Devastating demyelination: A catastrophic presentation of acute disseminated encephalomyelitis in an adult

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Abstract

***Background*: Acute disseminated encephalomyelitis (ADEM) is a rare, immune-mediated demyelinating disorder of the central nervous system (CNS). It is typically monophasic and most frequently affects children, but adult presentations, though uncommon, can be fulminant and associated with poor outcomes. Diagnosis in adults is often challenging due to overlapping clinical features with infectious and autoimmune processes.**

***Case presentation*: We report the case of a previously healthy 37-year-old woman who presented with one week of right upper limb numbness and severe brachial pain radiating to the neck. Initial suspicion was brachial neuritis or thoracic outlet syndrome. The following day, she developed high-grade fever (>39 °C), chills, sore throat, and dyspnea. On admission, she was hypotensive with an absent carotid pulse, requiring cardiopulmonary resuscitation, intubation, and inotropic support. Arterial blood gas analysis showed hypercarbic respiratory failure with pH 7.19, arterial carbon dioxide partial pressure ($PaCO_2$) 58 mmHg, and a lactate level of 6.2 mmol/l. Chest radiograph revealed bilateral fluffy opacities. Neurologically, she developed refractory generalized seizures. The electroencephalogram (EEG) showed diffuse epileptiform activity. Magnetic resonance imaging (MRI) of the brain revealed subtle bilateral parietal T2 hyperintensities with diffusion restriction, while spinal MRI showed longitudinal hyperintensities from C1 to D3. Laboratory workup revealed markedly elevated procalcitonin (>100 ng/dl), normal thyroid-stimulating hormone (TSH) (0.383 µIU/ml), and negative viral and bacterial serologies. Serum myelin oligodendrocyte glycoprotein (MOG) antibody was weakly positive at 1:10. Despite administration of intravenous immunoglobulin over three days and escalation of antiseizure therapy, her sensorium did not improve. Progressive sepsis with metabolic acidosis, dyselectrolytemia, coagulopathy, and diffuse cerebral edema ensued. Repeat imaging showed early uncal herniation. Despite maximal intensive care, she suffered cardiac arrest and expired.**

***Conclusion*: This case underscores the fulminant course of adult-onset ADEM, which may initially mimic peripheral or systemic illness, delaying diagnosis. Even with timely immunotherapy and intensive supportive care, outcomes can be fatal. The weak MOG antibody positivity highlights potential overlap syndromes and the limitations of current therapeutic strategies. Further research into advanced immunomodulatory therapies and approaches to manage malignant cerebral edema is warranted.**

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Keywords: Acute disseminated encephalomyelitis (ADEM), central nervous system, demyelinating disease, fulminant neuroinflammation, myelin oligodendrocyte glycoprotein (MOG).

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Introduction

Acute inflammatory conditions of the central nervous system (CNS) are uncommon but can progress rapidly, sometimes with devastating outcomes in otherwise healthy adults. Their clinical course often overlaps with infectious, autoimmune, and parainfectious processes, making early recognition difficult. Among these disorders, acute disseminated encephalomyelitis (ADEM) is a monophasic, immune-mediated demyelinating condition of the CNS, more frequently encountered in children but occasionally reported in adults, where it may follow a more fulminant trajectory. (1–4) We describe the case of a previously healthy 37-year-old woman who initially presented with focal neurological complaints but deteriorated rapidly into a fatal neuroinflammatory syndrome despite timely initiation of therapy.

Case report

A 37-year-old female schoolteacher, previously healthy, presented with one week of right upper limb numbness and severe progressive pain radiating to the neck, worsened by movement. Initial evaluation suggested brachial neuritis or thoracic outlet syndrome, and nerve conduction studies were planned.

The following day, she developed high-grade fever with chills, sore throat, odynophagia, dyspnea, and chest pain. She later became hypotensive with loss of carotid pulse and shifted to intensive care unit, requiring cardiopulmonary resuscitation, intubation, and inotropic support. Chest radiography revealed bilateral fluffy opacities, and arterial blood gas analysis confirmed hypercarbic respiratory failure with lactic acidosis. Patient was started on broad spectrum antibiotics due to the increasing trend of white blood cells and fever.

Neurologically, the patient developed involuntary eyelid flickering progressing to generalized seizures and refractory status epilepticus. EEG showed diffuse epileptiform activity. Magnetic resonance imaging (MRI) of the brain revealed subtle gyriform T2 hyperintensities with diffusion restriction in bilateral parietal lobes. MRI spine demonstrated longitudinal hyperintensities extending from C1 to D3 without mass lesions. Intravenous immunoglobulin was administered over three days, and antiseizure therapy was escalated. Seizure activity ceased, but her sensorium did not improve.

Progressive sepsis with multiorgan dysfunction ensued, including metabolic acidosis, dyselectrolytemia, and coagulopathy. Repeat neuroimaging showed diffuse cerebral and cerebellar edema, effacement of cisterns and ventricles, and early uncal herniation. Despite maximal intensive care and supportive measures, she developed cardiac arrest and expired.

Laboratory investigations included bacterial cultures, viral markers (including severe acute respiratory syndrome coronavirus 2 [SARS-CoV-2] and influenza), parasitic serologies, and antiphospholipid antibodies. Procalcitonin was markedly elevated. Serum myelin oligodendrocyte glycoprotein MOG antibody was weakly positive at 1:10 (**Table 1**).

Discussion

ADEM is generally considered a single-episode autoimmune demyelinating condition of the central nervous system, occurring most often in children but also documented in adults. (1,2) The disorder is usually preceded by infection or vaccination, with an aberrant immune reaction against myelin antigens believed to drive the pathology. (1,3) In pediatric populations, outcomes are often favorable with corticosteroid therapy, but in adults, disease onset can be atypical and sometimes progresses rapidly with high morbidity and mortality. (3,4) Rare fulminant forms have been described, characterized by explosive neurological decline and life-threatening cerebral edema. (5,6)

In the present case, the patient's early symptoms—unilateral sensory loss with severe brachial pain—resembled peripheral nerve pathology, particularly brachial neuritis. (7) This highlights the diagnostic challenge in adult ADEM, where initial focal or radicular features may delay recognition. Subsequent systemic manifestations including fever, respiratory failure, and hemodynamic collapse were consistent with a parainfectious presentation, which has been linked to viral triggers such as adenovirus and influenza in prior reports. (8,9)

MRI demonstrated new onset bilateral cortical signal changes together with longitudinal lesions across the cervical-thoracic cord, findings strongly indicative of inflammatory demyelination. (10–13) An additional challenge was the weakly positive MOG antibody. Low titres alone are not sufficient to establish MOG antibody-associated disease; however, overlap between adult ADEM and MOG seropositivity has been documented, raising the possibility of a broader disease spectrum. (12,14) Recognition of such overlap is clinically relevant, as MOG antibody-associated demyelination may differ in prognosis and relapse risk.

Despite treatment with intravenous immunoglobulin (IVIG), antiseizure medications, and intensive supportive measures, the patient's condition progressed to severe cerebral edema with herniation, a

complication noted in fulminant cases. (5,6,10,15) Corticosteroids were deferred due to suspected infection and pending culture results. Although decompressive craniectomy and other rescue therapies have been attempted in isolated reports, outcomes have remained inconsistent. (6,15)

This case illustrates that adult-onset ADEM can present as peripheral or systemic illness in the early stages, complicating timely diagnosis. Also disease trajectory can be abrupt and catastrophic, even with appropriate immunotherapy and intensive care. Radiological and antibody findings are helpful but not definitive, and diagnostic uncertainty remains in cases with overlapping features. Finally, the limitations of current therapies highlight the need to investigate additional strategies, such as plasma exchange, B-cell–targeting biologics, or approaches to malignant cerebral edema.

To conclude, fulminant ADEM in adults represents a rare yet often fatal manifestation of an otherwise usually monophasic illness. Prompt recognition, rapid initiation of immunotherapy, and coordinated intensive care remain critical, but further research is needed to identify effective treatments for aggressive neuroinflammatory syndromes.

Conclusion

Fulminant ADEM in adults is an extraordinarily rare but catastrophic presentation of an illness more commonly seen as a monophasic, self-limiting disorder in children. In contrast, the adult fulminant phenotype is distinguished by precipitous neurological decline, severe cerebral edema, and high mortality despite maximal supportive and immunomodulatory therapy. For intensivists, such cases highlight the dual challenge of achieving rapid diagnostic certainty amid diagnostic overlap with other fulminant demyelinating or infectious encephalitides, while simultaneously initiating timely neuroprotective and disease-modifying interventions.

Critical care support is central to management: meticulous intracranial pressure control, prevention of secondary brain injury, seizure management, and organ support remain essential components of care. Yet, even with aggressive corticosteroid therapy, adjunctive use of IVIG or plasma exchange, and coordinated neurocritical care, outcomes remain dismal in many patients. This underscores the urgent need for prospective studies to define prognostic markers, evaluate emerging biologic or targeted immunotherapies, and establish standardized treatment algorithms for fulminant neuroinflammatory syndromes.

This case reinforces the devastating nature of fulminant adult-onset ADEM and illustrates that improved recognition, early immunotherapy, and optimized multidisciplinary intensive care are imperative. However, altering the natural history of this entity will ultimately depend on translational research that bridges neuroimmunology with critical care practice.

Table 1. Important investigations

Viral markers	
- HBsAg	Negative
- HCV Ab	Negative
- HIV p24, I and II antibody	Negative
Infectious etiology	
- Dengue serology	Negative
- Malaria	Negative
- Scrub typhus serology	Negative
- Leptospirosis serology	Negative
Respiratory viral panel for Influenza A subtype H1N1, Influenza B, SARS CoV-2, respiratory syncytial virus A and B	Negative
Blood, urine, and endotracheal tube cultures	No Growth
Others	
- Procalcitonin	>100 ng/dl
- Thyroid stimulating hormon	0.383 micro IU/ml
- Anti thyroid peroxidase	7.22 IU/l (normal <34)
- Anti nuclear antibody	Negative
- Anti neutrophil cytoplasmic antibody	Negative
Antiphospholipid antibodies	
- Anticardiolipin antibodies (IgG, IgM)	Negative
- Anti beta 2 glycoprotein (IgG, IgM)	Negative
- Lupus anticoagulation	Negative

Legend: HbsAg=hepatitis B surface antigen; HCV Ab=hepatitis C virus antibody; HIV=human immunodeficiency virus; SARS CoV-2=Severe Acute Respiratory Syndrome Coronavirus 2; IgG=immunoglobulin G; IgM=immunoglobulin M.

References

1. Pohl D, Alper G, Van Haren K, Kornberg AJ, Lucchinetti CF, Tenembaum S, et al. Acute disseminated encephalomyelitis: Updates on an inflammatory CNS syndrome. Neurology 2016;87:S38–45.
2. Anilkumar AC, Foris LA, Tadi P. Acute Disseminated Encephalomyelitis. Treasure Island (FL): StatPearls Publishing; 2024 [cited 2025 Jan]. Available from: https://pubmed.ncbi.nlm.nih.gov/28613684/
3. Li K, Li M, Wen L, Wang Q, Ding X, Wang J. Clinical Presentation and Outcomes of Acute Disseminated Encephalomyelitis in Adults Worldwide: Systematic Review and Meta-Analysis. Front Immunol 2022;13:870867.
4. de Seze J, Debouverie M, Zephir H, Lebrun C, Blanc F, Bourg V, et al. Acute fulminant demyelinating disease: a descriptive study of 60 adult patients. Arch Neurol 2007;64:1426–32.
5. Sekula Jr RF, Marchan EM, Baghai P, Jannetta PJ, Quigley MR. Central brain herniation secondary to fulminant acute disseminated encephalomyelitis: implications for neurosurgical management. Case report. J Neurosurg 2006;105:472–4.
6. Azab MA. Neurosurgical intervention in an unusual case of extensive acute disseminated encephalomyelitis - A case report and literature review. Surg Neurol Int 2023;14:176.
7. Ferrante MA, Wilbourn AJ. Lesion distribution among 281 patients with sporadic neuralgic amyotrophy. Muscle Nerve 2017;55:858–61.
8. Campbell D, Wong GS, Park H, McLeod G. An adult case of adenovirus-associated acute disseminated encephalomyelitis. Case Rep Infect Dis 2023;2023:5528198.
9. Tenembaum S, Chitnis T, Ness J, Hahn JS; International Pediatric MS Study Group. Acute disseminated encephalomyelitis. Neurology 2007;68:S23–36.
10. Hochmeister S, Gattringer T, Asslaber M, Stangl V, Haindl MT, Enzinger C, et al. A Fulminant Case of Demyelinating Encephalitis With Extensive Cortical Involvement Associated With Anti-MOG Antibodies. Front Neurol 2020;11:31.
11. Young NP, Weinshenker BG, Lucchinetti CF. Acute Disseminated Encephalomyelitis: Current Understanding and Controversies. Semin Neurol 2008;28:84–94.
12. Mirbagheri S, Eckart Sorte D, Zamora CA, Mossa-Basha M, Newsome SD, Izbudak I. Evaluation and management of longitudinally extensive transverse myelitis: a guide for radiologists. Clin Radiol 2016;71:960–71.
13. Miki Y. Magnetic resonance imaging diagnosis of demyelinating diseases: An update. Clin Exp Neuroimmunol 2019;10:32–48.
14. Zang W-Z, Yang H, Li D, Zhao Z-D, Sun Y-J, Xia M-R, et al. Clinical comparative analysis of monophasic and multiphasic acute disseminated encephalomyelitis in adults. Arch Med Sci 2020;19:687–93.
15. Ide T, Ebashi R, Eriguchi M, Aishima S, Abe T, Hara H. Fulminant demyelinating disease of the central nervous system effectively treated with a combination of decompressive craniectomy and immunotherapy: A case report and literature review. Clin Case Rep 2024;12:e9059.

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