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**Case Report**

# Acute hemorrhagic encephalitis in pediatric patient: a case report

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## ABSTRACT

Acute hemorrhagic encephalomyelitis (AHEM) is considered a rare presentation of acute disseminated encephalomyelitis (ADEM) and is challenging to manage. ADEM is a rare illness with an incidence of 0.2–0.4 per 100,000 children annually. Here we discuss a case report of a 3-year-old patient who presented with the clinical features of ADEM. On neuroimaging patient had a rare form of ADEM as hemorrhagic encephalitis. AHEM incidence is very low in the pediatric age group with poor prognosis. With the use of immunosuppressants and immunoglobulin, the survival of the patient was possible.

**Keywords:** ADEM, AHEM, Encephalitis, Immunoglobulins

## INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an acute demyelinating disorder of the central nervous system characterized by multifocal white matter involvement. Some forms have atypical presentations having peculiarities and might be challenging to diagnose and manage.[1] ADEM is a rare illness with an incidence of 0.2-0.4 per 100,000 children annually. Acute hemorrhagic encephalomyelitis (AHEM) is considered a rare form of ADEM and is usually fatal whereas ADEM patients show full recovery. It is very difficult to distinguish AHEM from ADEM based on clinical presentation and neuroimaging.[2] There are very few cases of AHEM reported worldwide and have poor outcomes.

## CASE REPORT

A 3-year-old female child born of non-consangious marriage, 1st by birth order was admitted with the chief complaints of fever and irritability for 2 days and focal convulsions in the form of facial twitching for 1 day.

The fever was high-grade, continuous, and was associated with decreased oral intake and irritability. The seizures were intractable and focal with secondary generalization.

There was no history of antecedent upper respiratory symptoms, diarrhea, rash, arthralgia, or conjunctivitis. There was no recent history of travel, vaccinations, or tuberculosis contact.

Past history was not significant.

In terms of family history, no history of medical or surgical illness was reported.

A per the birth history, the child was full term normal vaginal delivered with a birth weight of 3 kg and no NICU stay.

As per the developmental history, child attained all the age-appropriate milestones for the age.

As per the immunization history, the child is vaccinated with the age-appropriate immunization as per NIS.

On examination, it was found that the child was febrile and irritable and the neurological examination at the first evaluation was suggestive of impaired consciousness with GCS 9/15 and brisk tendon reflexes.

***Management***

The recurrence of symptoms associated with rapidly progressive refractory status epilepticus required multiple antiepileptic support along with mechanical ventilation support. After the initial supportive care, a lumbar puncture was done, and intravenous vancomycin, ceftriaxone, acyclovir, and 3% hypertonic saline along with the supportive care were started. Cerebrospinal fluid (CSF) analysis was done suggestive of elevated proteins 75 mg/dl, sugars 86 mg/dl, and no pus cells. CSF cultures were negative for bacterial, fungal as well as viral growths. The coagulation profile was normal. Biochemical tests for liver and renal function tests and serum electrolytes were normal.

Non-contrast computed tomography (CT) scan was done and was not suggestive of any significant abnormality.

Within 48 hours of admission, the patient became fully unconscious, unresponsiveness to deep pain, and decerebrate posturing with GCS 4/15.

Magnetic resonance imaging (MRI) was suggestive of multifocal hemorrhages involving bilateral frontoparietal as well as temporal lobes, in bilateral thalami, posterior limb of the internal capsule, pons, and occipital lobe suggestive of acute hemorrhagic leukoencephalitis (Figure 1).

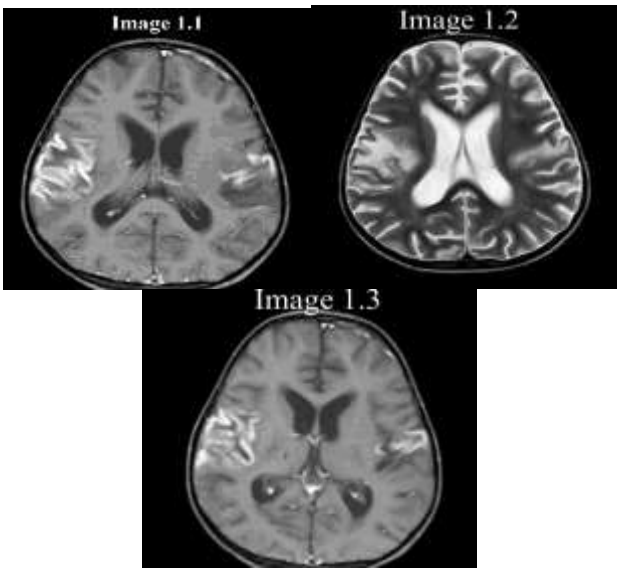


**Figure 1: Magnetic resonance imaging.**

Hence, patient was treated with IV methylprednisolone (30 mg/kg pulse therapy), and intravenous immunoglobulin (IVIG) (2 gm/kg).

***Recovery***

After a period of 35 days of prolonged ventilation developing complications such as ventilator-associated pneumonia and sepsis along with CNS recovery, the patient was extubated and managed accordingly. She was discharged after 2 months with rehabilitation for 15 days and with right-sided hemiparesis.

## DISCUSSION

ADEM is an acute demyelinating disorder of the central nervous system with multifocal white matter involvement. The main characteristic of the disease is diffuse neurological signs with variable outcomes. Nevertheless, some forms of presentation have an atypical presentation and they might be a challenge to diagnose and manage. Acute hemorrhagic encephalomyelitis (AHEM) is considered a rare form of ADEM, frequently seen among adults.[3] AHEM is usually fatal, whereas full recovery is the rule for patients with ADEM. Both are usually preceded by viral infections or vaccination. Nevertheless, no etiological factor was identified in CSF or the brain in the case of ADEM. Although many infectious and vaccine antecedents have been associated with both conditions, an active search for such causes is often unrevealing.

As per the clinical presentation initially, the main differential diagnosis of the illness was infectious meningitis, encephalitis, vasculitis, fulminant multiple sclerosis, venous sinus thrombosis, or hemorrhagic cerebrovascular disease. Most patients have a fever at the time of presentation, differentiating the illness from cerebrovascular accidents.[5] But infectious meningitis or encephalitis has a similar presentation to AHEM as they are more commonly preceded by respiratory tract infections. But on detailed investigations, none of the infectious agents was isolated from CSF and also the initial imaging finding was not suggestive of any suppurative enhancement. And also, it is very difficult to distinguish ADEM and AHEM based on the MRI findings and diagnosis mainly relies on the biopsy then.[6]

In our patient, several clinical findings, acute fulminant presentation, and no infectious cause are detectable, and MRI findings strongly support the diagnosis of AHEM. Previous studies have demonstrated promising results with IV methylprednisolone, IVIG.[7,8] Hence following the same protocol, our patient stabilized and showed signs of improvement.

## CONCLUSION

Even though the incidence of AHEM in children is rare, mortality has been higher in these cases. The diagnosis is challenging in the pediatric age group. However, prompt clinical suspicion and imaging studies will help in diagnosis and effective management with improved outcomes.

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