

Diagnostic dilemmas in fulminating sub-acute sclerosing pan-encephalitis (SSPE) with atypical presentation

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Abstract

Subacute sclerosing panencephalitis (SSPE) is a progressive, fatal disease of the central nervous system caused by a persistent measles virus in the brain. It is clinically characterized by insidious onset of intellectual deterioration and behavioral changes followed by myoclonic jerks and eventually complete neurologic deterioration. In about 10% of patients, the clinical manifestations of SSPE are not typical and that may cause a delay in the diagnosis. We report here 3 cases of SSPE with atypical presentations. Bilateral vision loss, hemiparesis, ataxia and acute encephalopathy with focal seizures were respectively the presenting symptoms at the onset of disease. The typical periodic EEG complexes in two patients and positive CSF measles IgG antibody led to the diagnosis of SSPE.

Keywords: SSPE; atypical presentation; periodic complexes; measles antibody

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) is a form of encephalitis that is reported to present years after infection with the measles virus. This complication occurs in children who are infected with measles virus at a young age. The risk of developing SSPE after childhood measles is estimated to be 1 in 25,000 in the general pediatric population and 1:5500 in children infected before 1 year of age. Previously it was considered as a disease of children but literature suggests that there is no age limit. It may be underdiagnosed due to atypical presentation.^{1,2} Patients with classical SSPE present with myoclonic jerks, behavior and motor symptoms along with periodic complexes in electroencephalography (EEG). But for atypical cases other differentials have to be ruled out.¹

We report here three patients of SSPE with atypical presentation, including hemi-dystonia, vision loss, ataxia and focal seizures.

CASE REPORTS

Patient 1

A 17-year-old boy presented in emergency department with sudden bilateral vision loss and hemi-dystonia. He was a previously healthy vaccinated child. He had Glasgow coma scale

(GCS) of 15/15, bilateral vision loss with intact pupillary and corneal reflex. He had left sided weakness with dystonia and exaggerated deep tendon reflexes in all four limbs with upgoing plantar response on the left side. Direct fundoscopic examination showed bilateral pale optic disc.

He was evaluated for possibility of stroke and encephalitis. Magnetic resonance imaging (MRI) brain with contrast showed hyperintense area at bilateral occipital region on T1W1 and FLAIR images without diffusion restriction (Figure 1), suggestive of posterior reversible encephalopathy syndrome (PRES). MR angiography and MR venography were normal. An EEG showed left centro-parietal spike, sharp and slow waves (Figure 2). Within 3 days, he became disoriented. Cerebrospinal fluid (CSF) analysis showed only mildly raised protein. Work up for Wilson disease, HIV, syphilis, autoimmune disease, paraneoplastic syndrome and porphyria was negative. The differential diagnoses were shortlisted to autoimmune encephalitis and SSPE. Repeated EEG showed diffuse slowing. Latter his investigations for autoimmune encephalitis was negative and measles CSF IgG antibodies came back as positive. He was diagnosed as SSPE with atypical presentation.

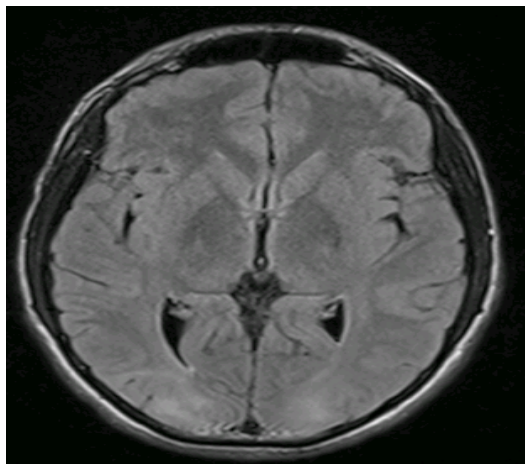


Figure 1. MRI brain FLAIR image of the 17 year old boy (Patient 1) presenting with sudden loss of vision and half body weakness at the time of admission. This image shows hyperintense signals in the bilateral occipital region suggestive of PRES.

Patient 2

This 15 years-old-girl came to emergency department with complaints of jerky movement of right side of body, deterioration in academic performance and emotional lability for the past four months. She had history of measles at age of one year. On examination, she was fully oriented and able to speak with labile mood. On examination, she had continuous myoclonic jerks at right side of body. There was right sided hemi-dystonia, power 2/5 on right side, with brisk reflex and up going plantar response. Her MRI brain, liver function test, antistreptolysin O (ASO) titer, thyroid stimulating hormone (TSH)

level and eye examination were all normal. EEG showed periodic complexes (Figure 3). CSF analysis was normal except positive oligoclonal band and measles IgG antibodies.

Patient 3

This seven years old child presented in emergency department with complaints of unable to walk for 20 days with history of one episode of loss of consciousness at school. He had history of measles at 1 year of age. On examination he had 4/5 muscle power, ataxic gait with brisk reflex and upgoing plantar response. EEG showed periodic complexes (Figure 4). His MRI brain and CSF analysis were normal except a positive CSF oligoclonal band and measles IgG antibodies.

DISCUSSION

Classic SSPE runs a stereotypic, four-stage protracted course over many months to years.² Our first patient with atypical presentation had acute bilateral sudden visual loss. His clinical features and EEG finding were atypical of SSPE. Most patients presenting with atypical signs and symptoms of SSPE had history of myoclonus and burst suppression in EEG at presentation or developed in the later part of their illness. But this was absent in our patient. This led to extensive work up. Previous report showed that cases of fulminating SSPE have atypical presentation and guarded prognosis.^{1,3-5} Sarkar *et al.* reported a case of SSPE with acute vision loss followed by vegetative state without myoclonic seizure and typical EEG finding.¹ Takayama *et al.* reported another case of SSPE presenting with sudden loss of vision, who progressed to vegetative state

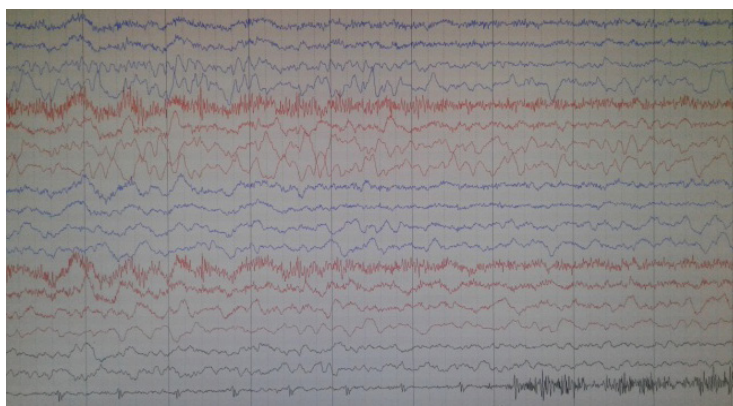


Figure 2. Electroencephalogram (EEG) findings of the same boy (Patient 1) showing Intermittent left centro parietal spike, sharp and slow waves and Slow posterior dominant rhythm (theta intermixed with delta activity). (Bipolar double banana montage, L then R)

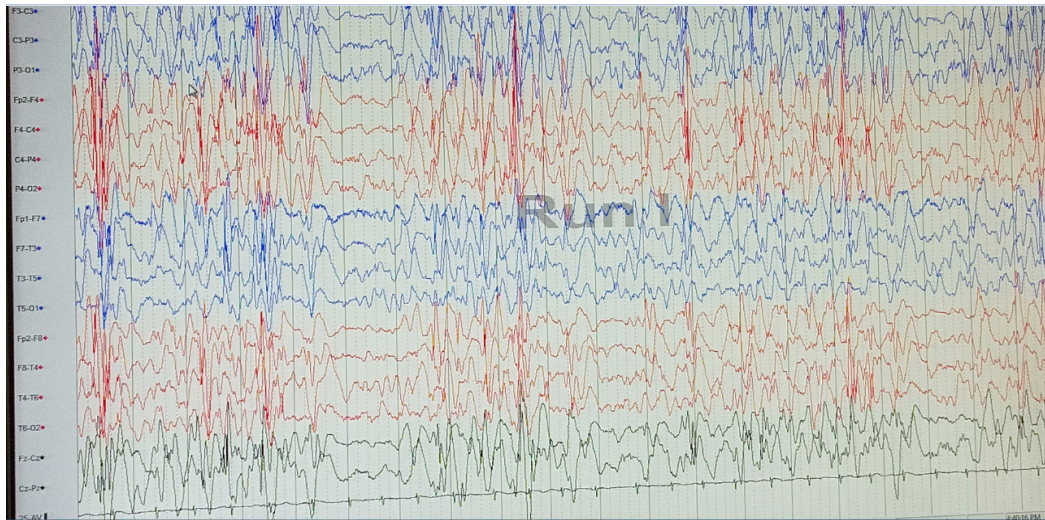


Figure 3. EEG of Patient 2 with hemidystonia showing periodic complex. EEG with sensitivity of 10 microvolt, HF filter 70 Hz, time base 20 sec/screen.

within two week without history of myoclonic jerks and periodic complexes in EEG.³ These two cases helped us to consider the diagnosis of SSPE. There were other reports of atypical EEG pattern of SSPE that were associated with adverse outcome suggesting that SSPE can present with atypical EEG.^{6,7} The negative investigations of autoimmune encephalitis, and the positive measles serology confirmed the diagnosis of SSPE. Literature review showed atypical form of SSPE occurring in about 10% patients with no defined clinical stages due to rapid clinical progression. Atypical features include unusual age of onset, visual loss, seizure and lack of SSPE-specific EEG

pattern with atypical fast progression of disease.¹⁻⁷

Our second patient was atypical as she presented with hemi-dystonia. However, she had myoclonic jerks and periodic complexes in EEG that favors SSPE. In our third patient, an EEG was performed because of suspected epilepsy. It showed the typical periodic complexes and this led towards the diagnosis of SSPE.

In conclusion, SSPE should be considered in any individual with unexplained neurological sign and symptom, especially in developing countries where measles is still a common problem.

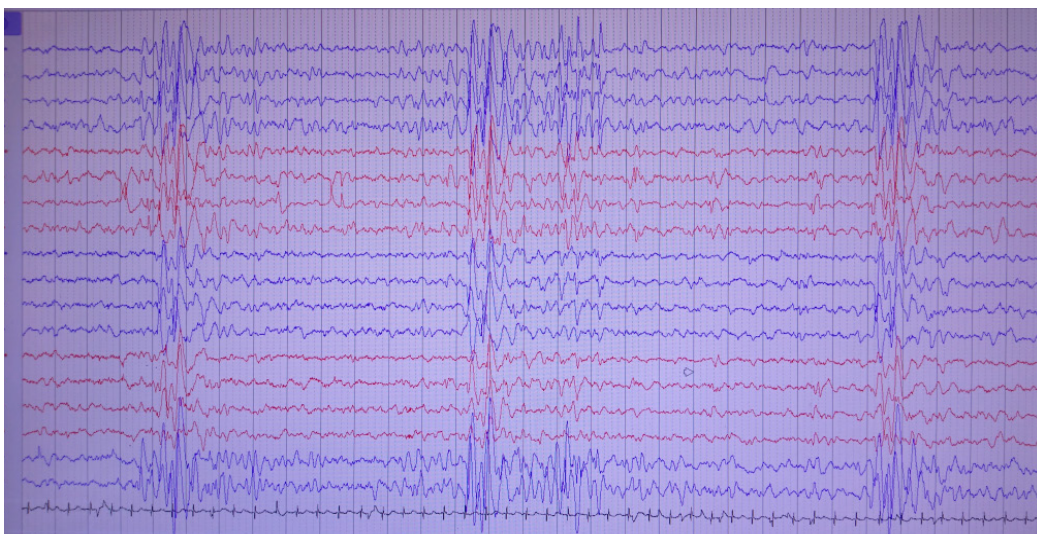


Figure 4. EEG of Patient 3 with ataxia showing periodic complex. EEG sensitivity of 15 microvolt, HF filter of 70 Hz, time base of 30 sec/screen. (Bipolar double banana montage, L then R)

Table1: Clinical spectrum of SSPE patients.

	Patient 1	Patient 2	Patient 3
Age of patient	17 years	15 years	4 years
P/H of Measles	No	1 year old	1 year old
Measles vaccination	yes	--	yes
Clinical Features	Vision loss	Focal fits	Walking difficulty
	Left sided hemiparesis	Emotional lability	LOC
	Hemi-dystonia	Hemi -dystonia	Ataxia
MRI Brain	Hyperintense signals	Normal	Normal
	In bilateral occipital region		
EEG findings	Centro-parietal spikes	Periodic complexes	periodic complexes
	Diffuse delta & theta slowing		
CSF OCB	Positive	Positive	Positive
CSF measles antibody	IgG +ve	IgG +ve	IgG +ve

P/H, Past history; --, not known; OCB, Oligoclonal bands

DISCLOSURE

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Conflicts of interest: None

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