CASE REPORT

Case of Acute Disseminated Encephalomyelitis

A. K. Simalti
Classified Specialist and Paediatric Intensivist, Department Of Paediatrics, Base Hospital, Delhi Cant

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ABSTRACT
Acute disseminated encephalomyelitis (ADEM) is characterized by Immune mediated inflammation of brain causing and gray matter. ADEM is an which usually follows a viral or bacterial infection, immunizations, and drug or serum administration. One such case is being reported here where a 3 year old girl was brought in status epilepticus as presenting feature of ADEM.

A. K. Simalti

INTRODUCTION

Immune mediated inflammation of brain causing demylination of white matter, gray matter is characteristic feature of acute disseminated encephalomyelitis (ADEM).[1] Typical setting is of abrupt development of irritability and neurological signs in children recovering from a viral prodrome. Neurological abnormalities include varying degrees of mental state changes ranging from drowsiness to coma.[2] We report a case of ADEM treated effectively with corticosteroid therapy

CASE REPORT

3 yr old girl was admitted with status epilepticus. She was apparently asymptomatic till 1 day back when she developed fever and cough for which she was given paracetamol and antihistaminic as cough suppressant. Fever had subsided after antipyretic and at time of seizure she was not having fever. Child was managed as per protocol for status epilepticus with midazolam, phenytoin and phenobarbitone. Seizures were controlled after giving phenobarbitone but child remained unconscious. In view of history of fever and seizure, diagnosis of meningitis was considered and. After initial stabilization and fundoscopy to rule out papiloedema,Cerebro spinal fluid (CSF) was collected which showed normal glucose, very high protein (more then reference range) and 2 cells both lymphocytes. This picture and absence of fever after the initial episode led us to think of other aetiologies. CT scan was ordered but findings were hypodensity bilateral thalamus and compression of cistern suggesting encephalitis with cerebral oedema. However as child remained comatose and afebrile, evaluation for metabolic encephalopathy in form of blood gas, ammonia, urea and sugar were done and found within normal limit. Diagnosis related to immune mediated encephalitis was considered in view of coma, high CSF protein, absence of fever and one day prior brief episode of febrile illness. MRI was ordered and it showed multifocal areas of altered intensity in white matter suggesting the diagnosis of ADEM.

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Corresponding Author:
Dr. A. K. Simalti, M.D., Classified Specialist and Paediatric Intensivist, Department of Paediatrics ,Base Hospital, Delhi, Cant. Mobile: 8475000296.
E-mail: ashishsimalti@rediffmail.com

Child was given IV methyl prednisolone at dose of 30 mg/kg for 5 days. Within 2 hrs of first dose there was noticeable improvement and child recovered significantly by 5th day. Irritability and lack of speech persisted along with episodes of excessive crying. Oral prednisolone at dose of 2mg/kg was continued and child showed gradual but sustained improvement over next 2 weeks.

DISCUSSION

ADEM is an autoimmune disorder which usually follows a viral illness.However it has also been reported after bacterial infection, immunizations, and drug and serum administration.[3] This illness has a seasonal trend with peaks in winter probably reflecting the prevalence of viral illnesses.[4] It is reported to be more common among boys before puberty.[5] Presentation ranges from lethargy to coma, seizures, suggesting generalized encephalopathy and localising signs reflecting cerebral (hemiparesis), brain stem (cranial nerve palsies), and spinal cord (paraparesis) involvement. Besides cerebellar ataxia may also be a presenting feature.[6]

Despite generalized involvement of brain, cerebrospinal fluid(CSF) is usually unaffected. Well-documented cases exist with totally normal CSF pressure, cell counts, and protein content. However CSF can also show a picture similar to purulent meningitis with high polymorphonuclear cell count and elevated protein. This is considered to represent a more necrotizing disease process. However if ongoing infective etiology is ruled out these CSF abnormality do not require specific intervention as they are self limiting. The levels of protein in the CSF may be increased, as it can be in many conditions in which myelin destruction occurs like cerebral infarction.[7] The albuminocytological dissociation shown in our case is not a typical feature of ADEM. Albuminocytological dissociation is considered a classical diagnostic feature of gullian barre syndrome.[8] Albuminocytological dissociation has also been reported with spinal cord tumors, intracranial tumor, Refsum's Syndrome and Osteosclerotic myeloma. asymmatric,bilateral areas of hyperintensities in white matter with ill defined margins is most common finding in MRI.demylination of white matter is the charecterstic involvement. The periventricular white matter is often spared. Gray matter lesions sometimes accompany the white matter abnormalities, especially in children.[6] The differential diagnoses of ADEM include other demylinating disorder like multiple sclerosis presenting for the first time although it is much more common among adults. Other relatively uncommon aetiolgies include macrophage activation syndrome and vasculitis of the CNS[9] Although ADEM is a monophasic disease, relapses have been described mainly as case reports. There is no standard treatment for ADEM though corticosteroid is the most frequently reported therapy; improving recovery in most patients Clinical response is usually evident within hours of initiation of treatment, particularly after pulsed IV corticosteroids. Corticosteroids have anti inflammatory action which also decrease the. blood-brain barrier permeability thus reducing further influx of active immune cells and humoral factors, contributing to demyelination. Besides corticosteroids other effective means to reduce inflammation include plasma exchange, and intravenous immunoglobulin.[10] Plasma exchange and intravenous immunoglobulins were not used in our patient because the patient showed dramatic improvement with corticosteroids.

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