Parainfectious Transverse Myelitis secondary to Varicella Zoster Virus

Nirmal Ghimire,1 Sanjib Dhungel,1 Krishna Dhungana,2 Srijana Pradhananga,1 Sushma Thapa,1 Ang Gelu Sherpa,1 Dinesh Koirala,1 Apurba Shrestha1

1Department of Internal Medicine, Nepal Medical College and Teaching Hospital, Jorpati, Kathmandu, Nepal,
2Department of Internal Medicine, Kathmandu Medical College and Teaching Hospital, Kathmandu, Nepal.

ABSTRACT

Varicella zoster virus is a human herpes virus that causes chickenpox and herpes zoster. Varicella zoster virus leads to numerous complications of the central and peripheral nervous systems. Transverse myelitis is a disorder characterized by focal inflammation of the spinal cord and results in loss of motor and sensory function below the level of injury. Transverse myelitis caused by Varicella zoster virus reactivation is rare in immunocompetent patients. Herein, we report a case of transverse myelitis caused by Varicella zoster virus in an immunocompetent young patient. A 33 years gentleman was admitted to our hospital with complaints of multiple pleomorphic skin lesions and fever for 10 days, unable to pass urine on his will for four days and weakness of bilateral lower limbs for three days. MRI spine showed lower thoracic vertebra. The patient was treated with steroids and Acyclovir. He recovered completely in one month after discharge. Prompt clinical diagnosis with early use of antiviral and anti-inflammatory treatment is important for good outcome of disease in resource limited country like Nepal.

Keywords: Immunocompetent; Nepal; Transverse myelitis; Varicella zoster virus.

INTRODUCTION

Varicella-zoster virus (VZV) is a human herpes virus that causes chickenpox and herpes zoster.1 It belongs to subfamily alfavirus. VZV leads to numerous complications of the central and peripheral nervous systems.2 These complications are rare in immunocompetent individuals.3 Central nervous system involvement in VZV infection has been reported as 0.1-0.75% in several studies.4,5 VZV infection presents with a variety of neurologic complications, including post-herpetic neuralgia, polyradiculoneuritis, Transverse Myelitis, vasculopathy, aseptic meningitis, Leukoencephalopathy, dorsal root or cranial nerve ganglionitis, ventriculitis, Necrotising angiitis and Meningoencephalitis.6 Transverse myelitis is rarely associated with VZV infection. The frequency of Transverse myelitis during or following varicella infection is reported as 0.3%.4 Transverse myelitis is a disorder characterized by focal inflammation of the spinal cord and results in loss of motor and sensory function below the level of injury. Herein, we report a case of transverse myelitis caused by VZV in an immunocompetent patient.

CASE REPORT

A 33 years gentleman was admitted to our hospital with complaints of multiple fluid filled skin lesions and fever for 10 days, unable to pass urine on his will for four days and weakness of bilateral lower limbs for three days. He developed multiple fluid filled itchy vesicles throughout the body predominantly on trunk. After six days of developing skin
lesions, he developed retention of urine and was unable to get off the bed. He didn’t feel sensations of hot and cold below the level of umbilicus. There were no symptoms in upper limbs. There was no history of loss of consciousness, altered sensorium, blurring of vision difficulty in swallowing and breathing.

There was no history of similar rashes in the past and recent vaccination. History was corroborated with his parents.

On examination, vitals were normal. General examination were normal except for multiple itchy skin lesions at different stages varying from clear fluid filled vesicles to crusts predominantly on trunk.

Initial neurological examination on admission showed normal levels of consciousness, cooperation, orientation, as well as intact cranial nerves and normal cerebellar function. Motor and sensory functions in the upper limbs were normal. Motor examination in bilateral lower limbs showed decrease tone and power (1/5). All sensory modalities below the level of umbilicus were absent. Plantar reflex was mute bilaterally and deep reflexes in bilateral lower limbs were absent. Anal tone was diminished with loss of anal reflex. Ophthalmoscopic examination was normal.

Owing to the above mentioned findings, provisional clinical diagnosis of parainfectious Transverse myelitis probably due to VZV was made.

Investigations showed Hemoglobin 13.6 g/dl, total WBC count 7020/cumm, platelet count 233000/cumm, ESR 10 mm/hr. Renal function tests, liver function tests, Thyroid function test and urinalysis were all normal. HIV ELISA, VDRL were negative. Cerebrospinal fluid(CSF) examination revealed total count of 63 cells/ul with all lymphocytes. Glucose and protein were 50 mg/dl and 116 mg/dl respectively.

A diagnosis of parainfectious Varicella zoster Transverse myelitis was made and he was started on intravenous steroids and Acyclovir. He was given Intravenous Methylprednisolone 1gm once a day for five days followed by Tablet Prednisolone 1 mg/kg once a day. Injection Acyclovir 10 mg/kg body weight in three divided doses were given for 14 days. By the end of first week of treatment, he regained power in the lower limbs (4+/5).
with regain of all sensory modalities. Patient was discharged with Foley's catheter in-situ due to persistence of impairment in bladder control.

Figure 3. Multiple healed scars over trunk.

Figure 4. Patient discharged with Foley self retaining. urinary catheter in situ.

Patient was followed in OPD with tapering of steroids weekly with discontinuation of steroids in six weeks. He recovered fully in one month after discharge including bladder control.

Figure 5. Patient came to OPD driving his car by himself.

DISCUSSION

TM is an inflammatory focal lesion of the spinal cord, characterized by the inflammation, edema and necrosis in one or several spinal segments, causing an alteration of the motor, sensory or autonomic function. Many cases of myelitis, termed postinfectious or postvaccinal, follow an infection or vaccination. Numerous organisms have been implicated, including Epstein-Barr virus (EBV), Cytomegalovirus (CMV), mycoplasma, influenza, measles, varicella, rubella, and mumps. Postinfectious myelitis often begins as the patient appears to be recovering from an acute febrile infection, or in the subsequent days or weeks. Approximately 25-40% cases of transverse myelitis are caused by viral infections with herpes viruses and poliovirus. Varicella zoster infection is not a common cause of transverse myelitis in immunocompetent patients.\(^8\)

The immune pathogenesis of TM is varied and reflects the rather diverse spectrum of this disease from idiopathic mechanisms to myelitis associated with the disease. There is evidence of perivascular infiltration by monocytes and lymphocytes in the lesion.\(^9\) Pathogenesis of the neurological complications associated with VZV infection is unclear. Allergic and vascular mechanisms have been suggested for some of these neurological complications that occur after infection with primary varicella and herpes zoster.\(^10\) The pathogenesis of parainfectious VZV myelitis has been thought to be a direct viral invasion, because VZV particles were found in glial cells, and the virus has been isolated from the spinal cord of patients with zoster myelitis. In a detailed report that included post mortem examination of the spinal cord, Hogan et al. presented evidence of direct invasion of VZV in a patient with transverse myelitis associated with VZV.\(^11\) Demonstration of the VZV antigen in CSF cells by immunofluorescence or isolation of VZV from the CSF is a confirmative evidence for viral central nervous system infection but it is rarely successful.\(^12\)

In our case, TM related to varicella infection was diagnosed on the basis of the development of pleomorphic rash, paraparesis, sensory and bladder dysfunction with MRI findings suggestive of TM. Patient’s serum showed high titre of anti-VZV antibodies (IgG) - 15.05 mIU/ml after a month.
Diagnosing VZV myelitis can be challenging. Physicians must maintain a high index of suspicion in atypical cases. It is important to rule out spinal cord compression due to epidural abscess or tumor, intrinsic bacterial or fungal infections of the spinal cord, and treatable vascular diseases with the help of other tests like blood tests, CSF examination, CT scan, MRI, and myelography. The detection of Anti-VZV antibodies and VZV DNA in CSF are confirmatory diagnostic tests. However, Rosenfeld et al. reported that patients showed clinical signs of severe VZV myelitis, despite negative Anti-VZV antibody tests and PCR results for VZV DNA. Imaging studies are useful for the diagnosis of VZV myelitis. MRI of VZV myelitis is likely to show T2-hyperintensity in the spinal cord.

There are no established treatment regimens for TM as a complication of VZV infection. Some researchers recommend high doses of Acyclovir and steroids. Although the standard treatment regimen for VZV myelitis is not yet established, there is anecdotal evidence for treatment of VZV myelitis with Acyclovir. Therefore, an early diagnosis and treatment with antivirals is essential for recovery from VZV myelitis and minimizing its complications. This treatment is even crucial to prevent the development of post-herpetic neuralgia. Though the additional benefit of the steroid is not clear we used steroids because the combination of high-dose Acyclovir and steroids have shown a good outcome in previous case reports.

In our case, TM related to VZV infection was diagnosed on the basis of clinical findings and MRI. We report this case because parainfectious Varicella zoster TM in an immunocompetent patient is very rare and he improved completely in a month and half with steroids and Acyclovir.

The spectrum of clinical outcomes in VZV myelitis ranges from spontaneous recovery to ascending progression and death. As many studies recommend high doses of corticosteroids and Intravenous Acyclovir in order to avoid progression and added complications, early antiviral and anti-inflammatory treatment in order to eradicate viruses and prevent future neurological events plays pivotal role in the management. This case report adds the importance of clinical diagnosis along with radiological assistance if available in the resource limited country like Nepal for early use of the drugs for the good outcome of disease.

CONFLICT OF INTEREST: None.

CONSENT: NMJ case report consent form was signed by the patient.

REFERENCES


