An 18-year-old man with recurrent oral and genital ulcers presented with uveitis, encephalopathy, headache, spastic paraparesis, and urinary retention. MRI demonstrated periventricular and hypothalamic lesions, sinus thrombosis, and longitudinally extensive spinal cord lesions (figure). Treatment with high-dose steroid medication resulted in dramatic neurologic and radiographic improvement.

Behçet disease is a multisystemic inflammatory disease characterized by recurrent oral aphthae, uveitis, and genital ulcers. Two patterns of CNS involvement have been described, including direct parenchymal involvement and cerebrovascular involvement. Simultaneous symptomatic parenchymal, vascular, and spinal cord lesions characteristic of neuro-Behçet disease in a single patient are highly unusual.

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