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

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**Case:**

A previously healthy, 2-year-old male presented with strabismus and ataxia after waking from his nap. His vital signs were normal at an outside ER, where he was treated with penicillin G for Streptococcus A. After transfer, he developed worsening ataxia, nystagmus, and progressive sleepiness. A 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. His preliminary CSF findings were unremarkable. MRI revealed T2/FLAIR hyperintensities with faint enhancement of the central midbrain lesion, suggestive of ADEM. All pending infectious and autoimmune studies of the CSF were ultimately 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). His somnolence persisted despite several rounds of PLEX. He did not withdraw from pain and had poor visual tracking with bilateral ptosis. Repeat MRI showed a decrease in previous abnormalities. Due to a lack of clinical improvement, his PLEX therapy was extended.

After 7 rounds of PLEX, his mental status began to slowly improve. He had improved tone in his upper and lower extremities with occasional responsiveness to voice, though he was still hypotonic overall with an inability to perform prior ADLs. The patient was eventually deemed medically stable for transfer to our inpatient pediatric rehabilitation service following 18 days of admission. He completed two weeks of inpatient rehab with significant improvement.

**Discussion**:

ADEM is a poorly understood disease most often seen in pediatric patients. Its etiology includes inflammatory demyelinating changes to the central nervous system, with encephalopathy and multifocal brain lesions, potentially triggered by a post-infectious response (1). This case highlights the importance of prompt diagnoses and treatment. The 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). 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.

**References**:

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