



Decompressive Hemicraniectomy for Severe Acute Disseminated Encephalomyelitis: A Case Report

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Abstract

This report presents a rare case of adult-onset severe Acute Disseminated Encephalomyelitis (ADEM) complicated by uncal herniation, which is a very rare complication of the disease, requiring lifesaving decompressive hemicraniectomy with complete recovery. This is a 27-year-old female, who presented with headache, right-sided body weakness, dysarthria, blurred vision, and progressive altered level of consciousness over a course of 2 days, preceded by an upper respiratory tract infection 7 days prior to the onset of the above-mentioned symptoms. Brain MRI showed extensive abnormal white matter lesions which appear hyperintense, involving occipital, parietal, and temporal lobes, more on the left side. CSF exam showed high protein. According to the clinical and MRI findings, she was diagnosed as severe ADEM. She was started on high-dose steroids and plasma exchange. Unfortunately, her condition deteriorated rapidly with sudden drop in her level of consciousness with fixed dilated pupils. Urgent imaging showed uncal herniation. Lifesaving urgent left decompressive hemicraniectomy was performed with subsequent resolution of herniation radiologically and clinically. She was kept on steroid and plasmapheresis, followed by Intravenous Immunoglobulin (IVIG) with gradual improvement. After one month, she completely recovered and became independent in her activities, with significantly improved MRI lesions. In conclusion, severe ADEM complicated by uncal herniation is an extremely rare presentation of the disease. Urgent and aggressive interventions, including decompressive surgeries, can save lives and lead to favorable outcomes.

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Keywords: ADEM disease; Decompression; Craniectomy; IVIG

Abbreviations

ADEM: Acute Disseminated Encephalomyelitis; IVIG: Intravenous Immunoglobulin; GCS: Glasgow Coma Scale; FLAIR: Fluid-Attenuated Inversion Recovery; ICP: Intracranial Pressure

Introduction

ADEM (Acute Disseminated Encephalomyelitis) is an autoimmune disease, characterized by demyelination of the CNS, which is commonly preceded by viral disease or vaccine administration, with a latent period of 4 to 21 days before the onset of symptoms, although the inciting event could not be found in many cases [1]. This disease occurs more commonly in children and adolescents than adults. It's a rare disease and its actual incidence is still unknown [2]. Typically, ADEM is monophasic, and patients usually present with an acute onset of multifocal neurologic symptoms with encephalopathy. Motor deficit is the main presenting symptom [1,2]. The diagnosis of ADEM disease is clinical and supported by neuroimaging. MRI usually shows demyelinating lesions in the white matter of the brain [1,2]. CSF analysis usually shows non-specific changes [2]. Management of ADEM disease includes high-dose steroid, Intravenous Immunoglobulin (IVIG), and plasmapheresis, with no definite guidelines regarding the optimal therapeutic approach [2-5].

Rarely, ADEM disease can be complicated by significant Increased Intracranial Pressure (ICP) due to massive brain edema, brain herniation and death. After reviewing the literature, we found that only very few cases of severe ADEM disease complicated by uncal herniation that required

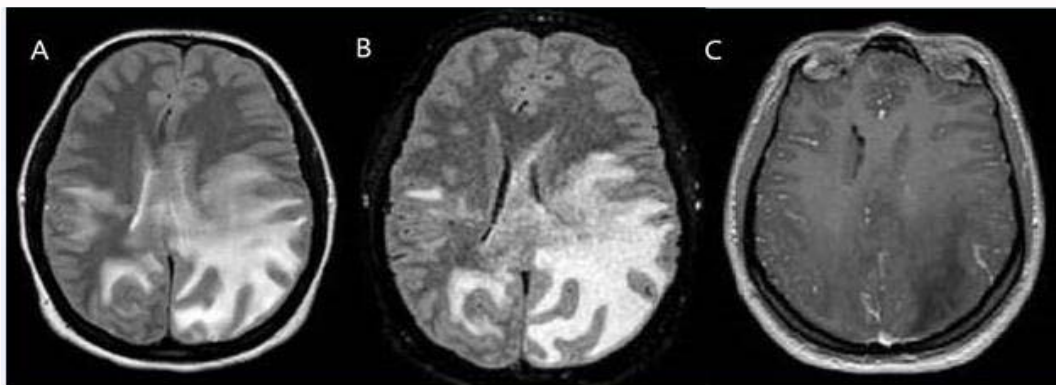


Figure 1: Initial brain MRI. A, B: shows hyperintense lesions on T2, Fluid-Attenuated Inversion Recovery (FLAIR), respectively, more on the left side, associated with local edema. C: Shows the same lesion appears hypointense on T1.

surgical hemispherectomy were reported [3-6]. Herein, we report a case of a 27-year-old female, who was diagnosed with severe ADEM, complicated by significant brain edema, and uncal herniation, that was managed by left decompressive hemispherectomy with full recovery within one month of therapy.

Case Presentation

A 27 year old female patient, with unremarkable past history, was admitted to our hospital with a history of headache, nausea, vomiting, right-sided body weakness, dysarthria, blurred vision, and progressive altered level of consciousness over a course of 2 days. These symptoms were preceded by upper respiratory tract infection (dry cough and general weakness) 7 days prior to the onset of the above mentioned symptoms.

At presentation, her examination was remarkable for altered sensorium with a Glasgow Coma Scale (GCS) of 13/15, preserved pupillary response, right sided hemiplegia with a power of 0/5, weakness of her left side with a power of 1/5, extensor plantar reflex bilateral. Brain MRI showed extensive abnormal white matter intensities which appear hyperintense on T2/Fluid-Attenuated Inversion Recovery (FLAIR), involving occipital, parietal, and temporal lobes, more on the left side, associated with local edema but with no herniation or significant midline shift (Figure 1). Laboratory investigations were unremarkable. CSF analysis was normal except for high protein levels. Coronavirus PCR test was negative. All other viral serology and PCR were negative.

Due to her acute presentation and the MRI findings, severe

ADEM disease was diagnosed. She was started on medical treatment with IV Methylprednisolone 1 gram daily for 5 days, and anti-edema medications (IV Mannitol and PO Acetazolamide) to decrease the ICP. As the presentation was severe, plasmapheresis was started as an additional therapeutic approach.

Unfortunately, on the next day, her condition deteriorated as she developed a sudden onset drop in her level of consciousness with fixed dilated pupils. Intubation was done to secure the airways; an urgent brain CT scan showed midline shifting to the right and uncal herniation. Urgent left decompressive hemispherectomy was performed. After the procedure, pupils returned to be reactive, and control brain CT scan showed resolution of the midline shift and uncal herniation, with the same finding of brain edema. She was continued on medical treatment that included 7 sessions of plasmapheresis, and 5 days of pulse steroid, followed by tapering oral prednisolone.

Two weeks after the procedure she was fully conscious, power in the left side improved to 3/5, but her right-sided hemiplegia did not improve. She was successfully weaned from the ventilator and extubated. Control Brain MRI done at that time showed improvement in the brain edema and the intensity of the previously mentioned lesions (Figure 2).

As patient's right sided hemiplegia did not improve, she was given IVIG 1 gm/Kg/dose for 3 days as a third - line medical treatment. The patient showed good clinical response after IVIG, manifested by a gradual improvement in her right side power (around 3 weeks after the decompression) with a gradual return to her activities of daily livings. After then, the patient was discharged home after 50 days

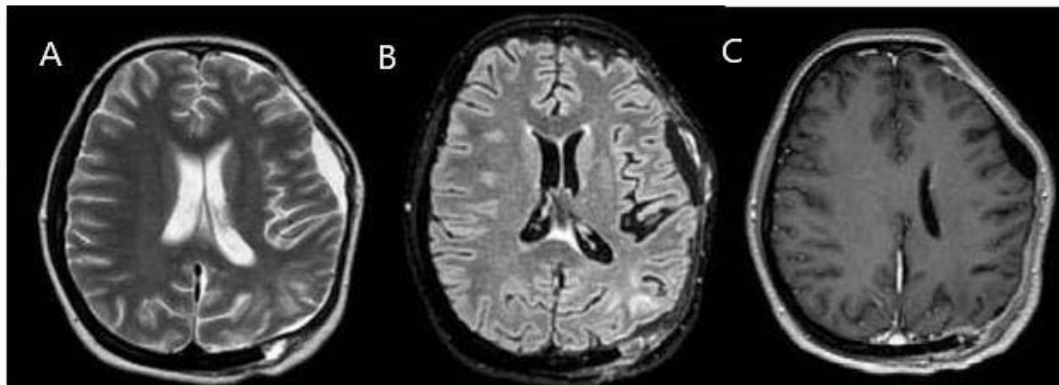


Figure 2: Control brain MRI. A, B: Shows improving hyperintense lesions on T2, FLAIR, respectively, C: shows improving hypointense lesion on T1.

of hospitalization, in good general condition, ambulating without assistance, and with full power in her four extremities.

Discussion

As ADEM is an autoimmune disease, immunosuppressive therapy is widely used as a first line management, including high dose steroid. Other treatment modalities include IVIG and plasmapheresis in patients who show poor response to steroid [2,5]. Severe forms of ADEM with a fatal outcome despite optimal medical therapy have been described. ADEM can rarely be complicated by massive brain edema, with life threatening increased ICP, and brain herniation that may not respond to the standard edema medical management (osmotherapy, hyperventilation, and sedation). The optimal management of such an extreme presentation is not yet clear.

Our patient had severe ADEM presentation according to the clinical and MRI findings. Her condition continued to deteriorate and she developed uncal herniation, despite optimal medical therapy with high dose steroid and plasmapheresis. Lifesaving decompressive hemicraniectomy was urgently performed to reduce the ICP, and the patient has fully recovered after that with no residual deficits. Surgical decompression for elevated ICP is a well-established neurosurgical intervention in other diseases like infarction, infection, and trauma [7]. In severe ADEM, such intervention was only reported as a successful therapy in a very few cases worldwide [3-6], and so long term outcome and quality of life of these patients remain a concern, especially if this intervention was performed in the dominant hemisphere of the brain.

This report emphasizes that aggressive intervention such as decompressive hemicraniectomy may be a lifesaving decision in some patients with severe ADEM who deteriorate despite standard immunosuppressive and anti-edema therapy, and this and support what was reported in other previous reports.

Conclusion

Severe ADEM disease complicated by significant brain edema and uncal herniation is an extremely rare presentation of this disease. Urgent and aggressive interventions, including decompressive craniectomy, can save lives and lead to favorable outcomes.

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