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Subacute Sclerosing Panencephalitis Presenting Like Parkinsonism: A Rare Case

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Abstract: ***Subacute Sclerosing Panencephalitis (SSPE) is characterized by a progressive decline in cognitive andmotor functions, seizures, and eventually death. The incidence of SSPE declined by at least 90 percent in countries that have practiced widespread immunization with measles vaccine. We present a case of a 14-year-old male who presented with an episode of seizure following which he developed hand tremors and slowness of movement, expressionless face, reduced tone of speech. He had a history of missed measles vaccination at the age of 9 months. On further evaluation there was a significant raised titre of antimeasles IgG antibodies in CSF with coinciding radiological and electroencephalogram supporting the diagnosis of SSPE. He was given trial with intrathecal interferon alpha and Isoprenosine . Unfortunately, there is no cure. Patients with SSPE usually die within 3 months to 3 years of diagnosis.***

Keywords: subacute sclerosing Panencephalitis, Measles, Parkinsonism, Vaccination

1. Introduction

Measles occurs worldwide and remains a leading cause of mortality especially among children ≤5 years of age. As of December 1, 2021, a total of 49 measles cases were reported by 5 jurisdictions. (1) Subacute sclerosing panencephalitis (SSPE) a serious complication of measles generally develops 7 to 10 years after a person has measles, even though the person seems to have fully recovered from the illness. (SSPE) is characterized by a progressive decline in cognitive and motor functions, seizures, and eventually death (2). The incidence of SSPE declined by at least 90 percent in countries that have practiced widespread immunization with the measles vaccine. Its pathogenesis is not well understood but may involve persistent infection with a genetic variant of the measles virus within the central nervous system (3)

2. Case Report

A 14-year-old male presented to outpatient department with resting tremors, slowness of movement, expressionless face, and hypotonia which was insidious in onset and progressed gradually over a period of 15 days. There was history of one episode of generalized tonic clonic seizure two months back, for which he was started on anti epileptics by general practitioner and wasn't evaluated for the same. Since 3 months the mother had been noticing him being lethargic, having difficulty in concentrating on studies and group activities. However, they did not seek any medical attention for it. The history does not reveal any deficit in scholastic activities prior to the onset of the current illness however he has been displaying gradual decline in cognitive function thereafter. He scored 18/30 in Mini Mental State Examination Hindi version which is suggestive of mild cognitive impairment. On clinical examination, power and reflexes were normal in all four limbs but there was rigidity in all four limbs, resting tremor of hand and micrographia was noted. History of missed measles vaccination and Parkinson's like symptoms in the absence of psychotropics without any significant familial history hinted towards SSPE. Hence, a detailed work-up for the same was warranted in this young boy. Among the pertinent investigations he was found to have elevated protein concentration and detectable significant titres of antimeasles IgG antibodies in CSF I. e 1: 100 (normal <1: 4), EEG showed periodic pattern with slow wave complexes, high fluid-attenuated inversion recovery signals (FLAIR) on Magnetic Resonance Imaging of the brain was consistent with diagnosis of SSPE.

He was given trial with intrathecal Interferon alpha twice a week and Isoprinosine daily. Although the patient was compliant with the medication, there was no improvement in the symptoms and eventually the parents discontinued interferon alpha. Patient is on follow up since 1 year. Currently the condition of the patient has deteriorated further.

3. Discussion

This young boy had atypical extrapyramidal features that are not commonly seen in SSPE. These are tremors, bradykinesia, mask like facies, reduced tone of speech, marked rigidity involving right upper and lower limb and micrographia. Uncommon presenting features have been described such as tremor, dystonia and parkinsonism like features.

This is a rare presentation of SSPE with juvenile parkinson like features. In SSPE, migratory basal ganglia lesions have been reported and axonal spread of virus from the substantia nigra has been implicated in producing parkinsonian symptoms. (4) Although SSPE has been divided into various stages. The rate of progression is variable. Stage I is characterized by development of neurologic symptoms such as personality changes, lethargy, difficulty in school, and strange behavior. Stage II is characterized by myoclonus, worsening dementia, and long-tract motor or sensory disease. Stages III and IV are characterized by further neurologic

deterioration with eventual flaccidity or decorticate rigidity and symptoms and signs of autonomic dysfunction. Myoclonus is absent. Stage IV is a vegetative state. Death usually occurs during stage IV but is possible in any of the stages. Stabilization at one stage for a period of time can occur, though patients tend to progress from one stage to the next. Some patients have a remitting and relapsing course. Seizures can occur in any of the stages. (5, 6)

4. Conclusion

Clinical presentation of SSPE widely varies ranging from progressive weakness, seizures, pyramidal and extrapyramidal symptoms, and coma. Thus, one must be aware of the atypical presentations of SSPE and a high index of suspicion is required for its diagnosis. SSPE should be considered in children and adolescents with parkinsonian symptoms, particularly in the absence of a history of vaccination against measles. Patients will die within 3 months to 3 years of diagnosis. No cure underscores the importance of vaccination**.**

References

[1] https: //www.cdc.gov/measles/cases-outbreaks. html
[2] Simkhada N, Adhikari P, Pathak BD, Dhakal B, Mahat K. A Rare Case of Subacute Sclerosing Panencephalitis Presenting As Generalized Seizure. Cureus.2021 May 18; 13 (5): e15100. doi: 10.7759/cureus.15100. PMID: 34159008; PMCID: PMC8212923
[3] Garg RK, Mahadevan A, Malhotra HS, Rizvi I, Kumar N, Uniyal R. Subacute sclerosing panencephalitis. Rev Med Virol.2019 Sep; 29 (5): e2058. doi: 10.1002/rmv.2058. Epub 2019 Jun 24. PMID: 31237061.
[4] Bozlu G, Cobanogullari Direk M, Okuyaz C. Subacute sclerosing panencephalitis with parkinsonian features in a child: A case report. Brain Dev.2015 Oct; 37 (9): 901-3. doi: 10.1016/j. braindev.2015.02.01.08. Epub 2015 Feb 28. PMID: 25736973.
[5] Raut, T. P., Singh, M. K., Garg, R. K., & Rai, D. (2012). Evolution of certain typical and atypical features in a case of subacute sclerosing panencephalitis. BMJ Case Reports, 2012 doi: http: //dx. doi. org/10.1136/bcr-2012-007052
[6] Kannan L, Jain P, Sharma S, Gulati S. Subacute sclerosing panencephalitis masquerading as rapid-onset dystonia-Parkinsonism in a child. Neurol India 2015; 63: 109-10