Ramsay Hunt Syndrome Complicated by Brainstem Encephalitis in Varicella-zoster Virus Infection

Yao-Yao Shen, Ting-Min Dai, Hai-Ling Liu, Wei Wu, Jiang-Long Tu

Department of Neurology, Second Affiliated Hospital of Nanchang University, Nanchang, Jiangxi 330006, China

To the Editor: Varicella-zoster virus (VZV) infection can lead to zoster or cranial nerve palsy such as Ramsay Hunt syndrome (RHS), or complications of central nervous system (CNS) such as myelitis, cerebellitis, encephalitis, and stroke syndrome.[1,2] However, the coexistence of RHS and VZV brainstem encephalitis is extremely rare to our knowledge. We, therefore, report a patient who presented with RHS and multiply cranial nerve palsies.

A 58-year-old immunocompetent man felt febrile and severe otalgia with a frequent dry cough. One week later, he gradually developed left facial paralysis, left ear hearing loss, and vertigo. After another 3 days, dysarthria, swallowing difficulty, and intractable hiccup presented sequentially. On the next day, he complained of diplopia with extraocular movement limitation and was admitted to the Second Affiliated Hospital of Nanchang University. On admission, his vital signs were stable. Neurological examinations showed the following: Left eye immobilization with right eye adduction limitation, left facial weakness and hypesthesia, left sensorineural hearing loss, paralysis of the left soft palate, and slurred speech. There were slightly decreased muscle power and hyperreactive tendon reflexes with a positive Barbinski sign contralaterally. Coordinate movement of four limbs was impaired, and the Romberg test was positive. Carefully, herpetic vesicles, erythematous ulcerative, and crusted scars were observed around the left external acoustic meatus. Brain magnetic resonance imaging (MRI), on admission [Figure 1], revealed a high signal intensity lesion involving basis pontis and medulla oblongata on both the T2-weighted image [Figure 1b] and fluid attenuation inversion recovery (FLAIR) [Figure 1d]. Magnetic resonance angiography showed normal findings. Moreover, contrast-enhanced MRI showed a spot-like enhancement in medulla oblongata as well as enhanced left facial nerve [Figure 1e and 1f]. On day 2 after admission, an electroencephalogram showed diffuse theta waves. Meanwhile, the cerebrospinal fluid (CSF) analysis showed an increased cell count (1360/mm^3, 80% lymphocytes) and protein level (1.37 g/L). CSF cultures for bacteria, fungus, tuberculosis, and herpes simplex virus DNA were negative. This patient was immediately administrated with intravenous acyclovir (10 mg/kg every 8 h) and methylprednisolone (40 mg/d) after hospitalization. Three days after admission the patient deteriorated with paralysis of bilateral limbs and pulmonary inflammation. He was treated with empiric antibiotics. On day 7, an elevated serum IgM antibody titer to VZV on enzyme-linked immunosorbent assay (ELISA) and the presence of CSF VZV DNA amplified by polymerase chain reaction (PCR) confirmed VZV infection. On day 14, his vertigo, swallowing and speaking function improved moderately, but his hearing loss and facial palsy remained unaltered. One month later, follow-up brain MRI demonstrated that the extent of the lesion had decreased [Figure 1g-1I] and PCR for VZV DNA in CSF became negative. Clinically, he could walk alone and hear high tone voice in his left ear.

VZV is a member of the family *Herpesviridae* with the ability to establish latency in dorsal root-, autonomic- and cranial ganglia. After reactivation, VZV causes herpes zoster in most cases. VZV might also infect CNS causing various neurological manifestations.[3,4] With the introduction of using PCR for detection of virus DNA in CSF, VZV has been reported to be the second most common viruses of encephalitis recently.[5] Based on herpes zoster with facial nerve palsy, our patient fulfills the criteria for RHS. Moreover, he can also be diagnosed with VZV brainstem encephalitis according to CSF evidence of VZV and pontobulbar involvement on brain MRI. However, reports about RHS accompanied by VZV encephalitis have been rarely documented in the literature. The reason for the low incidence of RHS complicated by VZV encephalitis is not well understood. Clinically, our case was initially diagnosed with RHS. Subsequently, he presented with multiple cranial nerve palsies (from III to X cranial nerves) and central spastic palsy (bilateral pyramidal tracts involvement), which may suggest a brainstem insult. Furthermore, transverse hyperintensities in central pons on T2-FLAIR obviously showed the lesions. Meanwhile, contrast-enhanced MRI showed enhanced left facial nerve. We speculate the possible mechanism of VZV spreading to CNS is the reactivated viruses, which establish latency in

Address for correspondence: Dr. Jiang-Long Tu, Department of Neurology, Second Affiliated Hospital of Nanchang University, Nanchang, Jiangxi 330006, China
E-Mail: Tujianglong85@126.com

Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.4103/0366-6999.170275

Received: 13-09-2015 **Edited by:** Xin Chen

How to cite this article: Shen YY, Dai TM, Liu HL, Wu W, Tu JL. Ramsay Hunt Syndrome Complicated by Brainstem Encephalitis in Varicella-zoster Virus Infection. Chin Med J 2015;128:3258-9.

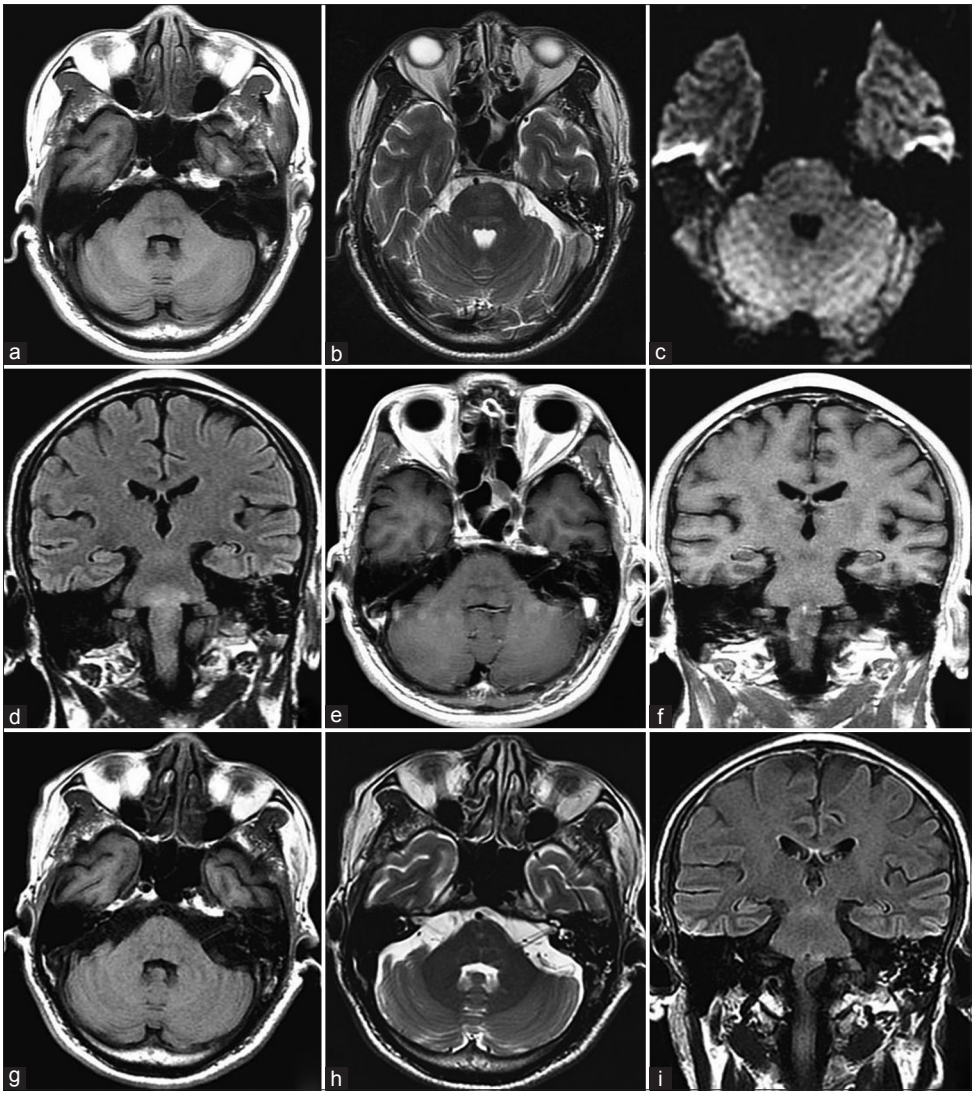

Figure 1: On admission, brain magnetic resonance imaging (MRI) shows the lesion in the basis pontis and medulla oblongata: T1-weighted hypointense (a), T2-weighted (b) and fluid attenuation inversion recovery (FLAIR) hyperintense (d), and slightly diffusion-weighted hyperintense (c). Contrast-enhanced T1-weighted MRI showed spot-like enhancement in medulla oblongata as well as enhanced left facial nerve (e and f). One month later, follow-up brain MRI demonstrates that the extent of the lesions has decreased on T1-weighted, T2-weighted images and FLAIR (g–i).

geniculate ganglia, upward thread porus acusticus internus along with facial canal, and eventually enter intracranially and firstly invade basis pontis. Meanwhile, VZV may also spread downwards along with general somatosensory fibers to the skin of external auditory canal resulting in herpes zoster formation. To the best of our knowledge, coexistence of RHS with VZV brainstem encephalitis is extremely rare. This case may widen our knowledge for the mechanism of VZV infection spreading into CNS.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Kim JH, Chung PW, Oh S, Hong SB, Chung CS, Jung CW, *et al.* Ramsay Hunt syndrome complicated by a brainstem lesion. J Clin Virol 2007;39:322-5.
2. Nagel MA, Cohrs RJ, Mahalingam R, Wellish MC, Forghani B, Schiller A, *et al.* The varicella zoster virus vasculopathies: Clinical, CSF, imaging, and virologic features. Neurology 2008;70:853-60.
3. Grahn A, Studahl M. Varicella-zoster virus infections of the central nervous system – Prognosis, diagnostics and treatment. J Infect 2015;71:281-93.
4. Koskiniemi M, Piiparinen H, Rantalaiho T, Eränkö P, Färkkilä M, Räihä K, *et al.* Acute central nervous system complications in varicella zoster virus infections. J Clin Virol 2002;25:293-301.
5. Mailles A, Stahl JP; Steering Committee and Investigators Group. Infectious encephalitis in France in 2007: A national prospective study. Clin Infect Dis 2009;49:1838-47.