Case Report

MONOPHASIC ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM) IN A 9 YEAR CHILD PRESENTING TO PEDIATRIC EMERGENCY WITH ABNORMAL MOTOR MOVEMENTS

Chauhan G1, *Bhatia P2, Bhardwaj AK1 and Sharma PD1

1Department of Pediatrics, M.M. Institute of Medical Sciences and Research, Mullana-133-207, Ambala
2Department of Pediatrics, Post Graduate Institute of Medical Education and Research, Chandigarh-160012, India
*Author for Correspondence

ABSTRACT

Introduction
ADEM is an uncommon monophasic inflammatory demyelinating disease. It needs to be differentiated from other demyelinating conditions like multiple sclerosis and neuromyelitis optica, due to different prognostic and treatment implications. Aims & Objectives: To highlight the clinical and radiological presentation of a case of ADEM in a 9 year old girl child. Case Summary: The child presented to emergency in altered sensorium with continuous abnormal movement of both lips and eyes. Clinical history revealed weakness in right upper and lower limb along with high grade fever and vomiting for past one week. On clinical examination, the patient had a GCS of E2V4M2, bilateral reactive pupil and extensor plantars. Investigations revealed mild CSF pleocytosis with MRI showing multifocal ill-defined areas of increased T2 signal intensity in white matter consistent with diagnosis of ADEM. Discussion: The present case of ADEM is highlighted due to its odd clinical presentation in form of abnormal motor movements, which are more common in adult variant. Moreover there was no preceding history of respiratory or GIT illness, though it is known to be present in at least 50-75% of pediatric cases. MRI is needed for diagnosis as well as to exclude other conditions.

Key Words: Abnormal Movements, Demyelinating, MRI, Pediatric, White Matter

INTRODUCTION

Acute Disseminated Encephalomyelitis (ADEM) is an uncommon acute inflammatory monophasic demyelinating disorder of central nervous system characterized by diffuse neurological signs and symptoms coupled with evidence of multifocal lesions of demyelination on neuroimaging. It is more common in children with equal sex distribution. The condition is estimated to account for 10-15% of acute encephalitis cases in the United States, Johnson (2005) however the true incidence of disease in India is undetermined and is likely to be more frequent than reported. The condition was originally described by Lucas in 18th century, but Mcalpine (1931) described three set of cases with ADEM: 1) Post vaccination 2) After infectious fevers and 3) Spontaneous.

The present case report is presented with an aim to highlight the odd clinical presentation of ADEM along with the characteristic radiological findings and a brief review of literature on the current subject.

CASES

Clinical Presentation
The child presented to Pediatric emergency wing of MMIMSR, Mullana, in a state of altered sensorium with continuous abnormal motor movements of lips and eyes along with momentary stiffening of all limbs.

History of Present Illness
Child was apparently well a week before the present complaints. He also had history of high grade fever for 5-6 days, weakness of right upper and lower limb for last 4 days, vomiting for 2-3 days and abnormal motor movements since last one day.
Case Report

Past History
There was no preceding history of any respiratory or GIT infection. No past or family history of any seizures and no history of any recent vaccination.

Clinical Examination Findings
The child had a Glasgow Coma Scale (GCS) score of 8/15 i.e. E₂ V₄ M₂; He was febrile (body Temp 102.4 F) with mild tachycardia (140b/mt).
CNS examination: Revealed bilateral reactive pupils and extensor plantars. There were no signs of meningeal irritation.

Other Systemic Examination
Chest, cardiovascular and abdominal examination was within normal limits and did not reveal any significant finding.

Investigations
Hemogram, Renal function tests, urine routine and Chest x-ray were all within normal limits.

Lumbar Puncture
CSF examination revealed mild lymphocytic pleocytosis with normal proteins. No oligoclonal bands were seen.

Neuroimaging (MRI)
MRI brain revealed relatively diffuse and symmetrical hyperintense areas of signal abnormality on T2 involving subcortical white matter supra and infra tentorially. Similar hyperintense areas (>1-2cm) were also noted involving deep grey matter of bilateral hippocampi, bilateral thalami and basal ganglia (Figure 1A-B).

Figure 1: T2 weighted images of MRI brain in the index case.  A- Relatively diffuse and symmetrical hyperintense areas involving sub cortical white matter. B- In addition to white matter lesions, deep grey matter involvement in form of bilateral basal ganglia and thalami noted

Considering the characteristic MRI findings and after correlating with clinical features, a diagnosis of Acute disseminated encephalomyelitis was established and patient was started on specific treatment. However, there were few odd points which are highlighted in table 1.
**Case Report**

**Table 1: Highlights the points for and against the diagnosis of ADEM in present case**

<table>
<thead>
<tr>
<th>For diagnosis</th>
<th>Against diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Acute onset polysymptomatic neurological</td>
<td>1. No preceding respiratory, GIT or other infection before onset of symptoms</td>
</tr>
<tr>
<td>involvement with encephalopathy</td>
<td></td>
</tr>
<tr>
<td>2. First Clinical event</td>
<td>2. No immediate prior vaccination exposure</td>
</tr>
<tr>
<td>3. Relatively Characteristic MRI findings</td>
<td>3. Abnormal motor movements, which are quiet uncommon in children</td>
</tr>
</tbody>
</table>

**Treatment Outline**

**At Admission**

For continuous abnormal activity of lips and eyes, the child was given Inj. Midazolam 1.5ml stat IV, followed by a loading dose of inj. Phenytoin 300mg IV in 20ml NS over 30 mts.

In addition, child was started on broad spectrum antibiotics, antiviral, inj. Dexamethasone, Oxygen and IV fluids.

**Following Diagnosis**

The child was put on inj. Methylprednisolone 450mg IV in 100ml NS over 1 hour and planned to be continued on same dose for next 4-5 days. However, she was lost to further treatment and follow up after 2 days of institution of specific therapy.

**DISCUSSION**

ADEM classically occurs within 2 days to 4 weeks following a viral infection. Approximately 70-93% of patients report a clinically evident antecedent infection during the prior few weeks, Leake et al., (2004) and Tenembaum et al., (2007). The infection or antigen exposure is suggested to trigger an autoimmune attack against self myelin of brain via possible molecular mimicry, resulting clinically in ADEM. However, it is not a universal finding and currently is not included in the new consensus clinical criteria. Present day vaccines are also unlikely to cause ADEM except for the animal brain derived rabies vaccine.

In the index case too, no prior infectious or vaccination history was evident. The first set of consensus diagnostic criteria for diagnosis of ADEM in children <10 years were proposed by the International Pediatric MS Study Group (IPMSSG) in 2007, Krupp (2007). These criteria are conservative by requiring encephalopathy in all cases as it is a major specific clinical feature that helps distinguish ADEM from Multiple sclerosis (MS). In our case too patient had an altered conscious state, at presentation, as evident by a low GCS. Recent prospective studies on pediatric ADEM by Mikealoff et al., (2007) and Tenembaum et al., (2002) have found presence of polysymptomatic neurologic symptoms and encephalopathy in 100%, 100% and 100% and 60% cases respectively. Our index case too had both specific clinical presentation features, though presence of abnormal motor movements was an odd feature as it is relatively uncommon in pediatric group as compared to adult ADEM.

The present consensus criteria also require presence of an abnormal MRI, which shows multifocal areas of T2, weighted signal abnormalities in CNS white matter and the lesions to be larger than 1-2cm with usually indistinct or non-sharp borders. Absolute and relative Peri-ventricular sparing was reported by Dale et al., (2000) in 78% cases. Our case too demonstrated extensive subcortical white matter involvement, with large lesions with indistinct borders. Deep grey matter involvement in form of hyperintense lesions in basal ganglia and thalami were also noted.

The treatment of choice is short term Methylprednisolone inj. with relatively good long term outcome; however 3 monthly MRI is advised to rule out development of new lesions. In non responsive cases, IV immunoglobulins are advised.

**Take Home Message**

1. ADEM should be suspected in all cases with acute onset monophasic multifocal neurological deficits and encephalopathy.
2. Presence of prior infectious or vaccination history is not a diagnostic or universal criterion.
3. MRI should be done both to establish diagnosis as well as to rule out other inflammatory demyelinating conditions, for treatment and prognostication purposes.

REFERENCES