International Journal of Contemporary Pediatrics
Ranabijuli PK et al. Int J Contemp Pediatr. 2024 Oct;11(10):1495-1498
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20242753

Case Report

Acute disseminated encephalomyelitis in a five-year-old girl: a case report

Pradeep Kumar Ranabijuli*, Komal Bhagwan Kamble, Nazparveen L. A., Sita Kumari

Department of Pediatrics, JagjivanRam Railway Hospital, Mumbai, Maharashtra, India

Received: 17 July 2024
Revised: 14 August 2024
Accepted: 21 August 2024

***Correspondence:**
Dr. Pradeep Kumar Ranabijuli,
E-mail: pradeepranabijuli@gmail.com

ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated demyelinating central nervous system disorder with a predilection to early childhood. ADEM is generally considered a monophasic disease. However, recurrence has been described and defined as multiphasic disseminated encephalomyelitis (MDEM). ADEM often occurs after infection or immunization and is clinically defined as acute polyfocal neurological deficits, including encephalopathy. Many times, ADEM is a diagnosis of exclusion, and early diagnosis and treatment are the keys to favorable outcomes. Magnetic resonance imaging (MRI) typically demonstrates reversible, large, ill-defined white matter lesions of the brain and often also the spinal cord, along with frequent involvement of thalami and basal ganglia. CSF analysis may reveal mild pleocytosis and elevated protein but is generally negative for intrathecal oligoclonal bands. We report a case of five years six-month- old girl who presented with fever, vomiting, headache, and cough for seven days. She had one episode of involuntary passage of urine. The child was lethargic at presentation and was unable to stand or walk. Clinical features and investigations, including MRI brain and spine, were suggestive of ADEM. She was started on Inj. Methylprednisolone followed by oral prednisolone in tapering dose for four weeks. Clinical improvement was seen in the form of improvement in activity and power in limbs. The child improved with normal neurological function on day three of inj methylprednisolone. On follow-up for six months, the child did not have any relapse and had complete neurological and radiological recovery.

Keywords: ADEM, Monophasic, Encephalopathy, Methylprednisolone

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is a monophasic, inflammatory, and demyelinating disorder of the central nervous system that typically follows a febrile infection. Etiopathogenesis is thought to be immune mediated, because in up to three-fourths of the cases; it follows an antecedent infection or immunization.[1] In view of the treatment differences between ADEM and other demyelinating disorders, being familiar with ADEM is essential for Pediatricians in managing acute neurological disorders. Currently, there are no specific biomarkers available to diagnose ADEM; hence, diagnosis is made after excluding clinical and laboratory findings and suggestive neuroradiological features of another disease.[2] At present, MRI plays a vital role in diagnosing ADEM, revealing characteristic demyelinating brain lesions. It helps distinguishing ADEM from other neurological conditions, guiding treatment for better patient outcomes.[2]

CASE REPORT

A five years, six month old female child presented to our hospital with a history of fever, cough, headache, and multiple episodes of vomiting for seven days. She complained of weakness in her lower limbs after five days of fever and later was unable to walk and stand. She had one episode of involuntary passage of urine.

There was no history of recent trauma or vaccination; however, twenty days ago, she was given an intramuscular Ondansetron injection by a private practitioner for vomiting. There was no history of abnormal movements or cranial nerve involvement such as visual loss, deviation of eyeball, swallowing difficulty, or breathing difficulty. There was no family history of similar complaints.

Clinical examination on admission revealed the child was drowsy, lethargic, irritable, dehydrated, and febrile with a heart rate of 156/min with feeble pulses in lower limbs, respiratory rate of 24/min, blood pressure 100/70 mmHg (50^{th} centile for age) and saturation 98% on room air. Nervous system examination-the child was drowsy and not responding to commands; tone and reflexes were normal, and power was 3/5 in upper limbs and 2/5 in lower limbs. No signs of meningeal irritation were present. Other systemic examinations were normal.

Routine blood investigations were normal, and CSF examination showed a total of 12 mononuclear cells and no oligoclonal band. Anti-MOG (IgG) antibodies and anti-aquaporin 4 (NMO IgG) antibodies were also negative.

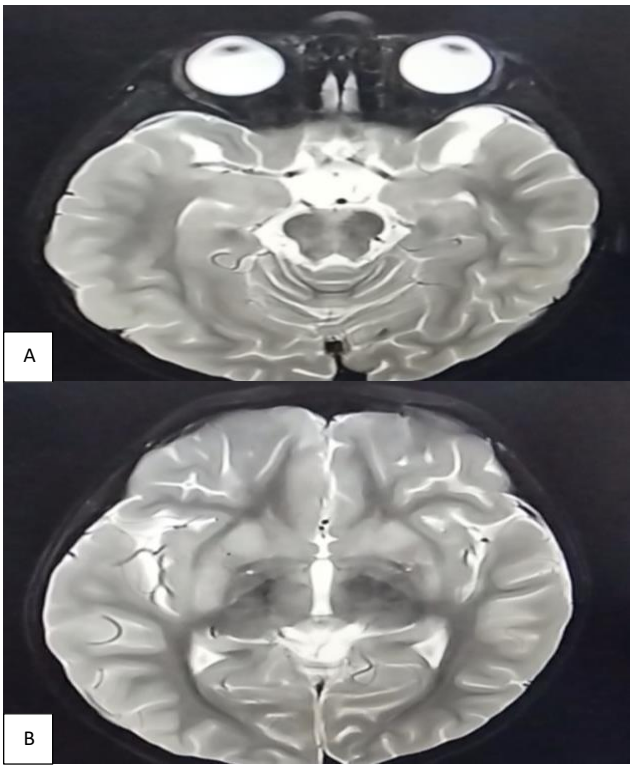

Figure 1 (A and B): MRI brain dated 07/11/2022 multiple bilateral asymmetric patchy ill-defined mildly hyperintense non-enhancing lesions are seen in the basal ganglia, thalami and caudate nuclei and the dorsal pons without any mass effect.

The imaging feature suggest possibility of encephalitis/ ADEM. There is mild asymmetric prominence of the sub arachnoid CSF space in the left anterior temporal region, suggesting an ill-defined arachnoid cyst.

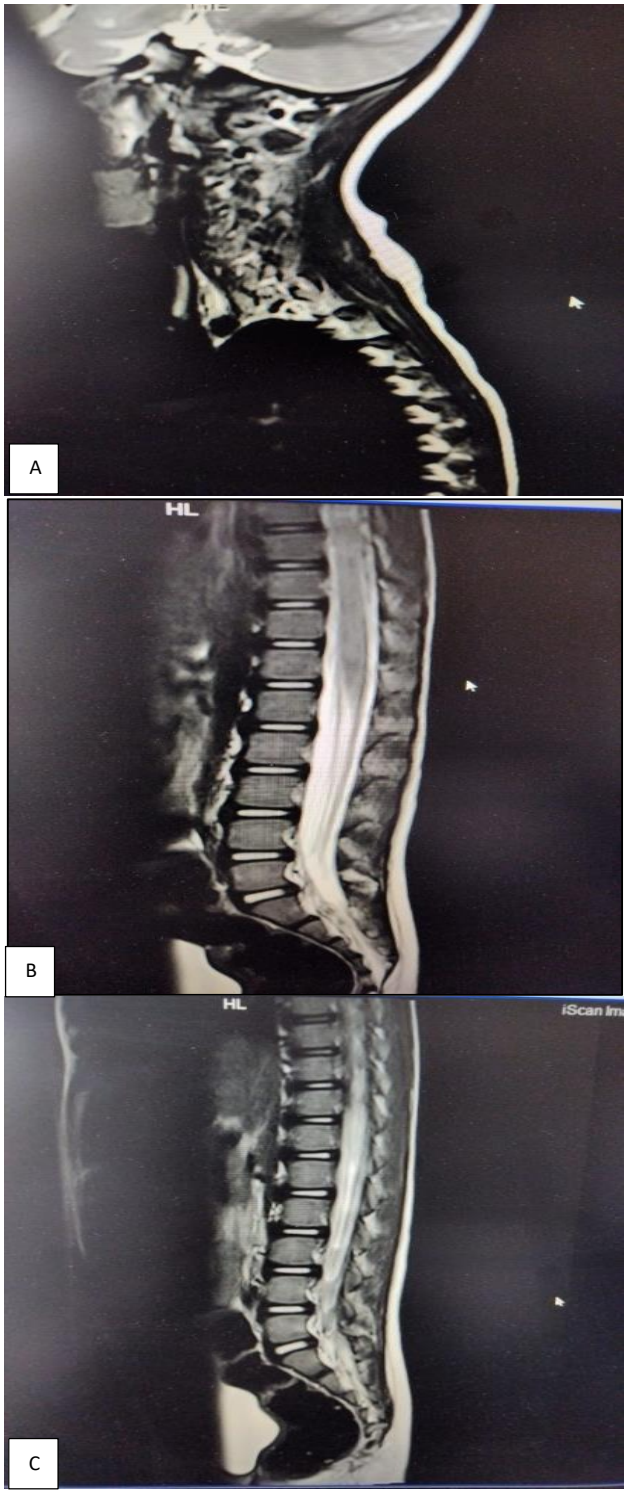

Figure 2 (A-C): MRI spine dated 07/11/2022 patchy ill-defined mild hyperintense non enhancing lesions are seen in spinal cord at C5- D2 and D7-D10 vertebral levels. Above features s/o myelitis.

Child was started on pulse therapy inj. methylprednisolone with a dose of 30 mg/kg/day for 5 consecutive days followed by oral prednisolone therapy with a dose of 1 mg/kg/day. Child showed good response in the form of improved sensorium and power in both lower limbs. By day three of inj. methylprednisolone

child started walking and power in upper and lower limbs gradually improved.

DISCUSSION

The annual incidence of ADEM is reported to be 0.4-0.8 per 100000 and the disease more commonly affects children and young adults in winter or spring. Most of the cases are reported post exanthematous infection or vaccination.[1,3-7] There seems to be no gender predominance.[8] The mean age at presentation is 6-8 years. In our child there is past history of fever with cold and cough 20 days back.

ADEM typically begins within six days to 6 weeks following an Antigenic challenge. It may be abrupt, acute, or may evolve over a period of a few days. ADEM typically presents as a monophasic illness but sometimes may have a biphasic or multiphasic course, depending on the neuraxis affected. Characteristic clinical features include sudden onset multifocal neurologic disturbances such as visual field defects, aphasia, motor and sensory deficits, ataxia, movement disorders, a depressed level of consciousness, focal or generalized seizures, and psychosis. In our case, the child presented with weakness in lower limbs and a depressed level of consciousness.

With the wider use of MRI, ADEM is now diagnosed more frequently. MRI T2 enhancing images shows disseminated multifocal lesions in the white matter, basal ganglia, thalamus and brainstem consistent with edema, inflammation and demyelination.[9] Sometimes during initial course of disease we may find a normal MRI brain. In our child MRI brain and spine findings are as mentioned along with photographs.

CSF analysis is usually normal, but sometimes, mild elevation of protein with lymphocytic pleocytosis can be found. Markers such as oligoclonal immunological bands, IgG, or myelin basic protein (MBP) are sometimes detectable but not diagnostic.[10] In our child, the CSF examination showed a cell count of 12 cells/cumm with 100% mononuclear cells, and other biochemical parameters were normal. In CSF, NMO-MOG antibodies were negative, and oligoclonal bands were absent. The electroencephalogram (EEG) often shows nonspecific features of an encephalopathic process, and visual evoked potential (VEP) responses may be delayed. In the absence of specific biologic markers, the diagnosis of ADEM is based on the clinical and radiologic features.

Due to lack of any pathognomic clinical feature or specific biomarker few differential diagnoses must be excluded before diagnosing ADEM. First priority should be to rule out infective cause of meningoencephalitis after ruling out infective causes demyelinating inflammatory process should be looked for.

Spontaneous improvement has been documented in patients with ADEM. However, recovery is incomplete in patients with ADEM who do not receive any form of immunomodulatory treatment. Treatment of ADEM includes supportive and immunomodulatory therapy.[11] In our child, inj methylprednisolone 400 mg in 100 cc of normal saline was given intravenously over a period of one hour once daily for five days, followed by Oral Prednisolone in the dose of 1mg per kg per day for two weeks and tapered gradually over next four weeks. Before giving inj. methylprednisolone, an X-ray chest and Mantoux test were done.

The outcome of ADEM is generally good, with 57-89% of children making a full recovery.[12,13] ADEM is considered to be a monophasic illness, but relapse may occur, and if it represents the same acute monophasic immune process, the term MDEM is used. However, it must be differentiated from a second attack of multiple sclerosis, which may take months to years and is more common in older age groups. A patient presenting with optic neuritis, ocular lesions, oligoclonal bands in CSF examination, disseminated in space and time, and periventricular lesion in MRI goes in favor of multiple sclerosis (MS). It is very necessary to differentiate ADEM/MDEM from MS as an early institution of therapy may alter the course of MS. Our child had clinical features and investigations suggestive of ADEM and showed complete clinical improvement after five doses of inj. methylprednisolone.

A regular follow up for six months revealed no relapse and the child remained neurologically normal.

CONCLUSION

ADEM is a rare autoimmune demyelinating disorder that mainly affects the CNS and is characterized by an acute inflammatory response targeting the myelin sheath surrounding fibers in the brain and spinal cord. This case report emphasizes that ADEM primarily relies on clinical and imaging findings. The early recognition and accurate diagnosis of ADEM facilitate timely management and minimize neurological damage. Prompt initiation of appropriate treatment is important to promote favorable outcomes in affected children.

ACKNOWLEDGEMENTS

Authors would like to thank the hospital administration of JagjivanRam hospital Mumbai and the scientific and ethical committee of the hospital.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Murthy SN, Faden HS, Cohen ME, Bakshi R. Acute disseminated encephalomyelitis in children. Pediatrics. 2002;110(2pt1):e21.

2. Kumar P, Kumar P, Sabharwal RK. Acute disseminated encephalomyelitis: case report and brief review. J Family Med Prim Care. 2014;3(4):443-5.
3. Ascherio A, Munger K. Epidemiology of multiple sclerosis: From risk factors to prevention. Semin Neurol. 2008;28(1):17-28.
4. Panicker JN. Bangalore: National Institute of Mental Health and Neurosciences, NIMHANS (Deemed University). Acute Disseminated Encephalomyelitis: Clinical Profile and Predictors of Outcome (dissertation). 2004.
5. Menge T, Hemmer B, Nessler S, Wiendl H, Neuhaus O, Hartung HP, et al. Acute disseminated encephalomyelitis: An update. Arch Neurol. 2005;62(11):1673-80.
6. Anlar B, Basaran C, Kose G, Guven A, Haspolat S, Yakut A, et al. Acute disseminated Encephalomyelitis in Children: Outcome and Prognosis. Neuropediatrics. 2003;34(4):194-9.
7. Tenembaum S, Chamoles N, Fejerman N. Acute disseminated encephalomyelitis: A long-term follow-up study of 84 pediatric patients. Neurology. 2002;59(8):1224-31.
8. Schwarz S, Mohr A, Knauth M, Wildemann B, Storch-Hagenlocher B. Acute disseminated encephalomyelitis: A follow-up study of 40 adult patients. Neurology. 2001;56(10):1313-8.
9. Singhi PD, Ray M, Singhi S, Kumar Khandelwal N. Acute disseminated encephalomyelitis in North Indian Children: Clinical profile and follow-up. J Child Neurol. 2006;21(10):851-7.
10. Marchioni E, Tavazzi E, Minoli L, Del Bue S, Ferrante P, Piccolo G, et al. Acute disseminated encephalomyelitis. Neurol Sci. 2008;29:S286-8.
11. Alexander M, Murthy JM. Acute disseminated encephalomyelitis: Treatment guidelines. Ann Indian Acad Neurol. 2011;14(1):S60-4.
12. Hynson JL, Kornberg AJ, Coleman LT, Shield L, Harvey AS, Kean MJ. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. Neurology. 2001;56(10):1308-12.
13. Dale RC, de Sousa C, Chong WK, Cox TC, Harding B, Neville BR. Acute disseminated encephalomyelitis, multiphasic disseminated encephalomyelitis and multiple sclerosis in children. Brain. 2000;123:2407-22.

Cite this article as: Ranabijuli PK, Kamble KB, Nazparveen LA, Kumari S. Acute disseminated encephalomyelitis in a five-year-old girl: a case report. Int J Contemp Pediatr 2024;11:1495-8.