

Recurrent Disseminated Encephalomyelitis

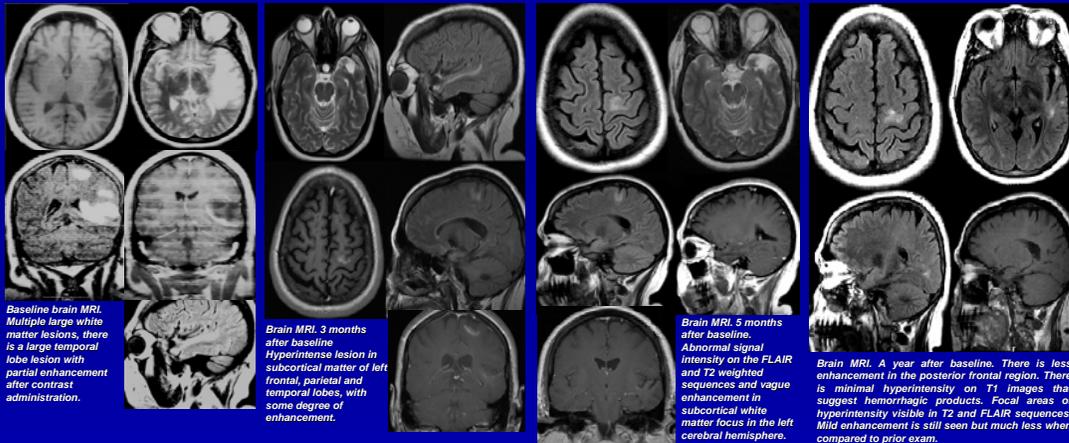
Report of 2 Cases

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Introduction: Acute disseminated encephalomyelitis is an inflammatory demyelinating disease with no clinical evidence based guidelines or biological markers for the diagnosis. The course is usually monophasic but rare cases of recurrence and multiphasic presentation have been documented. We describe the clinical and radiological features of 2 adult patients with Recurrent Disseminated Encephalomyelitis (RDEM) evaluated at our institution.

Case 1. 42 year old woman, developed distractibility, dull headaches, dysnomia and dysgraphia. Brain MRI showed several white matter hyperintense foci including a large left parieto-temporal lesion. Metastatic disease work up was all negative. Cerebrospinal fluid analysis showed P 43, WBC 2, RBC 0, 2 oligoclonal bands and normal IgG index. Brain MR spectroscopy performed 3 months after the initiation of symptoms was consistent with a demyelinating process. She improved after treatment with IV steroids. 3 months later she developed incoordination and marked dysarthria. A brain biopsy was performed. The pathology showed findings suggestive of a demyelinating process.

Case 2. 27 year old woman with headaches, nausea, vomiting and blurred vision. Brain MRI showed an enhancing dominant right basal ganglia lesion, extending to the caudate and cerebral peduncle, with other non enhancing lesions. CSF showed WBC 40, P 31, no oligoclonal bands, increased IgG index. She was hospitalized and treated with IV steroids with improvement of symptoms and resolution of the lesion enhancement on MRI. She was discharged from the hospital and remained asymptomatic. Four months later, she developed generalized tonic-clonic seizures and was admitted to the hospital where she developed sudden respiratory distress and required intubation. Her neurological exam showed bilateral tremor in upper extremities with no dysmetria and was otherwise normal. A new brain MRI was done which showed abnormality in right basal ganglia, thalamus, medulla which increased from the study done 3 months prior. During her hospitalization she developed anisocoria with dilation of left pupil that resolved. She received IV steroid treatment and recuperated.

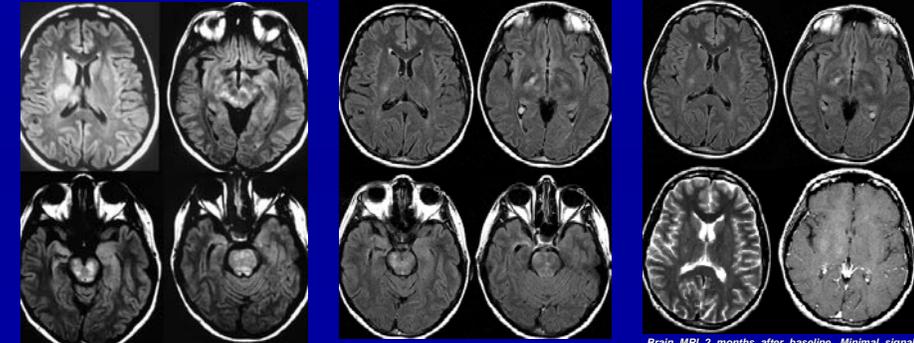


Baseline brain MRI. Multiple large white matter lesions, there is a large temporal lobe lesion with partial enhancement after contrast administration.

Brain MRI 3 months after baseline. Hyperintense lesion in subcortical matter of left frontal, parietal and temporal lobes, with some degree of enhancement.

Brain MRI 5 months after baseline. Abnormal signal intensity on the FLAIR and T2 weighted sequences and vague enhancement in subcortical white matter focus in the left cerebral hemisphere.

Brain MRI. A year after baseline. There is less enhancement in the posterior frontal region. There is minimal hyperintensity on T1 images that suggest hemorrhagic products. Focal areas of hyperintensity visible in T2 and FLAIR sequences. Mild enhancement is still seen but much less when compared to prior exam.



Brain MRI FLAIR at baseline. Hyperintense lesions in thalamus, basal ganglia, corona radiata, periventricular WM, cerebral peduncles, posterior limb of internal capsule, midbrain and medulla.

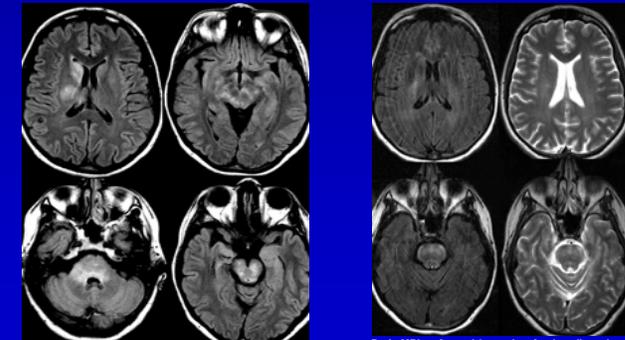
Brain MRI a month after baseline. Significant improvement in right basal ganglia, right temporal lobe and thalamus, compared with previous study. No focal abnormal enhancement or significant mass effect.

Brain MRI 2 months after baseline. Minimal signal abnormality and swelling associated with the left optic tract and scattered through the left basal nuclei and upper right brainstem, with minimal abnormal enhancement, improved compared to prior exam.

Conclusions: RDEM is defined as a new demyelinating event fulfilling diagnostic criteria for ADEM, occurring at least 3 months after the initial ADEM event and at least 4 weeks after completing steroid therapy, showing the same clinical presentation and affecting the same areas on MRI as the initial ADEM episode.

Multiphasic ADEM: Refers to one or more ADEM relapses, including encephalopathy and multifocal deficits, but involving new areas of the CNS on MRI and neurologic examination. Relapses take place at least 3 months after initial ADEM attack and at least 4 weeks after completing steroid therapy.

ADEM in its various presentations must be considered in the differential diagnosis of Multiple Sclerosis. Radiologic characteristics can help distinguish the two, including extent of the lesions, involvement of gray matter, preference for basal ganglia, thalamic and midbrain involvement. Early diagnosis and initial management may improve the patient's outcome. In addition, appropriate identification as ADEM will obviate the need for long-term immunomodulatory therapy.



Brain MRI performed 4 months after baseline. Signal abnormalities are seen in basal ganglia on the right hemisphere as well as thalamus and brain stem.

Brain MRI performed 6 months after baseline, shows abnormal signal intensity throughout the pons and portions of the midbrain, in the posterior limb of the internal capsule and the right frontal periventricular white matter.

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