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*Case Report*

# Neurosurgical intervention in an unusual case of extensive acute disseminated encephalomyelitis – A case report and literature review

Mohammed A Azab

Department of Biomedical Sciences, Boise State University, Boise, United States.

E-mail: *Mohammed A Azab - mohammed.azab@kasralainy.edu.eg

***Corresponding author:**
Mohammed A Azab,
Department of Biomedical Sciences, Boise State University, Boise, United States.

mohammed.azab@kasralainy.edu.eg

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## ABSTRACT

**Background:** The clinical presentations of demyelinating diseases are variable and can range from mild symptoms to fulminant presentations. Acute disseminated encephalomyelitis is one of those diseases which usually follow an infection or vaccination.

**Case Description:** We report a case of extensive acute demyelinating encephalomyelitis (ADEM) with massive brain swelling. A 45-year-old female presented to the emergency room with status epilepticus. Patient has no history of any associated medical problems. Glasgow coma scale (GCS) was 15/15. CT brain was normal. Lumbar puncture was done and cerebrospinal fluid showed pleocytosis and increased protein content. About 2 days after admission, the conscious level rapidly deteriorated and GCS was 3/15, with the right pupil fully dilated and unreactive to light. Computed tomography and magnetic resonance imaging brain were done. We performed an urgent decompressive craniectomy as a life-saving procedure. Histopathological examination was suggestive of ADEM.

**Conclusion:** Few cases of ADEM with brain swelling were reported, but there is no solid consensus about the appropriate management of these cases. Decompressive hemicraniectomy is a possible choice, but further research is needed to evaluate the proper timing, and indication of surgery.

**Keywords:** Acute demyelinating encephalomyelitis (ADEM), Brain edema, Decompressive craniectomy, Demyelinating, Herniation

## INTRODUCTION

Primary demyelinating diseases include multiple sclerosis and less frequently encountered diseases as acute demyelinating encephalomyelitis (ADEM), neuromyelitis optica, tumefactive demyelinating disease, Marburg's disease, Balo's disease, and Schilder's disease. ADEM is an inflammatory demyelination process of the central nervous system that usually occurs after a viral or bacterial infection or vaccination. The presentation varies from mild neurological deficit to severe fulminant disease.[9] The implicated pathophysiology includes an autoimmune response to certain myelin-associated antigens.[6] Immunosuppression and supportive therapy is the mainstay of treatment.[15]

The acute deterioration could be caused by seizures, disturbed conscious level, and brain herniation. In rare occasions, ADEM can cause severe life-threatening brain edema and increased

intracranial pressure (ICP).[19] This brain swelling may be refractory to brain dehydrating measures and requires surgical decompression.[22] The prognosis of ADEM has been noticed to be favorable,[4,11] while there is not much evidence about the role of surgical management in cases of fulminant ADEM with raised ICP.[4,22]

Herein, we describe a case of fulminant ADEM, where the rapid development of severe cerebral edema and brain herniation was treated with decompressive hemicraniectomy and temporal polectomy. We tried to report similar cases in the literature to determine the optimal management in those cases and to highlight the heterogeneity of clinical presentations and outcome.

## CASE DESCRIPTION

A 45-year-old female presented to the emergency room with status epilepticus. The patient has no relevant history for any associated medical problems. Initial resuscitation was done and midazolam and loading phenytoin was given to the patient. The patient improved and glasgow coma scale (GCS) was 15/15. CT brain was normal. The patient was admitted to the intensive care unit (ICU) for further examination. By history reviewing, she had a fever 4 days before admission. Electroencephalogram (EEG) was done and revealed generalized slowing of electric activity. Lumbar puncture was done and cerebrospinal fluid (CSF) was sent for analysis which showed pleocytosis and increased protein content. About 2 days after admission, the conscious level rapidly deteriorated and GCS was 3/15, with the right pupil fully dilated and unreactive to light. CT brain is shown in Figure 1. Magnetic resonance imaging (MRI) with contrast is requested urgently Figure 2.

Brain dehydrating measures were started. The patient was given IV mannitol, IV dexamethasone, and deep sedation which were applied. IV acyclovir was given empirically. The patient did not improve and the condition remained the same. Urgent decompressive craniectomy was done. Intraoperatively, the brain was markedly swollen, so we performed frontal and temporal polectomy. Postoperatively, the patient's conscious level did not improve. The patient is still in the ICU. Histological features are characteristic of ADEM, as shown in Figure 4.

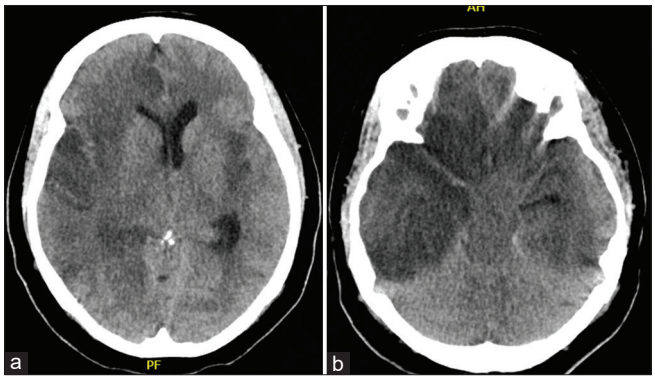


**Figure 1:** (a and b) Preoperative computed tomography brain shows massive cerebral edema with midline shift.

## DISCUSSION

ADEM is an inflammatory demyelinating disease that is relatively rare in adults.[11] Although immunosuppressant therapy is the mainstay of treatment of ADEM, the response time of available therapies may allow for progression of brain edema and herniation. Few reports are described in the literature about the management of ADEM causing massive brain edema.[5,13,14] There is no general consensus about the best management strategy in these cases.[8]

Diagnosis of ADEM is largely dependent on clinical presentation and imaging. Earlier in the disease, CT may be normal, or it may show multifocal white matter changes in clinically advanced cases.[10] MRI is considered the main investigation for a suspected ADEM. Hyperintense confluent white matter changes in T2-weighted, and non-contrast fluid-attenuated inversion recovery imaging scans

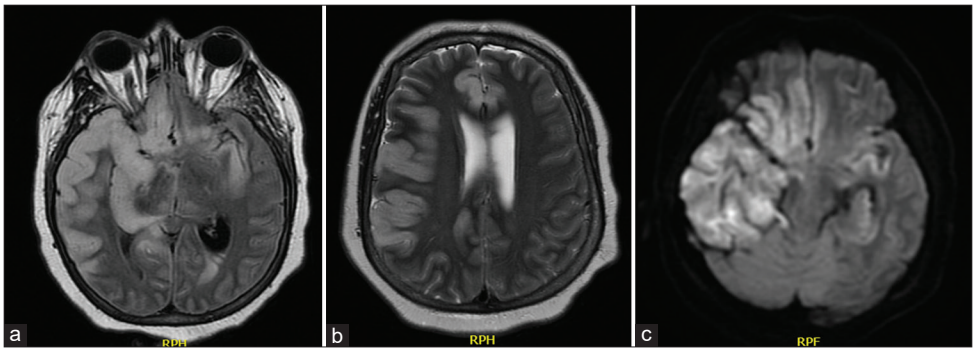


**Figure 2:** (a-c) Magnetic resonance imaging (MRI) fluid-attenuated inversion recovery, T2MRI, diffusion-weighted imaging MRI, brain showing diffuse scattered patchy hyperintensities affecting mainly the right temporal and parietal lobes, and left hippocampus. RPH: Relative peak height, RPF: Relative peak flow.

**Table 1:** Similar case reports of extensive ADEM requiring emergency decompressive craniectomy.

| Reference | Age | Sex | H/O recent infection | Clinical presentation | Preoperative GCS | Medical treatment | EVD inserted or not | ICP monitor bolt | Conservative methods used to reduce ICP | Duration from onset to decompressive | Lobectomy was done or not | Outcome |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Dombrowski *et al.*[4] | 41 | M | Yes | Dysarthria and aphasia | 4 | Broad-spectrum antibiotics, IVIG, steroids | Yes | No | Hyperosmolar therapy, sedation and hyperventilation | 5 days | No | Conscious level improved |
| Refai *et al.*[13] | 51 | F | Yes | Headache, left sided hemiparesis | NM | Acyclovir and broad-spectrum antibiotics. High-dose methylprednisolone | No | Yes | Hyperventilation, mannitol, moderate hypothermia and sedation | 2 days | No | Improved |
| Sekula *et al.*[14] | 32 | M | Yes | Left central facial weakness and left hemiplegia | 3 | Acyclovir | Yes | No | Mannitol, and intravenous dexamethasone | 4 days | Yes | Conscious level improved |
| Granget *et al.*[5] | 18 M | F | Yes | Fever and seizures | | Acyclovir, IVIG | Yes | No | Mannitol and barbiturates | Few hours | No | Improved |
| Ahmed *et al.*[1] | 38 | F | Yes | Left sided weakness | 9 | Acyclovir | No | No | Mannitol, intravenous dexamethasone | 10 hours | No | Improved |
| Von Stuckrad-Barre *et al.*[22] | 34 | F | No | Low grade fever, headache | 3 | NM | No | No | IV mannitol, sedation with midazolam, mild hyperventilation | 9 days | No | Improved |
| Abid *et al.*[7] | 32 | F | Yes | Left hemiparesis | NM | Steroids | NM | NM | NM | 5 days | Yes | Death |

IV: Intravenous, NM: Not-mentioned, GCS: Glasgow coma scale, ICP: Increased intracranial pressure, EVD: External ventricular drain, ADEM: Acute demyelinating encephalomyelitis, IVIG: Intravenous immunoglobulin, M: Male, F: Female

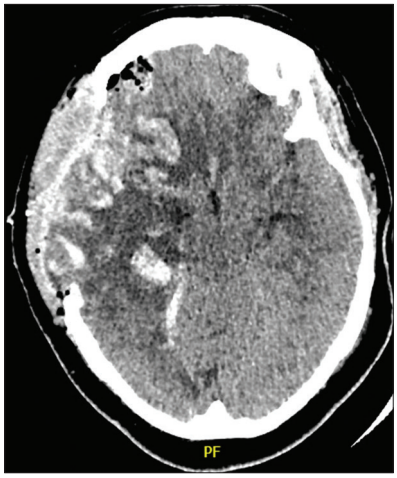


**Figure 3:** Postoperative computed tomography brain. PF: Plain Film.

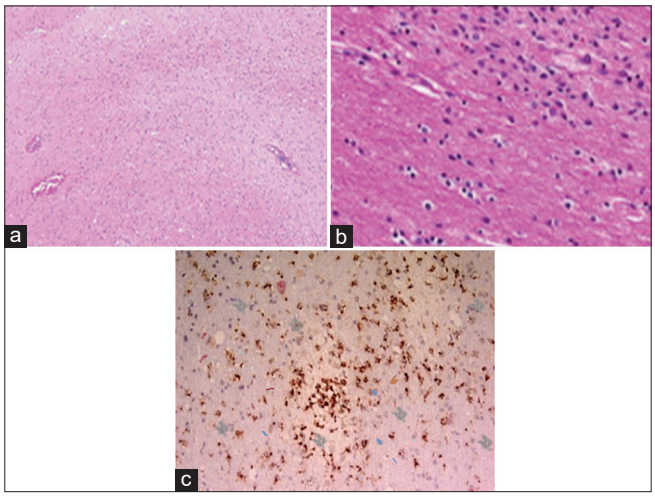


**Figure 4:** (a and b) Diffuse demyelination of the cortical white matter with abundant mononuclear inflammatory cells most of them are lymphocytes with reactive pink gliosis. (c) CD68/SR-D1 staining is positive showing monocytes and macrophages distribution in the perivascular areas.

are very common.[16,21] Brainstem and spinal cord lesions are frequently encountered.[16] CSF analysis may show non-specific changes in the form of increased cell count and protein content, especially the immunoglobulin level.[18]

High dose of intravenous steroids, intravenous immunoglobulin, and plasmapheresis is the main treatment lines in the acute stage.[2] A good prognosis and proper functional recovery is anticipated in cases treated aggressively.[12] However, in cases of increased ICP, the optimal treatment is still undefined and the effect on prognosis is not documented. Conservative management in the form of brain dehydrating measures, hyperventilation, and hypothermia can reduce the ICP in most cases.

In cases of persistent ICP elevation, decompressive craniectomy is the last option. This can reduce the intracranial pressure, but the effect on the outcome is still undefined. There are no solid criteria for the exact timing and patient selection for decompressive craniectomy for ADEM. The use of decompressive hemicraniectomy has been well described in cases of herpes simplex encephalitis.[3,5,14,17,20] However, few reports describe decompressive hemicraniectomy as a life-saving method in cases of ADEM with massive brain edema.[4,22] There is not much evidence about the long-term outcome and prognosis in those cases.

In this case, we tried to reduce the ICP with conservative measures, but the patient's condition did not improve. Therefore, decompressive hemicraniectomy with frontal and temporal polectomy was done. The patient's condition did not improve postoperatively. Hossem *et al.* treated a case of ADEM with decompressive hemicraniectomy, and lobectomy and the patient died.[7] While Ahmed *et al.* reported marked improvement of their case after decompressive craniectomy.[1] Table 1 lists the case reports of fulminant ADEM in the literature that were managed by decompressive craniectomy.

## CONCLUSION

The reported literature describing the role of decompressive craniectomy in the management of severe cases of ADEM is still lacking. Few cases of ADEM with brain swelling were reported, but there is no solid consensus about the appropriate management. Decompressive hemicraniectomy is a possible choice, but further research is needed to evaluate the proper timing and indication of surgery.

### Ethical approval

Consent was obtained by the patient in this study. The Hospital Research Board approved the Institutional Review Board (IRB) for this work.

### Author Contributions

Dr. Mohammed A Azab has contributed to designing the idea, drafting, manuscript writing, and the final review.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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