Case Report

Acute Disseminated Encephalomyelitis: Case Report and Brief Review

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Abstract

Acute disseminated encephalomyelitis (ADEM) is a rare disease of central nervous system with myriads of presentation. It is a diagnosis of exclusion and relies on neuroimaging which may be normal at the onset. It is a diagnostic challenge at its first attack. Here we present a case of ADEM which initially presented with atypical feature and normal neuroimaging but later turned out to be a case of ADEM. Early diagnosis and treatment holds the key for favorable outcome.

Keywords: Acute disseminated encephalomyelitis, central nervous system, neuroimaging

Introduction

Acute disseminated encephalomyelitis (ADEM) is an autoimmune inflammatory disorder of the central nervous system (CNS). Etiopathogenesis is thought to be immune mediated, because in up to three-fourths of the cases; it follows an antecedent infection or immunization.[1] Currently for ADEM, magnetic resonance imaging (MRI) is the imaging modality of choice to demonstrate lesions in white matter of brain. There are no specific biomarkers available currently to diagnose ADEM; hence, diagnosis is made after excluding clinical and laboratory findings and suggestive neuroradiological features of other disease.

Case Report

A 4-year-old boy was brought to our Hospital with complaints of slurring of speech and difficulty in swallowing and fever for 2 days. He was evaluated at emergency and his GCS was 10/15 (E4V2M4). On examination, he was in altered sensorium; tone was increased in upper and lower limb with brisk deep tendon reflexes. Plantars were upgoing and cranial nerve examination was normal. No meningeal sign was present. He was started on with IV antibiotic (ceftriaxone), acyclovir and supportive measures. Prior to this he had fever for 5 days, 8 days back along with mild cough and cold. On Day 4 of fever child had one episode of seizure. He was admitted in the intensive care unit of a local hospital and managed. MRI of brain was done which was unremarkable except for mildly dilated lateral and third ventricle. CSF cytobiochemistry was normal. He was managed with IV Ceftriaxione and IV Acyclovir IV Phenytoin and other supportive measures. Gradually he improved and was discharge in a stable condition after 6 days. Two days later patient developed slurring of speech and difficulty in swallowing with fever for which he was admitted in our hospital. Initial investigations revealed Hb - 11.8 gm/dl, TLC -15200/mcl, platelet - 3.13 lac. RFT and LFT were normal. CRP was 17 mg/L, Typhi dot, Widal test, Blood C/S and Malarial antigen test was negative. MRI of brain was done which showed inhomogeneous area of increased signal in T2 weighted images in basal ganglia, pons, midbrain, medulla, thalami, and frontal cortex [Figures 1-3]. Fundoscopy was normal. Lumbar puncture revealed CSF cell count was 3 cells (100% lymphocytes), Protein - 32 mg/dl and Glucose - 56 mg/dl (capillary blood glucose was 86 mg/dl) LDH - 12 U/L. CSF Bacterial antigen test causing encephalitis *(Streptococcus Group B, H. influenzae b, S. pneumoniae, N. meningitidis, E. coli K1)*, herpes IgM test and herpes PCR were negative. EEG revealed generalized cerebral dysfunction with no definitive irritable foci. Diagnosis of ADEM was considered. Child was started on IV methylprednisolone (500 mg) pulse therapy for 3 days, following which he was started on oral steroid therapy. He showed good response and speech improved. Gradually his sensorium became better. By Day 5, he started responding to commands. On day 6 visual evoked potential was done which revealed perception

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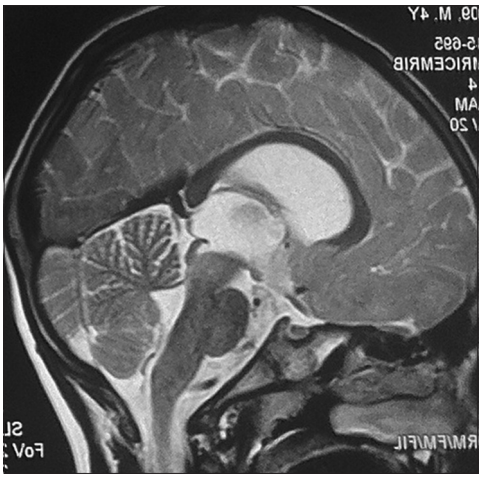

Figure 1: MRI T2 weighted image showing areas of hyper intensity involving brainstem

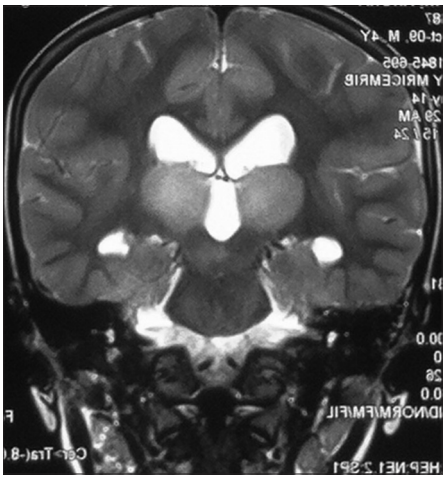

Figure 2: MRI T2 weighted image showing areas of hyper intensity involving caudate nucleus and thalamus

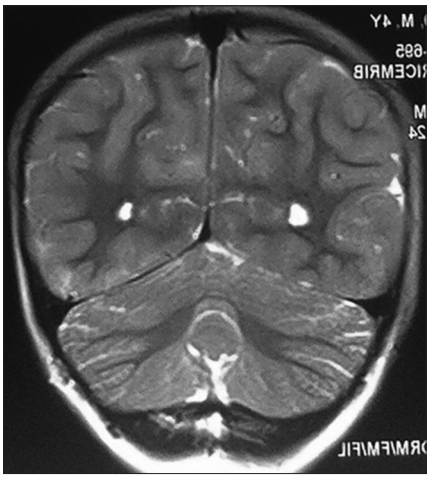

Figure 3: MRI T2 weighted image showing areas of hyper intensity involving white matter

of light bilaterally. Power in upper and lower limb gradually improved. He was discharged and at the time of discharge he was conscious oriented with GCS of E4 V5 M6. He was seen on follow up after 7 days and neurological examination revealed alert child with mild slurring of speech.

Discussion

The annual 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 case are reported post-exanthematous infection or vaccination.[2-6] There seems to be no gender predominance.[7] The mean age at presentation is 6–8 years.[8]

ADEM typically begins within 6 days to 6 weeks following an antigenic challenge. It may have abrupt, acute, or may evolve over a period of 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. As in our case, child presented with slurring of speech with difficulty in swallowing, altered sensorium and seizure.

CSF 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.[9] The electroencephalogram (EEG) often shows non-specific 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. In our case CSF and VEP studies were normal.

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.[8] Sometimes during initial course of disease we may find a normal MRI BRAIN. Initial MRI scan in our case had no evidence of ADEM but later MRI revealed finding suggestive of ADEM.

Spontaneous improvement has been documented in patients with ADEM. However, the recovery is incomplete in patients with ADEM not receiving any form of immunomodulatory treatment. Treatment of ADEM includes supportive, and immunomodulatory therapy. There is no controlled trial on its treatment. Most of the literature is in consensus with the use of high-dose intravenous methyl prednisolone, intravenous immunoglobulin (IVIg), and plasmapharesis as various modality of treatment. Intravenous methyl prednisolone is the first-line drug (10–30 mg/kg/day, up to a maximum of 1 g/day) for

3–5 days followed by oral corticosteroid treatment continued with gradual tapering over 6 weeks to reduce the risk of relapses. Intravenous immunoglobulin (IVIg) (0.4 gm/kg/day for 5 days) is another option. Either plasma exchange or IVIg, could be the second-line treatment, when corticosteroids fail.[10]

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

The outcome of ADEM is generally good, with 57–89% of children making a full recovery.[11,12] ADEM is considered to be monophasic illness but relapse may occur and if it represents same acute monophasic immune process, the term multiphasic disseminated encephalomyelitis (MDEM) is used. But it must be differentiated from second attack of multiple sclerosis which may take months to years and more commone in older age groups. 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 early institution of therapy may alter course of MS.

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