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Hyperintense Basal Ganglia on T1-Weighted MR Images in a Patient with Central Nervous System Lupus and Chorea

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Summary: We describe a patient with central nervous system lupus and choreatic movements in whom both basal ganglia showed high signal intensity on T1-weighted MR images, while the signal on T2-weighted images remained low. Within 8 months after onset, the choreatic movements had disappeared, with a corresponding decrease in the hyperintense T1 signal. The emergence of the choreatic movement disorder in this patient might have been related to the T1 hyperintensity of the basal ganglia, which, in turn, might have resulted from a vascular insult associated with central nervous system lupus.

Central nervous system (CNS) involvement has been reported in approximately 40% of patients with systemic lupus erythematosus (SLE) (1). Seizures, psychosis, and stroke are the most common clinical manifestations of CNS lupus (1, 2). Because the CNS involvement can be associated with significant morbidity (1), early recognition of the neuropsychiatric symptoms of CNS lupus is important. The diagnostic value of magnetic resonance (MR) imaging of the CNS in this regard has been documented (1, 3, 4). We describe a case of reversible T1 hyperintensity of the basal ganglia at MR imaging in a patient with CNS lupus and chorea.

Case Report

A 32-year-old woman was hospitalized for recurrent episodes of right-sided hemiplegia and altered mental status. SLE had been diagnosed when she was 25 years old, and the patient's symptoms had been managed with prednisolone. Ten weeks before the present admission, she had experienced sudden onset of numbness and weakness of the right upper and lower extremities, and confusion, which had resolved within 1 hour. Similar episodes recurred 3 and 6 weeks before the present admission. The patient also reported the development of involuntary movements of the upper and lower extremities and the mouth.

On the day of admission, the patient had become confused and had experienced right-sided hemiplegia. Although these symptoms resolved within 4 hours, irritability developed, along with loss of concentration. Blood analysis revealed decreased red blood cells (3.5×10^{12}/L) and normal white blood cells and platelets. Serum level of C-reactive protein was elevated (1.1 mg/dL), while that of complement was reduced (24.1 CH_{50} U/mL; normal, 30 to 40 CH_{50} U/mL). Serology revealed the presence of increased anti-DNA antibody (113 U/mL; normal, <7 U/mL). Serum tests for both the antinuclear antibody and the lupus anticoagulant were positive, whereas those for the cardiolipin IgG and IgM antibodies were negative. Hepatic and renal functions were normal. Analysis of cerebrospinal fluid revealed pleocytosis (20/3 mm^3), with a supranormal protein concentration of 41 mg/dL and an IgG level of 8.4 mg/dL. Symptoms and laboratory data were compatible with SLE (5).

A presumptive diagnosis of CNS lupus was made, and the patient was given oral (30 mg/d) and intravenous high-dose (1000 mg/d for 3 days) methylprednisolone. Subsequently, the psychotic symptoms worsened, and computed tomography (CT) of the brain revealed mild atrophy.

Two months later, the patient had a sudden onset of left hemiparesis. Deep tendon reflexes were increased on the left side. Choreatic movements were not present in the left limbs. The level of consciousness was normal. MR imaging of the brain revealed high signal intensity in the left caudate, putamen, globus pallidus, and right putamen on T1-weighted images (Fig 1A). These areas had a relatively low signal intensity on T2-weighted images (Fig 1B). Small areas of low and high signal intensity on T1- and T2-weighted images, respectively, were detected in the left ventrolateral putamen and lateral to the anterior horn of the left lateral ventricle (Fig 1A and B). No contrast enhancement was present. Single-photon emission CT (SPECT) using *N*-isopropyl [^{123}I]-P-iodoamphetamine disclosed an area of low perfusion in the right putamen.

The choreatic movements were alleviated with bromperidol, which was tapered off within 6 months without recurrence of the involuntary movements. The patient has had no further clinical symptoms of CNS lupus. Follow-up MR imaging 1 year later showed a decrease in the size of the T1 signal abnormality of the basal ganglia (Fig 1C), while the T2 signal intensity remained low (Fig 1D), with an interim appearance of small areas of high T2 signal intensity, most likely representing lacunar infarctions in the ventrolateral putamen bilaterally. MR images obtained 2 years later showed normal T1 signal intensity and absence of the high T2 signal intensity of the basal ganglia (Fig 1E).

Discussion

Our patient presented with recurrent episodes of hemiplegia, altered mental status, choreatic movements, and psychosis during the course of SLE. The former two symptoms may be

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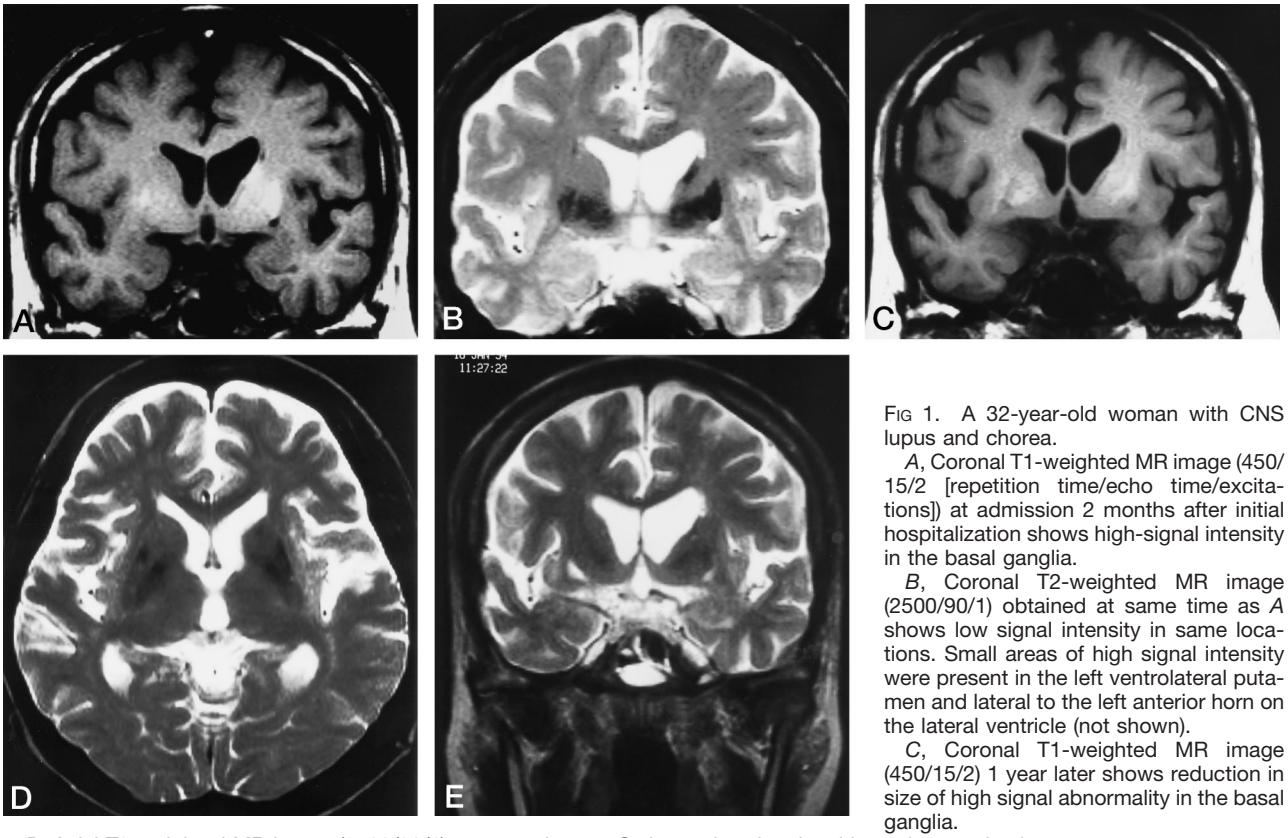

FIG 1. A 32-year-old woman with CNS lupus and chorea.

A, Coronal T1-weighted MR image (450/15/2 [repetition time/echo time/excitations]) at admission 2 months after initial hospitalization shows high-signal intensity in the basal ganglia.

B, Coronal T2-weighted MR image (2500/90/1) obtained at same time as *A* shows low signal intensity in same locations. Small areas of high signal intensity were present in the left ventrolateral putamen and lateral to the left anterior horn on the lateral ventricle (not shown).

C, Coronal T1-weighted MR image (450/15/2) 1 year later shows reduction in size of high signal abnormality in the basal ganglia.

D, Axial T2-weighted MR image (2500/90/1) at same time as *C* shows that the signal intensity remains low.

E, Coronal T2-weighted MR image (3100/93/1) 2 years after *A* and *B* shows the hypointensity in the basal ganglia is reduced.

related to ischemic changes in the basal ganglia, which were seen on MR images as lacunae and on SPECT scans as an area of hypoperfusion. Chorea can be a manifestation of CNS lupus (6). Because the chorea occurred bilaterally in our patient, this symptom may have been related to the hyperintense T1 signal abnormality seen in the bilateral basal ganglia. The psychosis may have been associated with methylprednisolone and prednisolone, because its emergence did not parallel the course of the other symptoms of CNS lupus.

According to several articles, 80% to 100% of patients with CNS lupus have abnormal findings at MR imaging (3, 4, 7, 8), including brain atrophy and focal lesions in the white matter beneath the corticomedullary junction and, less frequently, in the periventricular regions (1, 3, 4). The latter lesions are characterized by increased signal intensity on T2-weighted images. One article describes increased signal intensity abnormalities on T1-weighted MR images in a patient with CNS lupus (9). Schott et al (9) reported a patient with CNS lupus whose MR images showed diffuse areas of high signal intensity on T1-weighted images within the white matter. These authors speculated that those lesions represented fatty changes in demyelinated cells. SLE and other collagen vascular disorders can produce demyelination (10, 11). Unique to our patient is that the high T1 signal intensity localized to the basal ganglia bilaterally and disappeared with the resolution of the choreatic movement disorder.

Nagai et al (12) reported a case in which the putamen appeared hyperintense on T1-weighted MR images following a hyperglycemic coma in a patient with chorea. Our patient, however, had no history of hyperglycemia. High-signal-intensity lesions in the basal ganglia on T1-weighted images have also been described in association with parenteral nutrition and liver damage (13, 14). Our patient had no evidence of these conditions. Moreover, the abnormal MR imaging findings in these two conditions are more prominent in the globus pallidus and are not associated with chorea.

Pathologic examination of brain tissue from patients with CNS lupus has revealed vasculopathy, infarcts, hemorrhages, and vasculitis (1, 15). Greenhouse (6) suggested that cerebral vasculitis may produce chorea in patients with CNS lupus. In our case, lesions with high T1 and low T2 signal intensity were coexistent with lacunae and were observed within the bilateral basal ganglia. These lesions may be a consequence of vasculitis associated with CNS lupus. Several authors have reported the MR imaging findings of CNS lupus with chorea (3, 9, 16, 17). Among those four cases, one had multiple lesions of high T2 signal intensity in the white matter, while the other three had no abnormalities. A variety of vascular abnormalities, such as thrombosis, minute hemorrhage, and/or inflammatory changes in the small vessels supplying these regions, may explain the different MR imaging findings.

Our patient had a positive serum test for the lupus anticoagulant. Recently, positive antiphospholipid antibodies have been reported in SLE patients with both choreatic movements and transient ischemic attacks (17). CT findings in these patients were normal. The symptoms may be the result of thrombosis of small vessels in the basal ganglia. Sydenham chorea is a disorder in which choreatic movements are associated with an abnormal autoimmune state. The histopathology of this disease is that of degenerative and inflammatory vascular changes that are more pronounced in the basal ganglia (18). MR imaging in patients with Sydenham chorea shows reversible abnormal T2 signal intensity in the basal ganglia (19–21).

In summary, we have described a patient with CNS lupus and choreatic movements in whom MR images showed high T1 and low T2 signal intensity in both basal ganglia. The choreatic movements disappeared within 8 months after onset and the signal intensity abnormalities on MR images likewise resolved.

The emergence of the choreatic movement disorder in this patient might have been related to these signal intensity abnormalities.

References

1. Van Dam AP. **Diagnosis and pathogenesis of CNS lupus**. *Rheumatology* 1991;11:1–11
2. Futrell N, Schultz LR, Millikan C. **Central nervous system disease in patients with systemic lupus erythematosus.** *Neurology* 1992;42: 1649–1657
3. Jacobs L, Kinkel PR, Costello PB, Alukal MK, Kinkel WR, Green FA. **Central nervous system lupus erythematosus: the value of magnetic resonance imaging**. *J Rheumatol* 1988;15:601–606
4. McCune WJ, MacGuire A, Aisen A, Gebarski S. **Identification of brain lesions in neuropsychiatric SLE by magnetic resonance scanning.** *Arthritis Rheum* 1988;31:159–161
5. Tan EM, Cohen AS, Fires JF, et al. **The 1982 revised criteria for the classification of systemic lupus erythematosus (SLE).** *Arthritis Rheum* 1982;25;1271–1277
6. Greenhouse AH. **On chorea, lupus erythematosus and cerebral arteritis**. *Arch Int Med* 1966;117:389–393
7. Vermess M, Bernstein RM, Bydder GM, Steiner RE, Young IR, Hughes GRV. **Nuclear magnetic resonance (NMR) imaging of the brain in systemic lupus erythematosus**. *J Comput Assist Tomogr* 1983;7:461–467
8. Aisen AM, Gabrielson TO, McClune WJ. **MR imaging of systemic lupus erythematosus involving the brain.** *AJNR Am J Neuroradiol* 1985;6:197–201
9. Schott AM, Colson F, Tebib J, Noel E, Bouvior M. **Imagerie en resonance magnetique et neurolupus**. *Rev Rheum* 1990;57:785–790
10. Allen IV, Miller JHD, Kirk J, Shillington RKA. **Systemic lupus erythematosus clinically resembling multiple sclerosis and with unusual pathological and ultrastructural features.** *J Neurol Neurosurg Psychiatry* 1979;42:392–401
11. Fulford KWM, Catterall RD, Delhanty JJ, Doniach D, Kremer M. **A collagen disorder of the nervous system presenting as multiple sclerosis**. *Brain* 1972;95:373–386
12. Nagai C, Kato T, Katagiri T, Sasaki H. **Hyperintense putamen on T1-weighted MR images in a case of chorea with hyperglycemia.** *AJNR Am J Neuroradiol* 1995;16:1243–1246
13. Mirowitz SA, Westrich TJ. **Basal ganglial signal intensity alterations: reversal after discontinuation of parenteral manganese administration**. *Radiology* 1992;185;535–536
14. Pujol A, Graus F, Peri J, Mercader JM, Rimola A. **Hyperintensity in the globus pallidus on T1-weighted and inversion-recovery MRI: a possible marker of advanced liver disease.** *Neurology* 1991;41: 1256–1257
15. Ellis SG, Verity MA. **Central nervous system involvement in systemic lupus erythematosus: a review of neuropathologic findings in 57 cases, 1955–1977.** *Semin Arthritis Rheum* 1979;8:212–221
16. Forrett-Kaminsky MC, Scherer C, Kaminsky D, Klein M, Duc M. **Imagerie par résonance magnétique dans une choree lupique.** *Rev Neurol* 1992;48:383–384
17. Asherson RA, Derksen RHWM, Bouma BN, Gharvi AE, Katar L, Hughes GRV. **Chorea in systemic lupus erythematosus and "lupus-like" disease: association with antiphospholipid antibodies.** *Semin Arthritis Rheum* 1987;16:253–259
18. Colony HS, Malamund N. **Sydenham's chorea: a clinicopathologic study**. *Neurology* 1956;6:672–676
19. Kienzle GD, Breger RK, Chun RW, Zupanc ML, Sackett JF. **Sydenham chorea: MR manifestations in two cases**. *AJNR Am J Neuroradiol* 1991;12:73–76
20. Heye N, Jergas M, Hotzinger H, Frahati J, Pohlau D, Przuntek H. **Sydenham chorea: clinical, EEG, MRI and SPECT findings in the early stage of the disease.** *J Neurol* 1993;240:121–123
21. Traill Z, Pike M, Byrne J. **Sydenham's chorea: a case showing reversible striatal abnormalities on CT and MRI.** *Dev Med Child Neurol* 1995;37:270–273