Unusual presentation of more common disease/injury

Where is reactivation after a long latency?

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Summary

The authors present a 90-year-old woman with unilateral glossopharyngeal, vagal and spinal accessory cranial nerve palsy along with pharyngeal and laryngeal vesicular eruptions. She was diagnosed with herpes zoster based on PCR testing on vesicular fluid for varicella-zoster virus (VZV). Reactivation of VZV in the head and neck region can cause life-threatening neurologic sequelae. Clinicians should be alert to the possibility of herpes zoster in a case of unilateral multiple cranial neuropathies and rapid combination therapy with acyclovir and corticosteroid should be initiated.

BACKGROUND

Herpes zoster, also known as shingles, results from reactivation of latent varicella-zoster virus (VZV) infection within the ganglia of the spinal cord and cranial nerve (CN).¹ In the head and neck region, the most common presentation is Ramsay Hunt syndrome (RHS), presenting with ipsilateral facial nerve (VII) palsy and vesicular lesions in the auditory canal and auricle, with and without involvement of the vestibulocochlear nerve (VIII).¹ In addition, it may also associate with various combinations of multiple cranial neuropathies including glossopharyngeal (IX) and/or vagal (X) nerve palsy.²⁻³ However, the possibility of herpes zoster in a case of laryngeal paralysis without VII CN involvement may be overlooked or diagnosed as idiopathic. Here, we report a case with VZV-induced laryngeal paralysis without RHS.

CASE PRESENTATION

A 90-year-old woman presented with a 2-day history of sore throat, odynophagia and hoarseness. She had no neurological or immunodeficiency condition including immunosuppressant drug use. On inspection, painful skin eruptions were noted in the left jaw and cheek without the presence of CN VII or VIII involvement. In addition, there were vesicular eruptions in the soft palate and posterior oropharynx (figure 1). The pharyngeal wall shifted to the right on phonation (curtain sign). Muscle weakness of the left-side sternocleidomastoid and trapezius muscles was evident. Laryngoscopy showed vesicular eruptions spreading from the left side of the posterior hypopharyngeal wall to the same side of epiglottis, aryepiglottic fold and piriformis sinus (figure 2). The left vocal cord was paralysed in the paramedian position with impaired pharyngeal wall contraction and laryngeal elevation. On the basis of these findings, she had the IX, X and spinal accessory (XI) CN palsy.

INVESTIGATIONS

Since the clinical presentation was suggestive of herpetic infection, serology of enzyme immunoassay (EIA) testing and PCR for VZV and herpes simplex virus (HSV) were

examined. Specimens of exudates for PCR were obtained from the posterior oropharyngeal lesions. EIA testing using paired sera showed a greater than fourfold increase in VZV IgG titer with a positive result of PCR for VZV. EIA testing for HSV showed a previous infection but a negative result of PCR for HSV. Brain MRI study with gadolinium enhancement revealed no remarkable abnormal findings of the central nervous system.

DIFFERENTIAL DIAGNOSIS

- ▶ Brainstem infarction
- ▶ Laryngeal neoplasm
- ▶ Laryngeal tuberculosis
- ▶ Laryngeal abscess
- ▶ Laryngeal granulomatous disease
- ▶ Laryngeal fungal infection

Figure 1 On inspection of the oral cavity, vesicular eruptions were noted in the soft palate and posterior oropharynx.

Figure 2 Laryngoscopy showed vesicular eruptions spreading from the left side of the posterior hypopharyngeal wall to the same side of epiglottitis, aryepiglottic fold and piriformis sinus.

TREATMENT

Just after the admission, intravenous administration of acyclovir of 750 mg for 7 days and corticosteroid of 60 mg with subsequent tapering the dose over 14 days was initiated empirically.

OUTCOME AND FOLLOW-UP

As a result of the treatment, the skin and mucosal lesions disappeared, and then the curtain sign and impaired pharyngeal wall contraction and laryngeal elevation were gradually resolved. However, after the onset, she suffered from repeated aspiration pneumonia and progressive nutritional disturbance because the vocal cord paralysis never recovered. She eventually died of respiratory insufficiency 1 year after the onset.

DISCUSSION

The relation between age and incidence of herpes zoster

Reactivation of VZV is presumably due to the decline in the host's cell-mediated immunity, usually in older and immunocompromised individuals.⁴ Of these, increasing age is the most significant risk factor for herpes zoster and its incidence progressively increases with age. For example, zoster incidence in persons aged 80 to 89 years is 10 times greater than that in children aged less than 10 years⁴ and approximately one-half of those who live to 85 years of age will experience an episode of zoster.⁴

The clinical presentation of herpes zoster in the head and neck region

VZV infection in the head and neck region could cause a spectrum of symptoms^{2-3 5}: RHS with multiple cranial neuropathies involving various combinations of CNs IX, X and others with herpetic mucosal eruptions in the pharyngolaryngeal region; RHS with skin eruptions in the C2-C3

cervical dermatomes; RHS with multiple cranial neuropathies but lacking any mucosal lesions; Multiple cranial neuropathies with neither RHS symptoms nor mucosal lesions; Multiple cranial neuropathies with mucosal lesions but lacking RHS symptoms; Mucosal lesions only, etc. Accordingly, clinicians should be alert to the possibility of VZV infection in a case of unilateral multiple cranial neuropathies regardless of whether herpetic skin or mucosal lesions are present.

The possible pathogenic mechanism of various CN involvements

The pathogenic mechanism of various combinations of CN involvements remains uncertain, several hypotheses have been proposed.^{1 5} First, multiple CN involvements may be due to the anatomical CN location. Hunt surmised that the gasserian, geniculate, petrous, accessory, jugular, plexiform and C2-C3 dorsal root ganglia comprised a chain in which inflammation of a single ganglion could extend to proximate ganglia. Numerous anatomical interconnections between CNs VII and VIII to XII may explain the simultaneous involvements of the CNs and cervical nerve by dissemination of VZV. Second, VZV infection may induce the selective vulnerability of blood supply to the CNs. The CNs IX to XII are supplied by the ascending pharyngeal artery, and the CN VII and maxillary branch and mandibular branch of the trigeminal nerve (V) are supplied by the middle meningeal artery.⁶ Third, reactivation of VZV may simultaneously occur in multiple ganglia, which is based on the evidence that the presence of VZV DNA was identified in multiple CNs, dorsal root ganglia and celiac ganglia.⁷

The diagnosis and treatment of herpes zoster

PCR to detect VZV in exudates is more sensitive and rapid diagnostic technique compared with conventional serologic tests.² Despite the lack of prospective randomised trials for RHS, a retrospective review showed a significantly neurological improvement in patients treated with combination acyclovir and corticosteroid within 3 days of onset.⁸

Our comments of the present case

Given the multiple CN involvements along with herpetic skin or mucosal eruptions, the present case was likely to develop herpes zoster by simultaneous VZV reactivation in each CN or dissemination of VZV via anatomical CN interconnections rather than simple extension of CN inflammation or selective insufficient blood flow to the CNs. Despite early combination therapy was initiated, the vocal cord palsy was unfortunately irreversible presumably because the recurrent laryngeal nerve travels a long distance from the ganglion of CN X to the innervated muscles. Our experience in the present case indicated that herpes zoster in the head and neck region could result in life-threatening cranial neuropathies, especially in an older patient.

CONCLUSION

Herpes zoster in the head and neck region can cause various CN palsies. Herpes zoster should be in the differential diagnosis in a case of unilateral multiple cranial neuropathies regardless of whether herpetic skin or mucosal lesions

are present. PCR to detect VZV in exudates is more sensitive and rapid diagnostic technique. An early combination therapy with acyclovir and corticosteroid may reduce the chance of the neurologic sequelae. However, vocal cord palsy can be refractory to the treatment and life-threatening, especially in older patients.

Learning points

- ▶ Herpes zoster in the head and neck region can cause various CN palsies.
- ▶ Herpes zoster should be in the differential diagnosis in a case of unilateral multiple cranial neuropathies regardless of whether herpetic skin or mucosal lesions are present.
- ▶ PCR to detect VZV in exudates is more sensitive and rapid diagnostic technique.
- ▶ An early combination therapy with acyclovir and corticosteroid may reduce the chance of the neurologic sequelae.
- ▶ Vocal cord palsy can be refractory to the treatment and life-threatening, especially in older patients.

Competing interests None.

Patient consent Obtained.

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