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Case Report

An uncommon complication of a common disease: varicella zoster myelitis

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Introduction

Primary varicella zoster virus (VZV) infection is called chickenpox. Neurological complications due to VZV infection are particularly seen in immune-deficient patients and rare among immunocompetent persons. We report a case of VZV infection with concomitant long segment dorsal spine transverse myelitis in an immunocompetent patient. Cerebrospinal fluid (CSF) was positive for VZV DNA. Significant improvement was observed following treatment with acyclovir.

Case report

A 77-year-old lady with previously good health presented to our neurology unit with backache for seven days and difficulty in walking for three days. She also complained of numbness of the right lower limb, difficulty in passing urine and constipation, along with difficulty in walking. She denied upper limb symptoms, visual impairment, double vision, swallowing difficulties or hiccups.

She was a known patient with hypertension. She denied high risk sexual behavior. She was afebrile at the time of examination. Her vital parameters were normal including blood pressure of 120/70 mmHg, pulse rate of 70 beats/min and a respiratory rate of 15 breaths/min. There was a vesicular rash distributed on her face, extremities and trunk. Some lesions were newly erupting lesions, and some were crusted (Fig. 1).

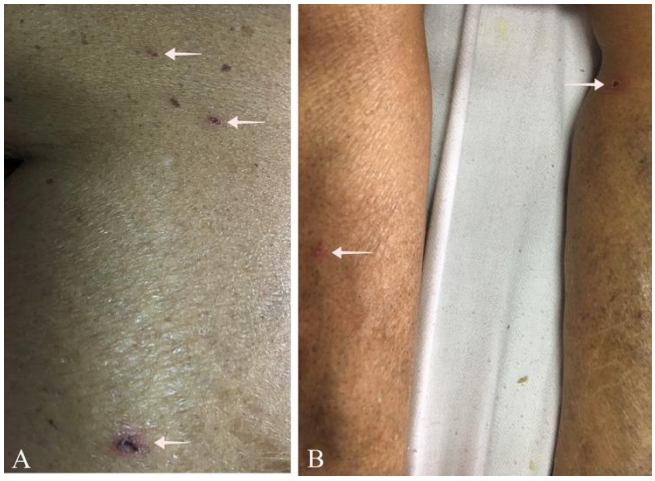

Figure 1: Vesicular rash involving trunk (A) and lower limbs (B) indicated by white arrow. Some lesions are crusted, some newly erupting.

There was no lymphadenopathy. Neurological abnormalities were confined to the lower limbs. The patient had grade 2/5 power in the left lower limb and grade 3/5 power in the right lower limb. Both knee and ankle jerks were absent on the left side, but the right knee and ankle jerks were normal. Both plantar responses were extensor. A sensory level was present at D6. Motor and sensory examination of the upper limbs were normal. Cerebellar signs were absent. Cranial nerve examination was normal. Eye examination did not reveal relative an afferent pupillary defect or papilloedema. Abdominal examination revealed a distended bladder. Cardiovascular system and respiratory system examination were unremarkable.

Investigations showed the following; haemoglobin 12g/dl, white cell count 9.9 x 10^3/µl (N-63.7%, L-17.3%, M-8.4%), platelet count 248 x 10^3/µl. Renal functions and liver functions were normal. Serum electrolytes were normal. CSF analysis revealed a white cell count of 523/cu mm (1% polymorphs and 99% lymphocytes), red blood cells 15/cu mm, protein 93.3 mg/dl and glucose 3.2mmol/l (random blood sugar 5mmol/l). CSF culture for bacteria, tuberculosis and fungi were negative. CSF VZV DNA was positive by PCR. She was negative on HIV testing. Magnetic resonance imaging (MRI) of the dorsal spine showed a T1 weighted hypo-intense and T2 weighted hyper-intense lesion involving more than 2/3^{rds} of the transverse diameter of the dorsal spine, extending from D2 to D9 level (Fig.2). The affected region of the cord was swollen. There was no significant contrast enhancement.

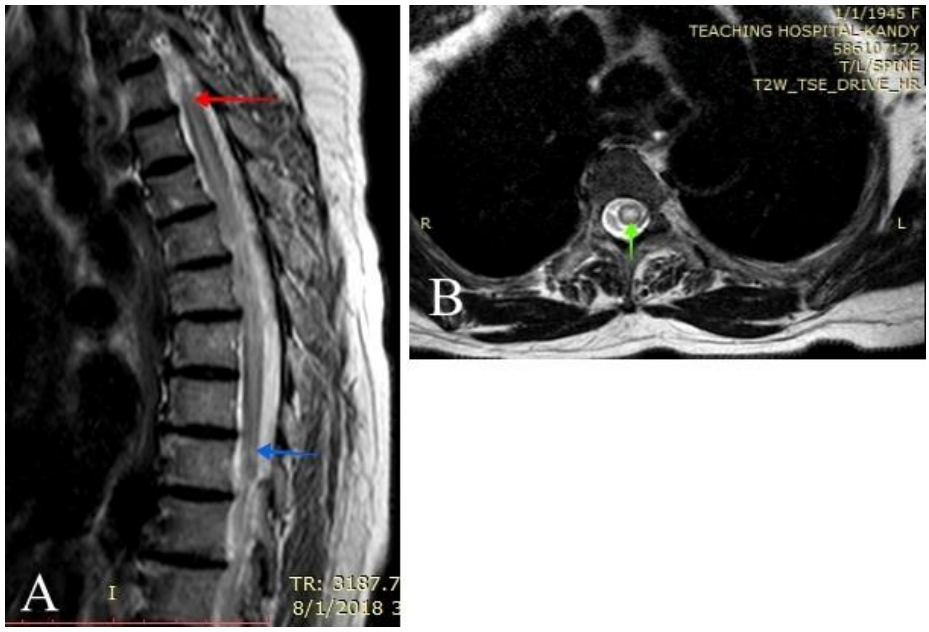

Figure 2: T2 weighted sagittal view of dorsal spine (A) shows hyper-intense lesion extending from D2 level (red arrow) to D9 level (blue arrow) and axial view (B) shows hyper-intense lesion (green arrow) involving more than 2/3 of the transverse diameter of the spinal cord.

She was treated with intravenous acyclovir for 21 days without steroids. One month after the completion of treatment she was able to walk without support and bowel and bladder functions became normal. She did not complain of post-herpetic neuralgia.

Discussion

This case report describes a long extensive transverse myelitis caused by VZV. Acute transverse myelitis has an estimated incidence of 1.34 to 4.6 per million population [1,2]. Acute transverse myelitis can be caused by demyelinating diseases, inflammatory or autoimmune diseases, infectious diseases and paraneoplastic conditions. Infectious or para-infectious myelitis can be caused by viruses, bacteria, parasites or fungi. Around 25%-40% of transverse myelitis are caused by infections with herpes viruses or polio virus [3].

VZV, an exclusively human, neurotropic, alpha herpes virus causes varicella (chickenpox). Infection is known to cause numerous complications in the central nervous system. These complications are rare among immunocompetent individuals. VZV induced neurological complications are reported as 0.1-0.75% [4,5]. Among these complications, transverse myelitis is one of the rarest. Our patient developed lower limb symptoms together with a chickenpox rash. However, cases of VZV myelitis without the typical rash have also been reported [6]. The pathogenesis of VZV myelitis is thought to be direct invasion of the spinal cord by the virus. This is evident by detection of the virus in the spinal cord of patients with varicella zoster myelitis [7]. The neurological findings of myelitis are characteristically bilateral and include a sensory deficit at a given level, motor weakness and abnormal bladder and rectal function. Our patient had asymmetrical neurological findings in the lower limb together with bladder and rectal involvement.

The diagnosis of varicella zoster myelitis may be challenging. It is necessary to rule out other possible aetiologies such as compressive myelopathies and demyelinating disease. The clinical history and systemic examination is important in gathering supportive evidence for VZV infection. Varicella zoster myelitis can be confirmed by detection of VZV DNA or VZV specific antibodies or both in CSF along with a VZV specific rash. CSF examination reveals a mild, predominantly mononuclear pleocytosis with elevated protein [8]. MRI of spine may be normal or show a T2-weighted hyper-intense lesion. Cord swelling and enhancement are occasional MRI findings. MRI of the spinal cord of our patient demonstrated long extensive transverse myelitis in the thoracic spinal cord. Neuromyelitis optica (NMO) typically cause long extensive transverse myelitis. In addition, multiple sclerosis, sarcoidosis, Sjögren syndrome and infectious diseases with CNS involvement are also known to cause long extensive transverse myelitis [9]. However, the typical skin rash and positive VZV DNA in CSF confirmed the diagnosis of varicella zoster myelitis in our patient.

The standard treatment regimen for varicella zoster myelitis is not yet established. However, anecdotal evidence suggests that treatment with high dose acyclovir and steroid is effective [10,11]. Early treatment with antiviral agents are important to prevent post-herpetic neuralgia [12]. Clinical outcome in varicella zoster myelitis may vary from spontaneous recovery to rapidly ascending myelitis and death [13]. Fatal cases of varicella zoster myelitis are reported among the immunocompromised patients. However, recovery is usually seen in immunocompetent patients. Our case is an example of varicella zoster myelitis with complete recovery following 21 days of acyclovir without steroids.

Conclusion

It is important to consider infectious aetiologies in the differential diagnosis of patients with long segment transverse myelitis. Detection of VZV DNA in CSF helps to confirm the diagnosis of varicella zoster myelitis. Early treatment with antiviral agents is associated with significant clinical improvement in varicella zoster myelitis.

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