Elsberg Syndrome: An Overlooked But Treatable Cause of Back Pain

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ABSTRACT

Introduction: Elsberg syndrome is an uncommon cause of lumbosacral radiculitis that presents as a constellation of symptoms, including urinary retention, bowel incontinence, severe constipation, impotence, and saddle anesthesia.

Case Presentation: A 32-year-old female presented to the emergency department with complaints of bilateral leg pain and urinary retention. Two weeks prior, she noticed new genital lesions and had a positive nucleic acid amplification test for herpes simplex virus (HSV) type 2. Magnetic resonance imaging of the lumbar spine showed extraforaminal enhancement and edema-like signal within all the lumbosacral nerve roots. Cerebrospinal fluid (CSF) studies, CSF culture, and meningitis panel were unremarkable.

Discussion: Elsberg syndrome likely accounts for 5% to 15% of patients with cauda equina syndrome. However, physicians often leave out Elsberg syndrome in the differential diagnosis of acute cauda equina syndrome and do not perform HSV testing to facilitate definitive diagnosis. As a result, Elsberg syndrome is underreported.

Conclusions: A high degree of clinical suspicion should be deployed when diagnosing patients with bilateral lumbosacral radiculitis, as early detection and treatment of Elsberg syndrome prevents long-term morbidity.

INTRODUCTION

Elsberg syndrome was first described by American neurosurgeon Charles Elsberg in 1931 as a self-limiting syndrome of acute urinary retention combined with spinal cord dysfunction and cerebrospinal fluid pleocytosis.[1] Elsberg syndrome encompasses a constellation of symptoms, including urinary retention, bowel incontinence, severe constipation, impotence, and saddle anesthesia due to acute lumbosacral meningoradiculitis.[2]

CASE PRESENTATION

A 32-year-old female with a past medical history of migraines, anxiety, and depression presented to the emergency department (ED) with complaints of bilateral leg pain and urinary retention. Two weeks prior, she started experiencing urinary frequency and was started on nitrofurantoin at an urgent care center after urine dipstick was concerning for a urinary tract infection. Three days later, she returned to urgent care because she noticed new genital lesions. Nucleic acid amplification test (NAAT) of the lesions returned positive for herpes simplex virus type 2 (HSV-2). She tested negative for other sexually transmitted diseases, including HIV. She was sexually active with 1 partner who takes daily valacyclovir, and she was prescribed a 7-day course of valacyclovir after being diagnosed with HSV-2. Prior to initiating treatment, she started experiencing new onset sacral pain that progressed to a burning sensation along the back of both thighs, extending into her heels. She denied any numbness or weakness in her lower extremities.

Despite completing treatment with nitrofurantoin, the patient continued experiencing dysuria, urinary hesitancy, and incomplete voiding. Her leg pain also was progressing, so she returned to urgent care 1 week later. Urinalysis at that time showed trace blood, 30 protein, and moderate leukocytes. Culture was positive for >100 000 colony forming units of Gardnerella vaginalis. Urology also evaluated her at urgent care and suspected her symptoms were due to continued HSV-2 infection, even though she did not have evidence of active lesions at that time. She was sent

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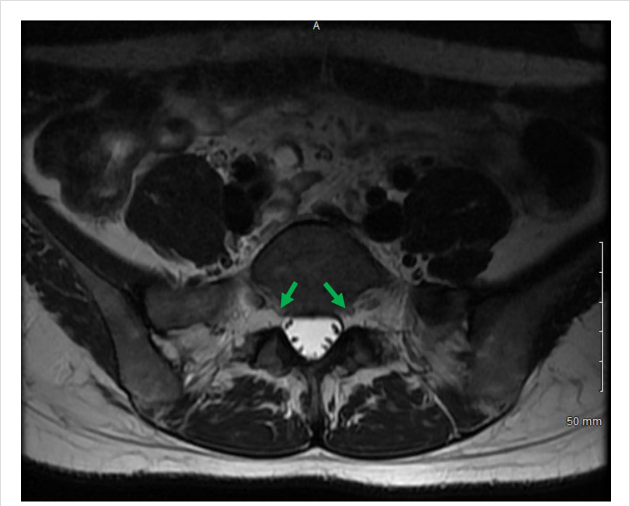
Figure. Magnetic Resonance Imaging Spine With and Without Contrast Demonstrating Extraforaminal Enhancement in the Lumbosacral Nerve Roots



home with a 5-day course of ciprofloxacin and a 10-day course of valacyclovir and prednisone and told to follow up with Neurology outpatient. One day later, she felt increased burning and itching in her vaginal area, along with new onset neck stiffness and headache. She presented to the ED for further workup of her symptoms.

In the ED, admission vitals and labs were within normal limits. Kernig and Brudzinski signs were negative on physical exam. Her basic metabolic panel, comprehensive metabolic panel, and urinalysis were unremarkable. Bladder scan showed 252 mL of urine. Given the clinical symptoms of headache and neck stiffness, lumbar puncture, head computed tomography (CT) and magnetic resonance imaging (MRI) of the brain were obtained to rule out an infectious etiology or an acute intracranial process. The patient's cerebrospinal fluid (CSF) protein, glucose, red blood cell count, and white blood cell count were within normal limits. CSF culture showed 3+ mononuclear cells with no organisms or evidence of polymorphonuclear neutrophils. The meningitis/encephalitis NAAT panel was negative for all pathogens, including HSV-2. Head CT and MRI brain showed no acute intracranial findings, but MRI of the lumbar spine showed extraforaminal enhancement and edema-like signal within all the lumbosacral nerve roots.

Based on the clinical presentation, recent HSV-2 infection, and abnormal MRI findings, the patient was diagnosed with Elsberg syndrome. Per Neurology's recommendations, she was started on intravenous (IV) acyclovir and methylprednisolone. Her leg pain improved significantly after 2 days of treatment. While she still experienced some urinary hesitancy, she no longer felt that she was voiding incompletely. She completed 5 days of IV acyclovir and methylprednisolone and was discharged on an oral prednisone taper and home IV acyclovir infusions to complete 14 days of treatment. She reported complete resolution of urinary symptoms and sacral pain following completion of the acyclovir course.

Table. Diagnostic Criteria for Elsberg Syndrome

Categories	Criteria
1. Laboratory-supported definite	(A1 OR A2) AND B5
2. Clinically definite	A1 OR A2; B1 AND two of B2–B4; B1 and B2 (if concomitant)
3. Clinically probable	A1 OR A2; B1 AND one of B2–B4
4. Clinically possible	A1 OR A2; one of B1–B4
5. Excluded	Neither of A1 nor of A2; any of D1–D3
A. Required	
A1. Clinical symptoms and signs of cauda equina involvement: urinary hesitancy or retention; bowel incontinence, or severe constipation (erectile dysfunction insufficient on its own)	
A2. MRI or electrophysiologic evidence of cauda equina involvement: enhancement of cauda equina; EMG evidence of radiculopathy	
B. Supportive but not required	
B1. Time course: acute/subacute onset; no relapse; progression over <3 months	
B2. Coexisting or recently preceding symptoms of genital herpes infection OR other clinical symptoms of herpes virus infection	
B3. Clinical (eg, exaggerated reflexes and Babinski signs) or MRI evidence of myelitis in conus	
B4. CSF pleocytosis	
B5. Documented herpes virus infection from CSF by PCR, culture, or detection of IgM serology	
C. Red flags	
C1. Relapses beyond 1 year from onset	
D. Exclusionary	
D1. Myelitis extending rostral to T9	
D2. Other neurologic symptoms suggestive of alternative etiology: optic neuritis, brain/brainstem syndrome	
D3. Other etiology proven/more likely for syndrome: NMOSD, dural arteriovenous fistula, viral transverse myelitis, other causes of myelopathy	

Abbreviations: CSF, cerebrospinal fluid; EMG, electromyography; IgM, immunoglobulin M; MRI, magnetic resonance imaging; N/A, not applicable; NMOSD, neuromyelitis optical spectrum disorder; PCR, polymerase chain reaction.

DISCUSSION

Elsberg syndrome is a rare neuroinflammatory disease that typically presents with lower extremity sensory impairment, weakness, saddle anesthesia, and urinary and/or bowel incontinence following HSV-2 infection.[3] Here we present a case of Elsberg syndrome secondary to HSV-2 infection in a young, immunocompetent, sexually active female. While the exact pathophysiology of Elsberg syndrome is unknown, it is thought to be related to latent viral infection resurfacing in the spinal nerve roots.

In cases of an identifiable trigger, Elsberg syndrome is most commonly preceded by HSV-2 or related viruses, including severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), West Nile virus, and varicella-zoster virus (VZV).[3] Reactivation of latent viral infection in spinal nerve roots and their sensory neurons is the posited mechanism of lumbrosacral myeloradiculitis. While many patients with HSV-2-related Elsberg syndrome are immunocompetent, immunocompromised patients are at greater risk for VZV-related Elsberg syndrome.[4]

Elsberg syndrome is fairly uncommon, accounting for up to

10% of cases with combined myelitis and lumbosacral radiculitis.[3] Retrospective review of lumbosacral myeloradiculitis evaluated at the Mayo Clinic from 2000 to 2016 identified 1035 patients with both myelitis and radiculitis, which are most suggestive of Elsberg syndrome.[5] Thirty of those patients were suspected to have Elsberg syndrome, but only 2 had been diagnosed at the time of clinical evaluation, indicating that Elsberg syndrome often goes unrecognized.

Diagnosis of Elsberg syndrome is based primarily on patient history, clinical symptoms, and diagnostic imaging. Salvodi et al published a tool to characterize Elsberg syndrome based on diagnostic certainty. Required symptoms include either clinical or MRI/electromyographic evidence of cauda equina involvement. Supporting symptoms include an acute onset (progression over less than 3 months), recent genital herpes infection, clinical or MRI evidence of myelitis in the conus medullaris, or CSF pleocytosis.[5]

According to the diagnostic criteria above, our patient had laboratory-supported definite Elsberg syndrome. She presented with dysuria and lower extremity pain around the time she developed primary HSV-2 lesions. She also presented with MRI evidence of cauda equina involvement within 1 month of HSV-2 infection confirmed via NAAT testing. Although CSF pleocytosis is a supporting symptom of Elsberg syndrome, it may be normal as it was in this case.

When treating Elsberg syndrome, confirmation of the causative virus is not always required, as medications are effective in mild cases without a definitive viral cause. However, in severe cases where meningitis is suspected, the causative virus is more necessary as higher doses of antiviral drugs are required to treat VZV infection than HSV infection.[6] Moreover, VZV-related Elsberg syndrome is associated with a more severe presentation.[7] Direct immunofluorescence assay, viral culture, and polymerase chain reaction (PCR) all can be used to distinguish HSV from VZV infections.[8] If antigen or PCR testing is unavailable, the patient's age and sexual history can provide clues for diagnosis because older age is a risk factor for VZV, whereas HSV is more common in younger, sexually active patients like our patient.[9]

Treatment of Elsberg syndrome consists of 10 to 21 days of acyclovir, which has been shown to decrease pain and improve symptoms.[5] While oral steroid tapers or high-dose IV steroids can help shorten the duration of symptoms, the use of steroids to treat Elsberg syndrome remains a topic of debate.[10] Our patient received 14 days of IV acyclovir and 5 days of IV methylprednisolone with an oral prednisone taper. She reported full resolution of her symptoms upon completion of treatment.

CONCLUSIONS

Because Elsberg syndrome is rarely reported and has a highly variable clinical presentation, it often goes unrecognized. There is a wide spectrum of neurological disorders associated with Herpesviridae, and clinicians should evaluate for preceding or ongoing symptoms of herpes infections when treating patients with unexplained multifocal neurological symptoms. We encourage clinicians to consider Elsberg syndrome as a differential diagnosis for patients with bilateral lumbosacral radiculitis, as treatment with acyclovir may shorten symptom duration and decrease pain severity. The diagnosis of Elsberg syndrome relies on clinical judgement, taking into account the patient's history, symptoms, and imaging findings. This case of Elsberg syndrome highlights the importance of early detection and treatment to prevent long-term morbidity.

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