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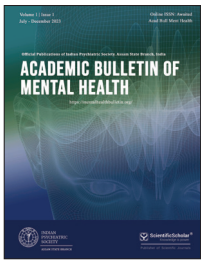

Case Report

Measles to Misfortune: A Case Report on Subacute Sclerosing Panencephalitis

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a progressive neurological disorder of children or young adults caused by measles infection. An abnormal immune response to the virus is said to cause brain inflammation, which may last for years. Mortality rate of the disease is around 95%. In this case report, we present the case of a 15-year-old unimmunized boy from rural Assam who presented with progressive sudden jerky movements of his limbs for 2 months, along with difficulty in doing his daily activities and forgetfulness. Although he was initially admitted with a provisional diagnosis of seizure disorder and treated with tab sodium valproate and tab clobazam, his mother, revealed a history of measles infection at 7 years of age. His Electroencephalogram (EEG) reports and CSF IgG measles antibody findings were suggestive of SSPE. He was immediately started on Tab Isoprinosine at adequate doses, but he later succumbed to his illness within 3 months of the onset of symptoms. Immunization of children with the measles vaccine plays a significant role in averting this deadly condition.

Keywords: Measles, Seizure, Forgetfulness, Isoprinosine, Immunization

INTRODUCTION

Subacute sclerosing panencephalitis (SSPE) or Dawson disease is a progressive neurological disorder in children or young adults caused by measles infection.[1] Commonly found in developing and underdeveloped countries, the worldwide incidence is 4–11 per 100000 cases of measles.[2] As per the latest literature, incidence rates were 21 cases per million population in India, 11 per million in Japan, and 0.06 per million in Canada.[3]

An abnormal immune response to the virus or possibly to a certain mutant form of the Virus, is said to cause brain inflammation, which may last for years.[4] The disease progresses through four stages, in which the initial stage is characterized by personality change, mood swings, fever, headache, and memory loss, followed by Stage 2, where there are jerky muscle spasms, seizures, and loss of vision. Stage 3 involves writhing movements and rigidity, and the last stage consists of loss of consciousness with a persistent vegetative state and death.[5]

Among the limited treatment options available, Isoprinosine is one of the first drugs used. It has immunomodulatory and antiviral properties. Beneficial effects of the drug are achieved in one-third of the cases when given at 50–100 mg/kg/day orally as a monotherapy or in combination with Interferon.[6] Ribavirin is found to inhibit the replication of subacute sclerosing panencephalitis (SSPE) virus strain and can be used intravenously or intrathecally.[7] The drug Interferon alpha, delivered directly into the spinal column with and without the addition of oral isoprinosine, is being tested, and approximately 50% of affected individuals experienced an improvement in their

symptoms.[8] Reports on the use of favipiravir and remdesevir are also available.[7]

The mortality rate is exceptionally high in the case of SSPE, about 95%, while the remaining cases undergo spontaneous remission.[9,10] Most individuals with SSPE will die within 1–3 years of diagnosis. In a small percentage, the disease will progress rapidly, leading to death within 3 months of diagnosis.[5] The average life span after the initial presentation is about 3.8 years, with a range of 45 days–12 years.[11]

Immunization of children with measles vaccine is the only definitive key step in the prevention of this deadly condition.

CASE REPORT

A 15-year-old boy hailing from rural Assam was referred to the department of psychiatry, Gauhati Medical College and Hospital (GMCH) from neurology outpatient department (OPD) on April 20, 2023. The chief complaints were sudden jerky movements of his limbs and difficulty in doing his daily activities for the past 2 months, along with forgetfulness for the past 10 days.

The onset of the symptoms was acute and the course of the illness was progressive. As his mother recalled, her child had returned from school one fine afternoon when he developed the first sudden jerky movement of his left upper limb. The patient, curious, started asking his mother about what had happened to him, as his movements were not under his voluntary control. The next day, he developed similar movement in the other upper limb, too. As days passed by, it started involving the lower limbs, too. After 2 weeks or so, the patient started having sudden jerky movements of all four limbs all at once. Initially, it occurred about once every 5 hours, progressing to once every hour, which increased to once every minute at presentation. Movements of lesser frequency would occur even during sleep. The movements crippled the boy, and he started needing the assistance of his family members in his day-to-day activities. He started talking less, and at later stages, communicated in either YES or NO. The patient started denying food and also had a mild degree of forgetfulness. There was never any history of loss of consciousness or involuntary passage of urine or stool. He was taken to multiple faith healers in the period of 2 months before being brought to Gauhati Medical College and Hospital .

On detailed questioning, a history of measles infection was found at the age of 7 years, for which some faith healing treatment was done. Birth history revealed normal vaginal delivery at home, with no immunization of the child.

On general examination, the patient was observed to have jerks of limbs every 60 seconds. On central nervous system (CNS) examination, the power of limbs was found to be 3/5, deep tendon reflexes 2+, and the plantar response was bilateral flexor. A mental state examination could not be done as the patient did not reply to most of the questions, but he could identify his parents.

With this, the patient was admitted to the Psychiatry ward with provisional diagnosis of seizure disorder. A routine work-up was done with concurrent administration of a sodium valproate tablet of 300 mg twice daily. Tab Clobazam 10 mg was also added along with supportive treatment with intravenous fluids and vitamins. An EEG was done. Neurology call was sent, who later diagnosed him as a case of SSPE based on the typical EEG findings (periodic R wave complexes suggestive of SSPE) and added Clonazepam and increased Tab Sodium Valproate to 1 g per day. Blood parameters and magnetic resonance imaging (MRI) findings were within normal limits. Cerebro-spinal fluid (CSF) analysis findings were not significant except that the Immunoglobulin G (IgG) anti-measles antibody in CSF was 450 IU/mL. Serum IgG anti-measles antibody was also done, which was found to be >300 IU/mL. With these available data and based on Dyken's criteria, a final diagnosis of SSPE was made.

The myoclonic jerks of the patient were controlled both in intensity and frequency, and Tab Amantadine was started at a dose of 100 mg twice daily, later increased to 100 mg thrice daily. Some degree of improvement was noticed, and the patient was accepting food and talking over the phone in a few words to his family members. Tab Isoprinosine was started at 500 mg once daily and later increased to 500 mg twice daily at 5 days interval, which was again increased to 1000 mg twice daily at the time of discharge. A week after starting Tab Isoprinosine, his serum IgG measles antibody decreased to 158 IU/mL. However, the patient's condition started to deteriorate. The poor prognosis of the illness was explained to the family members. On the insistence of the family members, the patient was discharged on May 20, 2023. He succumbed to his illness the next day at home.

DISCUSSION

Children developing SSPE generally have a measles infection history at a very young age, which is followed by a period of latency of about 8 years before the disease manifests.[1] The disease is seen to be affecting males more than females and is more common in the children and adolescents of rural background, as seen in this case.[8]

CONCLUSION

The above case is a rare manifestation of measles infection with rapid progression and it is an eye-opener on the presentation of a patient of SSPE in the Neurology and Psychiatry OPD. Most importantly, immunization of children with the measles

vaccine to avert such deadly conditions is of utmost public health concern.

Ethical approval

The research/study complied with the Helsinki Declaration of 1964.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

Dr. Deepanjali Medhi is on the editorial board of the Journal.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Available from: www.ninds.nih.gov [Last accessed on 2023 September 10]
2. Rocke Zoe, Belyayeva Mariya, Subacute Sclerosing Panencephalitis. [Updated 2022 Jul 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan.
3. Campbell H, Andrews N, Brown KE, Miller E. Review of the Effect of Measles Vaccination on the Epidemiology of SSPE. Int J Epidemiol 2007;36:1334–48
4. Gershon AA. Measles Virus (Rubeola). In: Benntt JE, Dolin R, Blaser MJ, eds. Mandell, Douglas, and Bennett's Principles and Practice of Infectious Disease. 9th ed: chap 160
5. National Institute of Neurological Disorders and Stroke (NINDS). "Subacute Sclerosing Panencephalitis Information Page | National Institute of Neurological Disorders and Stroke"
6. Gascon GG. International Consortium on Subacute Sclerosing, Randomised Treatment Study of Inosiplex Versus Combined Inosiplex and Intraventricular Interferon-alpha in Subacute Sclerosing Panencephalitis (SSPE): International Multicenter Study. J Child Neurology 2013;18:819–27
7. Samia P, Oyieke K, Tunje D, Udwadia-Hegde A, Feemster K, Oncel I, *et al*. Options in the Treatment of Subacute Sclerosing Panencephalitis: Implications for Low Resource Areas. Curr Treat Options Neurol 2022;24:99–110
8. Available from: www.rarediseases.org "Subacute Sclerosing Panencephalitis Information Page" [Last accessed on 2023 September 10]
9. Nathan J, Khedekar Kale D, Naik VD, Thakker F, Bailur S. Substantial Remission in Subacute Sclerosing Panencephalitis by Following the Ketogenic Diet: A Case Report. Cureus 2019;11:e5485
10. Gutierrez J, Isaacson RS, Koppel BS. Subacute Sclerosing Panencephalitis: An Update. Dev Med Child Neurol 2010;52:901–7
11. Jafri SK, Kumar R, Ibrahim SH. Subacute Sclerosing Panencephalitis – Current Perspectives. Pediatric Health Med Ther 2018;9:67–71

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