Subacute Sclerosing Panencephalitis Presenting as Multiphasic demyelination: A Diagnostic Challenge

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BACKGROUND AND AIM

Subacute sclerosing panencephalitis (SSPE) is a late complication of Measles infection.

The incidence of SSPE is approximately 4–11 cases per 100 000 cases of measles.

Traditionally, the latent period between measles infection and onset of symptoms of SSPE is 6–8 years and the classical age of presentation is 8–11 years.

It results from persistent measles virus infection, often due to mutations in the viral matrix (M) protein that prevent normal viral budding and clearance.

Adult cases present with a more atypical presentation and aggressive course compared with childhood SSPE.

Here we describe an adult SSPE case which presented with features of relapsing and remitting pattern and an aggressive course.

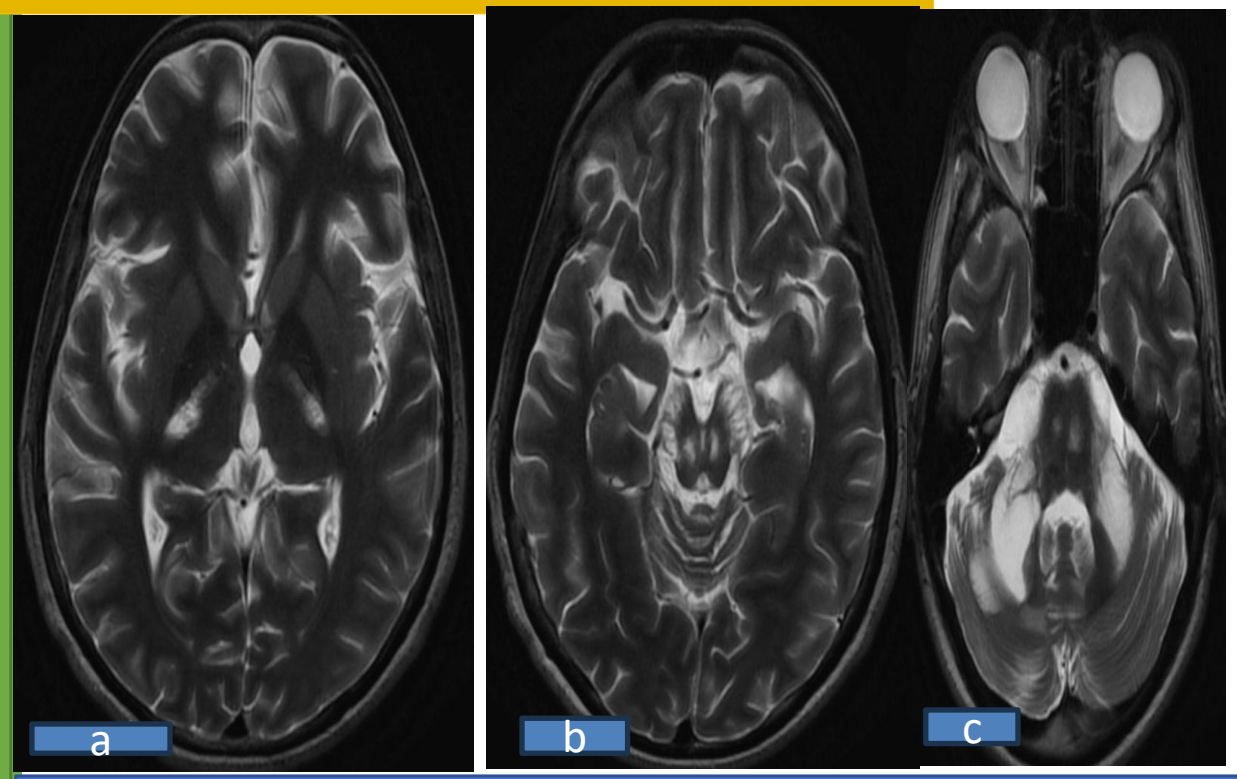

FIGURE-1
The axial T2 (a-c) images shows symmetrical hyperintense signal involving bilateral internal capsule, crus cerebri, medial longitudinal fasciculus and bilateral middle cerebellar peduncle.

CASE DETAILS

A 29-year-old healthy male developed acute right-sided ataxia, diplopia, and hemiparesis after a febrile illness. MRI showed right cerebellar and brainstem lesion , and CSF revealed lymphocytosis with mild protein elevation. Serum Mumps IgM and nasopharyngeal Rhino/Enterovirus PCR were positive. He was diagnosed with acute disseminated encephalomyelitis (ADEM) at another hospital and was treated with Methylprednisolone, and improved with minimal residual deficit.(mRS-2)

After 2 months, the patient relapsed with left hemiparesis and visual symptoms, which progressed over one month and then stabilized (mRS-3). MRI showed right cerebellar encephalomalacia/gliosis, bilateral corticospinal tract hyperintensities, and midbrain/peduncular changes . No treatment was given during this phase.

Patient presented to our hospital ,5 month after onset of second symptoms During hospital stay , symptoms further progressed with visual blurring, dysphagia, dysphonia, and bilateral limb weakness (MRS-5, with feeding tube and mechanically ventilation) and was treated with Methylprednisolone and later Plasmapheresis . Repeat MRI shows axial T2 weighted symmetrical hyperintense signal involving bilateral internal capsule, crus cerebri, medial longitudinal fasciculus and bilateral middle cerebellar peduncle. The diffusion weighted images and corresponding ADC shows faint peripheral diffusion restriction. The post contrast scans does not reveal any obvious post contrast enhancement. (FIGURE-1,2)

JC virus and extended viral panels were negative. CSF study suggestive of 6 WBC , with lymphocyte (100%) , protein (33.7) and glucose 57 , with corresponding plasma RBS of 110, gram stain, bacterial culture ,fungal element and India ink negative.

Autoimmune, vasculitis, and sarcoid workup (HLA-B54, ANA, ACE, pathergy, Lyme, NMO/MOG/MS) were unremarkable. However, CSF was positive for measles antibody (1:22), with CSF : serum ratio nearing equivalence(FIGURE-3,4), PCR was negative. EEG showed diffuse theta slowing without any periodic complexes (Figure 5). Final impression of atypical SSPE was kept and treated with Ribavirin and Levamisole.

OUTCOME at follow up: Patient came after 2 month with a static disease course, was bed-bound, and required tracheostomy and Ryle's tube for airway and nutritional support (mRS 5).

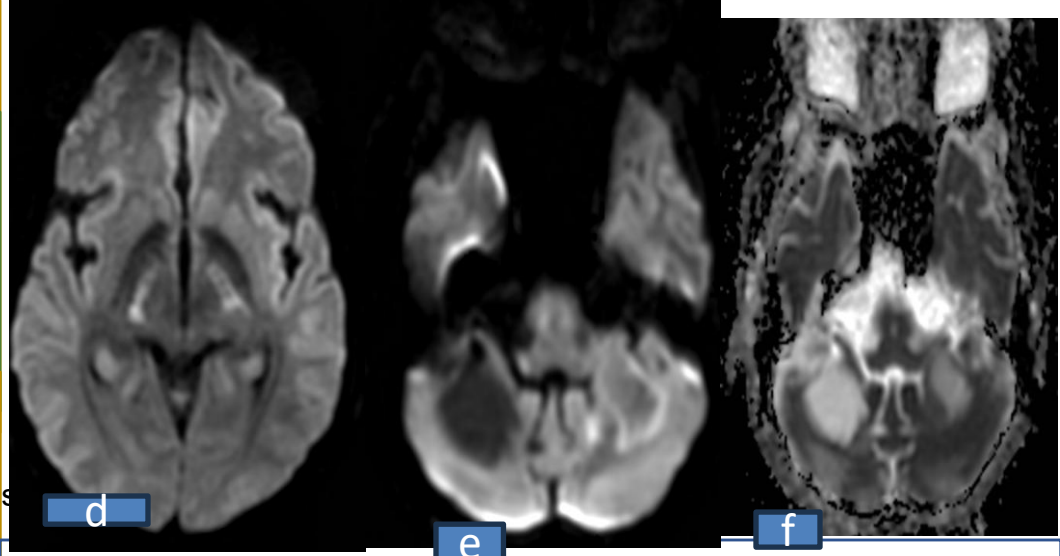

FIGURE-2
The diffusion weighted images (d-e) and corresponding ADC (f) shows faint peripheral diffusion restriction

S. No.	Test	Result	Result value	Reference Range: Non-Reactive	Reference Range: Reactive	Kit Reference
25.	Anti- Measles IgM (Serum)					
26.	Anti- Measles IgM (CSF)					
27.	Anti- Measles IgG (Serum)	Positive				
28.	Anti- Measles IgG (CSF)					
29.	Mumps IgM (Serum)					
30.	Mumps IgM (CSF)					
31.	Varicella Zoster IgM					
32.	Varicella Zoster IgG					
33.	Epstein Barr Virus IgM					
34.	Epstein Barr Virus IgG					
35.	Fecal Rotavirus Antigen					
36.	Adeno Antigen					
37.	Adeno IgM					
38.	Adeno IgG					
39.	Astro Antigen					
40.	Noro Antigen					
41.	Parvo B-19 IgM					

FIGURE-3

DISCUSSION

Our patient initially presented with acute focal neurological deficits following a febrile illness, leading to a diagnosis of ADEM. Initial steroid responsiveness and radiological feature further supported this diagnosis.

However, the relapse with new tract-based MRI changes, persistent progression despite immunotherapy, and the emergence of visual and bulbar symptoms prompted reconsideration of the diagnosis.

Unlike classical SSPE, this case lacked hallmark features such as myoclonus or periodic EEG patterns. The CSF measles antibody was positive but with a near-equivalent serum ratio, which is atypical compared to the high intrathecal synthesis seen in classical SSPE.

CONCLUSION

This case highlights an atypical adult-onset presentation of SSPE, initially mimicking post-infectious ADEM and autoimmune demyelination. The progressive relapsing course, evolving MRI changes, and eventual demonstration of intrathecal measles antibody synthesis confirmed the diagnosis. It emphasizes the need to suspect SSPE even in adults with unexplained progressive encephalopathy with posterior white matter changes

REFERENCE.

1- Wendorf KA, Winter K, Zipprich J et al. Subacute sclerosing panencephalitis: The devastating measles complication that might be more common than previously estimated. Clin Infect Dis 2017.

2-Goraya J, Marks H, Khurana D, et al. Sub acute sclerosing pan encephalitis (SSPE) presenting as acute disseminated encephalomyelitis in a child. J Child Neurol 2009.

Test Report

Test Name	Results	Units	Bio. Ref. Interval
MEASLES INTRATHECAL IGG ANTIBODY TEST, CSF, EIA (EIA)			
CSF	22.96	U/mL	
Serum	17.85	U/mL	
CSQ (units CSF/units SERUM)	1.29	U/mL	<1.3

FIGURE-4

FIGURE-5