

Unihemispheric Acute Disseminated Encephalomyelitis: A Case Report

Tek Hemisferi Tutan Akut Dissemine Ensefalomyelit: Olgu Sunumu

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is a rare idiopathic inflammatory demyelinating disease of the central nervous system (CNS), which usually occurs following a viral or bacterial infection, or vaccination. Characteristically, ADEM presents with acute encephalopathy with accompanying multifocal neurological symptoms and signs. Due to unavailability of a specific biological marker, neuroimaging is very important in establishing the diagnosis of ADEM. The typical magnetic resonance imaging findings of ADEM are large, multiple and almost symmetrical white matter lesions. Herein, we report a 31-year-old woman presenting with headache, confusion, left hemiparesis and hemihypoesthesia preceded by an infection. The patient fully recovered after intravenous methylprednisolone treatment. MRI revealed white-matter lesions that were typical for ADEM, but the unihemispheric involvement broadened the spectrum of disseminated encephalomyelitis. (*Archives of Neuropsychiatry* 2010; 47: 166-8)

Key words: Acute disseminated encephalomyelitis, unihemispheric, demyelination, magnetic resonance imaging

ÖZET

Akut dissemine ensefalomyelit (ADEM), nadir görülen, çoğu zaman viral veya bakteriyel bir enfeksiyonu ya da aşılantıyı takiben gelişen, merkezi sinir sisteminin (MSS) idyopatik inflamatuvar demiyelinizan bir hastalığıdır. Karakteristik olarak, ADEM akut ensefalopatiye eşlik eden multifokal nörolojik belirti ve bulgular ile kendini gösterir. Özgül bir biyolojik belirtecin yokluğu nedeniyle, görüntüleme yöntemleri ADEM tanısında önem kazanmaktadır. ADEM'in tipik MR görüntüleme bulguları; büyük, birden çok, neredeyse simetrik ak madde lezyonlarıdır. Bu yazıda; 31 yaşında, bir enfeksiyonu izleyen dönemde, baş ağrısı, konfüzyon, sol hemiparezi ve hemihipoestezi başvuran bir kadın hasta sunulacaktır. Hasta, intravenöz metilprednizolon tedavisi sonrası tamamen normale dönmüş ve izleyen beş yıl boyunca yeni bir yakınması olmamıştır. MR görüntülemelerinde gözlenen ADEM için tipik ancak tek hemisferi tutan lezyonlar dissemine ensefalomyelit spektrumunu genişletmektedir. (*Nöropsikiyatri Arşivi* 2010; 47: 166-8)

Anahtar kelimeler: Akut dissemine ensefalomyelit, tek yanlı, demiyelinizasyon, manyetik rezonans görüntüleme

Introduction

Acute disseminated encephalomyelitis (ADEM) is a rare idiopathic inflammatory demyelinating disease of the central nervous system (CNS), which usually occurs following viral/bacterial infections or vaccinations (1). Despite their resemblances, two autoimmune demyelinating CNS diseases, multiple sclerosis (MS) and ADEM, are considered mostly as distinct entities with their clinical, genetic, imaging and histopathological characteristics (2). However, these features might considerably overlap, especially in atypical cases, causing difficulties in the differential diagnosis. Occasionally, infectious, inflammatory and neoplastic disorders of the CNS might also present with ADEM or MS-like features.

Absence of definitely specific and sensitive biological markers renders neuroimaging extremely critical in the diagnosis of all these disorders. Different than many other inflammatory CNS disorders, ADEM typically affects widespread CNS locations at the same time point and thus, ADEM patients usually present with acute encephalopathy findings such as confusion, loss of consciousness, seizures, and the accompanying multifocal neurological symptoms and signs. Accordingly, typical ADEM lesions are large, multiple and semi-symmetrical foci of demyelination that are usually localized in bilateral supra- and infratentorial white matter regions (3). Here, we report a patient presenting with headache, confusion and left sensory-motor hemiparesis, with multiple large contrast-enhancing magnetic resonance imaging (MRI) lesions only in the white matter of the right hemisphere and fulfilling the criteria for ADEM.

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Case Report

A 31-year-old woman presented with headache, confusion and numbness of the left arm and leg. She had been suffering from headache, malaise and numbness on the left arm and leg for the last six days. During the next several days, her mental status had deteriorated and she had also developed a rapidly progressing weakness of the left arm and leg. Her past medical history was unremarkable, except for acute gastroenteritis that started three weeks prior to the hospital presentation. At admission, she was confused, agitated and disoriented. She was dysarthric and she could only obey simple commands. Her physical examination was normal with no fever and her neurological examination revealed left hemiparesis (4/5 on MRC scale) and hemihypoesthesia. Routine blood chemistry, complete blood count and sedimentation rate were normal. Her first MRI with a 3T unit (Siemens, Erlangen, Germany) showed multiple large (>2 cm), confluent and poorly marginated, T2/FLAIR hyperintense lesions with perilesional oedema and diffuse gadolinium enhancement in the central white matter of the right hemisphere (Figures 1 and 2). Periventricular white matter and corpus callosum were spared. The cerebrospinal fluid (CSF) contained 12 lymphocytes, normal protein level (34 mg/dl, normal: 15-60 mg/dl) and elevated IgG index (1.85, normal <0.7) with several oligoclonal bands (OCBs). An extensive serological testing for neurophilic viruses, systemic vasculitis markers and neuromyelitis optica (NMO) IgG were negative. Gruber-Widal test for salmonella infection was positive at 1/40 dilution, which

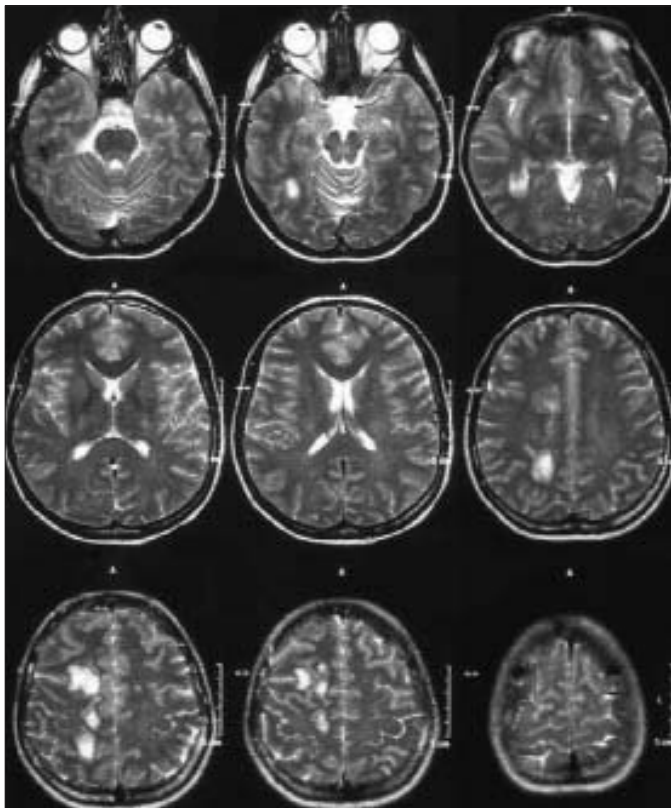


Figure 1. Acute disseminated encephalomyelitis with unilateral lesions. Axial T2-weighted MRI sections show unilateral, multiple, large, poorly marginated and hyperintense lesions with perilesional edema in central white matter

was compatible with the history of recent gastroenteritis. The patient was diagnosed with ADEM based on clinical and imaging findings. Following treatment with intravenous high-dose methylprednisolone (1g once a day) for 7 days, she completely recovered within 2 weeks. Control MRI at the sixth month of follow-up was almost normal (Figure 3). After 5 years of follow-up, she still remains without any neurological signs, symptoms or MRI lesions.

Discussion

ADEM is a rare immune-mediated inflammatory demyelinating disease of the CNS, which can occur at any age, but more commonly in children (1). It is typically preceded by an infection or vaccination. Salmonella infection has been reported as an immunological trigger for ADEM, as in our case (4).

Diagnosis of ADEM is based on clinical and radiological features. A differential diagnosis with the clinically isolated syndrome (CIS) should be done, considering the recent literature, which recommends early immunomodulatory treatment for CIS (5). While ADEM and MS/CIS are considered distinct diseases, they might occasionally imitate each other's features. It might be particularly difficult to differentiate ADEM from CIS since they are practically both isolated monophasic demyelinating disorders. The criteria for ADEM and its variants proposed by The International Pediatric MS Study Group include (i) a first

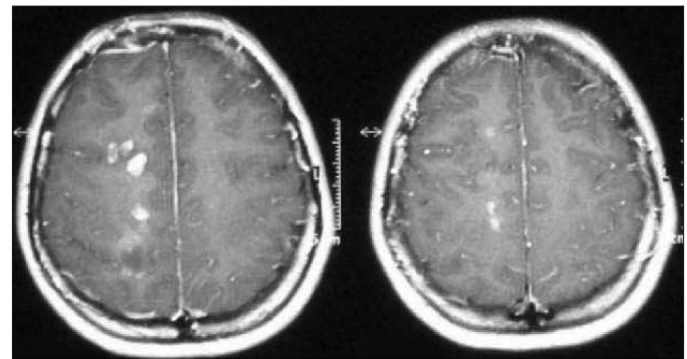


Figure 2. Axial T1-weighted MR images show diffuse gadolinium enhancement in the lesions. Note that all lesions show similar enhancement pattern indicating concurrency

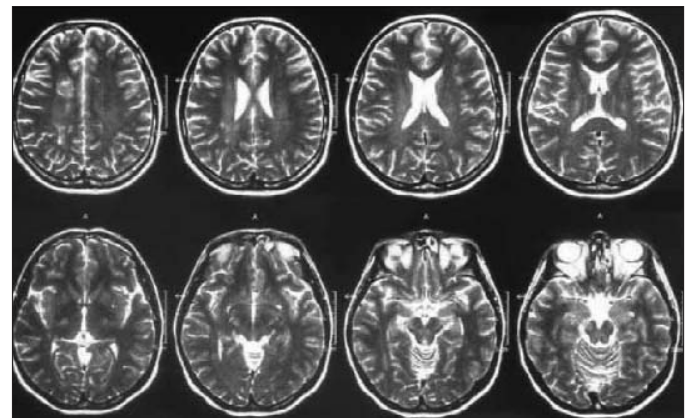


Figure 3. Axial T2-weighted MRI sections obtained at the sixth month of the follow-up show that the previous lesions have almost completely disappeared

clinical event with a polysymptomatic encephalopathy, with acute or subacute onset, showing focal or multifocal hyperintense lesions predominantly affecting the CNS white matter; (ii) no evidence of previous destructive white matter changes; and (iii) no history of a previous clinical episode (1).

These criteria reflect the typical and distinctive pathological features of ADEM, which influence widespread CNS locations in a usually, but not necessarily, monophasic disease course. Accordingly, symmetrical, bilateral, large (>1-2 cm), contrast-enhancing, supra- and infratentorial white matter lesions of the same age are observed on MRI of typical ADEM patients. These lesions usually quickly resolve and do not recur (6). Features that are unusual for MS, such as grey matter involvement (especially basal ganglia and thalamus) and sparing of periventricular white matter, are frequently seen in ADEM cases (7). Indistinct and poorly marginated lesions are also characteristic features of ADEM (8). While ADEM patients presenting with typical or atypical tumefactive MRI lesions with right or left predominance have been reported (9), in all these cases, lesions have been detected in both hemispheres and usually in both supra- and infratentorial locations. In contrast, the MRI lesions in our patient were not only strictly confined to one hemisphere, but the infratentorial CNS regions were spared as well.

Nevertheless, the clinical presentation of our case is fully compatible with the ADEM criteria, mentioned above. Absence of old MRI lesions, presence of same-age MRI lesions, all enhancing contrast, and resolution of these lesions shortly after the attack, support this diagnosis and decrease the likelihood of MS. Furthermore, our patient did not show any new neurological symptom or MRI findings in a 5-year follow-up period. ADEM is considered mostly a monophasic disease and the prognosis in adult patients has been reported to be favorable (1), as in our case. Moreover, prodromal symptoms as headache and malaise, followed by a rapid onset of encephalopathy and neurological deficits are typical for ADEM unlike MS. The CSF analysis of our patient showed lymphocytic pleocytosis, elevated immunoglobulin index and OCBs. While the CSF analysis in ADEM is usually expected to show no cells or mild pleocytosis, as in our case (10), presence of OCBs in the CSF is more typical for MS. However, this finding is not specific for MS and OCBs can also be detected in 58% of adult ADEM cases (11).

In summary, the presented case does not only constitute an interesting example for selective immunological vulnerability of the CNS, but it also expands the spectrum of disseminated encephalomyelitis with the unihemispheric involvement. In patients with restricted CNS involvement, the diagnosis of ADEM should not be immediately abandoned, especially if the clinical and radiological features are consistent with the ADEM criteria. Prior to starting long-term treatment methods, these patients should be closely monitored for re-emergence of new CNS findings or lesions to avoid unnecessary use of immunomodulating drugs.

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