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HOPE IN THE FACE OF SUBACUTE SCLEROSING PANENCEPHALITIS: A CASE REPORT OF COMPLETE REMISSION WITH ISOPRINOSINE

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ABSTRACT

Subacute Sclerosing Panencephalitis (SSPE) is a rare and relentless neurodegenerative disorder linked to persistent measles virus infection. We present a case of a 16-year-old female with gradually worsening rigidity, cognitive decline, and myoclonic seizures, minimally responsive to valproic acid. A history of measles infection at the age of two years emerged. Positive cerebrospinal fluid (CSF) IgG and IgM anti-measles antibodies, along with an elevated CSF/serum antibody index, confirmed SSPE. Treatment with Inosine pranobex and clonazepam yielded significant motor and seizure control improvements. This case highlights the importance of early SSPE diagnosis and tailored interventions, shedding light on clinical progression and therapeutic responses.

Keywords: Subacute sclerosing panencephalitis; Measles; Inosine pranobex

INTRODUCTION

Subacute Sclerosing Panencephalitis (SSPE) is a rare and relentless neurodegenerative disorder caused by persistent infection with the measles virus.[1] The disease has a gradual progressive course leading to death within 1-3 years.[2] Despite the global reduction in measles incidence due to vaccination, SSPE continues to present clinical challenges, especially in regions with low vaccination rate.

The etiology of SSPE is closely related to a mutant form of the measles virus that lies dormant in the central nervous system (CNS) for years before reactivating and causing extensive inflammation and demyelination.[3, 4] It is particularly difficult to manage SSPE because of this latency period, which frequently lasts for several years and complicates early detection and intervention.[5-7]

Furthermore, the varied characteristics of SSPE can cause delays in treatment and diagnosis because it might mimic other neurological conditions. Inosine pranopex is a drug that has inconsistently shown promise in its treatment.[8, 9] The four regions with limited healthcare infrastructure and low vaccination rates are particularly impacted, emphasizing the continuous need for attentive surveillance, increased immunization outreach, and public health education.

This case report offers a poignant illustration of SSPE's clinical complexities, shedding light on the importance of timely recognition, comprehensive diagnostic evaluation, and tailored therapeutic interventions.

CASE PRESENTATION

A 16-year-old female was admitted to the Neurology Department of Bolan Medical Complex Hospital (BMCH), Quetta, with a history of gradually progressive symptoms. She presented with diffuse rigidity, cognitive deterioration, and generalized myoclonic seizures that displayed minimal responsiveness to valproic acid treatment. The symptoms had commenced several months prior, and her condition had deteriorated progressively.

The patient's clinical course was characterized by a gradual loss of social abilities and daily living skills, such as feeding and self-dressing. She exhibited a once age-appropriate cognitive ability and language expression, which rapidly regressed after the onset of her illness. Notably, her past medical history included a measles infection at age two years, and her family history was unremarkable for neurological or metabolic diseases.

Upon examination, the patient demonstrated generalized stiffness and rigidity in her limbs, accompanied by ankle clonus and hyperactive deep tendon reflexes. She experienced continuous myoclonic jerks, and her neurological status was marked by uncontrolled rigidity, swallowing difficulties, and bowel and bladder incontinence. Although alert, her communication was limited, and she exhibited confused mentation at the time of admission.

Initial brain magnetic resonance imaging (MRI) yielded unremarkable results, prompting further investigation (Figure 1). Given her history of measles infection and

clinical manifestations, a cerebrospinal fluid (CSF) analysis was performed. The CSF analysis revealed normocytosis and normal glucose and total protein levels. Notably, the CSF displayed positive IgG and IgM antimeasles antibodies, and the CSF/Serum antibody index was elevated at 3.12 (normal value <1.3). Her EEG showed a burst suppression pattern.

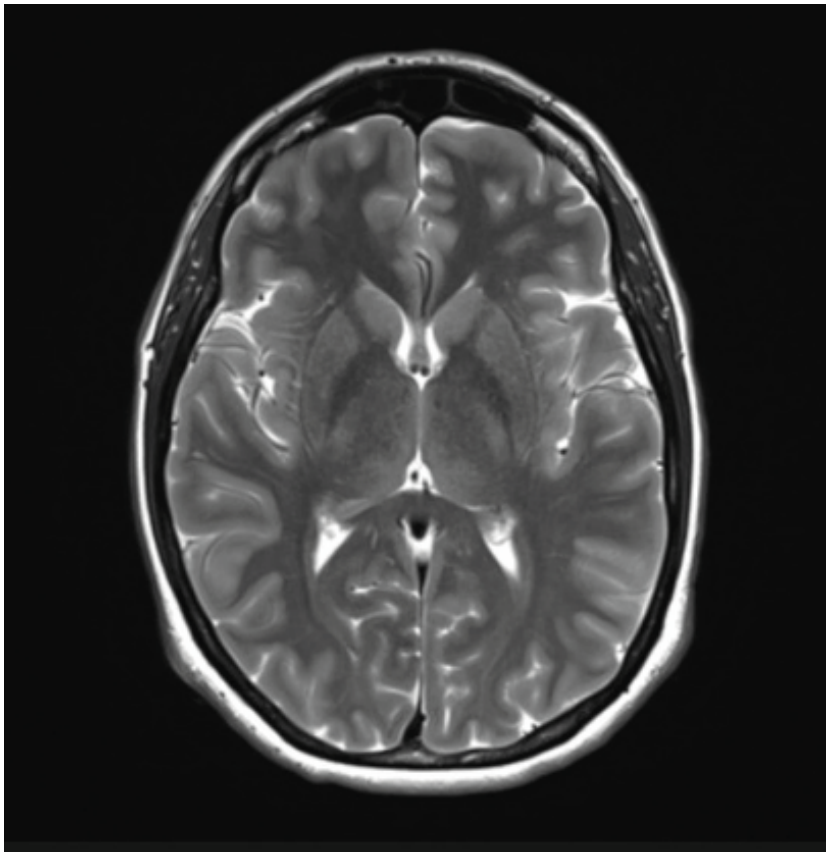

Figure 1 : MRI Brain T2 Scan Normal Study.

Based on the CSF and EEG findings and in conjunction with her clinical history, the patient was diagnosed with SSPE. A therapeutic regimen was initiated, encompassing oral Inosine pranobex 100mg/kg/day in divided doses and supportive clonazepam and levetiracetam therapy. Notably, her generalized stiffness gradually improved during treatment, culminating in its complete resolution a month after therapy initiation.

Follow-up evaluations revealed positive outcomes over time. At the one-year mark after SSPE treatment commencement, the patient demonstrated improvements in hypertonicity, cessation of myoclonic jerks, and regained communication abilities. She achieved significant functional gains, resuming daily activities and achieving freedom from seizures.

DISCUSSION

Incidence of measles during infancy represents an established risk factor for the development of SSPE. Consequently, it is advisable to administer early doses of the MMR (Measles, Mumps, Rubella) vaccine to young children identified as being at a heightened risk of measles infection.[10] An expert committee convened by the World Health Organization conducted a comprehensive review of SSPE epidemiology, revealing an estimated incidence of four to 11 SSPE cases per 100,000 measles cases. Notably, the risk of SSPE escalates significantly when measles infection transpires during infancy, particularly in children under the age of two. Southeast Asian nations like India and Pakistan have exhibited notably elevated SSPE incidence rates.[11]

Pakistan is among the five nations where nearly one million children have not received their initial measles vaccination. The vaccination coverage rates in Pakistan for both the first dose and booster dose of the measles vaccine are notably deficient.[12] The research conducted in Karachi revealed that 78% of the children had received the single measles vaccine, while 12% had

received both doses of the vaccine. Nevertheless, only 55% of the children exhibited detectable levels of measles antibodies, indicating immunity to the disease. This disparity underscores a substantial gap between the percentage of vaccinated children (90%) and the proportion who have developed effective immunity against measles (55%).[13] During the period spanning from January to August 2022, Pakistan documented a total of 6,749 reported cases of measles, reflecting an incidence rate of 29 cases per million. In the preceding year, 2021, Pakistan recorded 10,399 measles cases, whereas the figure for 2020 stood at 2,747 reported cases.[14]

Currently, there are no established or standardized treatment protocols available.[15] The primary approach to treatment entails the administration of antiepileptic medications to manage seizures, complemented by antiviral agents, often in conjunction with immunomodulatory therapy. Notably, Inosine pranobex has demonstrated therapeutic efficacy in multiple studies as the most beneficial intervention for individuals afflicted with SSPE[16]. In a specific case, cessation of treatment resulted in patients succumbing to respiratory illnesses. Cruzado et al.[17] documented the case of a female patient who received a daily dose of 100 mg/kg and subsequently entered a vegetative state at 20 months, eventually passing away at 28 months. Likewise, Gokcil et al[18]. reported a similar outcome when administering doses ranging from 50 to 100 mg/kg of body mass daily, resulting in deteriorating patient conditions. In contrast, Nasirian et al.'s[19] observations yielded more favorable outcomes. Among the 16 cases studied, they noted a cessation of disease progression in four cases, while six cases displayed a slower rate of deterioration. Remarkably, of these six patients with slower progression, three extended their lifespan by an additional four years, two survived up to seven years and one remarkably lived for an additional ten years. The exact rationale behind this prolonged survival among the six patients with slower progression remains uncertain, though genetic factors may play a role.[20] In a multinational study encompassing 500 cases of SSPE, Inosine pranobex emerged as the most widely employed treatment option across participating countries. Nevertheless, concerns have arisen regarding its accessibility in regions where SSPE is prevalent, primarily due to its associated high cost. Inosine pranobex has demonstrated a beneficial impact on both survival rates and the mitigation of neurological deficits in approximately one-third of SSPE cases when administered at a dosage range of 50–100 mg/kg per day, with a maximum daily dose of 3 g, taken orally in three to five divided doses. This therapeutic approach can be utilized either as a standalone intervention or in combination with Interferons (IFN).[21]

In our specific case, following the confirmation of SSPE through the presence of positive anti-measles antibodies in CSF, the patient was initiated on a regimen of Inosine pranobex at a dosage of 100 mg/kg daily, administered in three divided doses. Following a consistent year-long course of treatment, the patient displayed unmistakable signs of full recovery. Notably, she became entirely free from seizures and regained the capability to engage in her daily activities. This remarkable recovery is unique; after an exhaustive examination of existing literature, we can confidently assert that no prior instances of near-complete recovery with this treatment have been documented. Consequently, this case study holds significant promise as a benchmark for future SSPE treatments. Further extensive observations and research endeavors must continue to explore the potential effects of Inosine pranobex in diverse scenarios, to facilitate the partial or complete recovery of patients, especially in low economic countries with low rates of vaccination.

One limitation of this case is the absence of repeated CSF analysis during treatment Certainly, early diagnosis and personalized SSPE treatment are crucial. However, due to cost constraints, repeated CSF analysis was not feasible in this case.

CONCLUSION

This case underscores the importance of early diagnosis and tailored SSPE management. The remarkable complete recovery observed with inosine pranobex treatment is a significant milestone, warranting further research. The complexity of SSPE highlights the need for ongoing comprehensive research and personalized therapies, offering hope for improved outcomes in this challenging condition especially in low economic countries with low rates of vaccination.

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Nimra Shafique; concept, case management, manuscript writing
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Amanullah Kakar; case management, manuscript revision

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