Progressive multifocal leukoencephalopathy

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A 58-year-old woman admitted to the hospital had a 3-month history of progressive neurologic deterioration characterized by severe global aphasia and bilateral limb weakness, greater on the right. The patient had a 10-year history of systemic lupus erythematosus (SLE). Polyarthritis had been present for 6 weeks before the onset of neurologic symptoms; she was treated with prednisone and methotrexate. A diagnosis of progressive multifocal leukoencephalopathy (PML) was established by a positive PCR for JC virus from CSF. The patient deteriorated and further active care was withdrawn.

Serial MRI showed an enlarging area of hypointensity on T1 and hyperintensity on T2-weighted images in the left parietal lobe. The lesion involved predominantly white matter and was without mass effect. A rim of high signal was present at the margins of the left parietal lesion on the diffusion-weighted image (figure) with a reduced apparent diffusion coefficient (ADC) on the ADC map. There was no enhancement after administration of IV gadolinium.

PML in SLE treated with immunosuppression is uncommon. The MRI findings in PML often include circumscribed lesions predominantly in white matter without mass effect or contrast enhancement. The MR sequences indicate a progressing area of cytotoxic edema surrounding prior areas of white matter damage.

Figure. Diffusion weighted imaging (echoplanar, repetition time = 5100 milliseconds, echo time = 137 milliseconds, $b = 1000 T$) showing a rim of high signal surrounding a lesion that does not demonstrate signal change or mass effect.
