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Case Report

MULTIPHASIC DISSEMINATED ENCEPHALOMYELITIS (MDEM) - A CASE REPORT

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ABSTRACT
Acute disseminated encephalomyelitis (ADEM) is an acute inflammatory demyelinating disorder of central nervous system characterized by multifocal white matter involvement. MDEM, a type of relapsing ADEM is a rare entity in pediatric age group. Magnetic resonance imaging (MRI) is useful to confirm the diagnosis. Treatment with intravenous high dose methyl prednisolone, usually leads to a rapid improvement. We report a case of MDEM of 6 year old girl diagnosed on basis of clinical presentation and new lesions on MRI on relapse after two years. She was treated effectively with corticosteroids. Clinical and radiological findings differentiated it from recurrent ADEM and on follow up from Multiple Sclerosis.

Keywords: *Demyelination, MDEM, white matter*

INTRODUCTION
ADEM is immunologically mediated inflammatory demyelinating disease of central nervous system and usually has a monophasic course. Though relapses have been described as recurrent or multipasic (Enrico *et al.,* 2008). In most patients it follows a viral illness but has been reported after bacterial infection, immunizations, and drug and serum administration. Clinical manifestations include abrupt development of irritability and neurological signs like changes in long tract signs in children we report a case of ADEM with relapse treated effectively with corticosteroids. Multiphasic DEM (MDEM) needs to be differentiated from recurrent ADEM and Mutiple Sclerosis.

CASES
A 6 years old female child presented with intermittent fever, headache and irritability for 15 days, altered sensorium, seizures and vomiting for 5 days. On examination vitals were normal, neck rigidity was present, pupils were bilaterally reacting, tone was increased and reflexes were brisk. Hemogram, renal functions, liverfunctions, Electrocardiogram and Chest X-ray were normal.

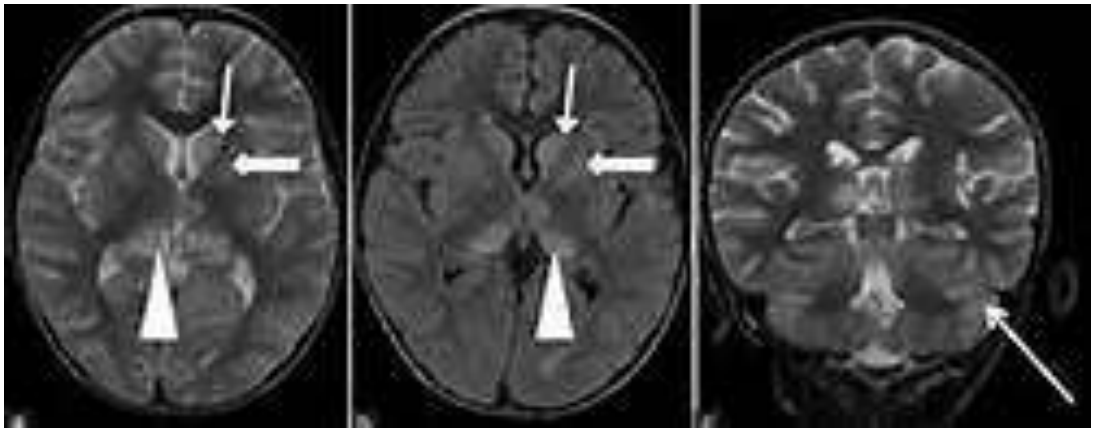

Figure 1: MRI brain showing few small hyperintense lesions in the sub cortical white matter of cerebral hemispheres, left caudate nucleus and rostrum of corpus callosum on right side

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CSF showed pleocytosis with increased proteins. MRI revealed few small hyperintense lesions in the sub cortical white matter of cerebral hemispheres, left caudate nucleus and rostrum of corpus callosum on right side suggestive of acute disseminated encephalomyelitis (figure 1). She was treated with intravenous methyl prednisolone for 3 days with which she showed dramatic improvement and was discharged on oral steroids. The child was readmitted after 2 years with complaints of bilateral progressive painless loss of vision for 15 days, headache and ittitability for 10 days. MRI brain revealed T2 hyperintensities inright middle cerebellar peduncle, posterior limb of right internal capsuleextending upto right corona radiata and right parietal grey white matter junction suggestive of relapse of ADEM (figure 2). USG and MRI orbit were normal. EEG was suggestive of abnormal record so child was put on antiepileptics. The child was given intravenous methyl prednisolone (30mg/ssskg/day) for three days and then started on oralprednisolone (1mg/kg/day) for two weeks. The child showed dramatic improvement to this therapy and led to resolution of all her neurological deficits. Follow up for 2 months revealed no neurological deterioration and no appearence of new lesions on follow up MRI.

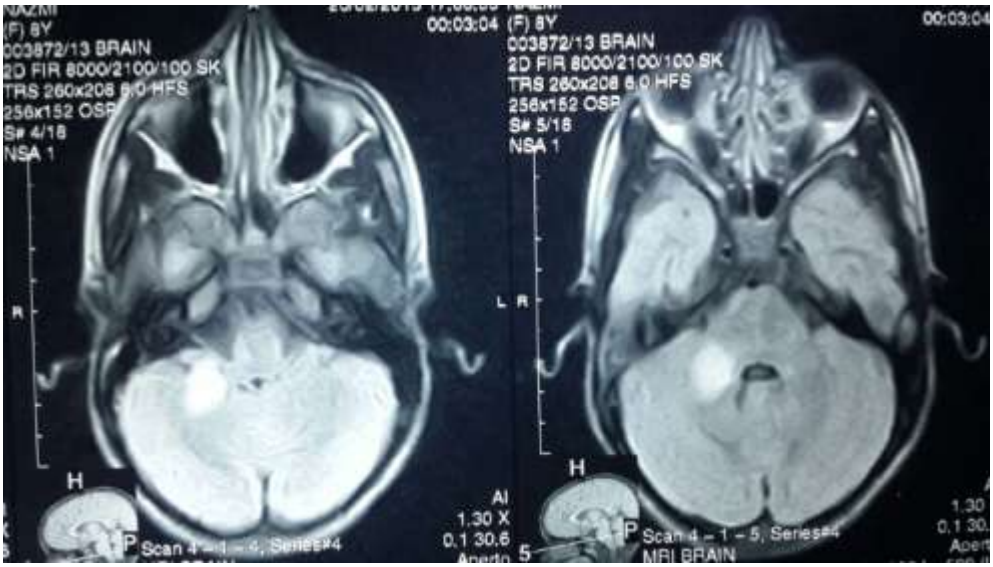

Figure 2: MRI brain showing revealed T2 hyperintensities in right middle cerebellar peduncle, posterior limb of right internal capsule extending upto right corona radiata and right parietal grey white matter

DISCUSSION

ADEM has a monophasic course but relapses can occur in between 5% and 25% of cases (Marchioni *et al.*, 2005; Tenembaum *et al.*, 2002) and have been described mainly as case reports (Rust, 2000; Suwa 1999). The relapsing forms can be subdivided into two categories: "recurrent" (80%), if the disease recurs at least 2 months after its onset and the lesions affect the same district that was involved in the first episode and "multiphasic" if the lesions present a dissemination in space and time (20%) (Enrico *et al.*, 2008; Revel *et al.*, 2000).

The clinical syndrome is variable but can range from focal neurological deficits to coma and death. Typically, ADEM is characterized by an abrupt onset of fever, obtundation, seizures, and focal neurological signs. Obtundation may progress to coma and up to 20% of patients die.

The diagnosis is often confirmed by MRI, which shows multifocal white matter lesions corresponding to the abnormal neurological findings (Bennetto and Scolding, 2004). Diagnosis in this case was made on the basis of clinical picture and MRI features. The presented case qualifies for a relapse as new clinical episode occurred at an interval of more than 2 month, with some different symptoms, and radiologic evidence of new lesions at new site.

Differential diagnosis of Multiple Sclerosis (MS) was also kept but age of presentation, bilateral optic neuritis, polysymptomatic presentation, presence of encephalopathy with loss of consciousness, only one relapse observed, and MRI findings of hyperintense lesions, grey matter involvement and appearence of no new lesions on follow up pointed more towards MDEM than MS (Bennetto, 2004).

The main therapeutic options for ADEM are corticosteroids, plasma exchange, and intravenous immunoglobulins. Corticosteroid is the most frequently reported therapy, improving recovery in most patients (Alexander and Murthy 2011). Clinical response is usually evident within hours of initiation of treatment, particularly after pulsed IV corticosteroids since they reduce inflammation, edema, and seal the blood-brain barrier, which further decrease the influx of active immune cells and humoral factors, contributing to demyelination (Straub *et al.,* 1997) Plasma exchange and intravenous immunoglobulins were not used in our patient because the patient showed dramatic improvement with corticosteroids.. The long-term prognosis of this entity depends on the etiology, with postmeasles patients having a higher mortality and significant morbidity and in nonmeasles cases ifull recovery occurs in 50%–75% of patients, in a period of 1–6 months after the illness (Hynson *et al.,* 2001; Dale *et al.,* 2000). The most common sequelae are focal motor deficits, and could range from mild ataxia to hemiparesis. A hyperacute onset, severe neurologic deficits as a result of aggressive disease, and unresponsiveness to steroids are poor prognostic indicators (Alexander and Murthy 2011).

We think our patient represents a case of multiphasic acute demyelinating encepahalomyelitis treated effectively with corticosteroids with no neurological sequale

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