Which “-itis” is it? A case of autoimmune encephalitis.

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Learning Objectives

1. Identify risks that lead to acute demyelinating encephalomyelitis (ADEM).
2. Differentiate ADEM from multiple sclerosis (MS) and infectious encephalitis.

Introduction

- ADEM is a rare autoimmune demyelinating process that affects the CNS.
- Onset of ADEM is acute and rapid in nature.
- ADEM often occurs post viral infection or post-vaccination, most often seen in children.
- Pathogenesis is believed to be linked due to an inappropriate immune cross reaction to myelin autoantigens.
- ADEM is difficult to diagnose, as it can masquerade its presentation similar to other demyelinating disorders, such as MS and infectious encephalitis.

Case Presentation

Initial Presentation

- 70-year-old woman with a past medical history of hyperlipidemia, HTN, and hypothyroidism
- Presented to the ED with a 12-hour history of aphasia and right facial droop
- She denied vision changes, hearing problems, dysphagia, or incontinence
- EMS reported the patient’s BP was 145/83mmHg and that she stopped her aspirin six months ago.
- Physical exam showed right-sided weakness, with the remainder of the exam unremarkable.
- A “brain attack” was called and a non-contrast head CT revealed no hemorrhage but demonstrated a hypodensity in the left frontal lobe.

Hospital Course

Follow-up MRI showed a large mass and white matter infiltration (Figure 1). The patient was admitted for further workup. SPECT MRI showed a demyelination process. Repeat LP showed resolution of the leukocytosis; samples were also sent for infectious workup that demonstrated negative results. Over time the patient improved on steroids and antibiotics. A discussion with the patient’s friends revealed she had a URI and received the influenza vaccine prior to her hospitalization. This information and demyelination on MRI led to a diagnosis of ADEM. The patient was continued on steroid therapy and discharged one week later following significant improvement. In summary, a 70-year-old female presented with multifocal neurological deficits and findings consistent to ADEM who demonstrated resolution over time.

Differentiating Etiologies of Encephalitis

ADEM	• Should be considered in patients with unexplained acute encephalopathy and multifocal neurologic signs • Encephalopathy is common with motor and sensory deficits, especially dysarthria, as seen in this patient • MRI and LP should be ordered to aid in diagnosis
MS	• Chronic autoimmune demyelinating disease • Follows the McDonald’s criteria • Plaques on MRI are characteristic findings • LP can show lymphocytic pleocytosis and oligoclonal bands
Infectious Encephalitis	• Infectious meningoencephalitis can have various causal organisms • Patients can often present with fever, headache, and meningismus • LP and CSF analysis would be useful, as cultures and PCR identify the pathogen

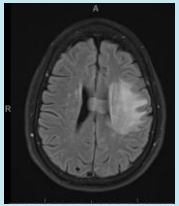

Figure 1. Follow-up T2 brain MRI demonstrating an 8.3 cm ill-defined lesion with FLAIR hyperintensity of the left frontoparietal lobe.

Conclusion

- This was an atypical presentation of ADEM in an elderly patient with recent URI and vaccination.
- Consider ADEM in patient presentations of acute encephalopathy.
- Ask patients or patient’s support regarding possible risk factors for ADEM, such as recent vaccination or viral illness.
- ADEM can present similarly to both MS and infectious encephalitis requiring a thorough history, physical, and diagnostic work-up.

References

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