Case Report

Acute Disseminated Encephalomyelitis—Masquerading as Pediatric Stroke: Case Report

Varsha H. Chauhan, Richa Chaudhary, Payal Meshram

Department of Pediatrics, Mahatma Gandhi Institute of Medical Sciences (MGIMS), Sevagram, Wardha, Maharashtra, India

Acute disseminated encephalomyelitis (ADEM) is an acute monophasic syndrome caused by immune-mediated inflammatory demyelination, often associated with immunization or viral illness. ADEM is associated with multiple neurological symptoms. We are presenting a case of ADEM with stroke, which responded very well to high-dose steroids. Here we report a case of ADEM, masquerading as pediatric stroke.

KEYWORDS: Acute disseminated encephalomyelitis, steroid, stroke

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an autoimmune inflammatory disorder of the central nervous system (CNS). It is thought to be immune mediated and usually follows an antecedent infection or immunization.[1] Magnetic resonance imaging (MRI) is the imaging modality of choice to diagnose ADEM, which shows lesions in white matter of the brain. No specific biomarkers are available currently to diagnose ADEM.

CASE REPORT

A 4-year-old girl presented with left-sided weakness, inability to talk, and urinary and bowel incontinence for 15 days and fever for 7 days. Fever was not associated with chills, and it rises mainly during evening time. Antenatal, natal, and postnatal histories were uneventful. Developmental history was normal.

Her general physical examination revealed heart rate as 120 beats/min regular, respiratory rate as 24 breath/min, and blood pressure as 110/70 mmHg, and pallor was present. On CNS examination, child was found to be conscious, oriented to time, place, and person. Higher functions were intact. Supranuclear type of facial nerve palsy was noted. The power of upper limb and left lower limb 3/5 and hypertonia with exaggerated reflexes was noticed. There was no sensory loss. She had motor weakness of left upper and lower limbs, but her bilateral deep tendons reflexes were exaggerated; bilateral planters were extensors.

On investigations, complete blood count, serum electrolytes, and kidney and liver function test results were found to be normal. Blood culture report was normal. Result of cerebrospinal fluid (CSF) examination was normal. MRI of the brain revealed bilateral, symmetrical, altered signal intensity lesions in bilateral periventricular deep white matter, centrum semiovale, bilateral thalami, and punctate areas were noted in pons, appearing hyperintense on T2-weighted (T2W) [Figures 1 and 2] and fluid attenuation inversion recovery sequence, and hypointense on T1-weighted (T1W) sequence [Figure 3]. The lesions in bilateral thalami and pons showed bilateral symmetrical diffusion restriction on diffusion-weighted imaging. Lesions in bilateral periventricular deep white matter and centrum semiovale showed peripheral diffusion restriction. Aforementioned findings were suggestive of ADEM. The patient was treated with intravenous methyl prednisolone for 5 days and shifted to oral prednisolone. Recovery—child responded to treatment, regained control over bladder, and showed improvement in movements of affected limbs.

Address for correspondence: Dr. Varsha Hitendra Chauhan, Department of Pediatrics, Mahatma Gandhi Institute of Medical Sciences, Sevagram, Wardha, Maharashtra 442102, India. E-mail: varsha@mgims.ac.in

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**Discussion**

The incidence of ADEM is reported to be 0.4–0.8 per 100,000, and the disease more commonly affects children and young adults in winter/spring. Most of the cases are reported postexanthematous infection or vaccination. The mean age at presentation is 6–8 years. Following an antigenic challenge, ADEM usually begins within 6 days to 6 weeks. ADEM typically presents as a monophasic illness, which is usually abrupt or acute in onset but may also evolve over a period of few days. Depending on the neural axis affected, ADEM may sometimes have a biphasic or multiphasic course. Characteristic clinical features include sudden onset of 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. As in our case, child presented with left-sided weakness, inability to talk, urinary and bowel incontinence, and fever.

The diagnosis of ADEM is based on the clinical and radiological features, and with the wider use of MRI, ADEM is now being diagnosed more frequently. MRI T2-enhancing images show disseminated multifocal lesions in the white matter, basal ganglia, thalamus, and brain stem consistent with edema, inflammation, and demyelination. Spontaneous improvement has been documented in patients with ADEM. However, the recovery is incomplete in patients with ADEM not receiving any form of immunomodulatory therapy. Treatment of ADEM is immunomodulatory therapy and supportive management. High-dose intravenous methyl prednisolone, intravenous immunoglobulin (IVIg), and plasmapheresis are the various modalities of treatment available. Intravenous methyl prednisolone is the first-line drug (10–30 mg/kg/day, up to a maximum of 1 g/day). It is given for 3–5 days followed by oral corticosteroid treatment continued, which is then gradually tapered more than 6 weeks to reduce the risk of relapses. Failure of corticosteroid therapy warrants...
the use of either plasma exchange or IVIg (0.4 g/kg/day for 5 days) as the second-line treatment.\[8\]

ADEM generally has a good outcome with 57%–89% of children showing full recovery following immunomodulatory therapy.\[9\]

**CONCLUSION**

Any child presenting with acute polysymptomatic encephalopathy with minimal CSF changes and MRI showing extensive white-matter changes should prompt the diagnosis of ADEM and steroid therapy initiated as long-term prognosis is favorable following steroid therapy.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Murthy SN, Faden HS, Cohen ME, Bakshi R. Acute disseminated encephalomyelitis in children. Pediatrics 2002;110:e21.
2. Ascherio A, Munger K. Epidemiology of multiple sclerosis: from risk factors to prevention. Semin Neurol 2008;28:17-28.
3. Panicker JN. Acute Disseminated Encephalomyelitis: Clinical Profile and Predictors of Outcome (dissertation). Bangalore, Indir: National Institute of Mental Health and Neurosciences, NIMHANS (Deemed University); 2004.
4. Menge T, Hemmer B, Nessler S, Wiendl H, Neuhaus O, Hartung HP, et al. Acute disseminated encephalomyelitis: an update. Arch Neurol 2005;62:1673-80.
5. Anlar B, Basaran C, Kose G, Guven A, Haspolat S, Yakut A, et al. Acute disseminated encephalomyelitis in children: outcome and prognosis. Neuropediatrics 2003;34:194-9.
6. Tenembaum S, Chamoles N, Fejerman N. Acute disseminated encephalomyelitis: a long-term follow-up study of 84 pediatric patients. Neurology 2002;59:1224-31.
7. 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;21:851-7.
8. Alexander M, Murthy JM. Acute disseminated encephalomyelitis: treatment guidelines. Ann Indian Acad Neurol 2011;14:S6.
9. Hynson JL, Kornberg AJ, Coleman LT, Shield L, Harvey AS, Kean MJ. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. Neurology 2001;56:1308-12.