

Herpes zoster oticus masquerading as lateral pontomedullary syndrome

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Abstract

Ramsay Hunt syndrome, also known as herpes zoster oticus is usually caused by reactivation of pre-existing varicella zoster virus in the geniculate ganglion. Here we report a case of herpes zoster oticus masquerading as lateral pontomedullary syndrome occurring during the pre-eruptive phase of chicken pox. The case is being reported for its rare and unusual clinical presentation.

INTRODUCTION

Ramsay Hunt syndrome was first described in 1907 by James Ramsay Hunt in a patient who had otalgia associated with mucocutaneous rashes, which he ascribed to infection of the geniculate ganglion by human herpesvirus 3. The syndrome comprises of an acute infranuclear facial neuropathy associated with erythematous vesicular rash of the skin of the ear canal, auricle (also termed herpes zoster oticus), and/or mucous membrane of the oropharynx.

Various clinical features reported with this syndrome include herpes auricularis, herpes facialis and herpes occipito-collaris with facial palsy. Cranial nerves VIII, IX, V, and VI are less commonly involved and when they do get involved, they are affected in descending order of frequency as listed above. The infection gives rise to vesiculation and ulceration of the external ear and ipsilateral anterior two thirds of the tongue and soft palate with ipsilateral infranuclear facial palsy and geniculate ganglionopathy.¹ Varicella zoster virus (VZV) infection causes two distinct clinical syndromes. Primary infection, also known as varicella or chickenpox, is a common erythematous disease characterized by a highly contagious generalized vesicular rash in a centripetal pattern. After chickenpox, VZV remains latent in the neurons of cranial nerves and dorsal root ganglia. Subsequent reactivation of latent VZV can result in localized vesicular rash, known as herpes zoster. VZV infection or reactivation involving the geniculate ganglion of the facial nerve within the temporal bone is the main pathophysiological mechanism of Ramsay

Hunt syndrome. Ramsay Hunt syndrome is a rare complication of latent VZV infection.² VZV has been detected by polymerase chain reaction (PCR) in the tear fluid of patients with Bell's palsy.³ Ramsay Hunt syndrome is rare below the age of 6 years and accounts for 16% of all causes of unilateral facial palsies in children and 18% of facial palsies in adults.⁴

CASE REPORT

A 38 years old normotensive, non-diabetic, immunocompetent, female presented to the emergency room with history of acute onset vertigo with gradual worsening during the initial 48 hours of its onset. It was associated with intractable vomiting, gait ataxia, weakness of left side of face and impaired hearing on left side. There was no history of preceding fever or sore throat. On examination the patient had gaze-evoked nystagmus with torsional component. It was associated with left infranuclear facial palsy, gait ataxia to left along with left palatal palsy. On the sixth day of admission, the patient developed vesicular rash in centripetal distribution over whole body including the left external auditory canal and soft palate which gradually resolved over the next four days. However, the rash over the left external auditory canal resolved after one week (Figure 1). Moreover, she had no risk factors for stroke or coronary artery disease. MRI (1.5 Tesla) of brain (with gadolinium) revealed subtle enhancement of vestibulocochlear complex. There was no radiological evidence of any infarct on diffusion weighted images. However, thin section T1 weighted images could not be done. Pure tone

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Figure 1. Vesicular rash of herpes zoster on external auditory canal

audiometry revealed left sided sensorineural hearing loss. Her serum chemistry was normal. CSF analysis revealed acellular fluid with mild elevation in proteins (86mg%) and positive PCR for varicella zoster virus. Smear from floor of the vesicle stained with Giemsa stain showed degenerating cells with multiple nuclei suggestive of VZV infection. Subsequently, confirmation was made by detection of IgM antibodies to VZV by ELISA test. HIV serology was nonreactive. With aforementioned clinical and laboratory parameters, a diagnosis of herpes zoster oticus masquerading as left pontomedullary syndrome was entertained.

DISCUSSION

Ramsay Hunt syndrome as a consequence of reactivation of VZV is well documented. Multiple cranial nerve involvement secondary to varicella infection too has been reported.^{5,6} Similarly cerebellitis is an uncommon complication of this infection.⁷ However, the occurrence of vestibulocochlear features prior to the appearance of vesicular rash is an uncommon occurrence.⁸ Our patient was conspicuous by predominant

vestibulocochlear features which preceded the appearance of VZV rash by six days. In addition, facial nerve and palatal palsy (9th and 10th cranial nerves) can be attributed to involvement of these structures or their nuclei by the same infection during their intracranial course. Elevated cerebrospinal fluid protein can result from meningeal inflammation following intracranial spread of VZV infection. Vestibulocochlear symptoms outweighed other symptoms of this syndrome, thereby suggesting VZV reactivation in the spiral and vestibular ganglion.⁹ It is argued that predominance of vestibulocochlear symptoms resulted from VZV transmission across the nerves in the internal auditory canal. Stroke due to varicella zoster induced vasculopathy is ruled out by normal neuroimaging. Moreover, patient had no risk factors for stroke. To conclude, VZV infection may sometimes pose diagnostic challenge by unusual presentation in the form of predominant vestibulocochlear features along with lower cranial nerve palsy, especially in the absence of vesicular rash thereby mimicking lateral pontomedullary syndrome.

DISCLOSURE

Conflict of interest: Nil

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