Cranial polyneuropathy caused by varicella zoster reactivation

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In May 2015, a 69-year-old woman presented to our hospital with a history of 1 week of dysphagia and dysphonia, 5 days of right sided otalgia, and electric shock-like pains from her right ear to forehead. Two days prior, she had been assessed at a different hospital, and diagnosed with trigeminal neuralgia. Aside from the neuralgia, neurological examination at that time was reported as normal, and no cause for her dysphagia was identified. She had no prior neurological deficits or reasons for immunocompromise. She had varicella infection during childhood.

On arrival at our hospital, nasendoscopy identified reduced right vocal fold movement, but no foreign body. The following day, she developed a right facial droop and vesicles were observed on her right palate. Neurological examination identified several signs, all right-sided: a lower motor neuron facial palsy, decreased facial sensation to light touch (V1–V3 distribution), uvula deviation to the left, reduced hearing and abducens palsy. The right corneal reflex was preserved. She had mild gait ataxia, but otherwise the remaining neurological examination was normal.

Head CT and MRI with gadolinium enhancement showed no evidence of stroke or cranial nerve abnormality. CSF examination revealed moderate lymphocytic pleocytosis with 71 x 10⁶/L white blood cells, all mononuclear, and VZV was detected by PCR. CSF glucose was 3.4mmol/L and total protein 0.46g/L. Flow cytometry and anti-nuclear antibody testing was normal. An audiogram confirmed right-sided, mild-to-severe sensorineural hearing loss. Videofluoroscopic swallow showed global reduction in the pharyngeal phase of swallowing.

We diagnosed VZV-induced cranial polyneuropathy, involving cranial nerves V, VI, VII, VIII, IX and X. The patient was commenced on intravenous acyclovir 10mg/kg, 3 times daily and prednisone 60mg daily for 2 weeks. We managed her right ocular symptoms with topical lubrication and an eye-patch. Given ongoing dysphagia, she required nasogastric feeding. One week later, repeat CSF analysis showed a decrease in white cells to 24 x 10⁶/L and VZV was negative by PCR.

She had significant hearing loss in the right ear and her post herpetic neuralgia persisted with diminished severity. Her post herpetic neuralgia persisted with diminished severity. Her gait normalised prior to discharge.

Discussion

The most common cranial nerve manifestation of VZV infection is Ramsay Hunt Syndrome (RHS). This characteristically involves herpetic skin lesions in the auditory canal, or auricle and unilateral peripheral facial palsy with or without auditory or vestibular symptoms. While RHS is the most common presentation, VZV infection in the head or neck can result in various combinations of cranial mono- or polyneuropathies.

VZV infection of the larynx or pharynx with multiple associated cranial neuropathies frequently starts with a mononeuropathy, the initial manifestations of which are typically dysphagia, odynophagia or dysphonia. In this case, the patient's dysphagia was originally considered in isolation from her other problems. While her infection remained untreated, she developed additional symptoms that indicated a clear progression of unilateral cranial nerve involvement, starting with CN IX and X,
extending to include CN V–VIII. Identifying palatal vesicles helped focus our investigations towards detecting VZV infection, but vesicles are not always present. Brain imaging was useful in excluding key differential diagnoses, such as brainstem stroke or malignancy.

This case is highly atypical. The last cranial nerve affected, the abducens nerve, is very rarely associated with VZV infection and we found no prior reports of the particular combination of cranial neuropathies observed in our patient. Diagnostic uncertainty and delayed presentation to the neurology service led to late initiation of treatment.

Evidence regarding the optimal treatment of cranial neuropathy caused by VZV is derived from treatment studies of RHS. Despite a lack of randomised control trials, clinicians generally favour combined treatment with acyclovir and corticosteroids. The lack of significant improvement in our patient at 2-month follow-up supports published evidence that treatment within 72 hours is more effective.

This case highlights the importance of considering atypical presentations of common diseases when a patient presents with an unusual constellation of neurological symptoms. A high index of suspicion is required to detect early signs of other pathology. Early neurological consultation is suggested to assist with these steps.

**Competing interests:**
Nil

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