

Multiple cranial nerve palsies caused by varicella zoster virus in the absence of rash

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Abstract

Varicella zoster virus (VZV) is a widespread human herpesvirus which causes chickenpox (varicella). VZV infection can produce a broad spectrum of neurologic disorders including multiple cranial nerve palsies. Among the cranial nerves, trigeminal and facial nerves are the most commonly involved. However, multiple lower cranial nerve palsies caused by VZV infection is very rare. It is a diagnostic dilemma for VZV infection if there are no skin or mucosa lesions, which are characteristic signs of VZV infection. We report the case of a 54-year-old man with sudden right hearing loss, sore throat, odynophagia, hoarseness, dysphagia and vertigo. Pure tone audiometry revealed right sensorineural hearing loss and laryngoscopy proved that there were paralyses of the right upper pharyngeal constrictor muscle and vocal cord. These findings were consistent with acute unilateral vestibulocochlear, glossopharyngeal and vagus nerve palsies. There were no skin or mucosa lesions noted in our patient. The diagnosis was assisted by the existence of anti-VZV serum IgM even if the polymerase chain reaction result was negative. The symptoms of the patient improved after receiving anti-viral therapy. We emphasize the role of VZV infection in multiple cranial nerve palsies and the importance of serologic test.

INTRODUCTION

Varicella zoster virus (VZV) is a widespread human herpesvirus. VZV infection can produce a broad spectrum of neurologic disorders including multiple cranial nerve palsies. Ramsay Hunt syndrome, the most common VZV-related cranial nerve palsy, is characterized by facial palsy, auricle vesicular eruptions and possible vestibulocochlear nerve palsy. Other combinations of multiple cranial nerve palsies caused by VZV infection based on different biomechanisms and anatomic positions have been raised.^{1,2} Skin or mucosa lesions such as vesicles may be accompanied as a characteristic sign but they may not occur (zoster sine herpette). Here we present a rare case with unilateral vestibulocochlear, glossopharyngeal and vagus nerve palsies due to VZV infection without any skin or mucosa manifestations.

CASE REPORT

A 54-year-old man developed sore throat, odynophagia and hoarseness and visited local clinic for help. Cold medication was prescribed but his symptoms persisted. One week later, dysphagia, choking, vertigo, right hearing impairment and right tinnitus occurred and he

consulted our emergency department two weeks later.

Initial vital signs revealed no fever or tachycardia. His conscious was clear. Physical examination revealed no skin rash or mucosal eruptions within the regions of his ears, face, or oral cavity. Upon neurologic examination, there was paralysis of the right soft palate. No meningeal signs or long tract signs was noted. Nasopharyngolaryngoscopy revealed that there were no mucosal eruptions on pharynx and larynx. There were paralyses of the right upper pharyngeal constrictor muscle and vocal cord (Figure 1). Pure tone audiogram revealed right sensorineural hearing loss (SNHL) (Figure 2a). Laboratory tests showed a white blood cell (WBC) count of 5900/mm³ with 60.1% neutrophils, 29.3% lymphocytes, and 9.4% monocytes. C-reactive protein was slightly increased (2.12 mg/dL). Cranial magnetic resonance imaging with gadolinium enhancement revealed there were no abnormal findings in his brain and posterior cranial fossa. These findings were consistent with acute unilateral vestibulocochlear, glossopharyngeal and vagus nerve palsies.

Serological testing was performed via enzyme immunoassay (EIA). It revealed positive results

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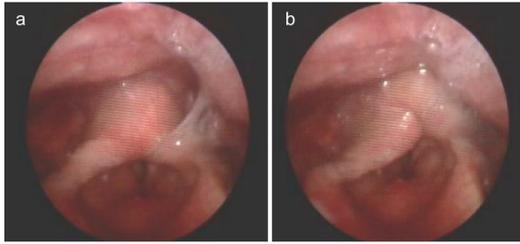


Figure 1. Laryngoscopy reveals paralysis of right vocal cord. **a** closure phase; **b** open phase.

for VZV immunoglobulin M (IgM), consistent with acute VZV infection. The cerebrospinal fluid (CSF) revealed a red blood cell count (RBC) of 14100/mm³, a WBC count of 325/mm³, a slightly elevated protein level of 173.3 mg/dl and a normal glucose level. The CSF result was thought to reflect a traumatic tap. VZV IgM (EIA) was negative in CSF. VZV was not found in CSF by fluorescent antibody stain. Polymerase chain reaction (PCR) did not detect VZV DNA in the CSF and in the serum.

After the serologic confirmation of acute VZV infection, the patient was treated with intravenous acyclovir(1500mg/day) for 7 days and then oral valacyclovir(4000mg/day) for 10 days. His symptoms were gradually improved after the administration of intravenous acyclovir and corticosteroid was added as an adjuvant therapy. The follow-up CSF test showed a decreased WBC count(7/mm³) and a reduced protein level(53.3 mg/dL). The follow-up pure tone audiogram revealed recovery of right SNHL (Figure 2b)

and the follow-up nasopharyngolaryngoscopy also showed recovery of paralysis of right vocal cord. Three months later, the follow-up serologic testing showed negative VZV IgM and positive VZV IgG, compatible with the previous diagnosis of acute VZV infection.

DISCUSSION

Several diagnostic tests are believed to be indications of active VZV infection: the detection of anti-VZV IgM antibody in serum or CSF or anti-VZV IgG antibody in CSF, or the presence of VZV DNA in blood mononuclear cells or CSF.³ Anti-VZV serum IgM antibody appeared 8-10 days after the appearance of skin or mucosal lesions and this indicated its limited use in very acute stage. However, anti-VZV serum IgM antibody can be detected more than 1 month after the beginning of the symptoms.⁴ In contrast, blood PCR for VZV DNA detection has high sensitivity and specificity at the start of the disease but its sensitivity decreases with time.⁵ Once symptoms start, VZV-DNA cannot be detected by PCR after several weeks. Anti-VZV serum IgM antibody has lower sensitivity; however, it is highly suggestive acute VZV infection when detected.

In our patient, the presence of anti-VZV serum IgM established the diagnosis of acute VZV infection. The improvement of the patient's symptoms after the administration of antiviral agent was also supportive of the diagnosis.³ A negative PCR in both the serum and CSF in our patient after more than two weeks of symptom

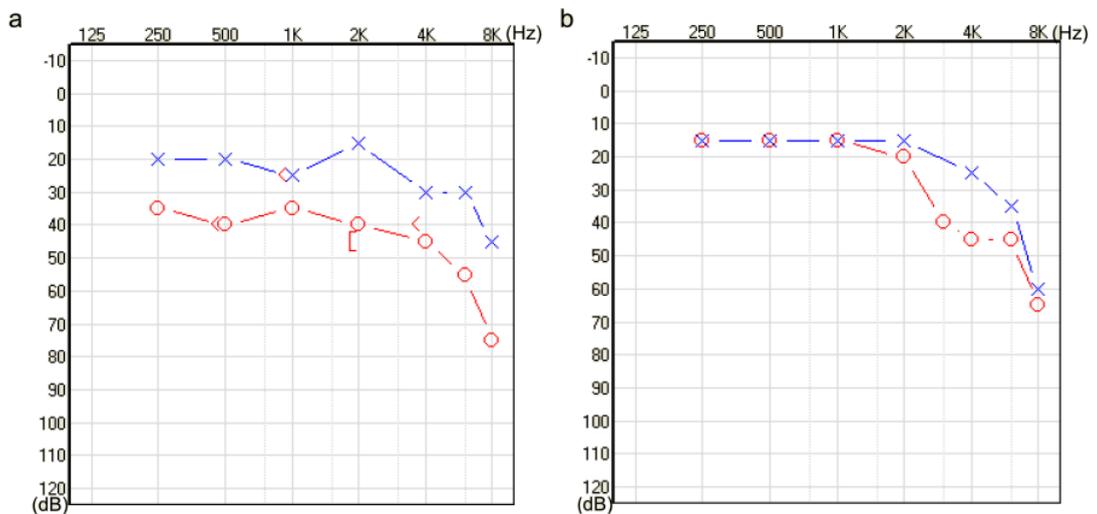


Figure 2. Pure tone audiometry. **a** normal hearing in the right ear and a moderate degree of sensorineural hearing loss with pure tone average of 48 dB in the left ear; **b** improvement of right ear sensorineural hearing loss during remission.

onset was not in conflict with the diagnosis, because the sensitivity of PCR for VZV DNA decreases with time. The examination of anti-VZV IgG was qualitative but not quantitative in our hospital, and patients with prior varicella infection will have a positive result. The fluorescent antibody stain to detect VZV was negative in our patient and it can serve as adjuvant diagnostic method due to its relative low sensitivity.

The cornerstone of treatment of VZV infection is antiviral agents, acyclovir, valacyclovir, and famciclovir. The use of antiviral agents is especially important for patients in immunocompromised status or other neurologic complications. They reduce the severity and duration of pain, limit complications and the risk of dissemination of VZV. Antiviral agents in combination with corticosteroid lead to a better outcome than antiviral agents alone.⁶ Besides, reports stated that there is better prognosis when treatment is initiated within 3 days after the start of the symptoms.

In conclusion, we reported here a rare case of VZV infection with unilateral eighth, ninth, tenth cranial nerve palsies. There was no skin or mucosal manifestations. The cranial nerve palsies was attributed to VZV infection, based on positive anti-VZV serum IgM.

DISCLOSURE

Conflict of interest: None

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