

## MRI changes in the central nervous system in a child with lupus erythematosus

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**Abstract.** We report on a 10-year-old girl with systemic lupus erythematosus who presented in status epilepticus as the only manifestation of central nervous system involvement. MRI of the brain showed diffuse gray and white matter lesions which almost completely resolved after treatment with methylprednisolone. MRI findings in this child are similar to those in adults with diffuse clinical manifestations. The study is essential in the initial evaluation of patients suspected of central nervous system lupus.

Lupus erythematosus (SLE) in children is accompanied by psychiatric and neurological complications in 20%–44% of cases [1]. There is not a single test specific for central nervous system (CNS) lupus. Usually a battery of tests is performed, including cerebrospinal fluid (CSF) examination, determination of various CSF and serum antibody titers and complement levels, and electroencephalogram (EEG) and neuroimaging studies. In contrast to adults, there is limited information on MRI pattern in childhood CNS lupus.

We report the MRI pattern in a child with CNS lupus who responded to corticosteroid therapy and emphasize the important role of MRI in the initial assessment and therapeutic decision.

### Case report

A 10-year-old black girl with established diagnosis of SLE presented with a new onset of mainly generalized tonic-clonic and some left-sided focal motor seizures lasting 25 min. She had no prior significant neurological or psychiatric history, but, in addition to SLE manifestations, she had steroid-induced hyperglycemia and hypertension secondary to renal disease. Her medications on admission were cyclosporine, nifedipine, glyburide, and prednisone. Upon admission, the patient's blood pressure was 156/107 mmHg, for which she was treated with intravenous (IV) hydralazine. Seizures were treated acutely with IV diazepam and then carbamazepine

as a maintenance therapy. After seizures stopped, the patient had no headache and the results of the neurological examination, including mental status, were normal.

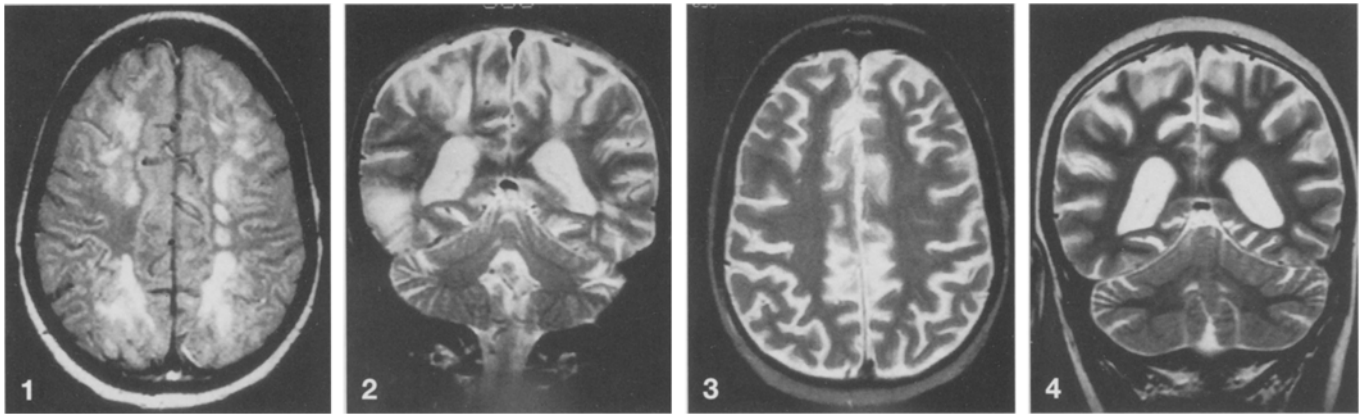
Abnormal laboratory test results were: glucose 212 mg/dl (normal 70–115 mg/dl); complement C<sub>3</sub> 64.9 mg/dl (normal 70–176 mg/dl); fluorescent anti-nuclear antibody screen 1:160; CSF complement C<sub>3</sub> and C<sub>4</sub> undetectable. MRI of the brain was performed on a 1.5-T system (General Electric). Axial and coronal images were obtained. Proton density (TR 2500/TE 30) and spin echo T2-weighted (TR 2500/TE 90) images showed multiple areas of increased signal intensity involving both white and gray matter, mainly in the frontal and posterior temporal lobes (Figs. 1, 2). The largest area of abnormal signal was localized to the right frontal region. EEG showed diffuse slowing of the cerebral activities and intermittent sharp waves, maximal over the right hemisphere.

The patient was treated with three pulse doses of IV methylprednisolone at 20 mg/kg and one bolus of cyclophosphamide IV at 500 mg/m<sup>2</sup>. One month later her SLE was in remission and a repeat brain MRI showed nearly complete resolution of the periventricular and frontal and temporal lobe lesions (Figs. 3, 4). She has been seizure-free on carbamazepine for more than a year.

### Discussion

Our case demonstrates CNS lupus with the following features: (1) diffuse gray and white matter MRI lesions in a child with SLE in the absence of focal neurological findings and CSF abnormalities other than undetectable C<sub>3</sub> and C<sub>4</sub> complements; (2) a positive correlation between left-sided partial seizures, focal EEG abnormalities, and the largest MRI lesion in the right frontal region; and (3) near-complete resolution of MRI lesions after immunosuppressive treatment.

Hypertensive encephalopathy, herpes encephalitis, and multiple sclerosis (MS) were excluded based on clinical presentation, neurological examination, and laboratory and MRI findings. Based on the MRI findings alone, the differential diagnosis would have included MS, hypertensive encephalopathy, and acute disseminated encephalomyelitis (ADEM). In MS the typical findings are of multifocal, asymmetrical lesions predominantly periventricular, but also involving the brain stem and cerebellum. In ADEM the findings can be identical



**Figs. 1, 2.** Multiple areas of increased signal in the periventricular white and gray matter extending to frontal lobes and posterior temporal lobes bilaterally. The largest lesion is located in the right frontal region. **1** Axial proton-density image (TR 2500/TE 30); **2** coronal spin echo T2-weighted image (TR 2500/TE 90)

**Figs. 3, 4.** One month after treatment: marked resolution of the high-intensity foci in the periventricular white matter and in the frontal and temporal lobes. **1** Axial proton-density image (TR 2500/TE 90); **2** coronal spin echo T2-weighted image (TR 3000/TE 85)

to those for MS, but in some cases symmetrical cerebral, basal ganglia or cerebellar lesions are present. The MRI features in hypertensive encephalopathy are variable, but may include patchy subcortical white matter lesions, most prominent at the gray-white matter junction.

Diagnosis of CNS lupus is based on high index of suspicion and a panel of laboratory studies, including CSF and serum examination for complement level and various antibody titers, tests which are neither sensitive nor specific.

MRI adds another dimension to the investigation of neuropathologic features of CNS lupus. Studies in adults using MRI show a diversity of findings and superiority of MRI over computer tomography [2, 3].

Different MRI patterns were reported in CNS lupus, depending on whether the disease was diffuse or focal [2, 4]. Attempts have also been made to correlate the MRI findings with other laboratory abnormalities and clinical outcome [4-6]. Our patient with generalized and focal seizures has an MRI pattern similar to that previously described in adults with diffuse CNS involvement presenting with generalized seizures or psychosis [2-4]. The dramatic improvement in MRI findings in our patient after treatment with corticosteroids suggests that the changes most likely represented edema. The improvement on MRI could also represent stabilization or improvement in vasculitis or the autoimmune process.

There is probably more than one pathophysiologic mechanism of CNS lupus in children. The pathology of SLE in the CNS includes foci of acute cortical encephalomalacia, neuronal loss, and demyelination. Prolifera-

tive changes in cerebral vessels occur less often in children than in adults. True vasculitis is relatively uncommon, and evidence of immune-mediated abnormalities are not found in the majority of patients with CNS lupus [7].

We conclude that MRI is a rapid and sensitive tool in detecting focal and diffuse lesions in SLE patients with acute neurological presentation. It is most helpful in the initial assessment of a patient and in therapeutic decision making when the results of other customarily performed tests are not readily available. Although MRI is sensitive in imaging CNS lesions, the findings are often nonspecific and, alone, not diagnostic of SLE. The MRI findings should thus be interpreted in the context of the patient's history, neurological abnormalities, and available tests. Follow-up MRI proved a more sensitive way of determining clinical outcome in our patient than a battery of tests, including CSF examination.

We recommend that every patient with SLE who develops neurological signs or symptoms should undergo MRI of the brain.

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