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Acute disseminated encephalomyelitis: a case report of effective early immunotherapy

K Ritarwan1, O R Ramayani2 and P Eyanoer3*

1Department of Neurology, Faculty of Medicine, Universitas Sumatera Utara, Jalan Bunga Lau No 17, Kemenangan Tani, Medan Tuntungan, Kota Medan, Sumatera Utara-20136, Indonesia
2Department of Pediatrics, Faculty of Medicine, Universitas Sumatera Utara, Jalan Bunga Lau No 17, Kemenangan Tani, Medan Tuntungan, Kota Medan, Sumatera Utara-20136, Indonesia
3Department of Community and Preventive Medicine, Faculty of Medicine, Universitas Sumatera Utara, Jl. Dr. Mansyur No.5 Medan, Indonesia
*Corresponding author: putrice00@yahoo.com

Abstract. Acute disseminated encephalomyelitis (ADEM) is a monophasic acute non-vasculitic inflammatory demyelinating disorder of the central nervous system characterized by diffuse neurologic signs and symptoms coupled with evidence of multifocal lesions of demyelination on neuroimaging. Despite the long-standing recognition of ADEM as a specific entity, no consensus definition of ADEM had been reached until recently. Historically, different definitions of ADEM have been in published cases of pediatric and adult patients, which varied as to whether events required (1) monofocal or multifocal clinical features, (2) a change in mental status, and (3) a documentation of previous infection or immunization. The treatment has been given to the patient such as supportive therapy and high dose corticosteroids.

1. Introduction
Acute disseminated encephalomyelitis (ADEM) is a monophasic acute non-vasculitic inflammatory demyelinating disorder of the central nervous system characterized by diffuse neurologic signs and symptoms coupled with evidence of multifocal lesions of demyelination on neuroimaging.[1]

The incidence of ADEM is 0.4–0.8 per 100,000 and the disease more commonly affects children and young adults, probably related to the high frequency of exanthematous and other infections and vaccination in this age group. There seems to be no gender predominance.[2] The diagnosis of ADEM quite often enforced in tropical countries where the incidence of infection is high. Acute disseminated encephalomyelitis is considered a monophasic inflammatory demyelinating disease with various clinical manifestations usually consist of a number encephalopathy syndrome and focal or multifocal inflammatory demyelinating CNS showed abnormalities, including optic neuritis and myelitis.[3,4] Approximately 50% to 75% of all cases preceded by a viral or bacterial infection after infection non-specific upper respiratory tract. Acute disseminated encephalomyelitis may also occur after vaccination (post-immunization encephalomyelitis).[5,6]

*Acute Disseminated Encephalomyelitis (ADEM) ADEM is more commonly preceded by a viral infection, with measles, varicella, rubella, mumps, and influenza being the more frequently reported
infections. Despite the availability of vaccines in many countries, measles virus remains one of the most common global infectious causes of childhood mortality and neurologic morbidity.[7] Loss of consciousness is a common clinical symptom, followed by fever and headache. Other clinical features include the rapid onset of multifocal neurological disorders, such as seizures, visual field disturbances, ataxia.[3,4,5,8] The Magnetic Resonance Imaging examination is the modality of choice in diagnosing ADEM imaging publishing. There is no standard treatment for ADEM. Many treatment approaches have used some form of specific immunosuppressive therapy similar to that used for Multiple sclerosis and other autoimmune diseases, including high-dose steroids, intravenous immunoglobulin (IVIG) or plasmapheresis.[9] The perfect cure was in 50-70% of cases, between 70-90% of cases were cured there are still residual symptoms, the average time required for recovery is 1 to 6 months. The mortality rate of up to 5%. The lousy state with no response to corticosteroid therapy, usually indicating a neurological attack suddenly.[10]

2. Case
Male patient, 18 years old, presented with sudden loss of consciousness seven hours before admission. The patient was doing his activity at the time. The patient looked weak. History of seizures was found, where the whole body was jerking, stiff, with a duration of five minutes/x spasms, frequency over ten times. History of vomiting was encountered. He began oral antibiotics for presumed pneumonia with subsequent resolution of the respiratory symptoms. However, his fever persisted. History of trauma was not found. History of hypertension, diabetes and heart disease were not found. A history of illicit drug used was not clear; there is no knowledge of the family, history of blood transfusion was not found. Neurological consultation documented encephalopathy (manifesting as lethargy and irritability).

The MRI result suspected Acute Disseminated Encephalomyelitis (ADEM). Repeat CSF analysis showed an improved lymphocytic pleocytosis (10 cells/μL). The view Procalcitonin result was 200μg/μL. Differential diagnosis at the time included acute or sub-acute CNS infection, post-infectious cerebellar ataxia, and ADEM. From the results patients diagnosed with acute disseminated encephalomyelitis (ADEM) and pneumonia and sepsis. Although the clinical picture raised concern for ADEM, the distinct lack of white matter involvement in brain imaging was more consistent with a viral encephalomyelitis. Although immunomodulatory therapy was given the presence of periventricular, and juxtacortical white matter, the treating team elected conservative management because his fever had resolved and his encephalopathy and ataxia had improved by the morning after the imaging was completed.

In the management of a given antibiotic Meropenem as first-line therapy, it given corticosteroids was methylprednisolone and anticonvulsant. Phenytoin was given. The Prognosis of patients with ADEM is usually good, but there are sepsis and pneumonia in this patient general condition. After being given treatment in the Intensive Care Unit (ICU), increased patient awareness of somnolence become apathetic. The patient can breathe spontaneously and has done extubation. But two days later, the patient must be re-intubated due to respiratory distress in these patients, which caused an infection in the lungs enumerated.

Assuming an accompanying para-infectious inflammatory process most likely ADEM, corticosteroid treatment was initiated despite ongoing pneumonia and procalcitonin >1μg/l. Steroids were given on day 14 (CSF leukocytes <50cells/μl) with an initial 5-day pulse of 1g methylprednisolone. The latter extended to 10 days after clinical improvement. Following slow steroid tapering over the next four weeks, both neurological symptoms and MRI findings substantially ameliorated.

3. Discussion
Acute disseminated encephalomyelitis (ADEM) was an inflammatory disease of immunologically mediated by the central nervous system (CNS) that produces multifocal demyelinating lesions affecting gray matter and white of the brain and spinal cord.[11] Acute disseminated
encephalomyelitis characterized a monophasic disease that commonly associated with antigen challenge (febrile illness or vaccination), which is believed to function as a trigger for the inflammatory response underlying the disease. It is most seen in the pediatric population but can occur at any age.[12]

It was in the early 18th century as a regular picture of measles and smallpox. Little is known about the events around the world, but in the United States may be about 1.5-3/100,000. ADEM is more common in winter, and 80% of cases occur in children in the first decade of life. 1931 McAlpine describes three sets of patients with ADEM: 1) post-vaccination, 2) after infection fever and 3) spontaneous. Some recent reports of ADEM in children have confirmed the observations of McAlpine.[9,13,14] In the diagnosis was ADEM, neurological deficits occur 3-6 weeks after the initial event. Onset may develop over a few days. The prodromal illness may precede neurologic symptoms. Acute disseminated encephalomyelitis can be on any part of the neuraxis, and thus the clinical presentation is variable and usually poly-symptomatic: altered mental status, pyramidal dysfunction, cerebellar ataxia, brainstem syndrome, optic neuritis, myelitis, and myeloradiculopathy rare and extrapyramidal syndrome.[14,15] Seizures are not uncommon, may be focal or generalized. Encephalitis more common in children younger than three years. Acute disseminated encephalomyelitis can rarely present with intracranial space occupying lesions picture, with demyelinating lesions.[16] There is no evidence of infection by viruses, bacteria, fungi, or parasites have been recorded.[17] Although ADEM usually described as demyelination with relative preservation of axons, axons damage in the brain has been in some patients. Lesions mostly involving the white matter, but also can involve cortical structure and gray matter deep.[5,9] Acute disseminated encephalomyelitis usually affects the white matter. However, lesions in the cortical gray matter and basal ganglia have also been with monositoid cell infiltration and demyelination perivenous area. The undisturbed demyelinating axons and neuronal affected less. Hyperemia and edema of periventricular followed by fibrosis in the late stage of the disease are also seen.[18]

Figure 1. Acute disseminated encephalomyelitis with small lesions. (A) Axial T2-weighted MRI shows bilateral hyperintense lesions in the central bounded ugly, periventricular, and juxtacortical white matter, (B) also involves both the thalamus and internal capsule, a child 18-month-old.

The Brain Imaging role in diagnosing ADEM. Typical non-specific MRI criteria are widespread, multifocal, asymmetric and extensive white matter lesions (typical “periventricular sparing” and lesions of similar inflammatory age) and possible affection of grey matter, with partial contrast enhancement and mainly a restriction in diffusion-weighted sequences.[19] MR-spectroscopy is allowed to reveal a reduced N-acetyl-aspartate (NAA) peak as a hint to reversible brain tissue damage,
as shown in our case.[20] Second, these diagnostic criteria also hold true for para-infectious as well as post-vaccinal-ADEM. Infectious diseases precede ADEM-onset in 70-90% of patients, whereas a post-vaccinal pathogenesis seems to be rare.[17,20] These preconditions are thought to trigger an overshooting immunologic response to ADEM with a fixed latency of days to weeks, as shown in our case. Upper respiratory tract virus-infections seem to be the most common causes for para-infectious ADEM, whereas Mycoplasma, Borrelia, Chlamydia and Legionella account splenectomized patient. Some aspects emerge from this instructive example of successful early immunosuppressive treatment in a rare autoimmune mediated para-infectious comorbidity.[20]

ADEM is an immune-mediated, monophasic inflammatory disorder of the central nervous system (CNS) and clinical as well as paraclinical differentiation among other primarily demyelinating CNS diseases, especially a first severe relapse of Multiple Sclerosis (MS) often is challenging.[3,19] Children or younger adults are affected by ADEM, but cases of adult-onset also have been described a monophasic course with a first-time demyelinating inflammatory CNS affection as well as a good response and favorable outcome after steroid therapy is characteristic for ADEM. However, some cases of recurrent or multiphasic courses.[7,19] A monophasic course with a first-time demyelinating inflammatory CNS affection as well as a good response and favorable outcome after steroid therapy is characteristic for ADEM. However, some cases of recurrent or multiphasic courses have also been reported (Tenembaum et al. 2007; Menge et al. 2005). Intravenous methylprednisolone is the first-line drug (10-30 mg/kg/day, up to a maximum of 1 g/day) for 3-5 days issued (Class IV). With this treatment modality, full recovery has been in 50%-80% of patients. Methylprednisolone-treated patients had a significantly better outcome concerning disability status when compared with those treated with dexamethasone.[2,6,10,19]

Oral corticosteroid treatment was continued with gradual tapering over six weeks to reduce the risk of relapse. However, this regimen is not in a randomized controlled trial. The role of corticosteroids in patients showed a delay in the course of the disease is questionable. Each type of vaccination should be avoided during the first six months after recovery.[7,20] Intravenous immunoglobulin (IVIG) (0.4 mg/kg/day for five days) is another option, but there are constraints on high costs and the evidence for this treatment modality in ADEM is Class IV. Improvements were within 2-3 days. In the absence of randomized controlled trials and the evidence available, either plasma exchange or IVIG could be a second-line treatment, when corticosteroids fail.[7,20]

4. Conclusion
It is the case of Acute Disseminated Encephalomyelitis in neurology patient. The treatment has been given to the patient such as antibiotics and supportive therapy, also planned high dose corticosteroids (immunotherapy) treatment.

References