Case Report

Acute Disseminated Encephalomyelitis with Measles

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Abstract

Acute disseminated encephalomyelitis is an inflammatory demyelinating illness usually associated with infections or antecedent immunization. Due to control of most vaccine preventable diseases in developed countries, most cases of acute disseminated encephalomyelitis occur in developing countries and are seen secondary to nonspecific upper respiratory tract infections. We report a case of acute disseminated encephalomyelitis associated with measles in a 2½-year-old male child despite having measles vaccination in infancy. The diagnosis was based on clinical findings and CT scan of brain. The patient was managed with high dose corticosteroids along with supportive measures. He recovered completely and follow-up for six months revealed no neurological deterioration.

Key words: *Acute disseminated encephalomyelitis, Measles, Postinfectious encephalomyelitis*

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Introduction

Acute disseminated encephalomyelitis (ADEM), also known as postinfectious encephalomyelitis, is an acute widespread demyelinating disease of the central nervous system that typically presents as a monophasic disorder associated with multifocal neurologic symptoms and encephalopathy.[1] The incidence of ADEM is about 0.4 per 100000 people per year.[2] Although ADEM occurs in all ages, most reported cases are in children and adolescents, with the average age around 5 to 8 years.[3] It usually occurs following a viral infection but may appear following vaccination and bacterial or parasitic infection.[2]

Clinical manifestations like abrupt development of irritability, fever, headache, myalgia, and drowsiness often precede the neurological symptoms of ADEM. The hallmark of clinical features of ADEM is the development of a focal or multifocal neurological disorder. Initial neurological features include encephalopathy ranging from lethargy to coma, hemiparesis, cranial nerve palsies, and paraparesis.[4] Although initially the symptoms are usually mild, they worsen rapidly over the course of hours to days, with the average time to maximum severity being about four and a half days.[5]

No controlled clinical trials have been conducted on ADEM treatment. The widely accepted first-line treatment is high dose intravenous corticosteroids such as methylprednisolone or dexamethasone, followed by oral doses of prednisolone tapering within 3–6 weeks. Patients treated with methylprednisolone have shown better outcomes than those treated with dexamethasone.[5,6]

Due to control of most vaccine preventable diseases in developed countries, most cases of ADEM occur in developing countries.[7] We report a case of ADEM associated with measles despite having measles vaccination in infancy, who was treated effectively with corticosteroid.

Case Report

A 2½-year-old male child presented with intermittent fever for five days, irritability, excessive crying and

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vomiting followed by unconsciousness for 2 days. He had history of measles 10 days back. He was vaccinated according to EPI schedule. On examination, he was febrile, mildly pale, non-icteric and had no lymphadenopathy. There was brownish discoloration with fine desquamation of skin all over the body (Fig 1). The Glasgow coma scale was 8/15. There were no signs of meningeal irritation and cranial nerves were intact. There were hypotonia, hyporeflexia and quadriplegia. Other systemic examinations revealed no abnormalities. The child progressively deteriorated in the first 2 days, was comatosed (GCS 5/15) and remained in this condition for 4 days. Complete blood count, renal and liver function tests, arterial blood gas analyses and electrolyte reports were normal. Chest radiography and ultrasonography of abdomen showed normal findings. Cerebrospinal fluid (CSF) was clear with 4 WBC/cmm of which all were lymphocytes. Protein and sugar values were normal. CSF culture revealed no growth. However, CSF antigen or antibodies for measles could not be assessed. CT scan revealed diffuse hypodensity in the subcortical white matter, periventricular white matter, centrum semiovale, corona radiata, thalamus and brain stem without perilesional edema (Fig 2). All these findings were suggestive of ADEM. The patient was treated with intravenous methylprednisolone 30 mg/kg/day, which had been given for 5 days followed by oral prednisolone (1 mg/kg/day) for another 7 days. The child showed dramatic improvement and recovered completely. Follow up for six months revealed no neurological deterioration.

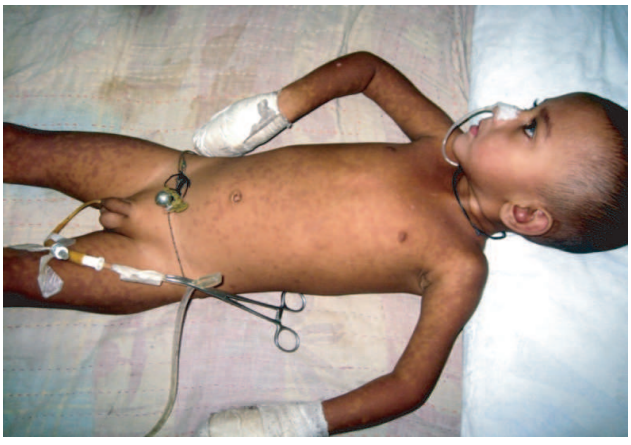

Fig 1. Brownish discoloration with fine desquamation of skin all over the body

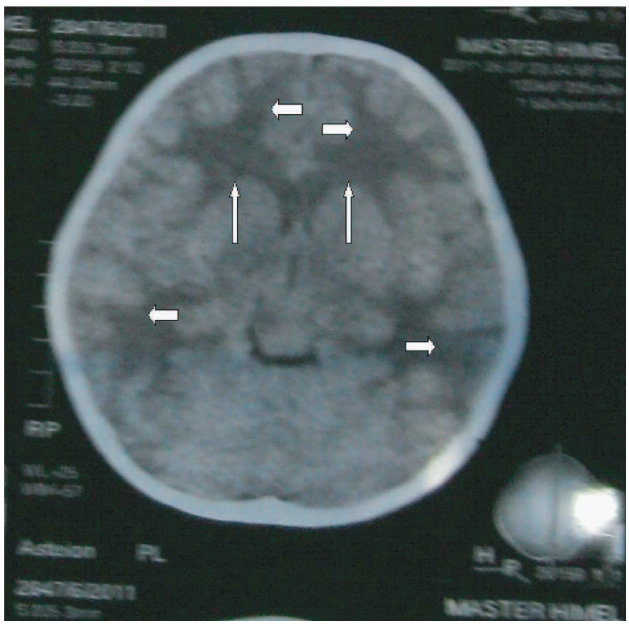

Fig 2. CT scan revealed diffuse hypodensity in the subcortical white matter (small arrows) and periventricular white matter (long arrows) without perilesional edema

Discussion

Acute disseminated encephalomyelitis is an acute demyelinating disorder of central nervous system characterized by diffuse neurological signs and multiple white matter lesions on neuroimaging.[6]

Postinfectious encephalomyelitis is associated with concomitant or antecedent infection, usually viral. Most notoriously, measles virus infection is followed by ADEM in approximately 1 in 1000 unvaccinated children and tends to produce more serious phenotype.[8] In developing and poor countries, because of poor implementation of immunization programs, measles and other viral infections are still widely prevalent and account for frequent occurrences of postinfectious demyelinating diseases. ADEM in developing countries is much more frequent than reported.[9]

Clinical features including fever, irritability, rapid neurological deterioration with history of measles, CT finding and subsequent improvement with high dose corticosteriod led us to the diagnosis of ADEM associated with measles. However, it is difficult to differentiate from subacute sclerosing panencephalitis (SSPE) presenting as ADEM, but presence of

myoclonic convulsions, latent period and no improvement with steroids can differentiate it from SSPE.[10] In myelitic form of ADEM, partial or complete paraplegia or quadriplegia and diminished or loss of reflexes also occur, which is consistent with our findings.[11]

ADEM produces multiple inflammatory lesions in the brain and spinal cord, particularly in the white matter. Usually these are found in the subcortical and central white matter and cortical gray-white junction of both cerebral hemispheres, cerebellum, brainstem, and spinal cord but periventricular white matter and gray matter of the cortex, thalami and basal ganglia may also be involved.[12] This is consistent with our findings.[12]

ADEM was associated with a mortality rate of 25% after measles infection with major residual neurologic sequel in 25 to 40% of survivors.[13] More recent data from two retrospective reviews of children diagnosed with ADEM found 100% survival, with more than 80% of children were neurologically normal in follow up.[2] Our case showed complete recovery. Follow up for six months revealed no neurological deterioration.

Conclusion

Acute disseminated encephalomyelitis is an acute yet treatable inflammatory and demyelinating disease with varying clinical presentation that responds well to immunomodulating therapy and the clinical outcome is favorable. Despite achieving and sustaining global measles vaccination coverage of about 80% over the past decade, measles remains the fifth leading cause of mortality among children aged <5 years worldwide. Because of contagiousness of measles, even sustained high coverage vaccination with single-dose strategy does not prevent large outbreaks of measles with significant morbidity and mortality. So, a second dose of measles immunization is essential for effective control of measles and prevention of neurological complications which is practiced in England and Wales, Albania, Romania, Oman, Shandong and Henan provinces of China and USA.[8]

References

1. Garg RK. Acute disseminated encephalomyelitis. Postgrad Med J 2003; 79: 11–17.
2. Leake JA, Albani S, Kao AS, Senac MO, Billman GF, Nespeca MP et al. Acute disseminated encephalomyelitis in childhood: epidemiological, clinical and laboratory features. Pediatr Infect Dis J 2004; 23: 756-764.
3. 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: 194–199.
4. Murthy JM**,** Yangala R, Meena AK, Reddy JJ. Clinical, electrophysiological and magnetic resonance imaging study of acute disseminated encephalomyelitis. J Assoc Physicians India1999*;* 47: 280–283.
5. Tenembaum S, Chamoles N, Fejerman N. Acute disseminated encephalomyelitis: a long-term follow up study of 84 pediatric patients. Neurology 2002; 59: 1224–1231.
6. Davis LE, Boss J. Acute disseminated encephalomyelitis in children: a changing picture. Pediatr Infect Dis 2003; 22: 829–831.
7. Samileh N, Hassan T. ADEM in children a descriptive study in Tehran Iran. Saudi Med J 2007; 3: 396–399.
8. Strebel P, Cochi S, Grawbosky M, Bilous J, Hersh BS, Okwo-Bele JM et al. The unfinished measles immunization agenda. Infect Dis 2003; 187: 1–5.
9. Murthy JM*,* Yangala R, Meena AK, Reddy JJ. Acute disseminated encephalomyelitis: clinical and MRI study from South India. J Neurol Sci 1999; 165: 133–138.
10. Comert S, Vitrinel A, Gursu HA, Deniz NC, Akin Y. Subacute sclerosing panencephalitis presenting as acute disseminated encephalomyelitis. Indian J Pediatr 2006; 73: 1119–1121.
11. Ropper AH, Brown RH. In: Adams and Victors principles of neurology. 8th edn. New York: McGraw-Hills, 2005: 790–792.
12. Wingerchuk DM. Postinfectious encephalomyelitis. Curr Neurol Neurosci Rep 2003; 3: 256–264.
13. Litvak AM, Sand IJ, Gibel H. Encephalitis complicating measles: report of 56 cases with follow-up studies in 32. Am J Dis Child 1943; 65: 265-295.