

## CASE REPORT

# A DEBILITATING COMPLICATION OF MEASLES VIRUS: SSPE

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## ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a rare and chronic neurodegenerative disease caused by an unceasing infection of the brain by an altered form of the measles virus acquired earlier in life. The initial symptoms usually involve regressive changes in intellect and personality. Over a span of several months, the psychological symptoms are augmented by neurological symptoms, which most often composes of myoclonic jerks. This disorder more often affects male children or adolescents as compared to females and adults. SSPE cases are now mostly seen in developing countries where measles virus is still highly prevalent. Here we report a case of a 6 years old male who presented to the Primary Health Care Centre in a small settlement area of Karachi; with complaints of gradual weakness of all four limbs associated with loss of power and mild fever. The boy was also having myoclonus seizures involving right upper arm and right facial jerks. Brain MRI showed no physical abnormality although the Electroencephalogram revealed abnormal signals which confirmed SSPE.

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## INTRODUCTION

Subacute sclerosing panencephalitis is the most devastating complication of measles infection, which infects earlier in life. However, development of SSPE occurs years later. Children are more often impacted by this progressive brain inflammatory condition as compared to adults. SSPE has a long latency period of development before specific psychological symptoms arise therefore it is often diagnosed late. Later symptoms include myoclonic seizures which may lead to paraplegia and thus becomes an incurable disease. Despite being incurable in nature it can be prevented by effective measles vaccinations which are now used worldwide. In our report we describe the case of a 6 year old boy who developed SSPE and was brought to the local primary healthcare center with complaints of inability to extend or flex limbs associated with high fever which subsided in 2 days.

## CASE REPORT

A 6 year old had low grade fever for 2 days that almost settled by the third and fourth day. The child

was only given paracetamol for relieving the fever. It was considered a viral fever as the child got better and continued his normal activities. However, 2 days later mother noticed that her son would fall after walking a few steps. This concerned the mother and just the same night mother was astonished to see that his son was not able to sit in the toilet. His condition deteriorated significantly by next day and the child was not able to sit and stand. His legs lost power and his speech became slurred. Upper arms movement was normal but below the lumbosacral spine no movement could be made. Following that the mother noticed a seizure involving the right arm and jerky movement turning face towards right side.

The child was taken to a pediatrician who advised Carbamazepine and Clonazepam for controlling the seizure. Physiotherapy of all 4 limbs was also advised. MRI Brain was normal however electroencephalogram (EEG) showed abnormal signals and a rare disease (SSPE) sub-sclerosing panencephalitis was diagnosed.



**A B C**  
**Figure 1- (A B C): Child unable to stand.**

## DISCUSSION

Subacute sclerosing panencephalitis is the most devastating consequence of a measles infection. Children infected with measles under the age of 1 year have a 16 times greater risk of developing symptoms of SSPE than those infected at age 5 years or later<sup>1</sup>. Children are more often impacted by this progressive brain inflammatory condition as compared to adults. According to research, the age range at the onset of the disease was observed to be 15 to 192 months (mean age: 80.02 months)<sup>2</sup>. A previously healthy 29-year-old pregnant woman has also been reported to develop subacute sclerosing panencephalitis<sup>3</sup> which is a rare case as it is more often found to occur in children.

Measles is a negative-sense RNA virus which is of the genus *Morbillivirus* within the family *Paramyxoviridae*. Humans are the natural hosts of the virus and are infected by it via host cell invasion. The World Health Organization (WHO) reports 189,929 cases in 2016 and 93,943 cases to date in 2017; nevertheless, measles is widely known to be underreported<sup>4</sup>. The measles virus truly infects the respiratory system, however; it presents as a generalized red maculopapular rash following the prodromal phase which includes high fever, malaise and a classic triad consisting of conjunctivitis, cough and coryza. The symptoms of measles usually appear about seven to fourteen days after a person is infected with the virus. Although an early diagnosis can be made if the Koplik's sign i.e. a white enanthem which manifests 2-3 days before the rash appears. Measles is a highly contagious infection and can be transmitted through respiratory droplets as the virus resides in the throat of its victim. There are a total of 23 subtypes of the virus that are recognized. A perilous one which is yet to be identified causes SSPE.

Although the pathophysiology of the SSPE is not fully understood; there is evidence that factors that favour humoral over cellular immune response are involved against the virus. Consequently, the virus is

able to infect the neurons and also survive in a latent form for years. Preceding development of the initial, nonspecific psychological symptoms which often lead to miss or late diagnosis. The symptoms of SSPE typically occur in a four-stage clinical course. With each stage, the symptoms are worse than the stage before. In Stage one there may be personality changes, mood swings, or depression. Fever and headache may also be present. This stage may last up to 6 months. The second stage presents with uncontrolled movement including jerking and muscle spasms. Other symptoms that may occur in this stage are loss of vision, dementia, and seizures. The jerking movements are replaced by writhing, twisting movements and rigidity in stage three, thus eventually leading to akinetic mutism. The last stage progresses as areas of the brain that control breathing, heart rate, and blood pressure become damaged, resulting in coma followed by death occurring due to complications. Management of the disease includes seizure control and prevention of secondary complications associated with the progressive disability. Trials of treatment with Interferon, Ribavirin, and Isoprinosine using different methodologies have been done and have shown beneficial results. However, the disease shows relentless progression; only 5% of individuals with SSPE undergo spontaneous remission, with the remaining 95% dying within 5 years of diagnosis<sup>5</sup>.

Electroencephalography (EEG) images and magnetic resonance imaging (MRI) are two major tools used for the clinical diagnosis of SSPE. Magnetic resonance imaging becomes increasingly effective with the progression of the disease, with primary lesions being noted on MRIs done at earlier disease stages having an incidence of only roughly 7.6%, with most patients in the early stages showing normal imaging studies<sup>6</sup>. Diffused or focal areas of high signal on T2-weighted magnetic resonance images become evident after 6 months of the disease, and progressive hemispheric, cerebellar and brainstem atrophies are seen in later stages<sup>7</sup>. With mortality ranging from 50-95% and cases being rare, the varied semiology and imaging presentations become very clinically valuable<sup>8</sup>. While antibodies of the IgM-class are seldom detectable, it is recommended that antibodies to the measles virus be tested for in blood and cerebrospinal fluid for further confirmation of diagnosis<sup>9</sup>.

## CONCLUSION

Globally, the incidence of measles infections is declining due to aggressive vaccination use. With the virus now essentially affecting regions with low vaccination rates such as in developing countries, where at least 90-95 % coverage is needed for elimination of this disease<sup>10</sup>.

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