Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Adult-onset acute disseminated encephalomyelitis: a rare case report in a 26-year-old female and review of literature

Mohammed D.M. Marsool, MBChB[a], Priyadarshi Prajjwal, MBBS[b], Pugazhendi Inban, MBBS[c], Ali D.M. Marsool, MBChB[a], Halla Tariq, MBBS[d], Omniat A. Hussin, MBBS[e,*]

Abstract

Acute disseminated encephalomyelitis is a demyelinating autoimmune condition that mostly affects the central nervous system. We present a case of a woman who presented with paralysis and speech deficits following an episode of upper respiratory tract infection. Brain and spinal cord imaging were used to confirm the diagnosis.

Keywords: acute disseminated encephalomyelitis, central nervous system, demyelinating disorders

Introduction

The central nervous system is largely impacted by the uncommon autoimmune illness known as acute disseminated encephalomyelitis (ADEM). It is distinguished by a sudden onset of neurological symptoms, such as fever, headache, vomiting, and mental confusion, which can develop into seizures, paralysis, and coma[1].

It is unknown as of yet as to what causes the disorder. It is believed that it occurs as a result of an autoimmune reaction to a certain component of bacterial or viral particles or even vaccination[2]. The cause of neurological impairment is the disease's inflammation, which causes demyelination of the white matter in the brain and spinal cord. Any person can be impacted by ADEM, although it is largely documented among children and young adults. It can have a major negative influence on these people's quality of life. Here, we present the case of a 26-year-old female who was diagnosed with ADEM.

HIGHLIGHTS

- A case of a 26-year-old woman who developed neurological symptoms following an upper respiratory tract infection.
- The diagnosis was acute disseminated encephalomyelitis (ADEM), which is an autoimmune condition that affects the central nervous system.
- The diagnosis was confirmed using brain and spinal cord imaging, along with cerebrospinal fluid and serum antibody testing.
- The paper highlights the importance of considering ADEM in patients with acute onset of neurological symptoms and a recent history of illness or immunization.
- The patient responded well to corticosteroid therapy. Corticosteroids are the mainstay of treatment for ADEM.

Case presentation

A 26-year-old female patient came to the emergency department with complaints of speech difficulties and abrupt onset paralysis in her lower limbs. She stated that 2 weeks before the commencement of her symptoms, she had experienced an upper respiratory tract illness. She denied having had any notable prior medical conditions, including multiple sclerosis or other autoimmune disorders. A physical examination indicated dysarthria as well as reduced reflexes and muscular strength in the lower limbs. Moreover, the patient developed a temperature of 38.5°C (101.3°F).

Although negative viral and bacterial cultures, laboratory testing (Table 1) revealed an increased white blood cell count of 12 000 cells/mm^3 with a left shift. White blood cell count in the cerebrospinal fluid (CSF) was 20 cells/mm^3, with high protein levels and lymphocytic pleocytosis (90% lymphocytes) (Table 2).

Magnetic resonance imaging (MRI) of the brain and spine revealed multifocal T2 hyperintensities consistent with ADEM (Figs 1 and 2) with open-ring enhancement. The patient was

[a]Neurology, University of Baghdad, Al-Kindy College of Medicine, Baghdad, Iraq, [b]Neurology, Bharati Vidyapeeth Medical College, Pune, [c]Internal Medicine, Government Medical College, Omandurar, India, [d]Multan Medical and Dental College, Multan, Pakistan and [e]Department of Medicine, Al Manhal University Academy of Science, Khartoum, Sudan

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Department of Medicine, Al Manhal University Academy of Science, Khartoum 1442, Sudan. Tel.: +249 99 263 3363. E-mail: omniatamir123@gmail.com (O.A. Hussin).

Annals of Medicine & Surgery (2023) 85:5242–5245

Received 12 July 2023; Accepted 17 August 2023

Published online 1 September 2023

http://dx.doi.org/10.1097/MS9.0000000000001239

Table 1

Lab results of a 26-year-old patient with ADEM

Laboratory test	Result	Reference range
White blood cell count	12.2×10^9/l	$4.5–11\times10^9$/l
Neutrophil count	7.8×10^9/l	$1.8–7.5\times10^9$/l
Lymphocyte count	2.9×10^9/l	$1.0–3.5\times10^9$/l
Monocyte count	0.7×10^9/l	$0.2–1.0\times10^9$/l
Eosinophil count	0.4×10^9/l	$0.0–0.5\times10^9$/l
Platelet count	220×10^9/l	$150–400\times10^9$/l
C-reactive protein (CRP)	2.4 mg/l	0–5 mg/l
Erythrocyte sedimentation rate (ESR)	12 mm/h	0–20 mm/h
Serum creatinine	0.8 mg/dl	0.6–1.2 mg/dl
Serum sodium	139 mmol/l	135–145 mmol/dl
Serum potassium	3.7 mmol/l	3.5–5.1 mmol/l
Serum calcium	9.1 mg/dl	8.6–10.2 mg/dl
Serum albumin	4.2 g/dl	3.5–5.2 g/dl

Note: Lab results were obtained upon admission to the hospital.
ADEM, acute disseminated encephalomyelitis.

diagnosed with ADEM and was started on intravenous methylprednisolone at a dose of 1 g per day for 5 days, followed by a gradual taper of oral prednisone over several weeks. Her symptoms improved gradually over the course of the treatment, and she was eventually discharged from the hospital with instructions to continue taking prednisone at a reduced dose.

A subsequent scan 5 weeks later showed the evolution of the previously demonstrated high T2 signal locations. Some of the bigger lesions had shrunk in size, while others that were only punctate had become more obvious (Fig. 3).

Discussion

ADEM is a disorder that is crucial to detect early despite having a low prevalence since it has the potential to seriously impair neurological function. While it is difficult to determine the exact incidence of ADEM, it is estimated to affect 1–2 per 100 000 people annually[1]. A quickly developing deficit in neurological function is how ADEM commonly manifests, and it is frequently preceded by an immune or infectious trigger, such as an upper respiratory infection (as in our patient), a vaccine, or another immune-related condition. Our patient reported a history of a viral-like episode 2 weeks before the onset of her symptoms, which goes along with similar findings published in the literature[2]. While the disease's progress is extremely unpredictable, some potential symptoms include polyradiculopathy, encephalopathy, and localized neurological abnormalities[3]. Because of the monophasic nature of ADEM, most patients make a full or nearly full recovery once their symptoms have subsided over weeks to months.

There are often nonspecific lab findings with regard to ADEM, despite the fact that some patients may have higher CSF protein or white blood cell counts[4]. The diagnosis of ADEM, which is primarily based on clinical and radiological symptoms, frequently depends on MRI of the spine and brain. The imaging features of ADEM include distinctive T2 hyperintense lesions that are often large, poorly defined, and contain both grey and white matter[5]. Edema or a mass effect may also be present, and the lesions are commonly ovoid or crescent-shaped.

The presence of three essential clinical characteristics is required for the International Pediatric Multiple Sclerosis Study Group (IPMSSG) criteria to establish the diagnosis of ADEM[6–8]. The first characteristic is the presence of a multisymptomatic neurological condition comprising encephalopathy, focal neurological impairments, and/or polyradiculopathy, with acute or subacute onset. The second one is that the brain and/or spine's MRI should reveal at least one distinctive lesion. The third essential characteristic is the appearance of several lesions that are spatially and/or temporally separated on an MRI, known as multifocal involvement. Other diagnoses that should be kept in mind when diagnosing ADEM include Acute Inflammatory Demyelinating Polyradiculoneuropathy, Aseptic Meningitis, Bell Palsy, progressive multifocal leukoencephalopathy (PML), cerebral autosomal dominant arteriopathy (CADASIL), and Brain Metastasis.

Table 2

CSF analysis of a 26-year-old patient with ADEM

Parameter	Result	Normal range
Appearance	Clear	Clear
Color	Colorless	Colorless
WBC count	20 cells/mm^3	< 5 cells/mm^3
Red blood cell count	0 cells/mm^3	0 cells/mm^3
Protein	85 mg/dl	15–45 mg/dl
Glucose	60 mg/dl	50–80 g/dl
Oligoclonal bands	Negative	Negative
Immunoglobulin G index (CSF/serum)	0.5	< 0.7

Note: Normal ranges may vary depending on the laboratory and the methods used for analysis.
ADEM, acute disseminated encephalomyelitis; CSF, cerebrospinal fluid; WBC, white blood cells.

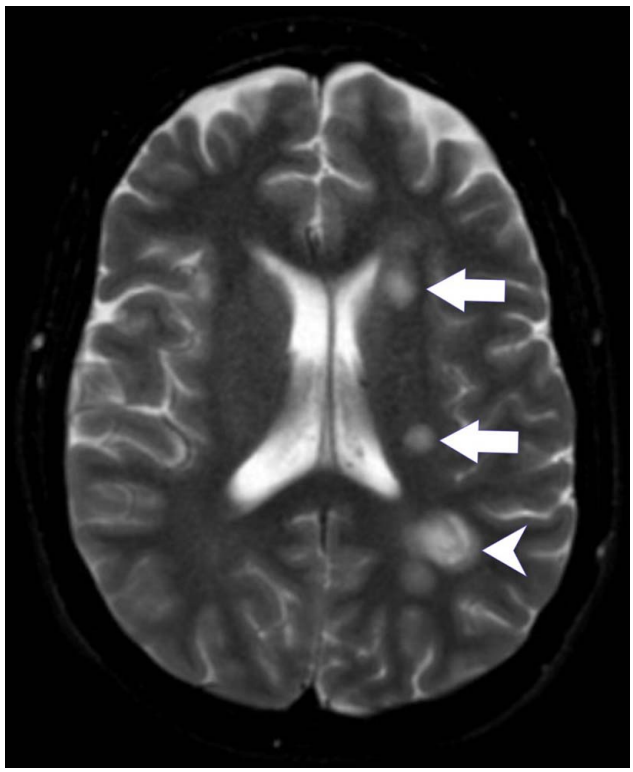

Figure 1. MRI of the brain shows multifocal high T2 signal in white matter regions (arrows) with ring enhancement (arrowhead).

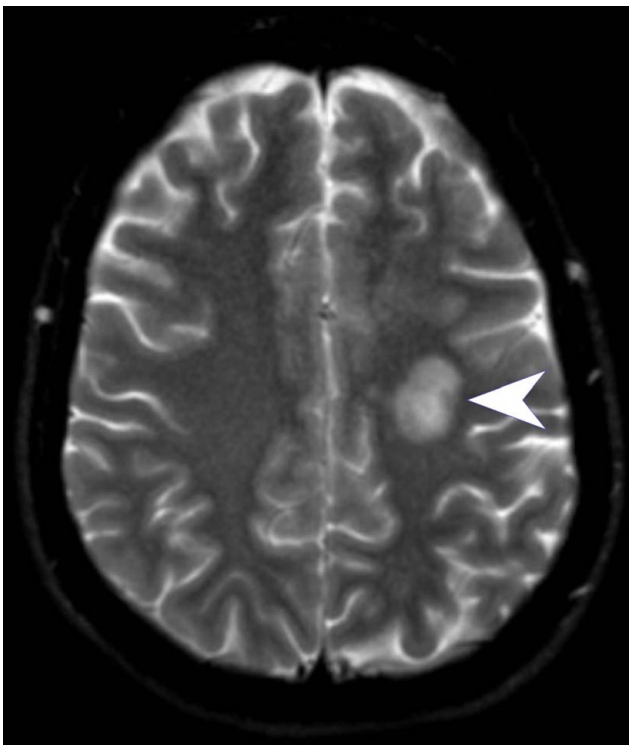

Figure 2. MRI of the brain shows multifocal high T2 signals in white matter regions. Also, open-ring enhancement (arrowhead) and leading-edge restricted diffusion are seen. No centrally restricted diffusion.

The International Pediatric Multiple Sclerosis Study Group criteria (IPMSSG criteria) calls for the exclusion of other diagnoses, such as multiple sclerosis (MS), neuromyelitis optica (NMO), and other inflammatory or infectious disorders that may mimic ADEM, in addition to these basic characteristics. Overall, the diagnostic criteria for ADEM emphasize the importance of clinical and radiological evidence of acute neurological dysfunction, multifocal involvement, and exclusion of other potential diagnoses. These criteria help ensure an accurate and timely diagnosis of ADEM, which is critical for the appropriate treatment and management of this rare but potentially debilitating condition.

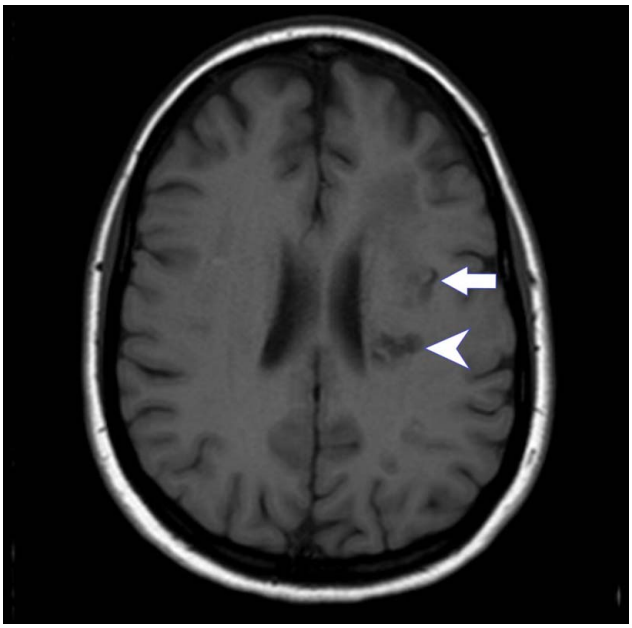

Figure 3. The follow-up scan shows the evolution of the previous T2 signal regions. Note that the left caudate (arrowhead) and lentiform (arrow) nuclei have significantly elevated T2 signals.

Typically, corticosteroids are used to treat ADEM, which can assist to lessen inflammation and prevent additional demyelination. Initial administration of oral prednisone is frequently followed by a progressive decrease of intravenous methylprednisolone[9]. If symptoms are severe or resistant to corticosteroids, some patients might need further immunomodulatory treatments, including intravenous immunoglobulin (IVIG), plasmapheresis, or rituximab. Although the majority of ADEM patients make a full recovery, some may suffer from permanent neurological disabilities, especially if the condition is severe or there are delays in its identification and treatment[10–12].

Conclusions

In this instance, a 26-year-old female patient with ADEM and a recent upper respiratory tract infection came with an abrupt onset of neurological symptoms. Clinical symptoms, test results, and imaging scans were used to make the diagnosis, and the patient responded favorably to corticosteroid therapy. Patients with acute onset of neurological symptoms and a recent history of illness or immunization should have ADEM taken into account when determining their differential diagnosis.

Key clinical message

The case emphasizes the significance of taking acute disseminated encephalomyelitis (ADEM) into account when making a diagnosis for a person who experienced severe neurological symptoms and recently had an infection. While being quite rare, ADEM can cause permanent deficits if untreated.

Ethical approval

Ethical approval was not required.

Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

No funding obtained.

Author contribution

M.D.M.M.: methodology, resources, writing – original draft preparation, review and editing, validation, and conceptualization;

P.P. and A.D.M.M.: writing – original draft preparation, review, and editing; P.I.: writing – original draft preparation, supervision, and validation; H.T.: writing – original draft preparation and visualization; O.A.H.: writing – original draft preparation and investigation.

Conflicts of interest disclosure

The authors declare no conflicts of interest, financial or otherwise.

Research registration unique identifying number (UIN)

1. Name of the registry: not applicable.
2. Unique identifying number or registration ID: not applicable.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not applicable.

Guarantor

Mohammed Dheyaa Marsool Marsool.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Data availability statement

No data is available for this case report.

Acknowledgements

None.

References

[1] Marchioni E, Tavazzi E, Minoli L, *et al*. Acute disseminated encephalomyelitis. Neurol Sci 2008;29:286–8.
[2] Menge T, Hemmer B, Nessler S, *et al*. Acute disseminated encephalomyelitis: an update. Arch Neurol 2005;62:1673–80.
[3] Pohl D, Alper G, Van Haren K, *et al*. Acute disseminated encephalomyelitis: updates on an inflammatory CNS syndrome. Neurology 2016; 87:38–45.
[4] Tenembaum S, Chamoles N, Fejerman N. Acute disseminated encephalomyelitis: a long-term follow-up study of 84 pediatric patients. Neurology 2002;59:1224–31.
[5] Kaunzner UW, Salamon E, Pentsova E, *et al*. An acute disseminated encephalomyelitis-like illness in the elderly: neuroimaging and neuropathology findings. J Neuroimaging 2017;27:306–11.
[6] Singhi PD, Ray M, Singhi S, *et al*. Acute disseminated encephalomyelitis in North Indian Children: clinical profile and follow-up. J Child Neurol 2006;21:851–7.
[7] Krupp LB, Tardieu M, Amato MP, *et al*. International Pediatric Multiple Sclerosis Study Group criteria for pediatric multiple sclerosis and immune-mediated central nervous system demyelinating disorders: revisions to the 2007 definitions. Mult Scler 2013;19:1261–7.
[8] Dale RC, de Sousa C, Chong WK, *et al*. Acute disseminated encephalomyelitis, multiphasic disseminated encephalomyelitis and multiple sclerosis in children. Brain 2000;123:2407–22.
[9] Kumar P, Kumar P, Sabharwal RK. Acute disseminated encephalomyelitis: case report and brief review. J Family Med Prim Care 2014;3:443–5.
[10] Anikumar A, Foris L, Tadi P. Acute Disseminated Encephalomyelitis. StatPearls [Internet]. StatPearls Publishing; 2021.
[11] Schwarz S, Mohr A, Knauth M, *et al*. Acute disseminated encephalomyelitis: a follow-up study of 40 patients. Neurology 2001;56:1313–8.
[12] Hynson JL, Kornberg AJ, Coleman LT, *et al*. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. Neurology 2001;56:1308–12.