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## **St. Louis Encephalitis and the Substantia Nigra: MR Imaging Evaluation**

Felix Cerna, Borna Mehrad, James P. Luby, Dennis Burns and  
James L. Fleckenstein

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# St. Louis Encephalitis and the Substantia Nigra: MR Imaging Evaluation

Felix Cerna, Borna Mehrad, James P. Luby, Dennis Burns, and James L. Fleckenstein

**Summary:** Neuroimaging findings in cases of St. Louis encephalitis (StLE) have yet to be reported despite the relatively high frequency of this entity. An epidemic permitted the documentation of isolated hyperintensity of the substantia nigra on T2-weighted images in two patients with StLE. This distribution of MR imaging abnormality in cases of StLE mirrors the reports presented in the literature that implicate the substantia nigra as peculiarly susceptible to the StLE virus. Isolated lesions of the substantia nigra revealed by T2-weighted imaging should suggest the possibility of StLE.

Brain imaging studies help in the diagnosis of some viral encephalitides. To date, however, modern neuroimaging techniques have not revealed findings in cases of St. Louis encephalitis (StLE) (1-4). An epidemic of StLE in Dallas, TX provided the opportunity to address this imaging deficiency. A series of 10 patients underwent CT examination (n = 10), and seven of those also underwent MR imaging; we were able to detect significant lesions in only two patients. We herein report these findings and discuss their relevance in the pathogenesis and diagnosis of StLE.

## Case Reports

### Case 1

A 21-year-old Hispanic man was admitted to Parkland Memorial Hospital on August 15, 1995, with a 1-week history of fever and headache. He had a history of homelessness and alcoholism. The results of the neurologic examination were normal, and the admitting diagnosis was aseptic meningitis. The patient left the hospital the next day against medical advice. He was brought back to the hospital on August 19 for continued fever and headache with new and worsening confusion, ataxia, nystagmus, and tremulousness. He was febrile

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From the Departments of Internal Medicine, Division of Infectious Diseases (F.C., B.M., J.P.L.), Pathology (D.B.), and Radiology, Division of Neuroradiology (J.L.F.), The University of Texas Southwestern Medical Center at Dallas, Dallas, TX.

Address reprint requests to James L. Fleckenstein, MD, Division of Neuroradiology, Department of Radiology, The University of Texas Southwestern Medical Center at Dallas, 5323 Harry Hines Boulevard, Dallas, TX 75235-8896.

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at 39.4° C. He was oriented only to person and was ataxic. He was unable to hold a glass of water because of diffuse, severe tremulousness. Lumbar puncture showed an opening pressure of 32 cm H<sub>2</sub>O, 180 nucleated cells, 10% polymorphonuclear leukocytes, 85% lymphocytes, and 5% monocytes. The protein concentration was 116 mg/dL. The CSF glucose was 50 mg/dL, and the plasma glucose was 93 mg/dL. The patient's confusion and tremors cleared during the next 4 days, and he was discharged. On September 21, 1995, he was doing well without any specific problems except for a mild headache. Tremulousness had disappeared. A serum StLE virus immunoglobulin G immunofluorescent antibody titer obtained on August 21 was 1:2048; the immunoglobulin M immunofluorescent antibody titer was 1:1280. CT findings of the brain at admission were normal. MR imaging then showed conspicuous T2-weighted hyperintensity in the substantia nigra bilaterally (Fig 1); the infusion of contrast medium failed to reveal enhancement of pathologic lesions.

### Case 2

A 37-year-old African-American man had been unable to work because of a history of paranoid schizophrenia. He was a homeless alcoholic and took no medication. He was admitted to Parkland Memorial Hospital on September 10, 1995, because of fever, altered mental status, and a history of a generalized seizure. His temperature was 38.5° C. He exhibited nuchal rigidity and was unresponsive to verbal commands or pain. He had diffuse, generalized hypertonicity, and his upper extremities were flexed. He had bilateral Babinski responses and an absent gag reflex. A lumbar puncture showed an opening pressure of 24 cm, 354 nucleated cells, 17% polymorphonuclear leukocytes, 72% lymphocytes, and 18% monocytes. The protein concentration was 143 mg/dL. The CSF glucose was 46 mg/dL, and the plasma glucose was 93 mg/dL. Mechanical respiratory ventilation was required because of the patient's inability to control his secretions. The patient remained hospitalized until November 21, 1995, being on the acute medical wards for the first 6 weeks and undergoing rehabilitation for the remainder. Serum StLE virus immunoglobulin G immunofluorescent antibody titers obtained on September 11, September 25, and October 11, 1995, were 1:128, 1:1024, and 1:1024, respectively. The immunoglobulin M immunofluorescent antibody titers were 1:320, 1:2560, and 1:2560, respectively. The findings of CT of the brain at admission were normal. MR imaging then showed conspicuous, asymmetric T2-weighted hyperintensity in the substantia nigra (Fig 2); the infusion of contrast material did not cause abnormal enhancement.

## Discussion

We report isolated involvement of the substantia nigra in two young men with StLE. These men were the youngest patients treated at our hospital

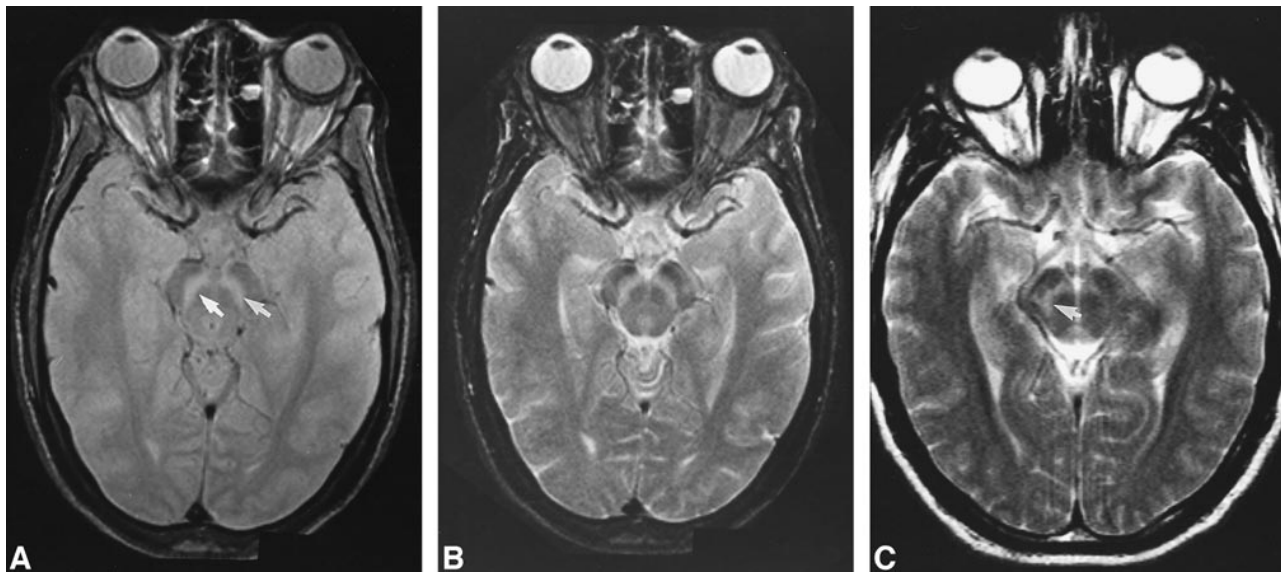


FIG 1. MR images of StLE.

A and B, Case 1: Balanced 2500/30 (TR/TE) image (A) shows hyperintensity in the substantia nigra bilaterally (arrows). T2-weighted 2500/120 image (B) obtained at same level as that shown in A shows hyperintensity in the substantia nigra more conspicuously. C, Case 2: T2-weighted 2500/120 image shows asymmetrical hyperintensity of the substantia nigra with the right side more involved (arrow).

among a group of 10 patients who were hospitalized with this disease during the summer and fall of 1995. They both had severe encephalitis clinically and diffuse tremulousness. One required extended ventilator support. Both men were alcoholic and homeless, factors that may have increased their exposure to the mosquito vector. Their tremors persisted through protracted hospitalization periods and were similar to those experienced by the other patients with StLE. The substantia nigra involvement was noted only with T2-weighted imaging. The abnormality was not apparent in the other five patients who had undergone MR imaging of the brain. The MR imaging was performed 2 and 9 days after the onset of illness. To our knowledge, this is the first and only report of such an abnormality.

An exhaustive review of the literature failed to reveal a similar distribution of focal signal changes of the substantia nigra in any brain disease, although the review included infectious, metabolic, and degenerative disorders. In recent reviews of Eastern equine encephalitis, Japanese encephalitis, and tick-borne encephalitis, it was reported that the involvement is more diffuse with areas of hemorrhage, infarction, and edema (5–8). Although the mid brain is mentioned in these diseases, there were no instances in which the substantia nigra was involved in such an isolated or selective fashion.

The literature on the pathologic features of StLE does, however, document selectively severe involvement of the substantia nigra. Reyes et al (10) studied the involvement of different sections of autopsied brains in 10 patients with StLE by

quantifying “cellular nodules” and “perivascular infiltrates” of inflammatory cells per  $\text{cm}^2$  (10). In all of their cases, the substantia nigra was always the region most involved. In terms of the percent of fractional volume affected and excluding mid brain measurements, the substantia nigra had at least eight times the quantity of brain involved than did the next most severely affected segment of the CNS (spinal gray matter). The substantia nigra had more than 250 times the fractional volume involved than did the cerebral cortex. Their results corroborated those of previous, more qualitative studies (11, 12).

The selective involvement of the substantia nigra fits also with the clinical pictures of the patients. Although not all patients with StLE are tremulous, the majority are, and this feature of the illness is particularly characteristic of the disease. Adults with Western equine encephalitis may be clinically indistinguishable from adults with StLE, so we cannot say whether the abnormality is specific or pathognomonic for StLE without further studies, although we found no report of such an abnormality in that condition. Although tremulousness in some patients may last longer than a year, there are no reports of postencephalitic parkinsonism (many years) after StLE. Why the virus so selectively and intensively involves the substantia nigra is not known, but investigation of the phenomenon may aid in our understanding of its pathogenesis. Severely ill and tremulous patients presenting with encephalitis who show isolated T2-weighted hyperintensity of the substantia nigra should be examined for StLE.

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