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Acute Disseminated Encephalomyelitis (ADEM) in Children: A Case Report

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Abstract

Acute Disseminated Encephalomyelitis (ADEM) is an inflammatory disease that causes demyelination and affects the white matter of the brain and spinal cord through an immune response. The patient, an eight-year-old Iranian Kurdish boy with a height of 125 cm and a weight of 24 kg, complained from fever for three days which didn't respond to acetaminophen medication. After three days, the symptoms developed into headache, vomiting, and decreased consciousness. The patient was admitted to the emergency department of Imam Hasan hospital in Bojnourd, Iran, in July 2019. LP diagnostic testing, Wright Agglutination test, Brain CT, and MRI examination were performed. Finally, a diagnosis of acute disseminated encephalomyelitis (ADEM) was confirmed. Following the ADEM diagnosis, methylprednisolone 500 mg was administered immediately and continued for five days. This case study suggests that MRI is the most effective method of diagnosis for ADEM, and high-dose methylprednisolone is the treatment of choice for this syndrome.

***Key Words*:** Acute Disseminated, Children, Encephalomyelitis, Methylprednisolone, MRI.

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1- INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is an inflammatory disease that causes demyelination and affects the white matter of the brain and spinal cord through an immune response. This syndrome has physiological and pathophysiological similarities to acquired demyelinating syndrome (ADS) of childhood syndrome and multiple sclerosis (MS) (1, 2). The incidence of this syndrome is about 0.07 to 0.4 per 100,000 people per year. However, the actual incidence of the disease in India and other countries is unclear due to lack of reported cases (3). Although ADEM occurs at all ages, most cases have been reported in children under nine and the hospital admissions have increased over the past decade (4, 5). Also, the results of different studies show that the prevalence of this disease is higher in males (6, 7). The disease has the highest occurrence in spring and winter, following the peak of viral infections (8). Clinical symptoms include fever, headache, nausea and vomiting, confusion, visual impairment, drowsiness, seizure, and coma, which often occur before neurological symptoms and appear four to 21 days after the predisposing agent. Symptoms are usually mild at the beginning but rapidly increase (5, 9). No independent diagnostic test can definitively diagnose the disease. However, magnetic resonance imaging (MRI) can be the most effective method in the early diagnosis of ADEM (6, 11). In this report, a case of Acute Disseminated Encephalomyelitis syndrome in an 8-year-old child is described.

2- CASE REPORTS

The patient was an 8-year-old Iranian Kurdish boy with a height of 125 cm and a weight of 24 kg, presenting with fever for three days which didn't respond to acetaminophen medication. After three days, symptoms developed into headache, vomiting, and decreased consciousness. The patient was then admitted to the emergency department of Imam Hasan hospital in Bojnourd, Iran, in July 2019. In his medical history, there were no significant cases of drug use and history of specific diseases. Vital signs at arrival were as follows: HR: 60, RR: 14, T: 39, BP: 106/70, and SPO2: 96%. Hematology results were as follows: ESR: 54.0 mm/h, WBC: 18.9×10^3/µL, RBC: 5.01×10^6/µL, hemoglobin: 13.9 g/dL, neutrophil: 81%, lymphocytes: 10%, and monocyte: 9%. Biochemistry results showed BS: 125 mg/dL, urea: 20 mg/dL, creatinine: 0.9 mg/dL, sodium: 137.4 mmol/L, and potassium: 4.96 mmol/L. After being visited by the emergency physician, the patient was treated with acetaminophen for hyperpyrexia but the fever didn't improve. Due to the patient's temperature not falling and the fever being accompanied by other symptoms, including severe pain in the head and neck, a consultation with pediatric infectious diseases specialist was requested. After the pediatrician's visit, LP diagnostic testing was performed promptly with the consent of the child's parents to test for signs of meningitis. Based on the results of cerebrospinal fluid analysis (**Table.1**), meningitis was ruled out.

Also, 2-mercaptoethanol and Wright Agglutination tests were performed to diagnose brucellosis, and the results were negative (Brucella Immunoglobulin G: 0.5 RU/ml, Brucella Immunoglobulin M: 0.6 RU/ml). Antibiotic treatment was initiated with ceftriaxone combined with vancomycin and SER-D/W 5%. Five days after starting the antibiotic treatment, the patient's condition improved, vomiting stopped, and headache and dizziness subsided. The patient then began a soft food diet. On the sixth day, despite continued treatment with antibiotics, the patient began vomiting again, and had fever three times in a day. On the seventh day of hospitalization, the patient's consciousness decreased and symptoms of mild nystagmus appeared. Following these symptoms, an emergency computerized tomography (CT) of the brain was

performed. The results showed no abnormality in ventricles, white matter, and cerebral gray matter. No space-occupying lesion and pathologic sign were observed on the collection and abscess. After the above examinations, a consultation request was sent to the neurology department. In the neurologic examination, no papillary edema was observed in the optic nerve, there were no signs of reflex abnormalities, the Babinski sign was negative, and the function of the cranial nerve pairs II, III, IV, V, and VI was normal. However, cerebellar tests showed some degree of balance disorders.

An MRI was then requested without emergency contrast. In the MRI examinations of the brain with standard sequences, signal changes were observed in the white matter (WM) on both sides, centrum semiovale (CSO), and periventricular (PV), internal capsule on both sides, posterior arm, brainstem, and cerebellar peduncles. No evidence of space-occupying lesion (SOL), hydrocephalus, and extracorporeal collection was observed. The 7-8 nerve complex was observed with symmetric signal size without SOL. Corpus callosum, pituitary gland, and thalamus were also found with signal and symmetric size without SOL. Opacity of the right and left ethmoid sinus and right sphenoid sinus were observed. Following the results, brain MRI was requested and after consulting with a radiologist for the interpretation of the results, the diagnosis of ADEM was confirmed (**Figure.1**). Following the diagnosis, pulse corticosteroid therapy was started immediately with methylprednisolone 500 mg and continued for five days. After five days, the patient was fully awake and the headache and Nystagmus were resolved. The patient was able to sit without help. Eventually, after the complete resolution of symptoms, the patient was able to walk and was discharged from the hospital.

Table-1*:* Findings from the cerebrospinal fluid analysis.

Test	Result	Unit	Normal Ranges
Hematology			
Appearance (Before Centrifuge)	Clear		Bacterial: Turbid Viral: Clear
Color (Before Centrifuge)	Colorless		
Appearance (After Centrifuge)	Clear		Bacterial: Turbid Viral: Clear
Color (After Centrifuge)	Colorless		
WBC (White blood cells) count	60	Cells/ mm^3	Bacterial meningitis higher than 1,000 per mm^3 Viral meningitis less than 100 WBCs per mm^3
RBC (Red blood cells) count	50	Cells/ mm^3	
Neutrophil	40	%	
Lymphocytes	60	%	
C-reactive protein	Negative		
Protein	67	mg/dL	Bacterial meningitis: 21 to 2220 (0.21 to 22.2) Multiple sclerosis: 13 to 133 (0.13 to 1.33)
Glucose	82	mg/dL	300> Normal(12)
Serology			
CSF PCR	Negative		
Bacteriology (CSF PCR)			
Neisseria Meningitidis	Negative		
Haemophilus influenzae	Negative		
Streptococcus Pneumoniae	Negative		
Entero Virus	Negative		
Mycobacterium Tuberculosis complex	Negative		
Herpes Simplex Virus	Negative		

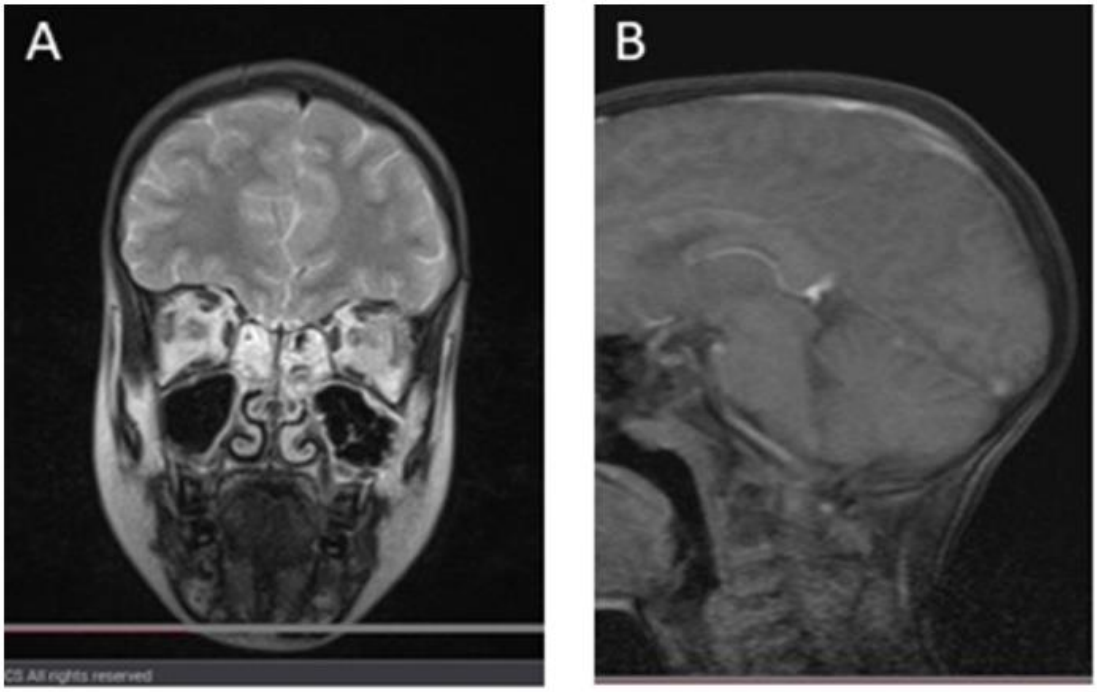

Fig.1: A. Coronal plane, **B**. Sagittal plane.

3- DISCUSSION

ADEM is a rare disease that triggers demyelination of the central nervous system (CNS) as a result of the immune response and mainly affects the white matter of the brain and spinal cord (3). ADEM is usually a childhood disease that often results from viral and bacterial infections or vaccinations (13). Studies have shown that neurological symptoms worsen after an average of 4.5 days (14). Brain MRI is the most effective way to diagnose the syndrome. The following observations in MRI may be evidence of the ADEM syndrome:

1. Multifocal lesions in white matter with or without involvement of basal ganglia.

2. Observation of single or multiple focal lesions, only in gray matter.

3. Localized lesions in the brainstem of the basal ganglia or cerebellum (15).

In most cases, evaluation of cerebrospinal fluid (CSF) is not helpful but should be considered before excluding other diseases (11). Studies have shown that people with ADEM may be more likely to develop Multiple Sclerosis (MS) when oligoclonal bands become positive, myelin oligodendrocyte glycoprotein antibody (MOG-Ab) be negative, and in case of abnormal MRI findings (7). It has also been suggested that repeated nerve imaging, even in relatively simple cases of ADEM with a duration of three to six months, can also be useful to evaluate the risk of multiple sclerosis or other immunological neurological disorders in these patients in the future (6). High-dose methylprednisolone (mPSL), one of the drugs that can improve inflammation of the central nervous system, is the main treatment option for ADEM (14). In a study by Rastogi et al., it was reported that patients' clinical symptoms improved after receiving methylprednisolone for four days (11). However, in cases of refractory or recurrent ADEM, other treatment options, including intravenous immunoglobulin (IVIG) and plasmapheresis, are also recommended (11, 14).

4- CONCLUSION

Our case study suggests that MRI is the most effective way to diagnose this syndrome and high-dose methylprednisolone (mPSL) is the main treatment option for ADEM.

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6- CONFLICT OF INTEREST: None.

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