On-line Table 1: Differential diagnosis of bilateral symmetric deep gray T2 hyperintensity[a]

Differential Diagnosis
Hypoxic-ischemic encephalopathy
Hypoglycemic encephalopathy
Viral encephalitis (especially flaviviruses: Japanese, West Nile, Murray Valley, and so forth)
Acute hyperammonemia (ie, acetaminophen [Tylenol] toxicity, acute decompensated liver failure)
Toxins (carbon monoxide, methanol, cyanide)
Creutzfeldt-Jacob disease (especially with restricted diffusion)
Venous thrombosis (ie, internal cerebral vein thrombosis)
Wernicke encephalopathy (thiamine deficiency)
Osmotic myelinolysis (ie, rapid correction of hyponatremia)
Posterior reversible encephalopathy syndrome (ie, central variant)
Dilated Virchow-Robin spaces (look for central FLAIR suppression)
Nonketotic hyperglycemia (diabetes mellitus; may have hemichorea-hemiballismus)
Lymphoma (especially with postcontrast enhancement; can be primary or secondary)
Primary brain neoplasm (ie, bithalmic glioma, gliomatosis cerebri)
Metastatic disease (especially with post-contrast enhancement and history of cancer)
Mitochondrial disorders (Leigh syndrome, MELAS > MERFF)
Wilson disease (especially with T1 hyperintensity)
Behçet disease (especially with oral/genital ulcers and visual disturbances)
Toxoplasmosis (immunocompromised patients such as HIV/AIDS)
Huntington disease (especially with family history and caudate abnormalities)
Fahr syndrome (look for evidence of calcification on MRI or CT)
Basilar artery thrombosis (unlikely to be a diagnostic dilemma on CTA/MRA)
Occlusion of artery of Percheron (ie, bilateral thalamic infarcts)

Note:—MELAS indicates mitochondrial encephalomyopathy, lactic acidosis, and strokelike episodes; MERFF, myoclonic epilepsy with ragged-red fibers.

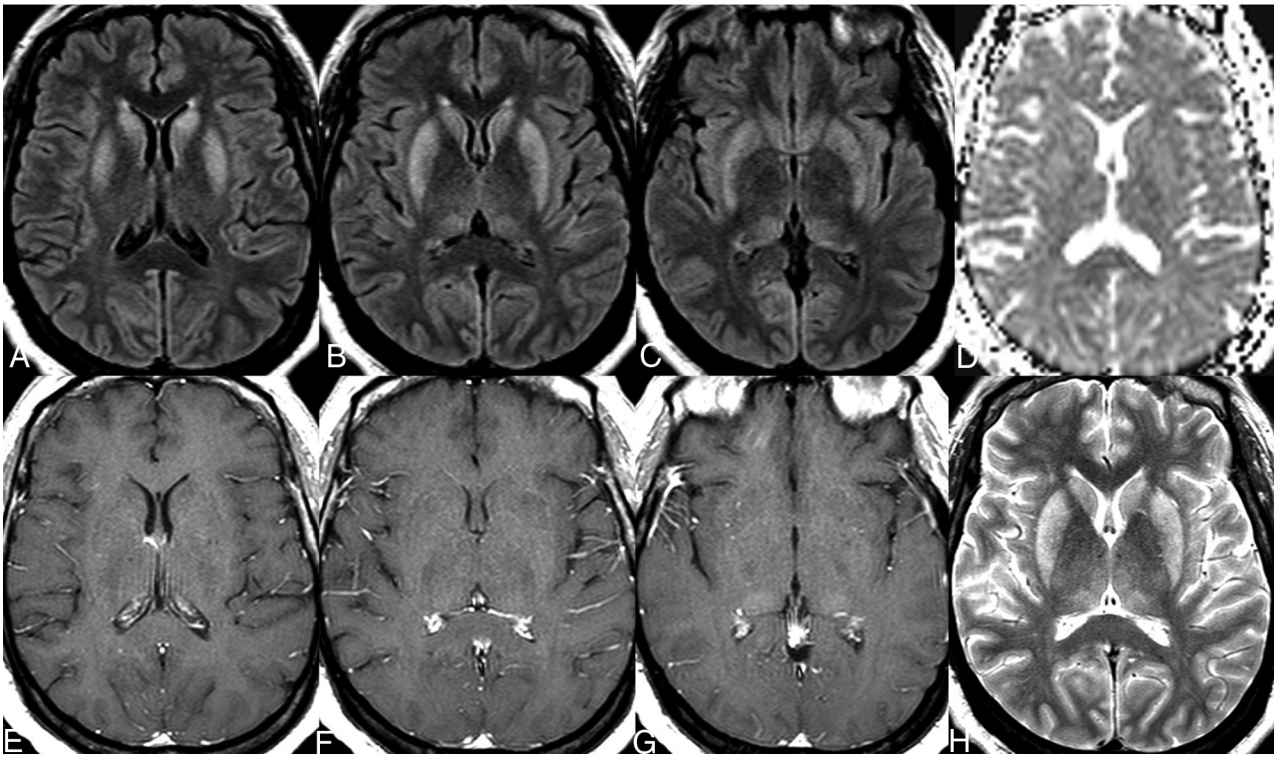
[a] Note that most of the entities on this list are ruled out during the clinical work-up of patients who are later given a diagnosis of autoimmune striatal encephalitis. Antibody testing in not always performed in these patients, or the selected antibodies that are tested may have negative findings, but the diagnosis can be established on clinical grounds in the absence of a more probable etiology and is supported by a positive clinical and imaging response to plasmapheresis. Also note that a pre-existing autoimmune disease such as lupus may be present and should increase the clinical suspicion for antibody-mediated disease in the setting of bilateral symmetric deep gray T2 hyperintensity on MRI brain examinations in these patients.

On-line Table 2: Clinical and laboratory tests in suspected autoimmune striatal encephalitis[a]

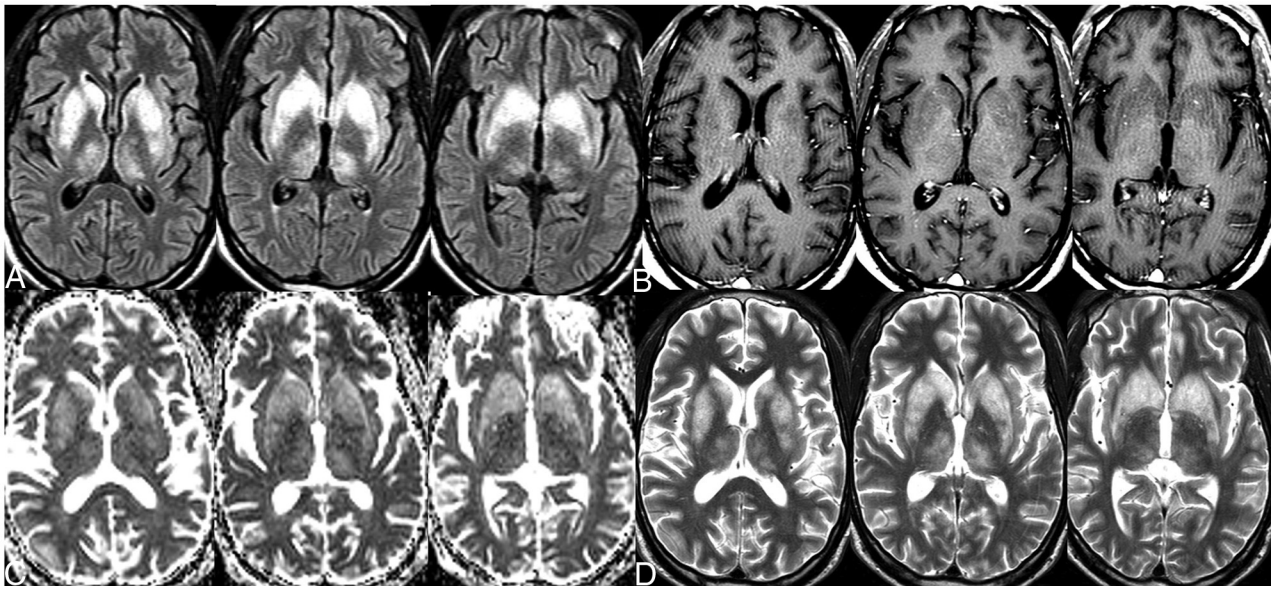
Clinical and Laboratory Tests
Clinical
Basic vitals: temperature, heart rate, blood pressure, respiratory rate, O^2 saturation
Clinical assessment: Glasgow Coma Scale, NIHSS
Serum
Basic labs: CBC w/diff, electrolytes, creatinine and glucose levels, erythrocyte sedimentation rate, C-reactive protein, urinalysis
Liver function tests, ammonia, acetaminophen, Vitamin B_1, B_{12}, cortisol, thyroid function
Coagulation panel, Gram stain, blood culture (bacterial, fungal), peripheral blood smear
Toxicology screen, CO, lead, mercury, copper, manganese, arsenic, methanol, cyanide
Hepatitis panel, HIV, mycoplasma, mononucleosis, aspergillus, other viral (see CSF)
Compliment (C3, C4), *HLA-B51*, ACE, SPEP, serum-free light chains, cryoglobulins
CSF
Physical characteristics: appearance, opening pressure
Basic labs: RBCs, WBCs w/diff, protein, glucose, cytology, Gram stain, and culture
Oligoclonal bands, IgG, myelin basic protein, C-reactive protein, lactate dehydrogenase
Cryptococcus, toxoplasmosis, pneumocystis, acid fast bacilli, syphilis (VDRL)
Virology: herpes simplex, varicella zoster, cytomegalovirus, Epstein-Barr, West Nile, influenza, coxsackievirus, parvovirus, JC virus
Autoimmune antibodies (serum and/or CSF)
Liver panel: mitochondrial, smooth-muscle, and liver-kidney microsomal
Antinuclear antibodies: dsDNA, SSA/Ro, SSB/La, Sm, nRNP, Scl-70
RF, CCP, C-ANCA, P-ANCA, cardiolipin, β2 glycoprotein (antiphospholipid)
Other: antithyroid antibodies, myositis-specific antibodies ($\pm$ muscle biopsy)
Autoimmune encephalitis antibodies associated with striatal involvement: NMDAR (NR1, NR2), Dr2, HU, LGI1.

Note:—CBC indicates complete blood count; w/diff, with differential; CO, carbon monoxide; ACE, angiotensin-converting enzyme; SPEP, serum protein electrophoresis; RBC, red blood cell; WBC, white blood cell; IgG, immunoglobulin G; VDRL, Venereal Disease Research Laboratory test; SSA/Ro, Sjogren syndrome type A antigen/Lupus Ro protein; SSB/La, Sjogren syndrome type B antigen/Lupus La protein; Sm, anti-Smith antibody; nRNP, nuclear ribonucleoprotein; Scl-70, antitopoisomerase I; CCP, anticyclic citrullinated peptide; RF, rheumatoid factor; C-ANCA, cytoplasmic antineutrophil cytoplasmic antibodies; P-ANCA, perinuclear antineutrophil cytoplasmic antibodies; Dr2, anti-Dopamine D2 receptor antibody; HU, hemolytic uremic syndrome; LGI1, leucine-rich, glioma inactivated 1; JC, John Cunningham.

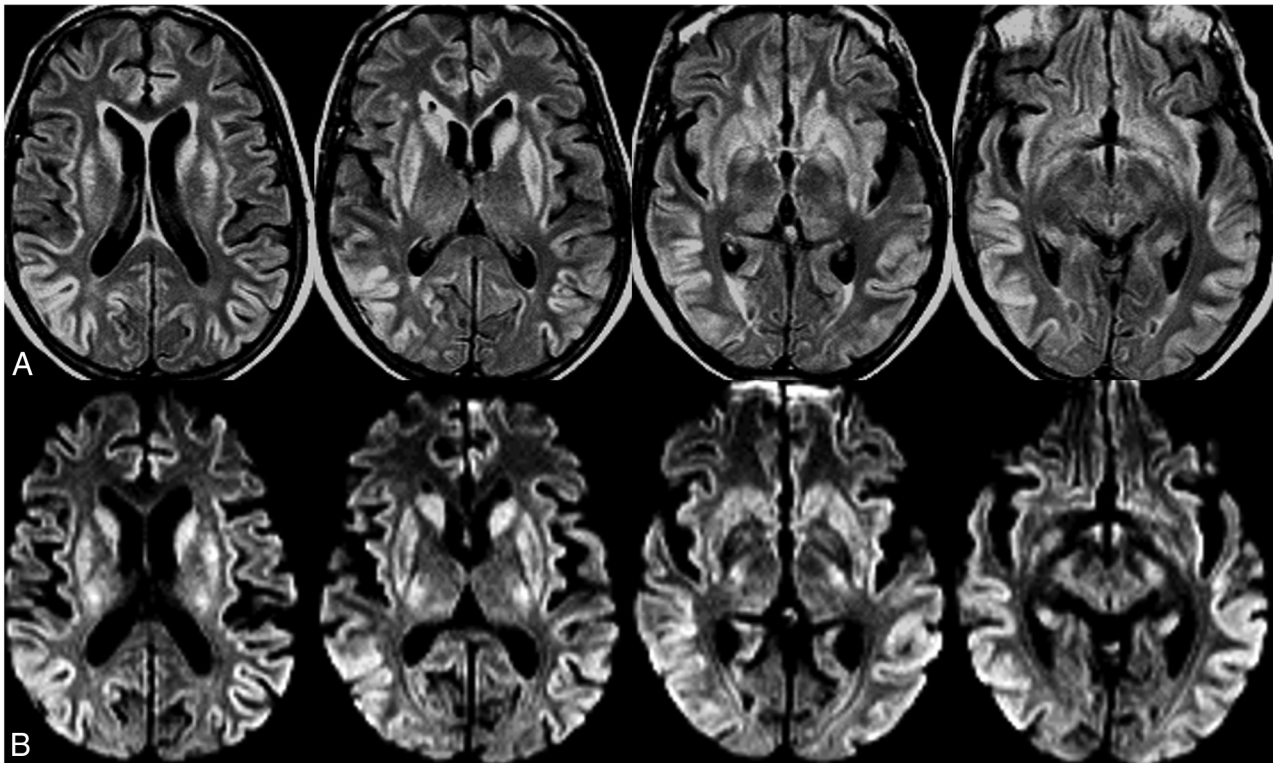
[a] In the age of electronic medical records, radiologists have full access to clinical data and can use this information to provide a more focused and appropriate interpretation of imaging findings. Note that not all the listed tests need to be performed on every patient, but review of the medical record for clinical context and relevant test results can help exclude important diagnostic considerations such as hypoglycemia, a hypoxic-ischemic event, carbon monoxide poisoning, and acute liver failure in the setting of bilateral symmetric T2/FLAIR hyperintense signal changes within the caudate and putamen on MRI brain examinations. Also note that in addition to these toxometabolic etiologies, atypical viral or fungal infections represent another major consideration and likely require lumbar puncture for further evaluation.



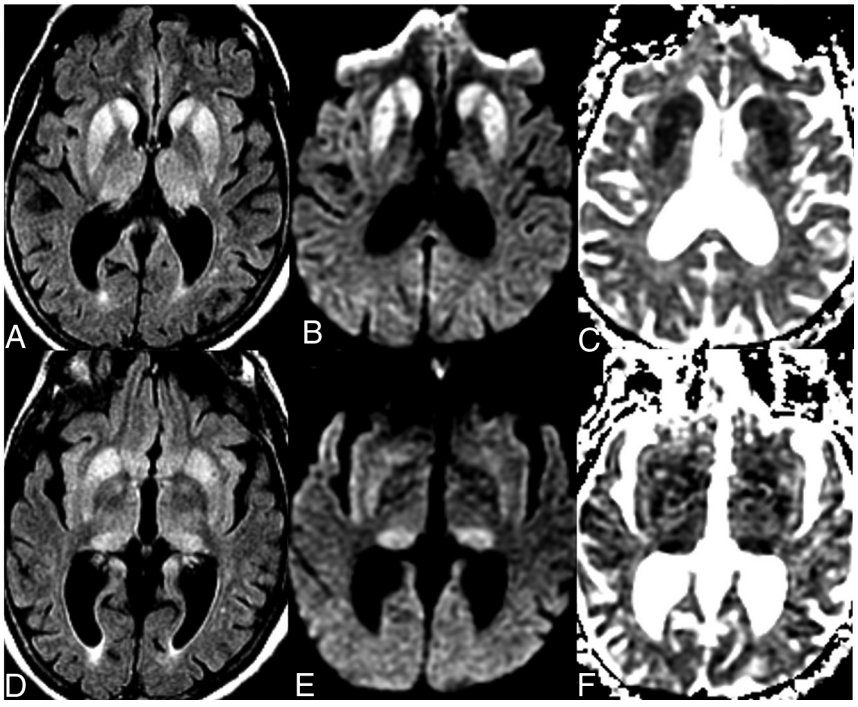
ON-LINE FIG 1. Anoxic brain injury 10 days after pulseless electrical activity (PEA), now with myoclonic jerks. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity of the dorsal striatum (caudate and lentiform nucleus) and thalamus, in addition to subtle gyriform T2/FLAIR hyperintensity throughout the cerebral cortex (*A–C* and *H*), without restricted diffusion (*D*) or postcontrast enhancement (*E–G*). Note the intrinsic T1 hyperintensity of the bilateral thalami (*E–G*), which was present on the precontrast sequence and suggests the development of coagulative necrosis.



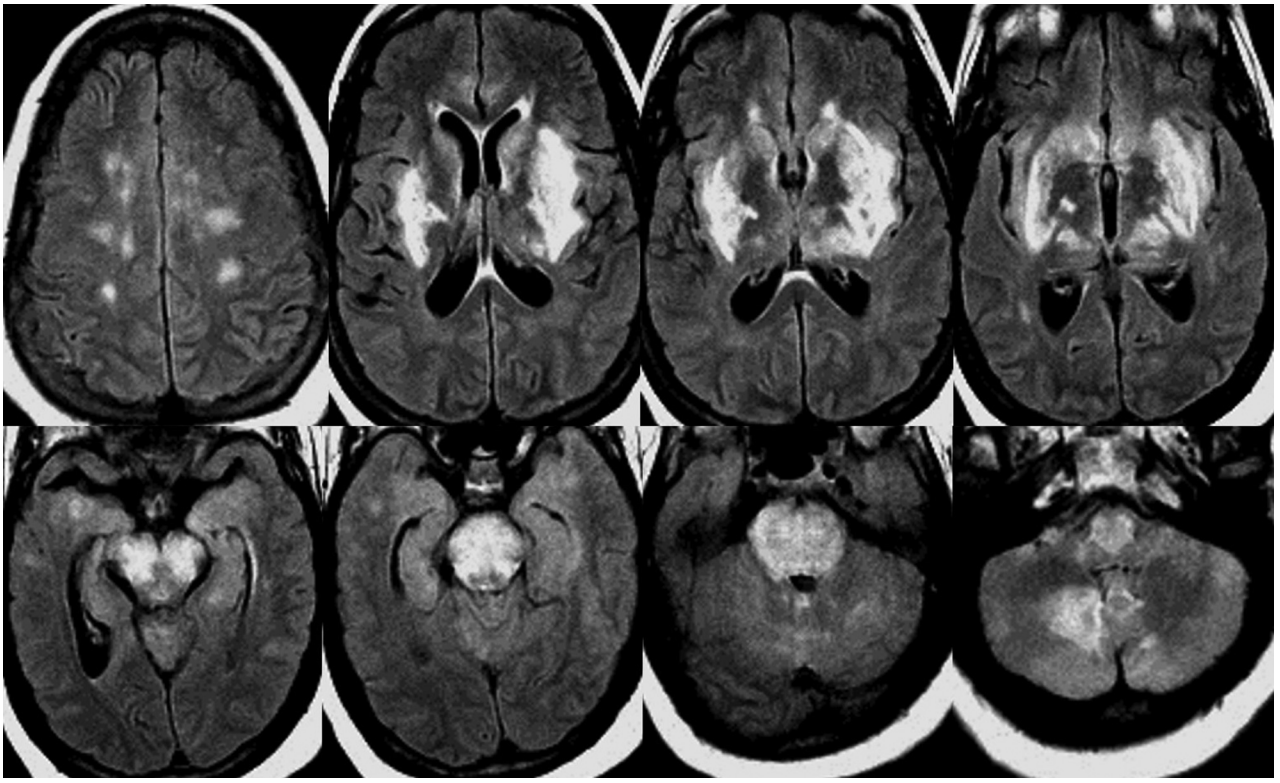
ON-LINE FIG 2. Acute hypoglycemia. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity of the dorsal striatum (caudate and lentiform nucleus) and thalami (*A* and *D*), in addition to subtle gyriform T2/FLAIR hyperintensity throughout the cerebral cortex (*A*), without restricted diffusion (*C*) or postcontrast enhancement (*B*).



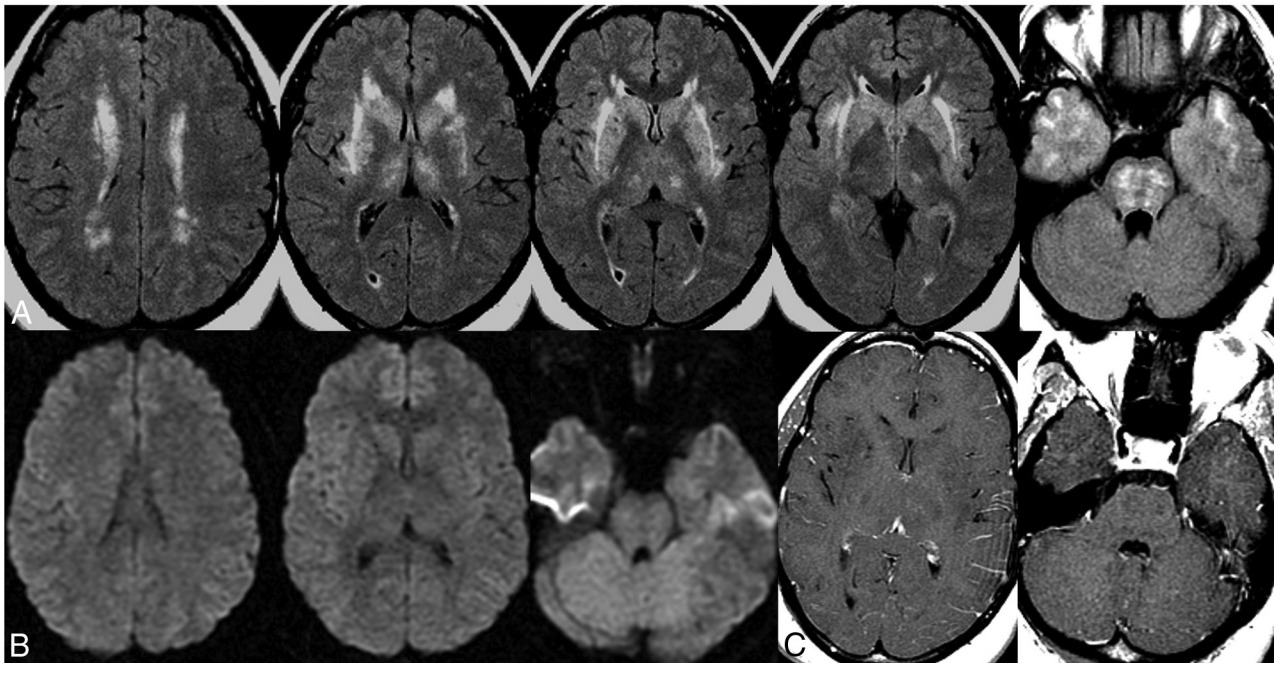
ON-LINE FIG 3. Prolonged status epilepticus. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity of the dorsal striatum (caudate and lentiform nucleus), thalami, and temporoparietal cortex (*A*) with associated restricted diffusion (*B*).



ON-LINE FIG 4. Creutzfeldt-Jacob disease. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity of the basal ganglia and posterior thalamus (pulvinar) (*A* and *D*) with associated high signal on $b=1000$ diffusion-weighted imaging (*B* and *E*) and dramatically reduced ADC values in these regions (*C* and *F*), consistent with intense restricted diffusion.



ON-LINE FIG 5. Central variant of acute hypertensive encephalopathy, also known as a central variant of posterior reversible encephalopathy syndrome. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity of the supratentorial and infratentorial white matter extending from the subcortical white matter into the deep white matter surrounding the basal ganglia with inferior extension along the corticospinal tracts into the midbrain, pons, and medulla. Note the additional patchy T2/FLAIR hyperintense signal changes within the basal ganglia, thalami, and cerebellum. These regions of T2/FLAIR signal abnormality did not demonstrate restricted diffusion or postcontrast enhancement.



ON-LINE FIG 6. Multiple sclerosis. MR imaging of the brain demonstrates bilateral symmetric T2/FLAIR hyperintensity in the periventricular white matter extending through the external/extreme capsules into the subinsular white matter, with additional involvement of the subcortical white matter of the temporal lobes and within the central pons, in addition to gray matter T2/FLAIR hyperintensity of the basal ganglia and thalami (*A*) without restricted diffusion (*B*) or postcontrast enhancement (*C*).