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Varicella Zoster Virus Related Myelo-Radiculo-Neuropathy: Case Report

Varisella Zoster Virus İlişkili Myelo- Radikulo- Nöropati: Olgu Sunumu

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Öz

Amaç: Varisella zoster virus (VZV) enfeksiyonları primer enfeksiyon veya reaktivasyon dönemlerinde çeşitli nörolojik sendromlara neden olabilmektedir. Burada VZV ile ilişkili, eş zamanlı transvers miyelit ve Guillain Barré sendromu gelişen olguyu sunmayı amaçladık.

Olgu: Yetmiş altı yaşında kadın hasta tüm vücutta suçiçeği ile uyumlu cilt lezyonları ve 10 gün önce başlayan yürüme güçlüğü şikayetleriyle başvurdu. Nörolojik muayenede paraparezi ve arefleksi görüldü. Spinal kord manyetik rezonans görüntülemede alt servikal ve torakal düzeyi tutan uzun segment transvers miyelit lezyonu izlendi. Beyin omurilik sıvısında, protein yüksekliği ve VZV DNA varlığı, serumda VZV IgM saptandı. İki gün sonra hastanın bacaklarındaki güçsüzlüğü arttı. Elektromiyografide akut aksonal motor polinöropati bulguları görüldü. Hasta yüksek doz steroid ve antiviral tedavi ardından intravenöz immunglobulin ile tedavi edildi. Kısmi iyileşme izlendi.

Sonuç: Transvers miyelit ve Guillain Barré sendromu, VZV enfeksiyonunun nadir görülen komplikasyonlarıdır. Olgumuz VZV ile ilişkili bu iki nörolojik durumun aynı hastada görüldüğü ilk vakadır.

Anahtar Sözcükler: Varisella zoster virüs; akut inflamatuar demiyelinizan polinöropati; Guillain Barré Sendromu; transvers miyelit

Abstract

Aim: Varicella zoster virus (VZV) infections can cause various neurological syndromes during primary infection or reactivation periods. We report two rare neurological complications of VZV, acute longitudinally extensive myelitis and acute motor axonal neuropathy due to primary chickenpox infection that occurred concomitantly.

Case: A 76-year-old female patient presented with skin lesions consistent with chickenpox all over the body and walking difficulty ten days ago. Neurological examination revealed paraparesis and areflexia. Spinal cord magnetic resonance imaging revealed a long-segment transverse myelitis lesion involving the lower cervical and thoracic levels. Elevated protein and the presence of VZV DNA were detected in the cerebrospinal fluid, and VZV IgM was detected in the serum. Two days later, the patient's weakness in her legs increased. Electromyography showed signs of acute axonal motor polyneuropathy. The patient was treated with intravenous immunoglobulin after high-dose steroid and antiviral therapy. Partial recovery was observed.

Conclusion: Transverse myelitis and Guillain Barré syndrome are rare complications of VZV infection. Our case is first case where these two neorological conditions related to VZV were seen in the same patient.

Keywords: Varicella zoster virüs; acute inflammatory demyelinating polyneuropathy; Guillain Barré syndrome; transvers myelitis

Bu olgu, 57. Ulusal Nöroloji Kongresi'nde tartışmalı poster olarak sunulmuştur.

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Introduction

Herpes zoster (shingles) and varicella (chickenpox) are two clinical forms of varicella-zoster virus (VZV). Both primary and reactivated VZV infections are associated with various neurological syndromes affecting central and peripheral nervous systems, such as encephalitis, meningitis, cerebellar ataxia, transverse myelitis, and Guillan Barré Syndrome (GBS) (1).
For the first time, we report two rare neurological complications of VZV, acute longitudinally extensive myelitis and acute motor axonal neuropathy due to primary chickenpox infection that occurred concomitantly.

Case

Seventy-six years old female patient was admitted to our clinic with 15 days history of a widespread rash and ten days history of difficulty walking. The maculo-vesiculo-pustular skin eruptions were typical for chickenpox. As far as she remembers, she was not vaccinated for varicella and did not experience chickenpox. In neurological examination, mild confusion, lower limb weakness (muscle strength 3/5), areflexia in lower limbs, and bilateral flexor plantar response were found. Other systemical examinations were within normal. Brain magnetic resonance imaging (MRI) and electroencephalography (EEG) were within normal limits. Spinal MRI showed acute longitudinally extensive myelitis (T6-T11) (Figure 1). Cerebrospinal fluid (CSF) examination showed increased protein level (156 mg/dl) and pleocytosis (45 leucocyte/mm^3). CSF polymerase chain reaction (PCR) for VZV was positive. CSF-PCR for CMV DNA was negative. Serological tests indicated primary VZV infection with positive IgM and negative IgG antibodies. Human immunodeficiency virüs (HIV) serology was negative. Anti-GM1 and Campylobacter jejuni lipo-oligosaccharide antibodies are not studied. The *serum* autoantibody marker, *neuromyelitis optica*-immunoglobulin G *(NMO-IgG)* was negative. The patient was treated intravenously with acyclovir 10 mg/kg/day and methylprednisolone 1 g/day. Two days later, the confusion improved, but weakness in the lower limbs progressed. Electromyography evidenced acute motor axonal neuropathy. Treatment with intravenous immunoglobulin (IVIG 0.4 g/kg/day for five days) was started. Two weeks after, symptoms improved with incomplete recovery.

Discussion

The most frequent VZV-related central nervous system (CNS) infections are meningitis and meningoencephalitis. The neuropsychiatric symptoms, headache, and altered consciousness are common in patients with encephalitis and meningitis. Cerebellar involvement may also occur. The most affected cranial nerves are the seventh and eighth cranial nerves (2, 3). VZV-related myelitis is seen only in 0.3% of varicella infections (4).

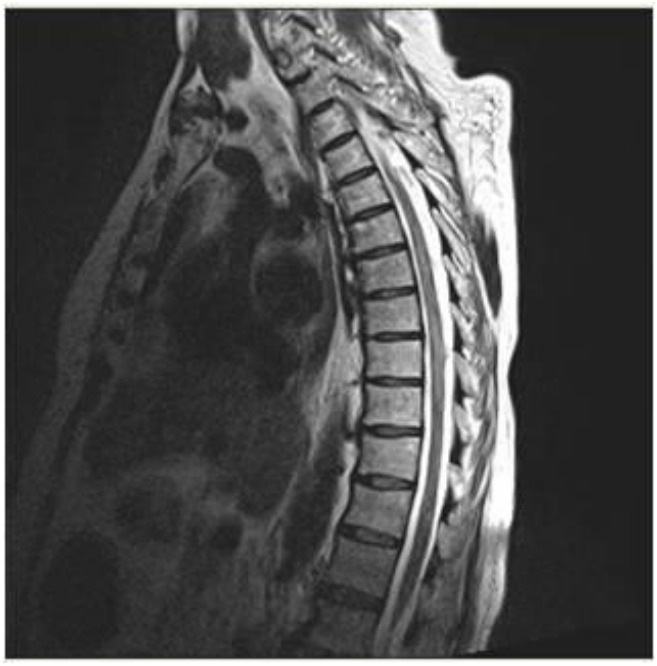

Figure 1. Thoracal spinal magnetic resonance imaging showing longitudinally extensive myelitis lesion hyperintense in T2 weighted images.

A study investigating clinical and laboratory features in patients with neurological involvement due to VZV infection found CNS infection in 34 of 282 patients (12%). Of 34 patients with VZV central nervous system infection, 18 had encephalitis, and 15 had meningitis. There was only one patient with myelitis (2). Paresthesia below the lesion level, paraparesis or quadriparesis, impaired sphincter functions, and vesicular lesions prior to neurological symptoms are typical (4). Myelitis is generally located at the cervical or thoracic level (5). The interval between rash and myelitis onset ranges from days to months (4). Most patients with VZV-related myelitis are immunocompromised (6). T2-weighted MRI is more sensitive than other MRI modalities. Multiple hyperintense lesions spreading from the dorsal ganglia with various degrees of edema and contrast enhancement in more than one segment have been reported (5). PCR showing positive VZV DNA and antibody in the CSF is highly pathognomonic for VZV-associated myelitis (7). There is no standard treatment for VZV-related myelitis. High-dose acyclovir and steroids are recommended (5).
In a series of VZV-related myelitis in immunocompetent patients, Wang et al. showed longitudinally extensive transverse myelitis (more than three segments) in 9/10 patients while neuromyelitis optica spectrum disorder in only three of them (8). They recommended the simultaneous use of glucocorticosteroid and antiviral drugs. They reported favorable outcomes in their patients (8). Our patient was not immunocompromised, and we also observed longitudinally extensive transverse myelitis. NMO-IgG was negative. The patient received antiviral and high-dose immunosuppressive therapy, but

peripheral nerve involvement followed despite this treatment.

Islam et al. found seven post-chicken pox GBS cases in a cohort of 536 consecutive patients with GBS from Bangladesh in addition to 15 previously reported cases of post-shingles GBS and 24 cases of post-chicken pox GBS. Most 46 cases were quadriplegic with sensory symptoms and cranial nerve dysfunction. The median time between skin lesions and weakness ranged between 8 to 11 days. Demyelinating sensory-motor type of GBS with favorable outcomes was seen in most patients. The VZV-related GBS was thought to occur by the activation of T cells by VZV, direct infection of peripheral nerves, or immune-mediated demyelination (9). However, the cellular immune system is involved in patients with acute inflammatory demyelinating polyneuropathy.

Nerve conduction studies and needle electromyography revealed an axonal form of GBS that differs from the characteristic demyelinating pattern. A limitation of this case report is the lack of the complete screening of infectious etiologies such as Campylobacter jejuni that may coexist with the VZV-related transverse myelitis and trigger the axonal form of GBS. However, the CSF PCR and serum antibody tests are highly confirmative of the VZV as the etiology of GBS in our patient. In contrast, the humoral immune systems play a role in the pathogenesis of other subtypes of GBS (9).

Conclusion

Transverse myelitis and GBS are both infrequent complications of VZV. There are only fewer than 50 cases of VZV-related GBS. Reports of VZV-related myelitis have only described a single or a few cases. Reported cases of VZV-related myelitis are frequently associated with immunosuppression and rarely confirmed by serologic tests. This case report is unique in presenting two rare conditions in an immunocompetent patient with acute varicella infection presented with longitudinally extensive transverse myelitis. Furthermore, despite the normal brain MRI and EEG, the confusion at the admission may also indicate meningoencephalitis.

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