



Varicella Zoster encephalo-myelitis: A rare presentation in an immunocompetent patient

Dr. Sudhir Kr Atri¹, Dr. Mohini², Dr. Vinay Singla^{3*}, Dr. Neeraj Bamel⁴, Dr. Gaurav Rathee⁵, Dr. Nishil Gowda⁶, Dr. Ekta⁷

¹ Senior Professor, Department of Medicine, Pt. B.D. Sharma PGIMS, Rohtak, Haryana, India

² Associate Professor, Department of Medicine Pt. B.D. Sharma PGIMS, Rohtak, Haryana India

^{3,5,6,7} Postgraduate Student Department of Medicine Pt. B.D. Sharma PGIMS, Rohtak, Haryana, India

⁴ Senior Resident, Department of Medicine Pt. B.D. Sharma PGIMS, Rohtak, Haryana, India

* Corresponding Author: Dr. Vinay Singla

Abstract

Varicella zoster encephalomyelitis is a rare complication of varicella-zoster virus (VZV) infection. Because of its nonspecific clinical presentation, VZV encephalomyelitis is under recognized. It was traditionally thought that varicella Zoster virus encephalomyelitis was a disease of the immunocompromised, however there have been increasing numbers involving immunocompetent patients as more clinicians utilize cerebrospinal (CSF) polymerase chain reaction (PCR) technique and antibody studies to diagnose VZV as a cause for neurologic disease. To date, there have been only a few case reports of VZV encephalomyelitis involving immunocompetent patients. Here we present the case study of a 15 year-old male patient who presented with altered sensorium in a tertiary care hospital in Haryana (India). Upon workup the patient turned up to be a case of VZV encephalomyelitis.

Keywords: encephalomyelitis, immunocompetent, technique, hospital

1. Introduction

Varicella zoster virus (VZV) is a neurotropic α herpes virus known to cause the primary infection manifesting as varicella or chickenpox. After the primary infection period, the virus may stay latent in the spinal and cranial ganglia neurons, and then become reactive after the latent term resulting in a herpes zoster infection. Neurological complications caused by chickenpox are rare, estimated as approximately 0.01%-0.25% [1]. These complications include postherpetic neuralgia, cranial nerve palsies, peripheral motor neuropathy, myelitis, encephalitis, thrombotic cerebral vasculopathy, acute ascending polyradiculitis, and aseptic meningitis [1, 2]. Varicella zoster encephalitis and transverse myelitis are uncommon complications of Varicella zoster infection [3, 4]. Immunosuppression is the principal risk factor for their development [5]. But case reports of VZV encephalomyelitis in immunocompetent patients are also coming up. Historically, the diagnosis of VZV encephalitis and/or myelitis was dependent upon the presence of the characteristic rash along with the temporal development of clinical encephalitis. With the advent of the polymerase chain reaction (PCR) technique for identifying VZV in the cerebrospinal fluid (CSF), VZV can be definitively diagnosed, thus allowing for directed therapy.

The case: 15 year old male, resident of Panipat, 10th std. student, presented with chief complaints of high grade fever since 15 days, rash since 15 days and altered sensorium since 2 days. The fever was associated with small, itchy, fluid filled blisters initially which later on become erythematous and crusted, gradually involving whole body. The patient took desi medications and got relieved in terms of fever and rash, only dry hyper pigmented scabs were left. After 2-3 days he started having frontal region headache.

Two days before admission, patient developed altered sensorium, associated with delayed to no response to verbal commands and decreased oral intake, associated with slight deviation of angle of mouth towards left side with drooling of saliva. On examination right sided bell's phenomenon was present. Bulk, tone, power and deep tendon reflexes (DTR) were normal.

On investigations, complete hemogram, LFT, RFT were normal. CSF tap was done which showed 100 cells/cu mm, predominantly lymphocytes with normal proteins and sugar and normal ADA. In view of history of chicken pox rash, CSF was also sent for IgM Anti varicella zoster virus (VZV) antibody by PCR, which came out to be positive. MRI Brain of the patient was also done which showed small ovoid and T1W hypointensity and T2W/FLAIR hyper intensity in right centrum semiovale. Diagnosis of Varicella Zoster encephalitis was made and patient was put on tablet acyclovir 500mg three times a day. Patient showed significant improvement in 6-7 days.

On 8th day of admission he started complaining of weakness of left lower limb and gradually of bilateral (B/L) lower limbs. Bulk was normal and tone was increased in B/L lower limbs. Power was 5/5 in B/L upper limbs, 3/5 in left lower limb and 4/5 in right lower limb. Deep tendon reflexes (DTR) were 3+ in B/L lower limbs. Proprioception, vibration senses were impaired in B/L lower limbs. Urinary retention was also present. MRI lumbar spine with whole spine screening was ordered which revealed patchy areas of altered signal intensity in spinal cord, most prominent in cervical cord (extending upto cervico-medullary junction) with similar patchy areas in dorsal cord and conus medullaris, suggestive of transverse myelitis. Patient was given injection methyl prednisolone 1g i.v OD for 5 days. There was significant improvement in general condition of the patient. Power in

B/L lower limbs returned to 5/5. Vibration sense, joint position sense and touch sensations also improved. Patient was put on tablet prednisolone 50mg once daily on discharge and it is being tapered off on outdoor follow up.

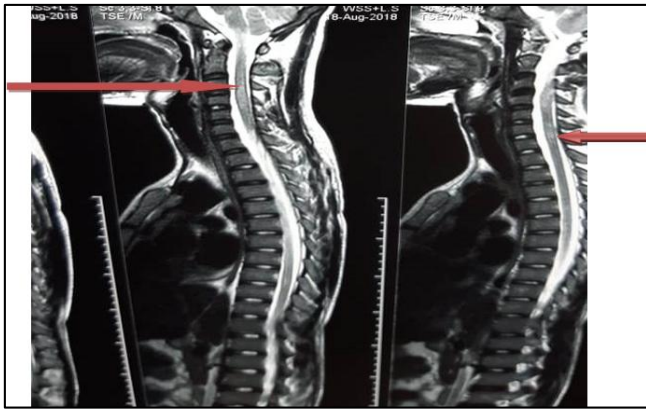


Fig 1: T2W sagittal section images of vertebral column showing hyperintense lesions in cervical spine (red arrows)

Discussion

Common CNS complications of chicken pox are cerebellar ataxia and encephalitis, and rare complications are transverse myelitis, aseptic meningitis, Guillian-Barre syndrome, meningoencephalitis, ventriculitis, optic neuritis, post-hepatic neuralgia, herpes zoster ophthalmicus, delayed contralateral hemiparesis, peripheral motor neuropathy, cerebral angitis, Reye syndrome and facial paralysis. The pathogenic bases for these complications have been thought to be many. The cause has been postulated as either direct viral invasion or through an immune-mediated allergic mechanism^[6]. Most pathologic studies have shown a picture more likely to be allergy-mediated injury^[7]. The virus is also known to cause vasculitis like episodes and even cerebrovascular accidents have been reported. In immunocompromised host, the virus can invade deeper tissues and the virus has been isolated from brain tissue or ventricles by polymerase chain reaction (PCR)^[8]. Central nervous system (CNS) complications of Varicella zoster infection are rare in immunocompetent patients. Simultaneous involvement of brain, cranial nerves and spinal cord in response to VZV reactivation is exceptional in an immunocompetent patient. Devinsky *et al*^[9] reported 13 patients with VZV myelitis, all were immunocompromised. To best of our knowledge only few cases of varicella encephomyelitis in immunocompetent persons have been reported in literature till date.

Acute transverse myelitis (ATM) is a condition of sudden weakness of lower extremities with sensory involvement, caused due to inflammation of the spinal cord. Viral disease is responsible for 20-40% cases of ATM such as epstein-barr, rubella, mumps, herpes simplex and VZV^[10]. The interval between chicken pox and development of ATM is variable. According to a report, it can occur with the rash or may be delayed for up to 2 weeks^[11]. The detection of anti VZV antibodies and VZV DNA in CSF are confirmatory diagnostic tests. Treatment is with corticosteroids, and methylprednisolone intravenous has been found to be effective^[12]. Antivirals have a controversial role and physiotherapy has a definitive supportive and rehabilitative role.

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