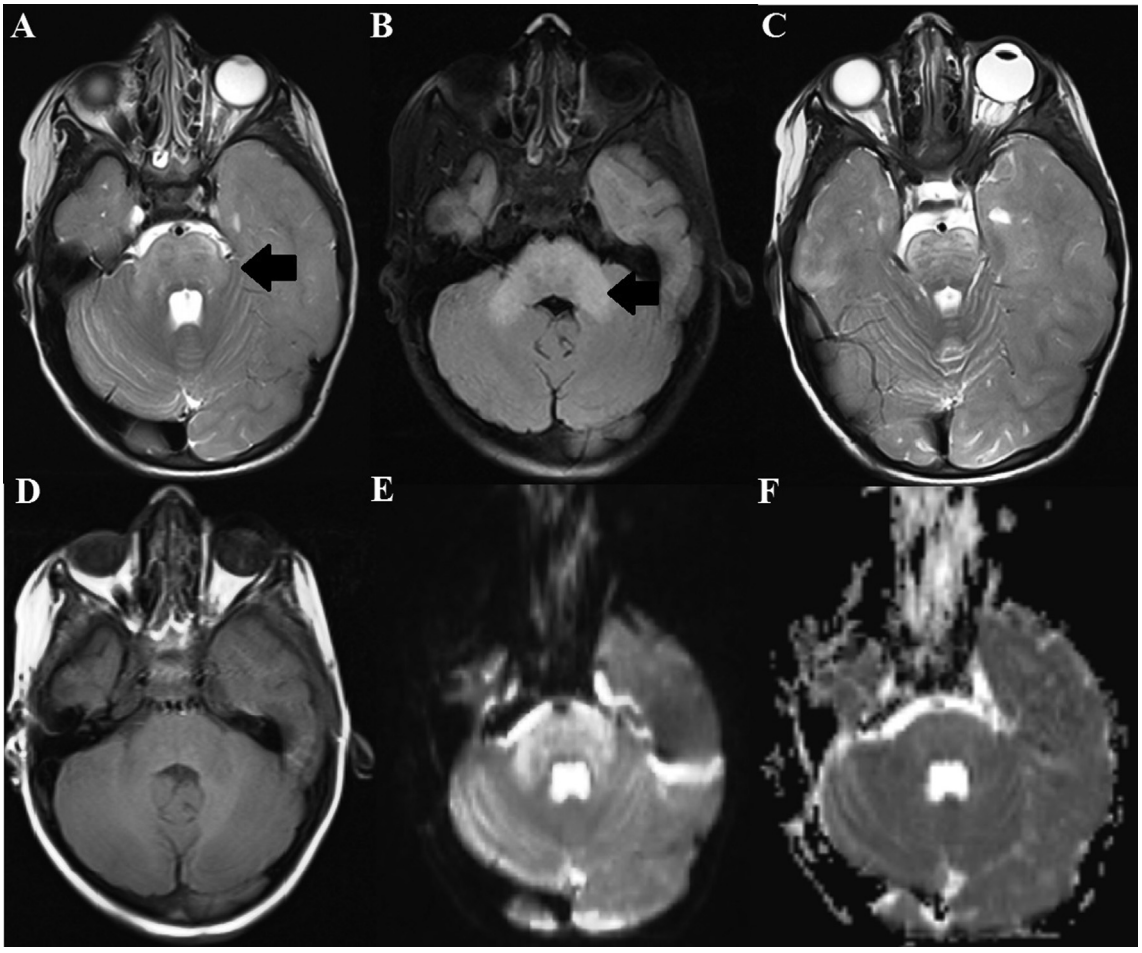

Fig. 1 – Axial T2-weighted (A and C) and fluid-attenuated inversion recovery (B) MRI images showing hyperintense lesion in the pons and middle cerebral peduncles (A and B, arrows). There is also involvement of ventral midbrain (C). The lesions are hypointense on T1-weighted images (D). Diffusion restriction is seen in the pons and middle cerebral peduncles (E, diffusion weighted image with b = 1000 and F, apparent diffusion coefficient image).