

# CASE REPORT

## ACUTE DISSEMINATED ENCEPHALOMYELITIS IN A 5-MONTH OLD INFANT

**N. Khosroshahi MD<sup>1</sup>,**  
**F. Mahvelati MD<sup>2</sup>,**  
**K. Kamrani MD<sup>3</sup>**

### Abstract

#### Objective

This article reports on an 5-month-old infant who was admitted to our hospital for prolonged fever, focal status epilepticus and encephalopathy.

Brain magnetic resonance imaging (MRI) demonstrated a single hyperintense lesion in right putamen in T2-weighted images.

A diagnosis of acute disseminated encephalomyelitis was made based on the clinical features, treatment response and clinical course. An executive literature review revealed a few cases under the age of one year. This patient is the youngest one who is reported from Iran.

**Keywords:** Acute disseminated encephalomyelitis, infant.

### Introduction

ADEM is the most common demyelinating disorder of childhood(1). It is a monophasic, polysymptomatic disorder involving the central nervous system white matter; this immune mediated inflammatory process commonly follows a viral infection or may occur after vaccination (2). The age group most commonly affected is between 1-20 years (2).

Reported here is a case of ADEM presenting with fever, partial status epilepticus and hyperintense lesions in right putamen revealed on MRI. The condition rarely occurs in infancy and hence this case is being reported for its unusual age, and indicates the likelihood of its occurrence at that early age. Despite its occurrence being rare in infants, this case emphasizes the need to consider a diagnosis of ADEM, even in infancy.

### Case report

A 5-month-old boy presented with two weeks history of intermittent fever. The patient was the first child of unrelated healthy parents. He was born at term by normal delivery after an uncomplicated pregnancy. Developmental milestones were acquired in an age-appropriate manner. Neither was there any family history of any neurologic disorder. He had had recent vaccinations.

On admission, he was unconscious, his body temperature was 38.7C and heart rate and respiratory rate were normal. Physical examination revealed bilateral sixth nerve palsy, and hyperactive deep tendon reflexes bilaterally in the lower extremities, but no abnormalities were seen in any other parts of the motor system. Blood examination including complete blood cell count, biochemistry electrolytes, coagulation system, complements, lactic acid, amino acids, blood gas analysis, and antinuclear and anti-

1. Assistant Professor of pediatrics, child neurologist, Tehran University of Medical Sciences

2. Assistant Professor of pediatric neurology, Shahid Beheshti Medical University

3. Assistant Professor of pediatrics, Tehran University of Medical Sciences

Corresponding Author:

N. Khosroshahi MD

Bahrami Hospital

Tel: +98 9121012861

Fax: +98 21 2220254

E-mail: nahidkhosroshahi@yahoo.com

DNA antibodies were unremarkable.

Cerebrospinal fluid analysis revealed no white blood cells, and protein and glucose concentrations of 20mg/dl and 49mg/dl respectively.

Electroencephalogram showed repeated burst of high voltage slow wave activity suggestive of focal slowing. Brain CT scan in the first day of admission was normal, but an MRI showed single hyperintense lesion in the right putamen in T2-weighted images, which was hypointense in T1-weighted imaging (Figure 1).

During the second day following admission he developed focal status epilepticus, consisting of internal

left eye deviation, neck and head rotation to the right and clonic movement in the left upper extremity. His seizure lasted six hours and was controlled with a high dose of phenobarbital and phenytoin.

He was discharged after two weeks, following control of his seizure and his being hypotonic. A repeat MRI of his brain, 3 months later, showed hyperintense lesion in the right putamen, the size of which was significantly reduced in comparison to the first imaging.

Follow up visits over the next 2 years, revealed normal development and neurological exam results. Brain MRI showed complete resolution of lesions.

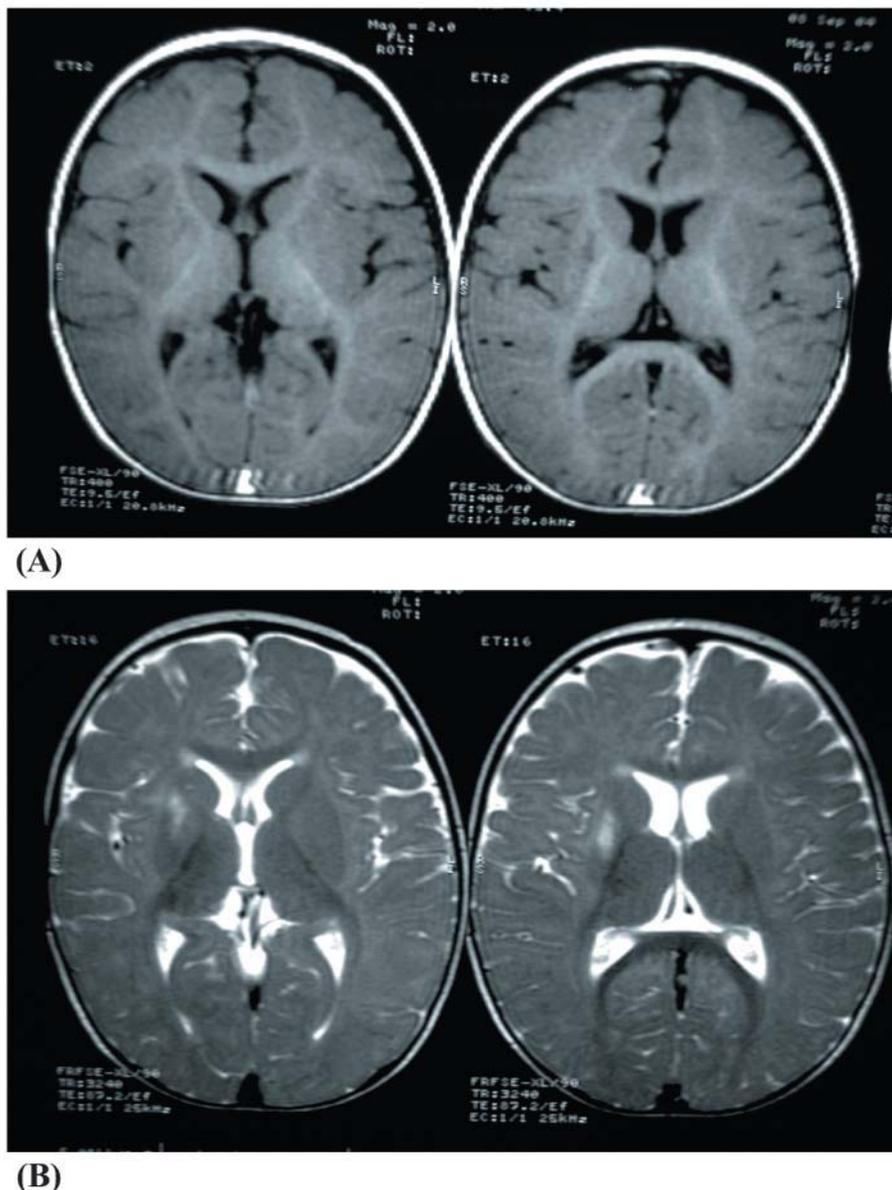


Figure 1. Five month-old infant with diagnosis of acute disseminated encephalomyelitis. A. T1 weighted imaging at onset of disease. B. T2 weighted imaging at onset of disease which shows single hyperintense lesion in right putamen.

**Discussion**

ADEM, a monophasic, polysymptomatic disorder involving the central system white matter, is an immune mediated inflammatory process that commonly follows viral infections or vaccination(2). ADEM accounts for up to one third of all known cases of encephalitis (2). As more and more magnetic resonance imaging studies are being performed (3), this potentially severe demyelinating disorder is likely to be increasingly diagnosed. The age group most commonly affected is between 1-20 years(2), with an onset during childhood ADEM reported to be usually around 3 or 4 years (4), or 5.3±3.9 years(5). The condition has rarely been known to occur before the age of one year(4). Mean ages documented in various reports differ, being 7.8 (6), 8(1) and 8.6 years(7).An extensive literature review revealed a few cases in infants, under the age of one year. The youngest patient, reported from Japan, was a 3-month old girl with symptoms of somnolence, poor feeding and vomiting (4). Another case, more similar to ours, was a 6-month-old female infant, who presented with new onset focal seizures involving the right upper extremity, irritability and low-grade fever preceded two weeks earlier by obits media and rash(8). The last case was an 8-month-old infant who presented with fever since the past two weeks ago, and had not been taking its feeds since the last 4 days, the infant was lethargic, had convulsions and altered sensorial status since the last two days (9). Such patients underscore the need to consider ADEM in the differential diagnosis of polysymptomatic neurological symptoms even in infants, especially in patients with a history of a preceding febrile illness or vaccination; brain MRIs are definitely required to confirm the diagnosis.

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