



## A case of acute disseminated encephalomyelitis (ADEM)

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### Abstract

**Introduction:** Acute disseminated encephalomyelitis (ADEM) is an uncommon monophasic idiopathic inflammatory demyelinating disease. ADEM typically presents with non-specific symptoms including nausea, vomiting, fever, and headache. This then progresses to neurological deficits such as motor weakness, altered sensorium, and often significant morbidity or mortality. Typically, the infectious insult is thought to be in transient viral infection or vaccination.

**Case Report:** We describe a case of ADEM in a 15-year-old girl that came to emergency department with dizziness and general weakness; she was unable to speak, and lethargic, with no positive past history. Her laboratory tests and spiral brain computed tomography (CT) scan were normal. She was admitted to neurology ward, and treated with high-dose corticosteroids. Our patient responded well to corticosteroids.

**Conclusion:** In patients with neurologic deficits in emergency department, the physician must think about ADEM although it is rare.

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### Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated demyelinating disorder of central nervous system (CNS), characterized clinically by new-onset polyfocal neurologic symptoms including encephalomyelitis, coupled with neuroimaging evidence of multifocal demyelination. ADEM is classically considered a monophasic illness, with the highest incidence in early childhood. The first descriptions of an ADEM-like disorder with recognition of a temporal relationship to infections (especially smallpox and measles) goes back to the 18<sup>th</sup> century.<sup>1</sup>

ADEM is a rare disease with an estimated annual incidence rate of 8 per 1000000.<sup>2</sup> It presents as an autoimmune demyelinating disease of the CNS triggered by a recent infections process in most but not all cases.<sup>3</sup>

ADEM typically presents with brain edema, perivenous infiltration of lymphocytes, macrophages, or neutrophils, and perivenous swelling with demyelination.

Patients typically present with rapid deterioration of symptoms including altered mental status, motor deficits, ataxia, nystagmus, seizures, and headache.

It is proposed to occur due to myelin autoantigens (myelin basic protein, proteolipid protein, and myelin oligodendrocyte protein) sharing antigens with those of an inciting pathogen.<sup>4</sup> These infectious antibodies cross-react with myelin autoantigens, and result in diffuse encephalomyelitis.<sup>4,5</sup>

We report a 15-year-old girl student, admitted to the emergency department of the Imam Reza educational hospital, Tabriz, Iran, complaining of lethargy feeling, gate imbalance, headache, dizziness, and diplopia.

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The diagnosis was ADEM.

### Case Report

The case was a 15-year-old girl student came to our hospital with her family, and her chief complaint was dizziness and general weakness; she was unable to speak. Her mother said that 2 hours before they came to the hospital, she was fully healthy; then slowly she began to weakened, then became generally weak, and had no speech communications; so her family got worried, and transferred her to the hospital.

In emergency department, she was on bed, did not move, and had no speech communications. Her mother said that she had a headache with dizziness, and examinations told that she was febrile (38.3 °C).

Generally, she was week and lethargic, her Glasgow coma scale (GCS) was 14-15, and had gait imbalance; she had fine nystagmus of both eyes.

Her past medical, familial, and social history was negative.

Her plantar reflex was ablated, and other neurologic examinations were normal; she did not have any sensitivity to folly catheter placement, and her vital signs were stable.

First, as we suspected drug toxicity, we checked routine laboratory test including electrolytes. Test results were as white blood cell (WBC) of 16800 per microliter with 84.9% neutrophils, and blood sugar (BS) of 133 mg/dl; but the urine toxicology was negative.

In emergency department, spiral brain computed tomography (CT) scan was done, that was normal without pathologic findings. Then lumbar puncture was done, and the result was as red blood cell (RBC) of 1350 per microliter, WBC of low than 10 per microliter, glucose of 61 mg/dl, lactate dehydrogenase (LDH) of 19 mg/dl, and protein of 35 mg/dl. The tests for human immunodeficiency viruses (HIV) and human papillomavirus (HPV) were all negative.

She was lethargic yet, because that her mental status and nystagmus, and given the patient's findings, the differential diagnosis

at the time was toxoplasmosis, fungal, or bacterial meningitis or abscess, CNS lymphoma, Cryptococcus, or demyelinating disease. She was admitted to neurology ward.

In neurology ward, brain magnetic resonance imaging (MRI) was done, and that was normal. Electroencephalography (EEG) had done, and the conclusion was some paroxysm of disorganization without significant epileptiform discharge (Figures 1 and 2).



Figure 1. Cervical magnetic resonance imaging (MRI)

The neurologist, with high suspicions to demyelination disorders, started the treatment for the patient as 5-day regime of high-dose methylprednisolone; and on the 3<sup>rd</sup> day of her hospitalizations, the neurologic deficit of patients began to improve.



Figure 2. Brain magnetic resonance imaging (MRI)

Patient's diagnosis was ADEM, and she was discharged from the hospital after 5 days; until 6 months after her sickness, she did not have any CNS signs or symptoms.

### Discussion

ADEM is a poorly understood neurological illness with significant long-term consequence.

White et al. present a case of ADEM post appendicitis in a middle-aged woman, with a positive response to intravenous immune globulin (IVIG) therapy. Significant cognitive and physical impairment is common sequel of ADEM.<sup>6</sup>

The post-infection and post-vaccination encephalomyelitis may make up about three quarters of cases with ADEM.<sup>7</sup> Post-vaccination ADEM is associated with several vaccines such as rabies, diphtheria-tetanus-polio, smallpox, measles, and mumps; the relationship with hepatitis B vaccine remains unclear.<sup>8</sup>

Patients typically present with a rapid deterioration of symptoms including altered mental status, motor deficits, ataxia, nystagmus, seizures, and headache. It is proposed to occur due myelin autoantigens (myelin basic protein, proteolipid protein, and myelin oligodendrocyte protein) sharing antigens with those of an inciting pathogen. These infectious antibodies cross-react with myelin autoantigens, and result in diffuse encephalomyelitis.<sup>9</sup>

After several days or weeks, clinical signs and symptoms usually include altered consciousness and multifocal neurological disturbance.

Typically, initial therapy for ADEM is Corticosteroids; intravenous steroids are usually given over a course of 5-7 days,

followed by course of oral steroids. Following steroid treatment, most patients start to recover over several days, and within 6 months of an ADEM attack, the majority of them will have complete or near to complete recoveries. Some patients may not respond to steroid, which should be treated with IVIG. Plasmapheresis (plasma exchange) is another treatment that can be used to reduce immune activity; chemotherapy drugs may be used in only very severe cases of ADEM.<sup>10</sup>

### Conclusion

In patients who came to the emergency department with neurologic deficit, the physician must think about ADEM although it is rare; it is important to rapidly initiate the treatment to control it easily.

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### Authors' Contribution

SNA, case presentation; SS, case management and treatment; SP, critic and writing; MP, supervise and manage.

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### Conflict of Interest

Authors have no conflict of interest.

### Ethical Approval

To report this case, we got inform consent from the patient and her father.

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