Progressive Multifocal Leukoencephalopathy Mimicking Central Nervous System Lupus

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Progressive multifocal leukoencephalopathy (PML) is a progressive demyelinating disease of the central nervous system (CNS) caused almost exclusively by JC virus. This opportunistic infection results in large multifocal areas of demyelination and must be strongly considered in immunosuppressed patients with new-onset neurological changes[1].

We describe a 47-year-old woman with systemic lupus erythematosus manifested by polyarthritis, Raynaud's phenomenon, autoimmune hemolytic anemia, positive antinuclear antibodies, dsDNA, and anticardiolipin antibody who developed acute mental status changes. Lupus cerebritis had been recently diagnosed and successfully treated with corticosteroids and monthly pulse intravenous cyclophosphamide. She was now uncooperative, somnolent, perseverating with asymmetric spontaneous limb movements, hyper-reflexia, with positive Babinski's sign and clonus. Laboratory results showed white blood cell count 1900/mm^3, absolute neutrophil count 1300, erythrocyte sedimentation rate 35 mm/h, creatinine 0.4 mg/dl, C3 complement 65 mg/dl (normal 90–180 mg/dl), C4 complement 8 mg/dl (10–40 mg/dl). Cerebrospinal fluid was colorless and clear, glucose 37 mg/dl (40–70 mg/dl), protein 104 mg/dl (15–45 mg/dl), white blood cells 40/mm^3 (0–6/mm^3), granulocytes 31%, lymphocytes 6%, and monocytes 63%. Cultures showed no growth. Pulse intravenous corticosteroids for lupus cerebritis were initiated, but she did not improve. Subsequent magnetic resonance imaging (MRI) of the head showed multiple non-enhancing white matter areas of demyelination, grossly different from her prior MRI showing T2 signal lesions in the white matter more consistent with Lupus cerebritis (Figure 1). Intravenous cidofovir was started empirically for PML; she died shortly thereafter. Post-mortem biopsy was consistent with PML.

In the 1950s PML was first described as a complication in lymphoproliferative and myeloproliferative disorders. PML became well known with human immunodeficiency virus in the 1980s. Cases associated with autoimmune disease were first reported in the 1970s[2]. All patients were immunosuppressed when diagnosed. Although laboratory results are sim-

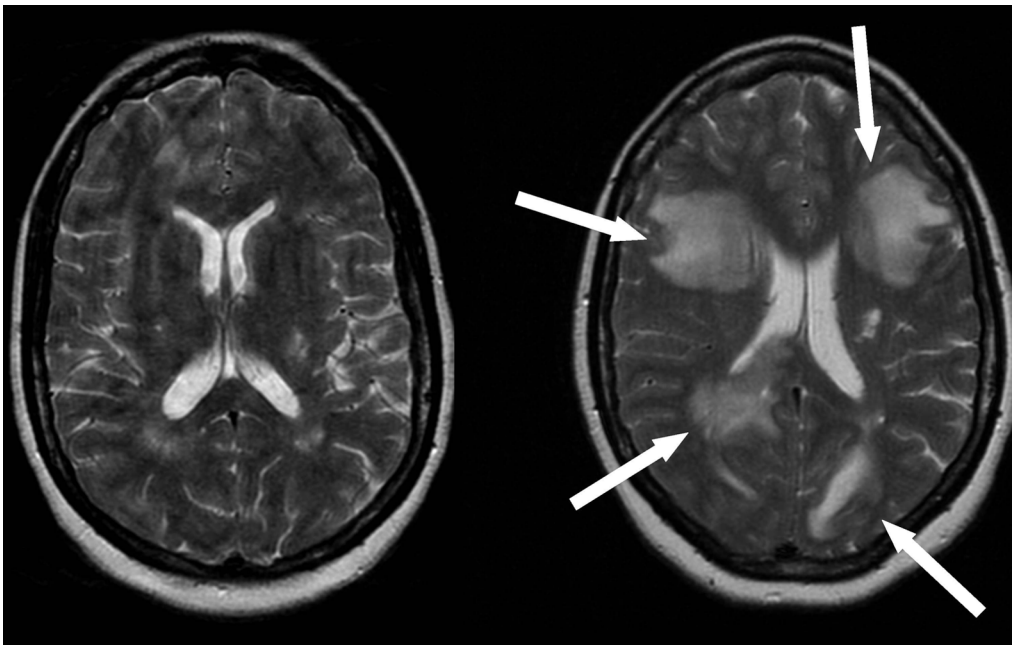

Figure 1. MRI (right panel) showing multiple non-enhancing white matter areas of demyelination (arrows) consistent with progressive multifocal leukoencephalopathy, grossly different from the previous MRI (left panel) showing T2 signal lesions in the white matter more consistent with lupus cerebritis.

ilar to those of CNS lupus, magnetic resonance imaging results reveal dramatic differences. PML demonstrates non-enhancing white matter lesions consistent with leukoencephalopathy; lupus cerebritis does not[3].

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