Varicella Zoster Virus Meningoencephalitis Presenting with Elsberg Syndrome without a Rash in an Immunocompetent Patient

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Abstract

Varicella zoster virus (VZV) infection usually manifests with a skin rash. To the best of our knowledge, this is the first report of a case of VZV meningoencephalitis presenting with Elsberg syndrome without a rash in an immunocompetent patient. Clinicians should consider the potential for VZV infection as well as herpes simplex virus infection in cases of aseptic meningitis accompanied by bladder and rectal disturbances, even in patients without any rash symptoms.

Key words: varicella zoster virus, meningoencephalitis, Elsberg syndrome


Introduction

Varicella zoster virus (VZV) is known to be responsible for a broad spectrum of neurological diseases, including post-herpetic neuralgia, meningitis and encephalitis (1, 2). The dermatological clinical manifestations of VZV range from primary infection with chickenpox to reactivated infection with shingles. VZV reactivation usually presents with a skin rash, although cases of VZV infection without skin rashes (zoster sine herpeta) associated with neurological diseases have been reported (3, 4).

Herpesvirus, especially herpes simplex virus (HSV)-2, is reported to be associated with lumbosacral radiculomyelitis, known as Elsberg syndrome (5). Other herpesviruses, including VZV, cytomegalovirus and Epstein-Barr virus, have also been shown to be associated with Elsberg syndrome, although there are few reports of cases of Elsberg syndrome (6-8). In addition, the onset of VZV meningitis associated with Elsberg syndrome has been previously reported, whereas no cases of VZV infection without a skin rash associated with Elsberg syndrome have been documented. We herein report the first such case, involving a previously healthy 57-year-old man who developed VZV meningoencephalitis associated with Elsberg syndrome without a skin rash.

Case Report

A 57-year-old man visited our emergency room complaining of a lasting fever with chills in addition to a progressively worsening headache with nausea and vomiting and intention tremors of the bilateral upper limbs, which had developed over a seven-day period. He had noticed bilateral knee pain at the time of onset of the fever and headache; however, it resolved spontaneously. He denied having a cough, abdominal pain, diarrhea, constipation, urinary symptoms or skin rashes. Moreover, there was no history of recent travel abroad, and he reported no contact with any diseased individuals. In addition, he had had no sexual intercourse during the six months prior to symptom onset, and his past medical history was not significant; his previous history of chickenpox was unknown. In the emergency room, he was not oriented in terms of place or date. His body temperature was 38.7°C, his blood pressure was 157/90 mmHg, his pulse was 115 beats per minute and regular and his respiratory rate was 20 per minute. Nuchal rigidity, jolt accentuation and Kernig’s sign were absent; however, there were bilateral intention tremors in the upper limbs, as well as myoclonic movement of the left lower limb and
mild dysarthria. Deep tendon reflexes were slightly increased in the bilateral upper limbs, although that of the lower limbs was normal. In addition, snout and jaw-jerk reflexes were positive. The patient exhibited no evidence of a rash, and other findings of the physical examination were unremarkable.

The laboratory data revealed a white blood cell (WBC) count of 7.3×10^9/L (neutrophils: 62.4%, eosinophils: 0.3%, basophils: 0.7%, monocytes: 8.9%, lymphocytes: 27.7%, no atypical lymphocytes) and platelet count of 186×10^9/L. There were no abnormal findings in terms of the hepatic or renal function. Lumbar puncture was performed after admission, which showed clear cerebrospinal fluid, with 341 WBCs/mm^3 (mononuclear cells: 98%, polynuclear cells: 2%), no red blood cells, a total protein level of 168 mg/dL and a glucose level of 47 mg/dL (blood glucose level: 97 mg/dL). Gram staining of the cerebrospinal fluid (CSF) showed no organisms, and there were no signs of fungi on India ink staining or acid-fast bacteria on acid fast staining. Polymerase chain reaction (PCR) of the CSF for Mycobacterium tuberculosis was also negative. The initial urinary analysis, chest X-ray examination and chest computed tomography (CT) and brain magnetic resonance imaging (MRI) scans without gadolinium-based contrast agents showed no abnormalities. The findings for serum immunoglobulin G (IgG) to HSV-1, VZV, cytomegalovirus (CMV), Epstein-Barr virus viral capsid antigen (EBV-VCA) and mumps virus were positive, while tests for serum immunoglobulin M (IgM) to each of these viruses were negative. Antibodies and antigens for human immunodeficiency virus (HIV) were negative, and HIV virus was not detected in the serum according to PCR testing.

The patient was therefore given a diagnosis of aseptic meningocerephalitis, the cause of which was suspected to be a virus, and empirically treated with acyclovir at a dose of 10 mg/kg intravenously every eight hours. After admission, he gradually defervesced, and his headache and consciousness level gradually improved. However, constipation and urinary retention appeared on day 15 of hospitalization, which showed clear cerebral spinal fluid, with 341 WBCs/mm^3 (mononuclear cells: 98%, polynuclear cells: 2%), no red blood cells, a total protein level of 168 mg/dL and a glucose level of 47 mg/dL (blood glucose level: 97 mg/dL). Gram staining of the cerebrospinal fluid (CSF) showed no organisms, and there were no signs of fungi on India ink staining or acid-fast bacteria on acid fast staining. Polymerase chain reaction (PCR) of the CSF for Mycobacterium tuberculosis was also negative. The initial urinary analysis, chest X-ray examination and chest computed tomography (CT) and brain magnetic resonance imaging (MRI) scans without gadolinium-based contrast agents showed no abnormalities. The findings for serum immunoglobulin G (IgG) to HSV-1, VZV, cytomegalovirus (CMV), Epstein-Barr virus viral capsid antigen (EBV-VCA) and mumps virus were positive, while tests for serum immunoglobulin M (IgM) to each of these viruses were negative. Antibodies and antigens for human immunodeficiency virus (HIV) were negative, and HIV virus was not detected in the serum according to PCR testing.

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Discussion

We herein describe a case of VZV meningocerephalitis without a rash in an immunocompetent adult man. VZV meningitis was previously considered to be a rare cause of aseptic meningitis due to the lack of appropriate detection tests; however, VZV is now well known to be a cause of aseptic meningitis in adults as a result of new developments in techniques, such as antibody and PCR testing. VZV infection is usually associated with skin rashes. In a previous article, skin rashes were reported to be observed in approximately 90% of VZV meningitis patients, with neck stiffness noted in only 38% of patients. In contrast, neck stiffness is usually observed in patients with HSV meningitis, although HSV meningitis is not associated with skin rashes (10). Neither neck stiffness nor skin rashes were observed in our patient. The absence of neck stiffness is not uncommon in patients with VZV meningitis, whereas the onset of VZV infection without an apparent skin rash is unusual, although cases of VZV meningitis without rashes have been reported (11, 12).

The current patient developed constipation and urinary retention during his hospital stay, symptoms that were clinically diagnosed as Elsberg syndrome. Elsberg syndrome was first reported by Elsberg in 1931. This condition is a self-limiting lumbosacral radiculomyelitis associated with clinical signs such as transient urinary retention and constipation and sensory symptoms (5). Human HSV-2 has been described to be a common etiology of Elsberg syndrome, whereas other herpesviruses, such as HSV-1, VZV, Epstein-Barr virus and cytomegalovirus, have been reported to be associated with lumbosacral radiculomyelitis (6, 8). Lumbosacral radiculomyelitis is commonly detected on spinal MRI imaging, which shows varying degrees of root or lower spinal cord edema with enlargement and hyperintensity on T2-weighted images, although there may be no spinal abnormalities in some cases (8). In the current case, spinal MRI did not show any abnormalities; however, we diagnosed the patient with Elsberg syndrome based on his clinical course. While the clinical manifestation of aseptic meningocerephalitis without a skin rash associated with Elsberg syndrome is common in patients with HSV-2 infection, the present case was due to VZV infection, which is an atypical clinical presentation. Another case report described Elsberg syndrome secondary to VZV infection with a skin rash complicated with rectal bleeding (13). In that report, the authors proposed several hypotheses for the rectal bleeding. The true pathogenesis of rectal ulcers has not been confirmed, performing a biopsy of the ulcer, which can detect VZV-related changes, may be useful for confirming the etiology of the...
lesion. Although Elsberg syndrome caused by VZV infection may be associated with an atypical clinical presentation, including rectal bleeding, there are few case reports describing Elsberg syndrome resulting from VZV infection. Therefore, more information from similar cases must be accumulated in order to clarify the clinical features of Elsberg syndrome associated with VZV infection. To our knowledge, this is the first report of a case of Elsberg syndrome occurring in a patient with VZV infection without a rash.

In summary, we experienced a case of VZV meningoencephalitis presenting with Elsberg syndrome without a rash. Clinicians should consider the potential for VZV infection as well as HSV infection in patients with aseptic meningitis accompanied by bladder and rectal disturbances, even those without a rash.

The authors state that they have no Conflict of Interest (COI).

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References