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Case Report

Acute/Multiphasic Disseminated Encephalomyelitis - A Case Report

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is an immunologically mediated inflammatory disease of the central nervous system resulting in multifocal demyelinating of the brain and spinal cord. With an annual incidence of 0.4 -0.6 per 100,000.ADEM represents diagnostic challenge for clinicians, as many disorders have a similar clinical and radiologic presentation.

Key words: Acute disseminated encephalomyelitis, MRI, multiphasic disseminated / postinfectious encephalomyelitis, Methyl prednisolone

INTRODUCTION

Acute disseminated encephalo-myelitis (ADEM), a monophasic disease process known as postinfectious encephalomyelitis, is an inflammatory demyelinating central nervous system (CNS) disorder that usually follows infection or more rarely vaccination.

The typical presentation is that of multifocal neurologic disturbance accompanied by change in mental status. [1,2] Multiphasic disseminated encephalomyelitis (MDEM) is a multiphasic /relapsing variant of ADEM.

We hereby report a case of ADEM/MDEM in a 5 year old girl.

CASE REPORT

A 5 year old girl was brought with history of high grade intermittent fever for 20 days. On 20th day of illness child had one episode of GTCS type convulsion, for about 30 minutes and followed by post ictal drowsiness for about 2 hours. Child had an episode of upper respiratory tract infection about 25 days prior. No significant birth or family history. At admission child was conscious with stable vitals. On CNS examination Higher Mental functions were normal with no cranial nerve deficit and no signs of meningeal irritation were present. Fundus study was normal. Power was 3/5 in both lower limbs, with bilateral extensor plantars. Haemogram and x-ray chest were normal.CSF analysis showed neutrophil predominant pleocytosis, (130-(25% L, 75% neutrophils)) CSF protein, sugar and chloride were within normal limits (Protein:44, Sugar:49, CL:118) CSF Gram stain , AFB and culture was negative. CT brain showed reversible postictal cerebral edema and signs suggestive of meningo – encephalities. In the ward she had fever spikes with altered consciousness, visual

hallucinations, inability to walk and was not able to recognise even parents for about 24 hours. Repeat LP showed lymphocyte predominant pleocytosis (Cell count:42(N:25 L:75)

MRI brain (Fig.1) on 4th day of admission showed near symmetric T2 & FLAIR hyperintensities in B/L basal ganglia and thalami. Also ill-defined asymmetric T2 and Flair hyperintensities in both cerebral hemispheres, which were features suggestive of Acute disseminated encephalomyelitis (ADEM) and child was started on high dose methyl prednisolone and gradually tapered.

Child was doing well for5 weeks after stopping steroids when had an episode of convulsion, lasting for about 2 minutes, GTCS type. Repeat MRI brain showed disappearance of all the previous lesions and appearance of ill defined T2 & FLAIR hyperintensities in right cerebellar hemisphere involving both grey and white matter. (Fig.2)

Patient was started on high dose steroid therapy and tapered.

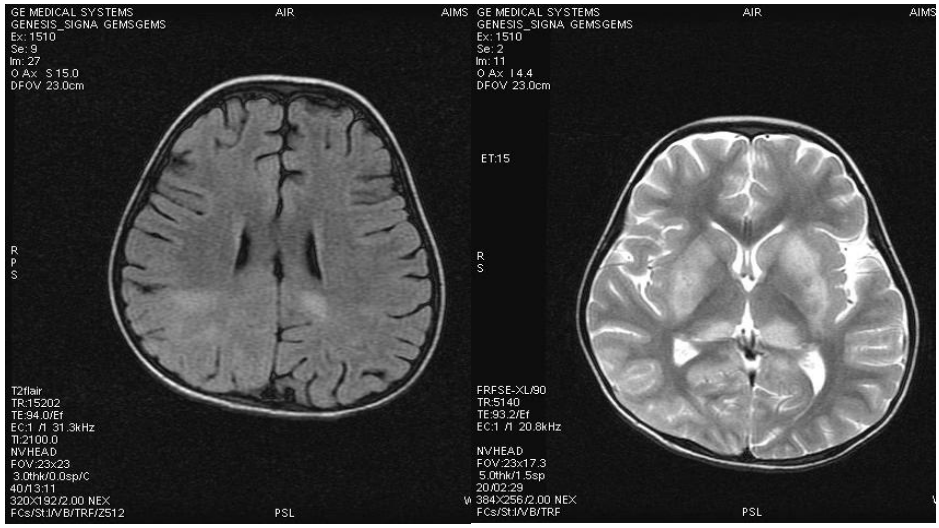

Fig1. Showing hyperintensities in B/L basal ganglia and thalami.Also illdefined asymmetric T2 and Flair hyperintensities in both cerebral hemispheres.

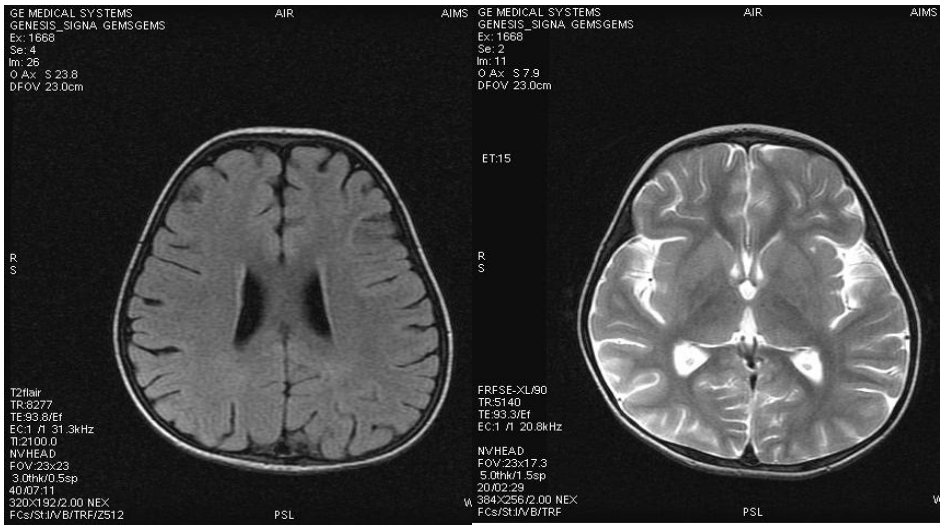

Fig.2 The previous lesions of the first scan had disappeared and appearance of ill defined T2 & FLAIR hyperintensities in right cerebellar hemisphere.

DISCUSSION

ADEM is an initial inflammatory, demyelinating event with multifocal neurological deficits , typically accompanied by encephalopathy .

Encephalopathy has been emphasized as a key distinguishing characteristic of ADEM in children.‘‘Encephalopathy’’ as currently applied is not precisely defined and may be confounded by postictal state or focal demyelinating lesions that may cause aphasia, frontal behavioral syndromes, or parietal-occipital visual spatial syndromes, which may be confused with a diffuse encephalopathy. [3,4]

ADEM is more frequent in pediatric age group. In one study of children with ADEM living in San Diego County,California, the incidence was estimated to be at least 0.4/100,000/y. [5] Another study from Fukuoka Prefecture, Japan reported the annual incidence to be 0.64 per 100,000. [6]

ADEM is frequently preceded by an infection or recent vaccination.Symptoms can follow almost 1-30 days later.Viruses that have been implicated in promoting the immune response responsible for ADEM include HSV, HIV,HHV-6, mumps, measles, rubella, varicella, influenza, enterovirus, hepatitis A, coxsackie, EBV, and cytomegalovirus. [7]

The most common preceding trigger is a nonspecific upper respiratory tract infection, which also is present in our case.
In ADEM children relapse seen within first six months of initial event is consider as MDEM.

MDEM is the recurrent ADEM in some cases where the premature cessationor tapering of therapy may lead to symptom recurrence.Hence, the monophasic nature of ADEM is defined as a lackof recurrence (within 3 months) in the absence of treatmentor while on appropriate treatment; and relapse that occursduring cessation or tapering of treatment should beconsidered as belonging to one monophasic episode. [8]

Another differential diagnosis considered is of Multiple Sclerosis, in which the child is usually an adolescent and relapse timing is more than 6 months.

Pathophysiology proposed for autoreactive immune responses to CNS white matter after an infectious trigger [5] is that it may trigger the subsequent autoimmune attack on the CNS, possibly via ‘‘molecular mimicry’’ i.e. similarities between viral epitopes and host myelin antigens such as myelin basic protein, myelin oligodendrocyte glycoprotein and proteolipid protein and the Inflammatory cascade theory. [9]

In a study by Leake et al majority of patients had exhibited multiple abnormalities including lethargy, ataxia, inability to walk, slow, slurred speech, cranial neuropathies and abnormal reflexes. Two-thirds of patients had altered mental status, including agitation,delirium, nonresponsiveness, moaning or inability to verbalize per baseline and/or inability to recognize parents or siblings.

In ADEM patients CSF may be normal in upto 61.5% or reveal a lymphocytic pleocytosis (usually between 50 and 180 cells/mm2) and elevated protein (commonly 0.5–1.0 g/dL)can be seen. [10] This child had leucocytosis of 130 cells/mm^3 with normal CSF proteins.

MRI is regarded as the imaging modality of choice in diagnosing ADEM. Which typically demonstrates widespread, bilateral, asymmetric patchy areas of increased signal intensity on T2-weighted imaging withindeep cerebral hemispheric and subcortical white matter as well as lesions in the basal ganglia, greywhite junction, cerebellum, and spinal cord. [11] Similar MRI findings have been observed in our child.

On follow up scan the initial lesions disappeared and new cerebellar lesions appeared, which were considered as MDEM, part of the monophasic episode. Disappearance of initial lesions,age group, relapse period less than 6 months from initial event, absence of black holes and no involvement of periventricular area on MRI differentiates from MS, which is more common in adoloscents, and T1 hypointensities occurring during clinical remission.

Treatment: Most commonly children respond to high dose of steroids, commonly used is methyl prednisolone, Response rate is good as also evidenced in this patient. Other modalities of treatment available for steroid resistant cases are IV immunoglobulins and most recently plasmapheresis which has been proved effective.

CONCLUSION

ADEM needs to be differentiated from Multiple Sclerosis with regular follow up imaging and should be treated adequately with steroids.

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