Myelitis Mistaken for Bladder Infection in SLE

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VANCOUVER, B.C. — Fever and urinary retention without obstruction in a patient with active systemic lupus erythematosus should be considered a medical emergency and treated immediately with high-dose intravenous corticosteroids, according to recent findings from Johns Hopkins University.

Those signs signal gray matter myelitis and spinal cord ischemia, and high-dose corticosteroids can prevent a cord infarct and permanent paraplegia, Dr. Michelle Petri said. Rheumatologists at the school have identified two previously unrecognized forms of myelitis in SLE patients: gray matter myelitis and white matter myelitis. Both are longitudinal and likely to span three vertebral segments.

Gray matter myelitis leads to rapid onset of permanent paraplegia and urinary incontinence in as little as 4 hours. Because it usually presents with acute urinary retention, it is often misdiagnosed and mistreated as a bladder infection.

But the "patient is announcing ischemia of the spinal cord and needs high-dose corticosteroids and to be admitted," said Dr. Petri, professor of rheumatology and director of the lupus center at Johns Hopkins in Baltimore.

If the syndrome—and how to treat it—were more widely recognized, "hundreds of young women would be saved from permanent paralysis," she said.

"When you have to place a catheter because the patient cannot urinate, treatment [with 1,000 mg IV methylprednisolone] should start," said Dr. Julius Birnbaum, a rheumatologist and neurologist at Johns Hopkins. "The overall message is, don't wait to provide treatment."

Gray matter myelitis, which the team considers a vasculopathy, presents with lower motor neuron signs: flaccidity and hyporeflexia, in addition to urinary retention and fever (Arthritis Rheum. 2009;60:3378-87).

On the other hand, white matter myelitis presents with upper motor neuron signs: hyperreflexia and spasticity. The onset is more gradual, antigravity strength is more likely to be preserved; attacks are less severe; and disability comes from repeated episodes that eventually lead to paralysis, in some cases. It is more likely an antibody-driven phenomenon; white matter myelitis shares features with neuromyelitis optica.

High-dose IV methylprednisolone is used to treat both forms of myelitis. Following that, patients at Hopkins are placed on steroid-sparing immunosuppressive regimens, which may include azathioprine, mycophenolate mofetil, or rituximab.

The findings are based on a record review of 22 SLE patients who presented with myelitis to the lupus center or transverse myelitis center at Hopkins in 1994-2007.

Dr. Birnbaum and his colleagues recognized the syndromes through an analysis of histories, physical exams, lab values, follow-up care, and MRIs.

The team discovered that 11 patients had gray matter myelitis, and 11 had white matter myelitis. There were no statistically significant differences between the two groups with regard to age, sex, or ethnicity. Most were women.

Of the 11 patients with gray matter myelitis, 10 presented for urinary retention. Because of the presence of fevers, "all of these patients were unfortunately and erroneously diagnosed as having urinary tract infections. By the time immunosuppressive treatment was initiated, there had likely already been irreversible injury," according to the study report. ■

VITALS

Major Finding: Of 11 SLE patients with gray matter myelitis, a newly recognized form in SLE, 10 presented with urinary retention and fever and were treated for bladder infections; the mistake likely led to permanent paraplegia.

Data Source: Retrospective study of 22 patients.

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