

# Neuroimaging Highlight

Editors: William Hu, Mark Hudon

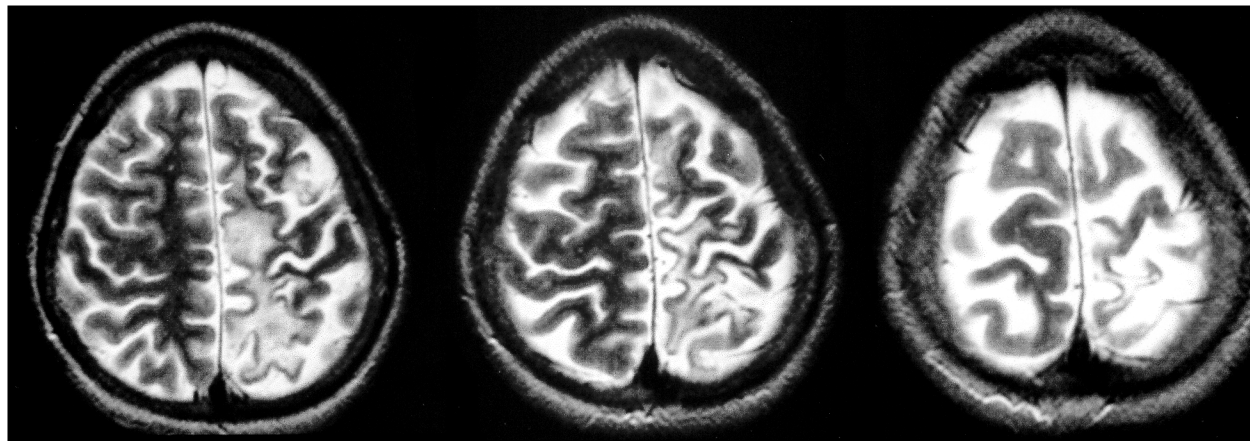
## Chronic (Rasmussen's) Encephalitis in an Adult

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A 24-year-old woman was referred because of incompletely-controlled complex partial seizures. Her seizures had started at age 21, after a mild head injury with brief loss of consciousness incurred in a biking accident, and were characterized by a sensation of bright flashing lights in the right visual field, followed by numbness and tingling in the right foot, spreading up the leg and to the arm, ultimately involving the entire right side, including the face. Occasionally they spread further to involve right facial twitching with jerking of the right arm and leg, loss of awareness and, at the onset of her epilepsy, rare secondarily generalized convulsions. Seizure frequency averaged three to four per month. She was initially treated with phenytoin and clobazam and subsequently changed to carbamazepine 800 milligrams per day. She also complained that her right side was no longer as strong as her left and that it was also numb, especially the leg, but felt that this weakness had stabilized or improved slightly over the past two years.

She had three brain MRI scans prior to this referral, reports of which described linear and curvilinear foci of increased T2 signal within the left temporoparietal and occipital white matter with some contrast enhancement. The reported radiographic differential diagnoses included demyelination, cortical dysplasia, atypical tuberosus sclerosis, neurosarcoidosis or gliosis related to inflammation, infection or trauma. Her neurological examination showed mildly increased tone and 4+/5 weakness in the right leg, a right pronator drift, right-sided hyperreflexia, right ankle clonus and a right Babinski sign. Sensation to pinprick was normal but joint position and vibration sensation were decreased on the right side. The findings appeared unchanged from those reported in a neurologist's examination three years earlier. Formal neuropsychological assessment revealed mild impairments in certain aspects of language processing (phonological and, to a lesser extent, lexical access) and in verbal working memory.

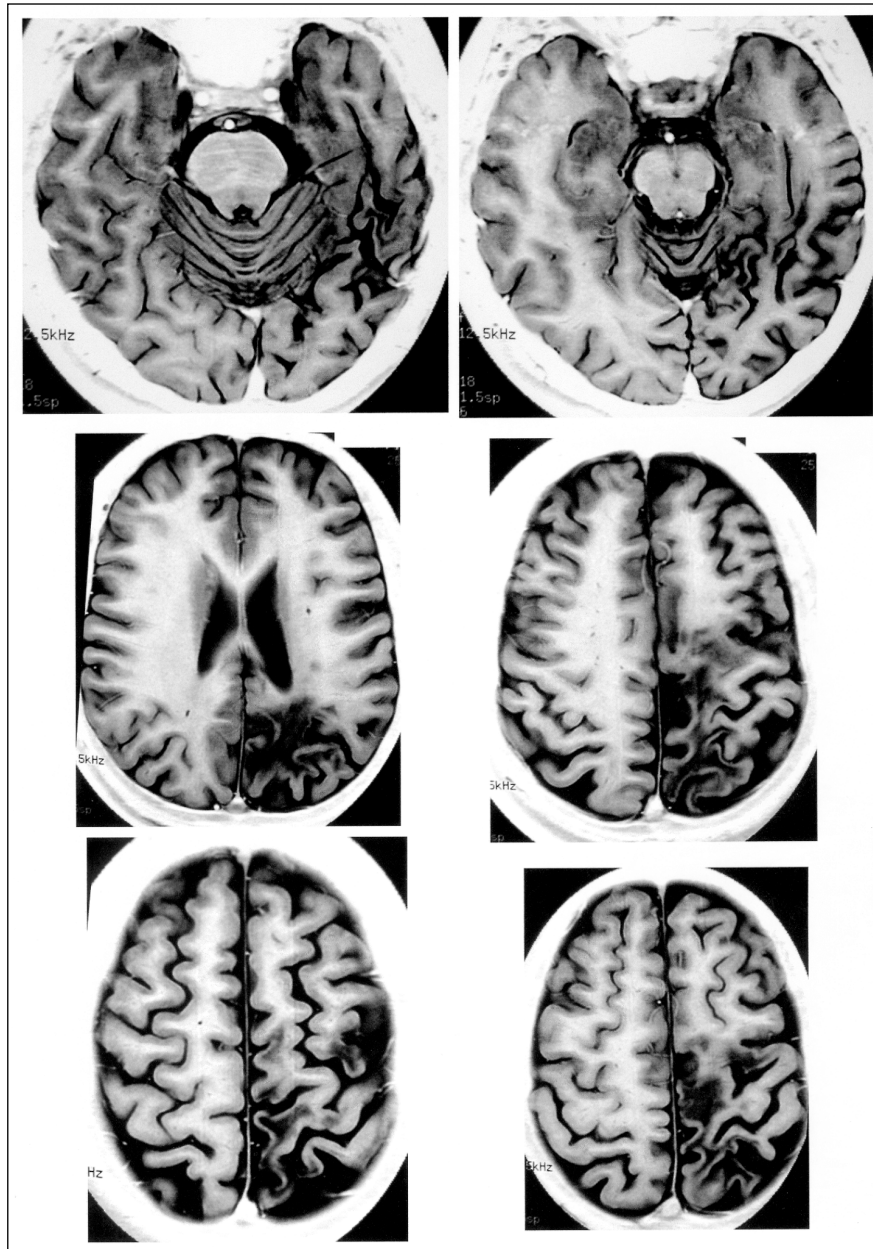


**Figure 1:** T2-weighted MRI showed increased signal through extensive region of left frontal and, especially, parietal and occipital lobes with associated parenchymal loss.

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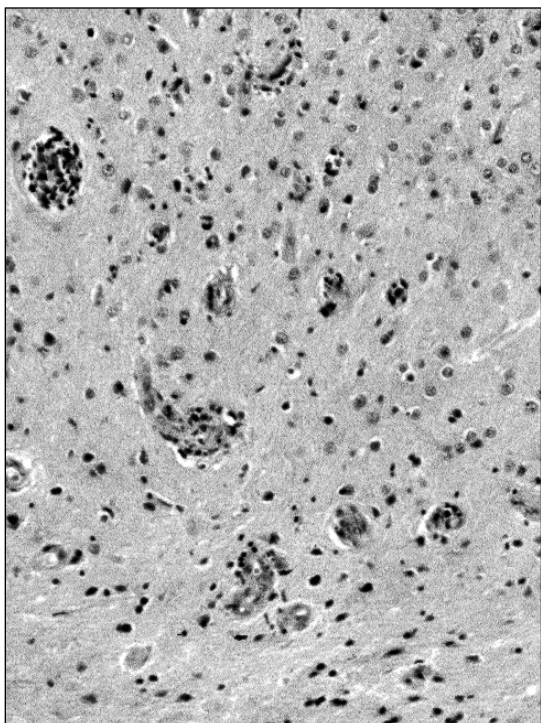
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**Figure 2:** Inversion recovery MRI demonstrated tissue loss in mesial and basal temporal gyri, and especially in parietal and occipital regions.

A repeat brain MRI and EEG were ordered. The EEG showed a mildly active interictal epileptiform abnormality over the left posterior temporal region. The MRI showed a large area of increased T2 signal intensity in the white matter of the left hemisphere, maximal in the parieto-occipital region with associated parenchymal loss and some extension of abnormal signal into the adjacent grey matter (Figure 1). Inversion recovery sequences demonstrated the extensive tissue loss in the parieto-occipital area as well as involvement of gyri in the left mesial and basal temporal lobe (Figure 2). The right hemisphere appeared normal.

A brain biopsy of the left parietal cortex and white matter was performed for diagnostic purposes, with the clinical and radiological suspicion of an atypical, adult onset chronic (Rasmussen's) encephalitis. Histological examination confirmed this, showing mild neuronal loss and gliosis in the grey matter with evidence of neuronophagia and few microglial nodules. Significant gliosis and rarefaction was evident in the adjacent white matter. Perivascular lymphocytic infiltrations (predominantly immunoreactive for CD3 T-cell marker) were evident in both grey and white matter. Focal collections of CD68 positive macrophages were present within the white matter.



**Figure 3:** Histopathology of biopsy specimen demonstrates mild neuronal loss and perivascular lymphocytic infiltrates in the cortex. The underlying white matter shows mild gliosis. H&E stain, 80X.

There were no viral inclusions and immunohistochemistry was negative for CMV, EBV, HSV-1 and 2 (Figure 3).

The patient has remained stable for over five years with no change in her seizure frequency, neurological examination or neuropsychological performance. Control MRIs have shown no interval changes over that time (Figure 4).

Rasmussen's encephalitis is typically a relentlessly progressive disease of childhood characterized by severe intractable epilepsy, intellectual deterioration and hemiparesis.<sup>1</sup>

However, rare patients with onset in adulthood have been described, occasionally noted to have a less severe course and a less severe epilepsy, often with a predominantly posterior localization.<sup>2</sup> As in this case, a head injury has often been remembered to precede the subsequent onset of the illness, although this remains an unproven etiologic link. Glutamate receptor GluR3 autoantibodies have been found in some, but not all, patients with proven Rasmussen's encephalitis, supporting the long-standing opinion that the disease has an immunological basis, possibly initiated by a focal insult such as head injury or repeated seizures, leading to breakdown of the blood brain barrier and initiation of an immune-mediated cascade of tissue injury leading to further seizures, further breakdown of the blood brain barrier, and so on.<sup>3</sup> The focally-initiated immune cascade hypothesis is offered as a possible explanation for the unilaterality seen in almost all cases of Rasmussen's encephalitis.

Transient benefits in progressive symptoms or seizure control after treatment with plasmapheresis or intravenous immunoglobulin therapy have been described in some patients with Rasmussen's encephalitis, irrespective of the presence or absence of measurable GluR3 antibodies.<sup>3-5</sup> Given this patient's clinical stability, and in accordance with her desires, antibody testing has not been performed and immunological treatments have not been given.

#### References

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**Figure 4:** Control MRI five years later shows no interval change. Biopsy tract and skull defect visible in superior parietal area.