Case Report

Elsberg syndrome caused by herpes zoster in the sacral region with preceding urinary retention

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Background: Elsberg syndrome (ES) is an acute-onset lumbosacral radiculitis with myelitis caused by a herpes virus infection.

Case Presentation: We present a case of a 77-year-old woman who was admitted with urinary retention prior to genital rash. The patient was diagnosed with ES and treated with intravenous acyclovir 250 mg every 8 h for 1 week.

Conclusion: Physicians should consider ES in patients with voiding dysfunction, as preceding neurological symptoms may lead to a misdiagnosis. Considering the adverse effects of the antiviral drug, its dosage should be according to the causative virus of the ES as well as the patient's age and medical history.

Key words: acyclovir, Elsberg syndrome, herpes zoster, urinary retention, VZV

INTRODUCTION

ELSBERG SYNDROME (ES) is an unidentified cause of acute lumbosacral neuritis and is related to recent herpes virus infection. Elsberg syndrome often presents with urinary retention. However, there are very few case reports of this condition, and it remains understudied.[1–3] Urinary retention may precede the appearance of skin rash, and ES should be considered in the differential diagnosis. Differential diagnosis between herpes virus type 1/2 (HSV-1/2) and varicella zoster virus (VZV) is sometimes necessary because the recommended antiviral treatment doses differ between the herpes types.[2]

CASE PRESENTATION

A 77-YEAR-OLD WOMAN who was previously healthy presented to the emergency department with a 3-day history of genital rash and pain that was preceded by voiding dysfunction, incomplete bladder emptying, and abdominal distention for 10 days. The patient was afebrile and had no headache, disturbance of consciousness, weakness, or paralysis of the lower extremities. The patient was sexually inactive. Physical examination revealed an erythematous rash with blisters over the left half of the pubic area and left buttock, consistent with the S2 and S3 dermatomes (Fig. 1). The patient showed no abnormal neurological findings, except for urinary retention. Ultrasound examination revealed an estimated 111 ml of post-void residual urine. Immunofluorescence testing for VZV antigen from the vesicle was positive, which led to the diagnosis of clinically probable ES. The patient was hospitalized, a urinary catheter was inserted, and she was treated with intravenous acyclovir 250 mg every 8 h (q8h) for 1 week. She was also treated with 500 mg acetaminophen, as needed. On hospital day 3, urinary discomfort and the voiding dysfunction reduced, with a residual urine volume of 70 ml on a bladder scan (radionuclide cystogram). On hospital day 6, neuralgia reduced, and the rash became crusted. The patient was discharged from the hospital on day 8. The skin rash and voiding dysfunction did not recur for 6 months, although postherpetic neuralgia persisted.

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DISCUSSION

ELSBERG SYNDROME, FIRST described in 1931,[1] is an acute lumbosacral radiculitis with myelitis that is caused by recent herpes virus infection.[2] The diagnosis in

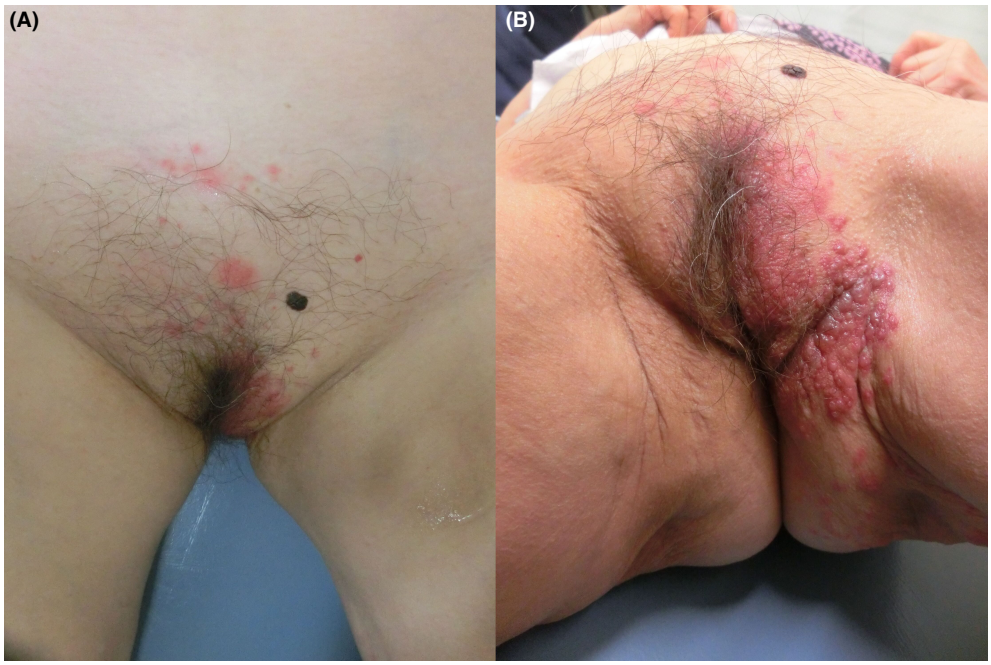

Fig. 1. Photographs of a 77-year-old woman with multiple vesicles in left sacral lesions (S2 and S3), which are compatible with genital herpes zoster infection.

our patient was "clinically probable," according to previous diagnostic criteria.[2] This case met the following criteria: A1, clinical symptoms and signs of cauda equina involvement, including urinary hesitancy or retention, bowel incontinence, or severe constipation; B1, time course indicating acute/subacute onset, no relapse, and progression over less than 3 months; B2, coexisting or recently preceding symptoms of genital herpes infection or other clinical symptoms of herpes virus infection, which implied "clinically probable" under the criteria. The details of Savoldi's criteria[2] and the assessment of the present case are shown in Table 1.

We consider this case novel as it describes the details of ES with urinary retention preceding the development of skin rash and pain. Elsberg syndrome is rare, but it has been described in several case reports or case series.[1,2,4] However, the occurrence of urinary retention preceding the development of a rash is rare and may make diagnosis difficult. Furthermore, some cases of ES with preceding urinary retention have been described, but details such as the definition of the diagnosis and the clinical course after treatment were not well described. Thus, this report will assist the diagnosis of ES in the emergency department setting. Early diagnosis of ES is important because the use of antiviral agents hasten the resolution of rash lesions and neurological symptoms, and decrease pain severity.[3]

Clinicians should consider ES in patients with voiding dysfunction because preceding neurological symptoms could lead to a misdiagnosis. In a previous study, 25% of patients with herpes zoster infection involving the lumbosacral dermatome developed voiding dysfunction. Among these patients, voiding dysfunction proceeded to a skin rash at a rate of 14.6%.[4] Thus, genital zoster should be considered a differential diagnosis when voiding dysfunction of unknown origin is observed.

There are several mechanisms that induce voiding dysfunction in patients, including neuritis and meningitis, but ipsilateral hemicystitis and reluctance to void due to pain have also been reported.[5] We believe that in this patient, the symptoms originated from neuritis because voiding dysfunction preceded the genital rash and pain, and there was no sign of cystitis on urinalysis.

Although magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) tests are not necessary to fulfill the diagnostic criteria,[2] these examinations are important when meningitis, encephalitis, myelitis, or other causes of cauda equina syndrome are suspected. In the present case, these diseases were unlikely because the patient had no related symptoms such as fever, headache, or limb paralysis; therefore, we did not undertake MRI or CSF tests.

Elsberg syndrome diagnosis is usually made clinically.[2,3] The differential diagnosis of the causative virus of ES is

Table 1. Diagnostic criteria for Elsberg syndrome (Savoldi's criteria) and assessment in the present case

Categories	Criteria	Assessment in the present case
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		N/A
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 (e.g., exaggerated reflexes and Babinski signs) or MRI evidence of myelitis in conus		N/A
B4. CSF pleocytosis		N/A
B5. Documented herpes virus infection from CSF by PCR, culture, or detection of IgM serology		N/A
C. Red flags		
C1. Relapses beyond 1 year from onset		N/A
D. Exclusionary		
D1. Myelitis extending rostral to T9		None
D2. Other neurologic symptoms suggestive of alternative etiology: optic neuritis, brain/brainstem syndrome		None
D3. Other etiology proven/more likely for syndrome: NMOSD, dural arteriovenous fistula, viral transverse myelitis, other causes of myelopathy		None

Note: A check mark (✓) means the findings in the present case met the criteria.
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.

sometimes necessary in severe cases, such as in cases of meningitis, because higher doses of antiviral drugs are required for VZV treatment (10 mg/kg q8h) than for HSV infection treatment (5–10 mg/kg q8h).[3,6] In mild cases, such as in the present case, confirmation of the causative virus is not always required; however, it is recommended for recipients of solid organ transplantation.[7]

For viral diagnosis, a direct immunofluorescence assay can distinguish HSV infections from VZV infections at a lower cost and more rapidly than a viral culture.[3] Polymerase chain reaction techniques (PCR) are also useful, if available.[3] The patient's age and sexual history could provide clues for diagnosis in a clinical setting where antigen or PCR testing is unavailable, because increasing age is a major risk factor for the development of herpes zoster, whereas HSV is more common than herpes zoster in younger patients.[8]

Antiviral drugs are recommended for the resolution of herpes zoster lesions and to decrease the severity of acute pain.[3] Although the correct acyclovir regimen for ES caused by VZV has not been established,[3] the usual dose that is administered is 5–10 mg/kg q8h for ES without meningitis and 10 mg/kg q8h for ES with meningitis or ES in immunocompromised hosts.[9,10] In the present case, because the only neurological symptom was voiding dysfunction, we administered 5 mg/kg q8h, considering the risk of adverse effects, such as renal toxicity or acyclovir-induced neurotoxicity, with increasing doses. According to the patient's neurological condition (consistent with meningitis, disseminated lesions, and weakness or paralysis of the lower extremities) and background (age and immunocompromised status due to HIV infection), an individualized dose of acyclovir was considered reasonable. Glucocorticoids remain controversial,[2,3] especially in

elderly patients, because of their adverse and nonestablished clinical effects in the treatment of VZV.[3]

In conclusion, we reported a case of ES caused by herpes zoster in the sacral region. Physicians should determine whether ES is present in patients with voiding dysfunction. Adequate treatment, including antiviral therapy, is mandatory.

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DISCLOSURE

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Registry and registration no. of the study/trial: N/A.
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