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 x 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 CH50 U/mL; normal, 30 to 40 CH50 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 showed 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 [123I]-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 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...
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 pred-
nisolone, 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 im-
ages. One article describes increased signal intensity abnormal-
ities 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 de-
myelinated 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 bilat-
erally 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-inten-
sity 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 antico-
agulant. Recently, positive antiphospholipid antibodies have
been reported in SLE patients with both choreatic movements
and transient ischemic attacks (17). CT findings in these pa-
tients were normal. The symptoms may be the result of throm-
bois 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
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