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Case Report

Subacute sclerosing pan encephalitis with different perspective: an atypical case

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ABSTRACT

Subacute sclerosing pan encephalitis (SSPE) is a rare type of brain disease that happens when a changed version of the measles virus keeps infecting the brain. The symptoms start with changes in behavior, then seizures, trouble thinking, stiff muscles, and eventually, the person might go into a coma and die. A 7-year-old male patient presented in OPD with complaint of single episode of generalized tonic clonic seizures lasting for 30 seconds, followed by left sided upper limb and lower limb weakness along with myoclonic seizures. Patient was active and on walking patient was swaying towards right side. Supportive management with anti-epileptics given but patient has poor prognosis. Our case showed three unusual signs of SSPE: starting at a young age, sudden loss of balance showing a fast disease progress, and happening in a child who had been vaccinated but didn't have a known measles infection. We think the child might have had a mild measles infection that wasn't noticed by the parents, which could have led to SSPE.

Keywords: SSPE, Atypical encephalitis, Myoclonus jerks, Neuroregression

INTRODUCTION

Prolonged exposure to a mutant strain of the measles virus can cause SSPE, a rare and gradually degenerative neurological illness. The first signs of SSPE are usually behavioral abnormalities and intellectual deterioration that appear years after the initial measles infection. As the illness worsens, individuals may develop dementia, coma, and widespread seizures, which can be fatal within three years after the onset of symptoms. Although they are uncommon, extended spontaneous remission has been documented. The diagnosis of SSPE is verified by a defined clinical course and certain markers, such as the presence of measles antibodies in the CSF, electroencephalogram (EEG), or histological results from a postmortem examination or brain biopsy. In order to accurately diagnose and treat SSPE, these factors help differentiate it from other neurological diseases. People in developing nations with little resources, like India, are disproportionately affected by SSPE. Per 100,000 occurrences of measles, SSPE is linked to between 4-11 cases, according to the world health organization.[1] Those who get measles at a very young age are more at risk of developing SSPE. Furthermore, after receiving a measles vaccination, risk of SSPE is significantly reduced, with an estimated 0.7 cases per 1 million vaccine recipients during a 6-year period.[2] Significance of measles vaccination in lowering the prevalence of SSPE is highlighted by these results, especially in areas with little access to healthcare.

CASE REPORT

A 7-year-old male patient presented in OPD with complaint of single episode of generalized tonic clonic seizures with frothing from mouth, unrolling of eyeballs and urinary incontinence, lasting for 30 seconds, followed by left sided upper limb and lower limb weakness along with myoclonic seizures. Patient was immunized for measles. Patient on examination vitals were stable; patient was active and on walking patient was swaying towards right side.

Management and outcome

Investigations

EEG showed predominant activity is of alpha type 8-9 Hz, 20-120 microvolts with generalized spike waves and slow waves over tempero-parietal region bilaterally (Figure 1).

MRI brain was suggestive of non-restricting asymmetrical hyper intensity involving subcortical, deep and periventricular white matter of bilateral fronto-parieto-occipital and right temporal lobes which was suggestive of SSPE (Figure 2).

CSF showed acellular on smear and glucose and proteins were normal, CSF for IgG measles antibody was positive.

Treatment

Treatment give were-IV anti epileptics, oral clobazam and oral inosine acedoben dimepranol.

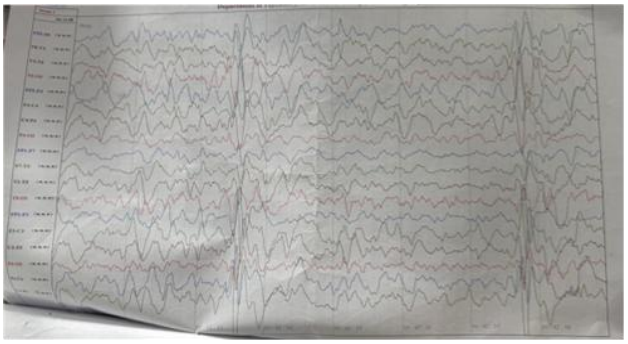

Figure 1: EEG showing spike waves and slow waves.

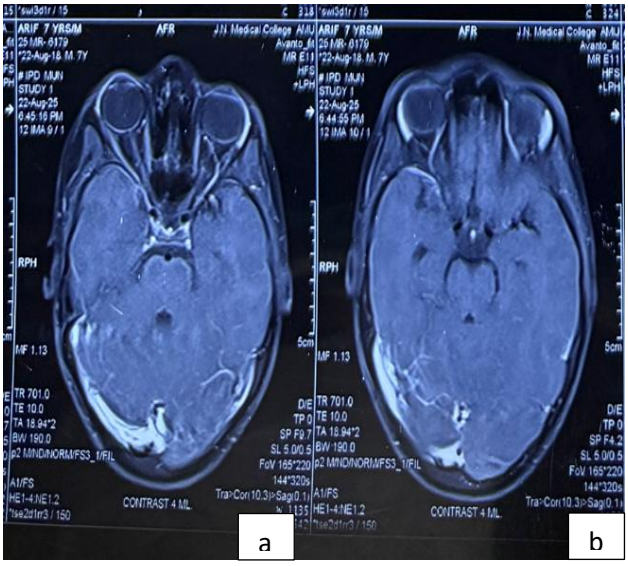

Figure 2 (a and b): MRI brain showing non restricting asymmetrical hyper intensity involving subcortical, deep and periventricular white matter of bilateral fronto-parieto-occipital and right temporal lobes.

DISCUSSION

SSPE is a rare type of brain disease that happens when a changed version of the measles virus keeps infecting the brain. The exact reason it happens isn't fully understood, but it's thought to be caused by the virus staying in the brain for a long time, which leads to damage in the white matter of the brain's outer parts and the brainstem. Even though many people in developed countries are vaccinated against measles, SSPE is still a big problem in poorer areas. It usually happens in children, and the symptoms often show up many years after they first had measles. The symptoms start with changes in behavior, then seizures, trouble thinking, stiff muscles, and eventually, the person might go into a coma and die. As the disease gets worse, patients often show signs like stiff muscles and difficulty moving. They might become paralyzed in all four limbs, and some muscle spasms might go away. In the final stages, breathing becomes strange and hard, and person might have unusual body positions like decerebrate or decorticate postures. The disease can be broken down into different stages based on how the symptoms appear, as shown in Table 1.[3]

Table 1: Staging of SSPE.

Stages	Clinical progression
Stage 1	Behavioral changes and cognitive decline
Stage 2	Myoclonus and motor deterioration
Stage 3	Pyramidal and extrapyramidal manifestations, the disappearance of myoclonus, and disorientation in sensorium
Stage 4	A vegetative state

The criteria for the diagnosis of SSPE were proposed by Dyken, as presented in Table 2.[3,4]

Table 2: Dyken's modified criteria for diagnosing SSPE.

Criteria	Diagnosis
Major	
Clinical	Progressive, subacute mental deterioration with typical signs such as myoclonus
Measles antibodies	Raised titers in serum (>1:256) and/or cerebrospinal fluid (>1:4)
Minor	
EEG	Periodic, stereotyped, high-voltage discharges
Cerebrospinal fluid	Raised gamma-globulin or oligoclonal pattern
Brain biopsy	Suggestive of panencephalitis
Special	Molecular diagnostic test to identify the measles virus mut

*Diagnostic requirement: two major plus one minor criterion are required, but if the features are atypical, then histopathological or molecular evidence may be required.

In our case, the patient showed signs of myoclonus and intellectual disabilities in stage 2, and then moved to stage 3, which shows how quietly and slowly the disease can progress, even in people who have been vaccinated. One study found that good measles vaccination programs protect people from SSPE and could even help eliminate it by getting rid of measles.[5] Data from both the disease and viruses suggest that the measles vaccine does not lead to SSPE. Another study stressed how important vaccination is in stopping disease from getting worse.[6] Our case shows that even though our country has good measles vaccination rates, a serious disease like SSPE still affects children who are fully vaccinated. This case reminds doctors to think about SSPE in younger, fully vaccinated kids who have cognitive problems and myoclonus. One big challenge with treating SSPE is that it's hard to spot early signs, especially when the changes in body are still reversible. Families of those with SSPE face a lot of stress, both physically and emotionally, and also financially, so they need a lot of help to deal with these problems. In the end, the best way to prevent this serious brain disease is through effective measles vaccination, which highlights how important these vaccination programs are for keeping public healthy.

CONCLUSION

Our case showed three unusual signs of SSPE starting at a young age, sudden loss of balance showing a fast disease progress, and happening in a child who had been vaccinated but didn't have a known measles infection. We think the child might have had a mild measles infection that wasn't noticed by the parents, which could have led to SSPE. This shows how tricky it is to diagnose SSPE in vaccinated people and reminds us to be more careful and check more thoroughly when such cases happen.

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