Acute Disseminated Encephalomyelitis  

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Abstract: Acute disseminated encephalomyelitis (ADEM) is acute multifocal white matter demyelination of central nervous system due to autoimmune response following viral infection or vaccination. We bring to your attention the case study of 11 year old boy reported with weakness of limbs and depressed level of consciousness. On examination there was increased muscle tone, decreased muscle power, exaggerated tendon reflexes and up going plantars. Magnetic resonance imaging (MRI) scan revealed asymmetrical multifocal hyper intense lesions with demyelination suggestive of ADEM.

Keywords: Acute disseminated encephalomyelitis, Demyelination, Central nervous system, Autoimmune, Cerebro spinal fluid, MRI scan, Steroids.

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is a demyelinating central nervous system disorder. It usually follows vaccination or infections like rubella, influenza, parainfluenza, mumps, infectious mononucleosis virus and with mycoplasma [1, 2]. Pathogenesis of ADEM is thought to result from an autoimmune response towards myelin via molecular mimicry [3]. Clinical features include signs of an acute meningoencephalopathy with meningsis such as fever, headache, nausea, vomitings, depressed level of consciousness, focal or generalized seizures & psychosis and multifocal neurological disturbances such as bilateral optic neuritis, visual field defects, aphasia, sensory & motor deficits, ataxia, and movement disorders [3, 4].

CASE REPORT

An 11 year old boy was brought to hospital by his father with chief complaints of weakness in both upper and lower limbs with depressed level of consciousness since three days, and two episodes of vomiting that subsided on medication. Over few days of admission drowsiness and weakness of limbs progressively worsened. There was no history of seizures, head injury, travel, or recent vaccination. Past history revealed an upper respiratory tract infection two weeks prior to admission.

Family history was unremarkable and there was no history of consanguineous marriage. Birth history consisted of delivery at full term normal vaginal delivery at tertiary care hospital without any post natal complications. Nutritional history, developmental history, anthropometry and his vitals were within normal limits. On physical examination he was afebrile, decreased alertness and sleepy. Motor system examination revealed normal muscle bulk, increased tone, decreased muscle power more in lower limbs (2/5) than upper limbs, and up going plantar in both limbs with unsteady gait. There was no sensory or autonomic system abnormality. There were no signs of meningeval irritation and increased intracranial tension.

Complete blood picture revealed hemoglobin of 13 gram/dl with normocytic normochromic RBCs, WBC counts of 7000 cells/mm3, and platelet counts of 60,000 cells/mm3. Other investigations complete urine examination, liver function test, renal function test, CRP, widal test, mantoux test and rapid malaria test were with in normal limits. Cerebrospinal fluid analysis was with in normal limits. MRI scan of brain showed asymmetrical multiple hyper intense lesions both in T2 weighed images and FLAIR images involving white matter of left temporal, parietal and occipital lobe, right temporal and parietal lobes of cerebral hemispheres (Fig. 1) and left cerebellum (Fig. 2). Findings suggestive of acute disseminated encephalomyelitis.

High dose of methyl prednisolone at 20 mg/kilogram per day was started. Patient showed marked clinical improvement after steroid therapy.
Patient was discharged after eight days of hospitalization with oral steroids and requested for follow up. Subsequent follow up revealed no neurological sequel or complication without any relapse of ADEM.

**DISCUSSION**

Acute disseminated encephalomyelitic (ADEM) is an uncommon inflammatory demyelinating disease of the central nervous system [5]. ADEM was first described in the early 18th century, typically following measles or chickenpox, and was associated with significant mortality and morbidity [6]. ADEM almost involves children with the incidence rate of 0.4/100000 yearly in patients less than 20 years-old. The peak incidence of ADEM is among 5 to 8 of age. Male to female ratio is the same [4].

It is thought to be triggered by an inflammatory response to either infection or vaccination. Various viral agents such influenza virus, enterovirus, measles, rubella, mumps, varicella, Epstein-barr virus, cytomegalovirus, hepatitis a virus and coxackie virus have been associated with ADEM. Bacterial triggers include Mycoplasma pneumonia, borrelia, and leptospira [7]. Pathogenesis of ADEM is thought to result from a transient autoimmune response towards myelin or other self-antigens via molecular mimicry, or by non-specific activation of auto reactive T-cell clones [3]. The sensitized T-lymphocytes by the peptides attaches to vascular endothelial cell in the help of adherence factor through blood circulation. Permeability of blood-brain barrier increases because of released inflammatory factors [1]. The hallmark of the pathological findings of ADEM is areas of inflammation predominantly in the Virchow Robin spaces, perivenous demyelination and infiltration of lymphocytes and macrophages. Other changes include hyperaemia, endothelial swelling, and vessel wall invasion by inflammatory cells, perivascular oedema, and haemorrhage [5, 8].

The hallmark of clinical features of ADEM is the development of a focal or multifocal neurological disorder. The onset of the central nervous system disorder is rapid with peak dysfunction in several days. Initial clinical features include encephalopathy ranging from lethargy to coma, and focal and multifocal neurological signs like hemiparesis, cranial nerve palsies, and paraparesis. Other commonly reported findings include meningismus, ataxia, and varied movement disorders [8].

Cerebrospinal fluid may be normal but frequently it shows some changes. Typical cerebrospinal fluid changes include increased pressure, lymphocytic pleocytosis (as much as 1000/ mm3, sometimes polymorphonuclearleucocytes initially), and raised protein (usually <1.0 mg/l).[8] Magnetic Resonance Imaging (MRI) is the diagnostic modality of choice [3]. Characteristic lesions seen on MRI appear as patchy areas of increased signal intensity on conventional T2-weighted images and on fluid attenuated inversion recovery sequence (FLAIR) they tend to be bilateral, but can also be asymmetric and are typically poorly marginated. Multiple lesions in the deep and subcortical white matter are common, which is characteristic of demyelination (gray matter lesions sometimes accompany white matter lesions, especially among children). While the number varies, multiple brain lesions are usually present. ADEM lesions are typically large (though smaller ones have also been seen) with diameters ranging from <5 mm to 5 cm. Additionally, brainstem and spinal cord abnormalities on MRI are common in ADEM [8]. Differential diagnosis includes early onset multiple sclerosis (MS).
Young age of a child, fever at the onset of disease, preceding illness, relapse occurring shortly after steroid discontinuation favors the diagnosis of ADEM [6].

High dose intravenous corticosteroids are approved as the first line of treatment, receiving intravenous immunoglobulin (IVIG); Plasmapheresis is also a useful remedy [4]. The lesions usually show a rapid response to steroid therapy, and a prolonged treatment with steroids (six weeks or more) is recommended in order to prevent recurrence of ADEM [7]. Prognosis of ADEM in the pediatric population is favorable, with overall good outcome. However, residual neurological deficit has been reported in approximately 20% of cases, and relapses up to 25% of cases after steroid taper has been seen [6].

CONCLUSION
Acute disseminated encephalomyelitis is uncommon inflammatory demyelinating lesion of central nervous system triggered by either infection or vaccination due to transient autoimmune response towards myelin. Clinically presents as acute meningoencephalitis with multifocal neurological involvement that shows good response to steroids. MRI scan shows asymmetrical, multiple lesions with characteristic demyelination of central nervous system.

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