

The Importance of Early Recognition: A Case of ADEM in a Toddler Presenting with Altered Mental Status

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Introduction

- Acute disseminated encephalomyelitis (ADEM) is a poorly understood disease most often seen in pediatric patients.
- Etiology includes inflammatory demyelinating changes to the central nervous system, with encephalopathy and multifocal brain lesions, which are potentially triggered by a post-infectious response (1).

Case Presentation

- A previously healthy, 2-year-old male presented to an OSH with acute strabismus and ataxia and otherwise normal exam and VS.
- He was found to be positive for Streptococcus, treated with Penicillin G, and transferred for further workup of neurological symptoms.
- After transfer, he developed worsening ataxia, nystagmus, and progressive sleepiness. Head and neck CTA revealed no acute findings.
- He subsequently became lethargic with nocturnal episodes of bradycardia and was transferred to the PICU for worsening somnolence, development of non-tracking pinpoint pupils, and diffuse hypotonia.
- MRI revealed T2/FLAIR hyperintensities with faint enhancement of the central midbrain lesion, suggestive of ADEM (Figure 1).
- All pending infectious and autoimmune studies of the CSF were normal.
- After initial treatment with high dose steroids followed by IVIG, his AMS did not improve.
- Due to increasing agitation, Neurology escalated management to include plasmapheresis (PLEX).

Case Presentation continued

- His somnolence persisted despite several rounds of PLEX, requiring extended PLEX treatment.
- Repeat MRI (Figure 2) 8 days later showed a decrease in previous abnormalities.
- After 7 rounds of PLEX, his mental status began to slowly improve, though he was still hypotonic overall with an inability to perform prior ADLs.
- The patient was eventually deemed medically stable for transfer to our IPR service following 18 days of admission.
- He completed two weeks of inpatient rehab with significant improvement.

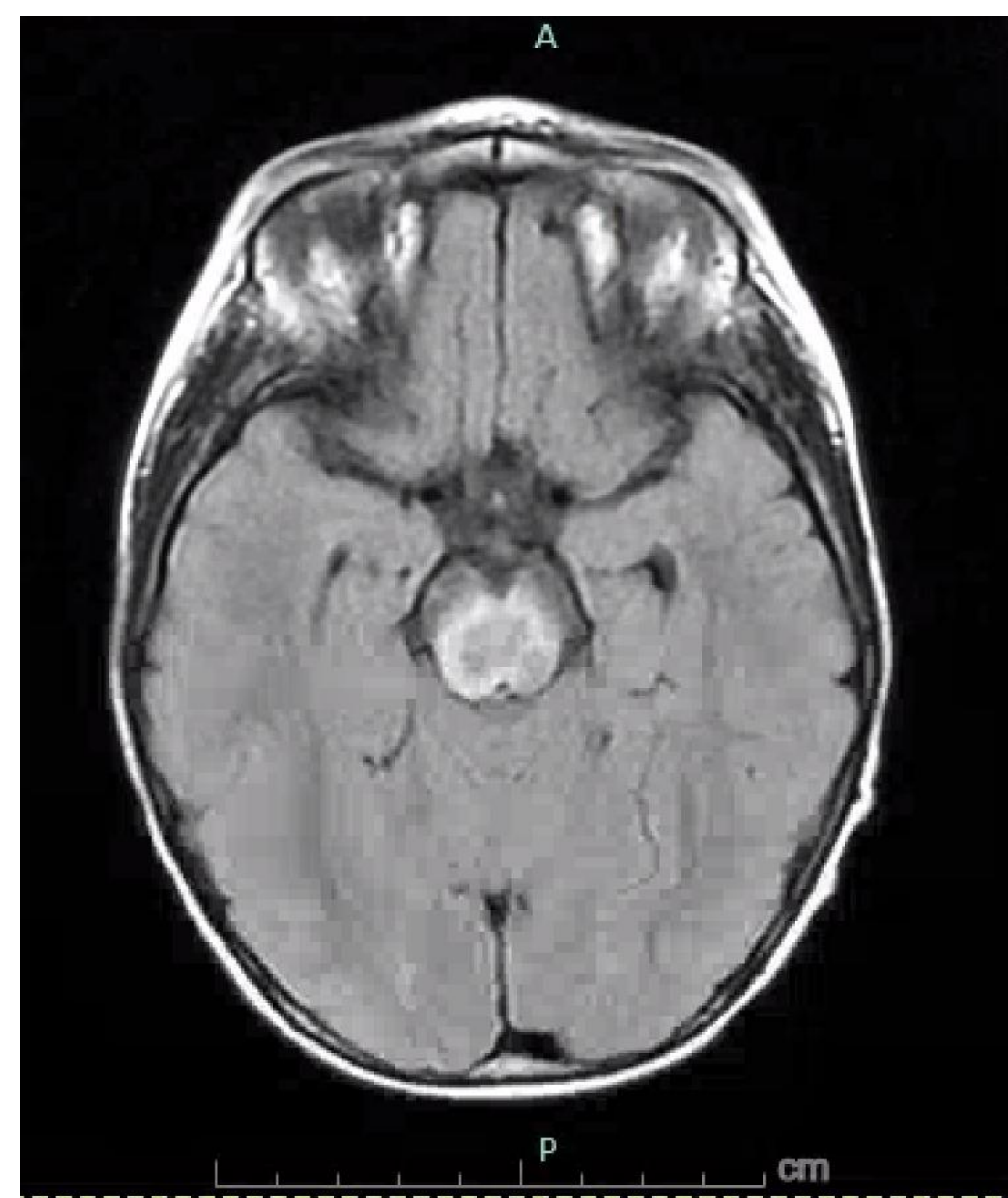


Figure 1. T2 FLAIR initial MRI.

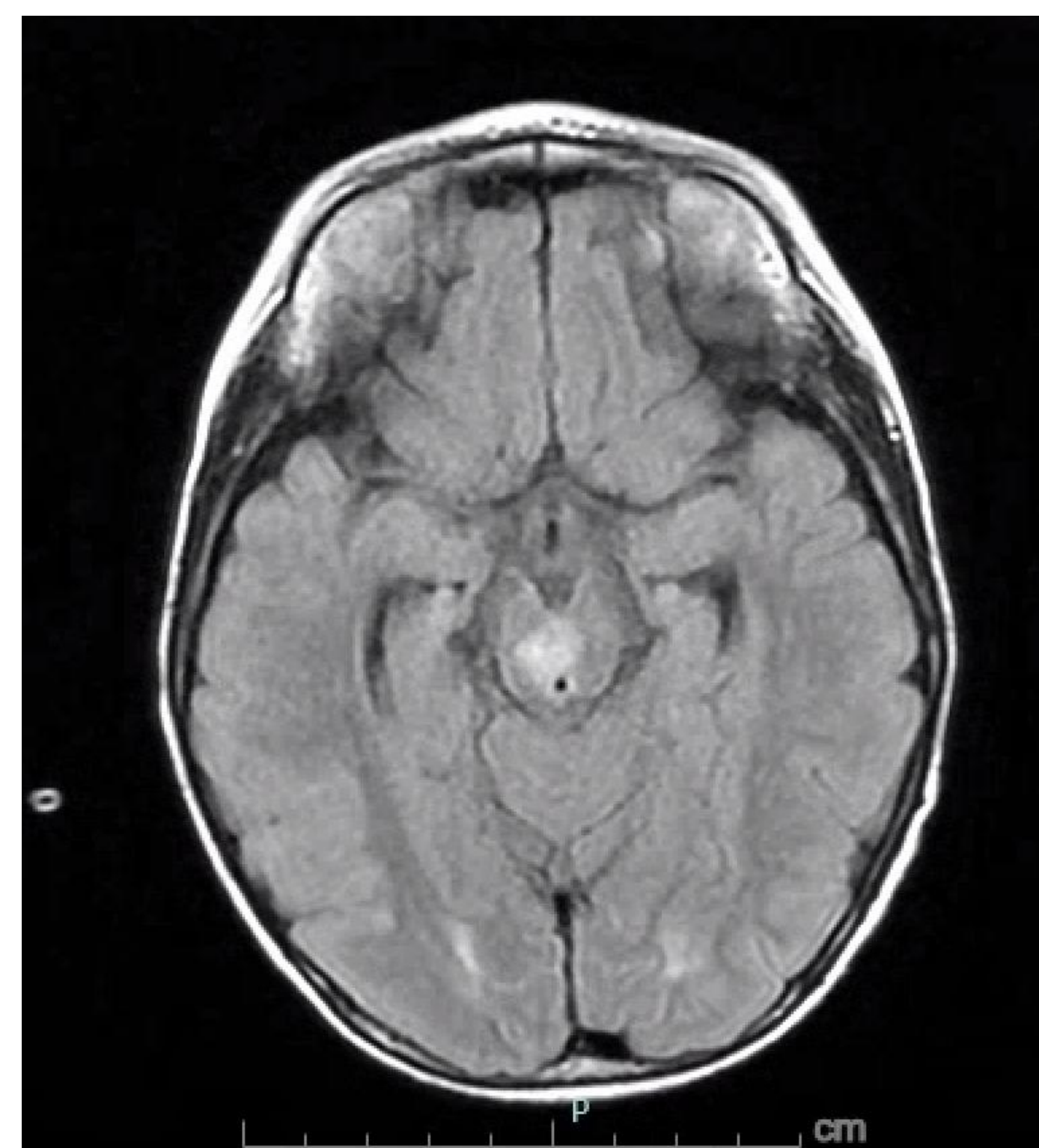


Figure 2. Repeat T2 FLAIR MRI.

Discussion

- This case highlights the importance of prompt diagnoses and treatment. This disease can possibly relapse and have long-term consequences with significant morbidity and poor neurologic outcomes (1,2).
- Younger patients (< 5yo) are particularly vulnerable to long term-effects of ADEM, such as severe behavioral and emotional problems (3).

Conclusion

- This unique case of ADEM required escalation to PLEX therapy, a refractory treatment for patients who fail to respond to steroids and IVIG (4,5).
- Because of the limited research on ADEM in younger pediatric patients, this case provides important clinical insight, particularly for younger patients who are refractory to initial treatments.
- Additionally, a clear infectious cause of this patient's condition was never fully identified.
- Though he was initially positive for Streptococcus A, this is an uncommon cause of ADEM.
- Because his positive result was identified at the time of his symptom onset, with swift treatment, the significance of this result is unclear.

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