Case Report

AN INFANT WITH A RARE ACQUIRED CNS DISORDER

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) or post-infectious encephalomyelitis is a rare neurological condition particularly affecting young children.

We report a case of 1-year 1-year-old boy with high grade fever, altered mental status, lower limb weakness and seizures. One week before these symptoms the child had complaints of loose stools and vomiting. His initial reports showed leukopenia, raised acute phase reactants and CSF showed elevated proteins. Clinical evaluation and MRI imaging confirmed the diagnosis of ADEM. Prompt treatment with high dose corticosteroids led to gradual improvement in neurological symptoms. The child was discharged with a tapering course of oral corticosteroids and followed up after 1 week, then after 2 weeks with improved neurological deficit and advised for further close follow-up.

This case emphasizes the importance of early recognition, aggressive treatment, and long-term monitoring in managing ADEM in children.

Keywords: Acute Disseminated Encephalomyelitis, ADEM, Pediatrics, IVIG

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INTRODUCTION

ADEM is an acute, immune-mediated inflammatory disorder of the central nervous system, principally involving the demyelination of white matter in the brain and spinal cord.[1.] This condition is precipitated by an acute viral infection like chicken pox, smallpox, Epstein-Barr virus, or herpes simplex virus. ADEM is considered a rare illness with an estimated incidence of 0.8 per 100,000 population per year.[2.] The mean age of clinical presentation in pediatric cohort's ranges from 5 to 8 years[3] with slight male predominance.[4.]

ADEM is a monophasic disease but can present with relapsing cases. Literature reported that treatment with oral corticosteroids is associated with a reduced relapse rate. In recent collaborative studies, treatment other than steroids, like B cell targeted treatment and intravenous immunoglobulins were also associated with a reduction in relapse frequency.[5.]

In Pakistan, for three years, almost 25 children with polysymptomatic monophasic ADEM were reported from 2006-2008.[6.]

Here we present a case of ADEM in 1-year-old boy to highlight the significance of prompt diagnosis and treatment for a better outcome of the disease course.

CASE DESCRIPTION

A 1-year-old boy presented with a complaint of high-grade fever for 2 weeks and seizures for 3 days followed by altered mental status and weakness in lower limbs. The patient also had a history of loose stools and vomiting 1 week

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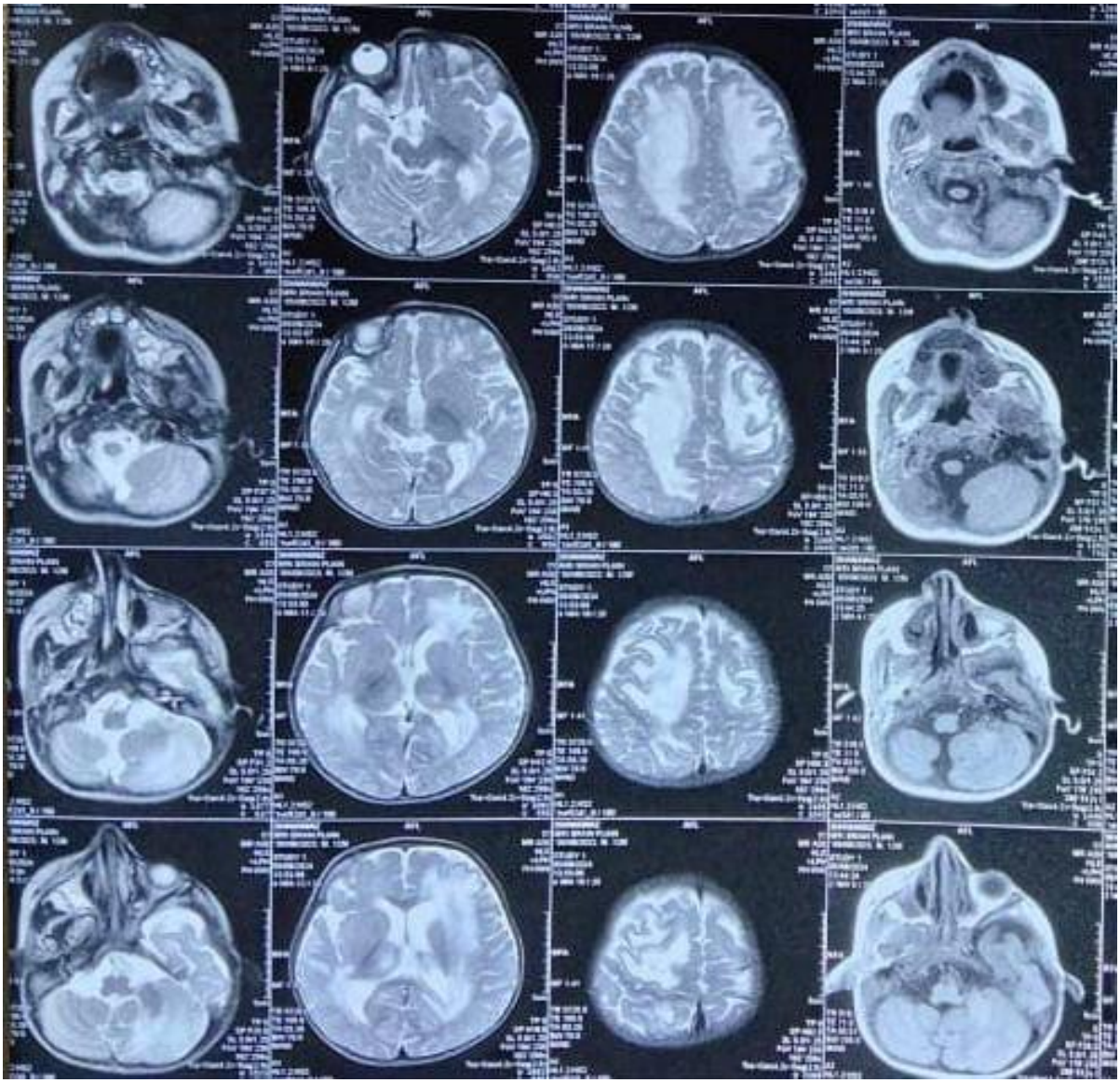
back. On admission, the child was febrile (102 F) and drowsy with vitals; Pulse Rate 142/min, Respiratory Rate 46/min, Oxygen saturation 96% at room air, and Basal Sugar Rate 100mg/dl. Neurological examination revealed signs of encephalopathy with GCS 5/15 (E2V1M2), brisk tendon reflexes, hypertonia, and bilateral extensor planter responses with Power 3/5 in both lower limbs. The patient was admitted to the Pediatric Department of Public Sector Tertiary Care Hospital. Laboratory work-up showed relative leukopenia 4.4×10^9/l(4-11×10^9/l) and raised acute phase reactants CRP 25.1mg/L (Normal Range less than 0.700mg/L). Cerebrospinal fluid (CSF) was clear and analysis showed Glucose 82 mg/dl, Protein 149mg/dl (20-40mg/dL), LDH 45U/l, WBC 4/mm3 and RBCs 10/mm3. On Cranial Ultrasound, there was bilateral symmetrically increased echogenicity of deep cerebral white matter with periventricular cystic changes. A provisional diagnosis of viral encephalitis was made. The patient was started on the anti-viral drug acyclovir along with anti-epileptics i.e. Phenytoin and Lerace. As there was no improvement in neurological status after initial management, it led to extended workup i.e. MRI Brain which showed diffuse periventricular white matter hyperintensities in T2 and FLAIR extending to subcortical region, diffuse symmetrical intensities in thalamus and abnormal signal intensities in left cerebellar hemisphere including margins of fourth ventricle, suggestive of ADEM.



Based on clinical presentation and MRI findings, the diagnosis of ADEM was confirmed. Definitive management started with high-dose intravenous methylprednisolone for 5 days along with neuroprotective care, nasogastric feeding, and bladder, bowel, skin, and mouth care. Throughout treatment, the child showed gradual improvement in neurological status with GCS 11/15(E4V3M4) and was discharged after 14 days of hospital stay on a tapering course of oral corticosteroids (Prednisolone) over 4-6 weeks and scheduled for close outpatient follow-up with MRI report. On follow-up after 2 weeks, the patient developed eye contact and has started to respond to commands as well with GCS 13/15 (E4V4M5).

DISCUSSION

ADEM is an immune-mediated central nervous system (CNS) disorder, characterized by multifocal symptoms, encephalopathy, and typical MRI findings. While the etiology is not fully understood, ADEM is commonly preceded by viral infection suggesting an autoimmune response to myelin basic protein. In some children, ADEM can occur in a recurrent pattern or can lead to chronic diseases, such as multiple sclerosis[7].

ADEM can occur at any age but usually affects children and young adults. Initial symptoms and signs of ADEM usually begin within 2 days to 4 weeks after a viral infection (influenza, EBV, CMV, measles, mumps, rubella) and include systemic symptoms such as fever, malaise, headache, nausea, and vomiting followed by rapid onset encephalopathy (behavioral change or altered consciousness) associated with a combination of multifocal neurological deficits. CSF examination shows lymphocytic pleocytosis and elevated proteins. Typical lesions on MRI are multiple, bilateral but asymmetric, and widespread within the CNS, predominantly involving the white cerebral matter. MRI brain identifies lesions of subcortical white matter in 93% of patients while the percentage of lesions identified in other parts of the brain are as follows: cerebral cortex 80%, periventricular white matter 60%, deep gray matter and brainstem 47%[8].

ADEM is treated with high-dose intravenous corticosteroids as first-line therapy. One common protocol is 10-30 mg/kg/d of methylprednisolone (maximum dose of 1g/d) for 3–5 days[9]. Improvement may be observed within hours but usually requires several weeks for full recovery. An oral steroid tapering for 4-6 weeks is recommended, however, if it is 3 weeks or less it may increase the risk of relapse[10]. Other treatment options include IVIG 2g/kg given over 2-5 days[11] or plasmapheresis (5-7 exchanges done every other day). Even in those children who acquire good neurological recovery; behavioral, visual, and motor impairments may be seen. In this case, neurocognitive testing may prove useful in recognizing these impairments.

CONCLUSION

ADEM is a rare but serious condition that affects young children. Early recognition and prompt treatment with corticosteroids are crucial for a favorable outcome. Long-term follow-up is essential to monitor for potential relapses and to assess the child's neurological recovery. In this case, our patient responded well to treatment, highlighting the importance of a multidisciplinary approach in managing ADEM in pediatric patients.

AUTHOR CONTRIBUTION

ASA: Supervision, Editing
MUA: Supervision, Editing, Discussion
AA: Discussion, References, Conclusion
SN: Abstract, Case Description
MM: Introduction, References

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