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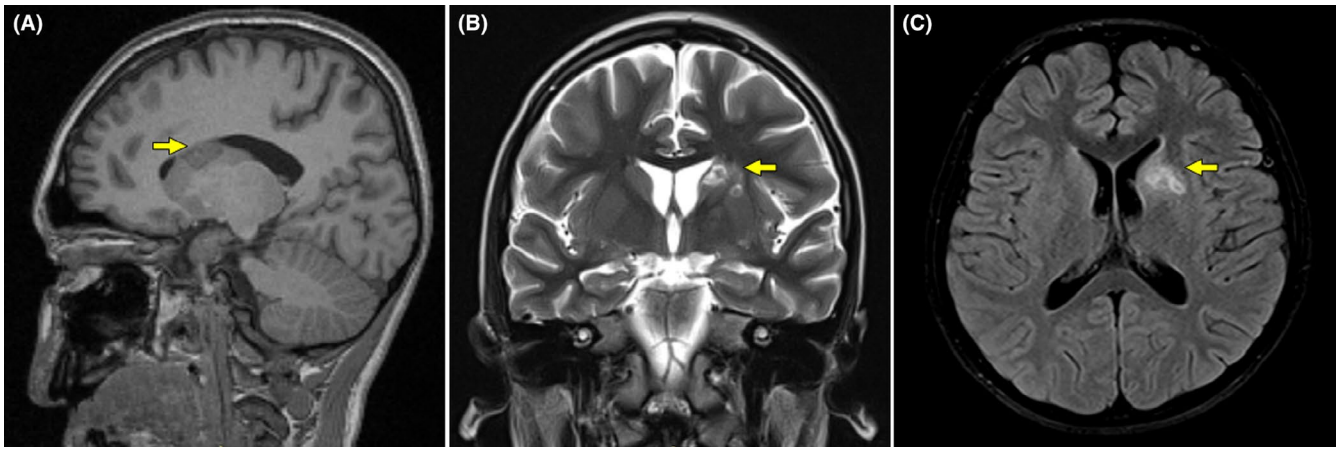
Clinical Images: Chorea in systemic lupus erythematosus: abnormal movements and ischemic lesions on magnetic resonance imaging



The patient, a 13-year-old boy, presented with a 3-day history of involuntary, stereotyped, purposeless movements that interfered with sleep and right upper extremity weakness. Three months before, he had been diagnosed with systemic lupus erythematosus (SLE) characterized by arthralgia, oral ulcers, and a malar rash, with antinuclear antibody titers of 1:160 (homogeneous pattern), negative results on the extractable nuclear antigen panel, anti–double-stranded DNA (anti-dsDNA) titers of 1:2560, and hypocomplementemia, for which prednisolone and chloroquine had been started. On admission, he had monoparesis of the right upper limb and chorea that prevented walking. There was no ocular or cranial nerve involvement, and sensory examination did not reveal any abnormalities. Clinical signs, including darting tongue, spooning, touchdown (Supplementary Video 1), and milkmaid grip, were present (Supplementary Video 2), confirming a diagnosis of chorea. Laboratory studies showed leukopenia, hematuria, and pyuria, without proteinuria. Findings of the transthoracic echocardiogram and cerebrospinal fluid were normal. Antiphospholipid antibodies (aPLs) were negative. Magnetic resonance imaging (MRI) showed subacute infarcts in the left caudate head/body junction (sagittal T1-weighted and coronal T2-weighted MRI, arrows) (**A and B**) and putamen (axial fluid-attenuated inversion recovery MRI, arrow) (**C**). Magnetic resonance angiography demonstrated absence of vasculitis. The renal biopsy showed class III lupus nephritis. Treatment with intravenous methylprednisolone and cyclophosphamide was started, and recovery from chorea was seen at the 6-month follow-up. Chorea is rare (0.7%-5%) in patients with SLE and is associated with aPLs and high disease activity (1). The classic signs of chorea in pediatric patients must guide the search for SLE and antiphospholipid syndrome, in addition to infectious, vascular, endocrine, or drug-related causes (2). In our case, the most likely etiology of chorea was a direct neuronal lesion in the basal ganglia mediated by antibodies because no stenosis, thrombosis, or vasculitis was observed on MRI and aPLs were negative. The high titers of anti-dsDNA and the favorable response to immunosuppression also support this hypothesis.

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