Multiphasic Acute Disseminated Encephalomyelitis Associated with Metapneumovirus Infection in a 7-year-old boy.

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Introduction

ADEM is a rare illness; annual incidence being 0.2 to 0.5 per 100,000 children. It usually affects 3-7 year old children. Approximately 3 to 6 cases are reported in the United States annually. Associated pathogens include several viruses and bacteria: HIV, influenza, coxsackie, CMV, EBV, MMR, varicella etc.

Case Presentation:

7-Year-old male presented with headache, slurred speech, drooling, ataxia and ptosis of 3 days duration. MRI brain revealed new demyelinating lesions. CT Head, MRA brain and Spinal MRI were normal. Of Note; He recovered from meta-pneumovirus URI 3 weeks prior. He was admitted for multiphasic ADEM. Patient had first episode of ADEM 4 months ago when he presented with a 6-weeks of headache, ataxia and facial droop. MRI showed patchy bilateral demyelinating disease, he was treated with IV Steroids and IV-Ig. His S/S improved and was discharged on steroid taper with close neurology follow up. At 2 months f/u he was clinically back to baseline and repeat MRI showed complete resolution of abnormal findings. Extensive workup including inflammatory markers, MOG, NMOab, ANA, RF, Ach Receptor ab,GABA-R ab, NMDAab were unremarkable.

Discussion:

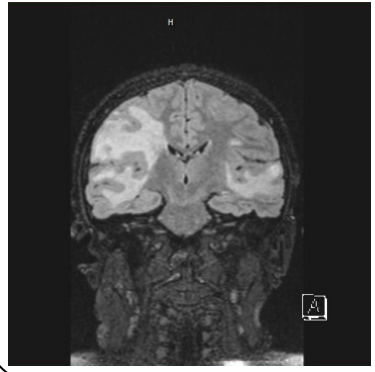
Pathogenesis of ADEM is not completely understood. It appears to be an autoimmune disorder of the central nervous system. It begins with activation of T cells by environmental triggers in genetically susceptible individuals, leading to production of cytokines and chemokines, TNF-alpha, complement activation, myelin phagocytosis, and oligodendrocyte apoptosis leading to demyelination.

Diagnosis: **Clinical Features** (all required for diagnosis):

1) A first polyfocal, clinical central nervous system event with presumed inflammatory demyelinating cause.
2) Encephalopathy that cannot be explained by fever, systemic illness, or postictal symptoms.
3) No new clinical and MRI findings emerge three months or more after the onset.
4) Brain MRI is abnormal during the acute (three-month) phase.

Lesion characteristics on brain MRI

1) Diffuse, poorly demarcated, large (>1 to 2 cm) lesions involving predominantly the cerebral white matter.
2) Deep gray matter lesions (eg, involving the basal ganglia or thalamus) can be present.
3) T1 hypointense lesions in the white matter are rare.



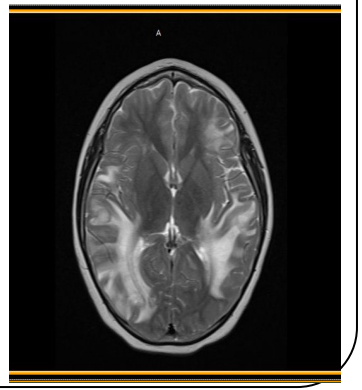


Figure 1 and 2: Show Diffuse grey and white matter lesions bilaterally, indicating demylating disease.

Treatment: Initial therapy with glucocorticoids rather than IVIG or plasma exchange is preferred; Frequent Assessment of response to initial therapy is the key in acute phase of disease.

Conclusion:There are no specific/confirmatory tests to establish the diagnosis of ADEM and it is considered a diagnosis of exclusion. Differentials that should be ruled out first include: Bacterial and viral meningitis or encephalitis, multiple sclerosis, NMOSD, and MOG Ab associated disease.

References

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