

ORIGINAL RESEARCH

Monophasic Acute Disseminated Encephalomyelitis (ADEM) in A 8 Year Child Presenting to Department of Pediatrics with Altered Sensorium and Weakness in Lower Limbs

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ABSTRACT

Introduction: ADEM is an uncommon monophasic inflammatory demyelinating disease. It needs to be differentiated from other demyelinating conditions like multiple sclerosis and neuromyelitisoptica, due to different prognostic and treatment implications. Aim of the case report is to highlight the clinical and radiological presentation of a case of ADEM in a 8 year old girl child.

Case Summary: The child presented to emergency in altered sensorium. Clinical history revealed weakness in bilateral lower limb along with high grade fever and difficulty in speaking, swallowing and walking for past one week. On clinical examination, the patient had a GCS of E2V3M4, bilateral reactive pupil, deep tendon reflexes brisk 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.

Conclusion: 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.

Keywords: Abnormal Movements, Demyelinating, MRI, Pediatric, White Matter

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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)¹ 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)² 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.

CASE REPORT

The child presented to Pediatric emergency wing of RMCH, Bareilly, in a state of altered sensorium. Child was apparently well 15 days before the present complaints. She also had history of difficulty in walking for last 15 days, high grade fever for 10 days, difficulty in speaking for last 5 days and altered sensorium since morning. 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. The child had a Glasgow Coma Scale (GCS) score of 9/15 i.e. E2 V3 M4; He was febrile (body Temp 102.4 F) with mild tachycardia (140b/mt). CNS examination: Revealed bilateral reactive pupils, deep tendon reflex brisk and extensor plantars. There were no signs of

meningeal irritation. Chest, cardiovascular and abdominal examination was within normal limits and did not reveal any significant finding. Hemogram, Renal function tests, urine routine and Chest x-ray were all within normal limits. CSF examination revealed mild lymphocytic pleocytosis with normal proteins. No oligoclonal bands were seen. 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).

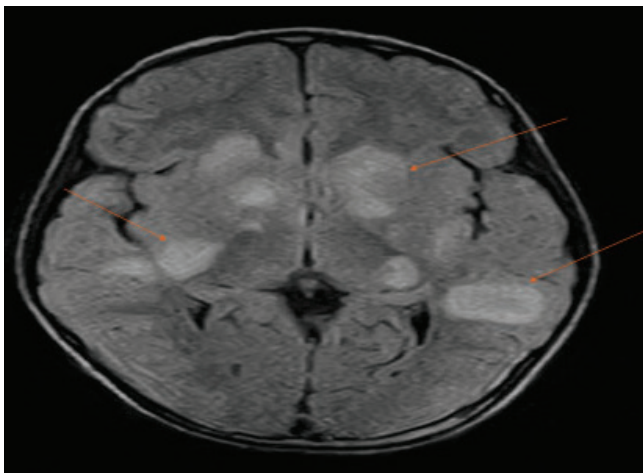


Figure-1: T2 weighted images of MRI brain in the index case. (A) Relatively diffuse and symmetrical hyperintense areas involving sub cortical white matter.

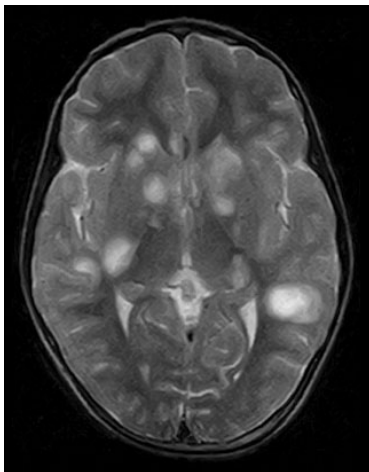


Figure-1: T2 weighted images of MRI brain in the index case. (B) In addition to white matter lesions, deep grey matter involvement in form of bilateral basal ganglia and thalami noted

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.

For diagnosis	Against diagnosis
Acute onset polysymptomatic neurological involvement with encephalopathy	No preceding respiratory, GIT or other infection before onset of symptoms
First Clinical event	No immediate prior vaccination exposure
Relatively Characteristic MRI findings	Abnormal motor movements, which are quiet uncommon in children

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

Treatment Outline at Admission

Initially child was managed on line of acute encephalitis and started on broad spectrum antibiotics, antiviral, inj. Dexamethasone, Oxygen and IV fluids.

Following Diagnosis

The child was put on inj. Methylprednisolone @ 30mg/kg/day in 100ml NS over 1 hour for 5 days. Then put on oral steroid for next 1 month and tapered and stop. Patient gradual improved to the treatment.

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., (2204) and Tenembaum et al., (2007).³⁻⁴ ADEM is triggered by infection or antigen exposure leading to an autoimmune attack against self myelin of brain via possible molecular mimicry. 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. 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).⁴⁻⁵ 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)⁶ and Tenembaum et al., (2002)⁷ have found presence of polysymptomatic neurologic symptoms and encephalopathy in 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)⁸ in 78% cases. Extensive subcortical white matter involvement with large lesions and indistinct borders were also seen in our case. Deep grey matter involvement in form of hyperintense lesions in basal ganglia and thalami were also noted. Short term Methylprednisolone inj. is treatment of choice available; however to rule out development of new lesions, 3 monthly MRI is advised. IV immunoglobulins are advised in non responsive cases,

CONCLUSION

Acute Disseminated Encephalomyelitis (ADEM) is a demyelinating syndrome which commonly follows a viral infection or immunisation. ADEM is mostly monophasic although recurrences have been reported in rare cases. Characteristic features are perivascular inflammation, edema and demyelination within the central nervous system. Clinical features vary from focal to multifocal neurological dysfunction

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

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