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# Lyme Encephalitis Complicated by Rapidly Progressive Acute Disseminated Encephalomyelitis (ADEM)

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## Background

- Lyme Disease is a multisystem illness caused by the spirochete Borrelia burgdorferi
- It is the most common vector born illness in the United States
- It can present as early localized, early disseminated, or chronic disease
- Early disseminated Lyme disease usually presents 3-10 weeks after inoculation with musculoskeletal and neurologic symptoms being the most common.
- Lymphocytic meningitis, cranial neuropathy, and radiculoneuritis constitute the triad of early neurologic Lyme disease.
- Nervous system involvement only occurs in the early disseminated phase.
- ADEM is an autoimmune demyelinating disease of the CNS commonly preceded by a viral infection or immunization.
- The incidence of ADEM is rare, with 0.8 cases per 100,000 population.
- Herein, we describe a rare case of Lyme encephalitis complicated by ADEM.

## Case Presentation

- A 55 year old immunocompetent Caucasian male presented with complaints of generalized head and neck pain, chills and photophobia for 4 days
- He denied any recent travel or infectious exposure.
- Physical exam showed meningeal signs with no neurological deficits or skin findings, and he was febrile, tachycardic, and had a leukocytosis
- CSF studies were consistent with bacterial meningitis, but CSF culture was negative.
- MRI displayed DWI in area of splenium and corpus callosum and FLAIR imaging revealed multiple lesions.
- Within 24 hours, he developed new-onset aphasia and became obtunded and required intubation with new imaging showing progressive cerebral edema.
- Seizure activity was ruled out with continuous EEG and patient was maintained on anti-epileptics.
- Vasculitis was also unlikely with nondiagnostic cerebral angiogram.
- Endocarditis was also unlikely with a negative transesophageal echo.
- Further diagnostic studies revealed positive Lyme antibodies, with a positive confirmatory western blot.
- Patient was continued on antibiotics and high dose steroids, however with minimal improvement and cerebral edema, a diagnosis of ADEM was made.
- Subsequent MRI’s done throughout his hospital stay remained unchanged, with no further improvement or new enhancing lesions.
- Plasmapheresis was initiated with a total of 5 treatments, which showed clinical and laboratory improvement.
- Due to persistent altered mental status and inability to protect airway, patient eventually required a tracheostomy and PEG tube placement prior to discharge for rehabilitation.
- Repeat LP done prior to discharge showed no WBC’s in the CSF.

Prior to discharge, the patient had completed 28 days of IV Rocephin, 7 days of 1 gram of Methylprednisolone, and was continued on Prednisone.

## Discussion

- At 2 months follow up, the patient and his family report continued progress. His tracheostomy has been reversed and he has been cleared for a diet.
- He is able to ambulate short distances with assistance and uses a wheelchair for long distances.
- He denies any pain or bladder/bowel dysfunction.
- He continues to have difficulty with visual spatial skills and tremors, and difficulty with sequencing, however overall has improved.

## Follow-up

- Acute disseminated encephalomyelitis (ADEM) is an autoimmune demyelinating disease of the CNS.
- It is commonly triggered by a preceding viral infection with acute onset and can be rapidly progressive.
- It is also thought to occur more commonly in children.
- Two theories are believed for the pathogenesis of ADEM, one being antigenic mimicry with antiviral antibodies cross reacting with myelin auto-antigens vs increased vascular permeability due to systemic inflammation.
- Common infectious pathogens thought to be associated with ADEM are MMR, Varicella, Smallpox, EBV, HSV, HHV-6, Influenza and HIV, although a cause is not always identified 25% of the time.
- Diagnosis of ADEM is a clinical diagnosis based on presentation, white matter lesions on MRI, and exclusion of alternative diagnosis.
- Neuroimaging with MRI usually reveals bilateral lesions, asymmetric, with lesions in the deep and subcortical white matter including corpus callosum, periventricular, and gray matter including the cortex, basal ganglia, and thalami.
- In ADEM, MRI lesions are hyperintense on T2 weighted and FLAIR sequences.
- Lumbar Puncture in ADEM typically reveals a lymphocytic pleocytosis and a mildly elevated CSF protein, and elevations in myelin basic protein.
- Differential diagnosis for ADEM includes MS, HSV encephalitis, Sarcoidosis, Vasculitis, PML.
- Lyme associated with ADEM has rarely been reported and is infrequent.
- Treatment is immunosuppression with high dose steroids, with alternative therapy consisting of IV Ig or plasmapheresis, although evidence based clinical trials of the treatment regimen are lacking.
- Clinical course is more severe in adults with less favorable outcomes.
- Complete recovery was reported in only approximately 33% of adult patients.
- Long term complications include a 35% chance of developing MS within 5 years.
- ADEM should be considered in patients with coma, CSF with protein and lymphocytes, negative CSF culture, and white matter lesions on MRI.

## References: