Ramsay Hunt Syndrome with Multiple Cranial Neuropathy and Meningitis: A Rare Presentation of Varicella Zoster Virus Infection

Naveen Seervi

Abstract: Ramsay Hunt Syndrome also called as herpes zoster oticus, is a rare complication of reactivation of varicella zoster infection in geniculate ganglion of facial nerve. It usually presents with painful vesicular eruption over pinna and external auditory meatus and lower motor neuron (LMN) type of facial palsy. However recently we came across a case of 51 years old female of Ramsay hunt syndrome with left sided 7th, 8th, 9th, 10th cranial neuropathy and VZV meningitis. This clinical presentation is rare to best of our knowledge. The patient was treated with acyclovir and oral steroids, and responded well.

Keywords: Ramsay Hunt syndrome, herpes zoster oticus, Varicella zoster virus, cranial neuropathy, meningitis

1. Introduction

Varicella zoster virus (VZV) is a DNA virus of the Herpesviridae family. Primary infection of VZV produces chickenpox, which generally occurs in childhood. After initial infection, VZV establishes lifelong latency in cranial nerve and dorsal root ganglia, and can reactivate years to decades later as herpes zoster (HZ) or “shingles”. Shingles are common presentation of VZV reactivation and present as painful vesicles following a segmental skin distribution. Ophthalmic division of the trigeminal nerve (Herpes zoster ophthalmicus) and facial nerve (Herpes zoster oticus) involvement are the two most commonly known clinical presentations regarding cranial nerves involvement.

Ramsay Hunt syndrome (RHS) is caused by reactivation of previous varicella zoster virus infection in geniculate ganglion of the facial nerve. It was first described by James Ramsay Hunt in 1907. It usually presents as pain in and around the ear and tonsillar region followed by vesicular eruption over pinna and in external auditory meatus and ipsilateral LMN facial palsy. Other common symptoms include ipsilateral loss of taste sensation, hearing impairment and vertigo.

Reports on this syndrome with multiple cranial neuropathy[2,3,4] and meningitis[5] are very rare in immunocompetent individuals. We hereby report a rare case of RHS with ipsilateral 7th, 8th, 9th and 10th cranial neuropathy and meningitis.

2. Case Report

A 51 years old lady presented with moderate to severe intensity pain around left ear for 10 days, multiple vascular eruption over left pinna and in external auditory meatus, deviation of angle of mouth towards right side, inability to close left eye for three days, headache and vomiting for three days. On the day of hospitalization she had developed hoarseness of voice and difficulty in swallowing.

On examination, she was conscious and oriented, her vital signs were stable. Multiple vesicular eruptions were present on left pinna and external auditory meatus (Figure 1). She had left sided LMN facial palsy (Figure 2, 3) (Grade V according to House-Brackmann grading scale of facial function). Along with this, there was left sided 8th, 9th and 10th cranial nerve palsies.

On systemic examination she had signs of meningeal irritation like neck rigidity and Kernig’s sign. Other systemic examination was normal. Her routine blood investigation like complete blood counts, fasting blood sugar, renal and liver function tests were normal. HIV was non reactive. MRI brain with gadolinium contrast was normal, except loss of pneumatisation of left temporal bone- probably due to inflammatory lesions (Figure 4). Her audiogram showed moderate to severe mixed hearing loss on left side.

CSF study revealed elevated opening pressure and lymphocytic pleocytosis (240cells/mm3; 80% lymphocytes). CSF protein was 43 mg/dL and CSF glucose was 53 mg/dL. CSF VZV DNA PCR was positive.

She was treated with intravenous acyclovir (30mg/kg/day) for 10 days followed by oral acyclovir (800mg five times a day) for additional four days. Along with it she was also put on oral prednisolone 60mg per day for initial 1 week with gradually tapering over next 2 weeks. Symptomatic treatment was given in form of analgesic, acyclovir ointment for local application, carboxy methylcellulose eye drops and physiotherapy. She showed improvement in dysphagia and hoarseness on 7th day of treatment and in facial palsy on 10th day. She was discharged on 11th day of admission. On follow up visit after next 15 days, her vesicles were completely healed. She had no dysphagia and hoarseness; on examination her uvula was central and palatal arches were symmetrical. Facial palsy was improved to grade 3. She was given symptomatic therapy and on 1 month follow up after discharge she was fully recovered.

3. Discussion

RHS forms the second most common cause of unilateral peripheral facial palsy after Bell’s palsy. Facial nerve involvement is initially due to inflammation caused by the
viral neuronitis and secondarily to the facial nerve edema.\(^5\)
The prognosis for facial palsy is poorer in RHS than in idiopathic forms.

Only few literatures reported uncommon clinical features. Chang et al, reported incidence of hearing loss in patients with RHS is 76\% and is more severe in the high frequency range than in the low frequency range.\(^6\) Adachi M reported glossopharyngeal and vagal nerve palsies due to herpes zoster.\(^7\) Lauridsen AG et al reported herpes zoster oticus with 5\(^{th}\), 7\(^{th}\), 8\(^{th}\), 9\(^{th}\), 10\(^{th}\), and 12\(^{th}\) cranial nerve palsy.\(^8\)

The involvement of multiple cranial nerves is thought to be due to anastomosis among 5\(^{th}\), 7\(^{th}\), 9\(^{th}\) and 10\(^{th}\) cranial nerves. It has also been explained as a consequence of an inflammation-induced infarction of a small vessel knowing that a small branch of the carotid artery supplies two or three contiguous nerves.\(^9\)

The coexistence of RHS and varicella zoster encephalitis and meningitis is rare in immunocompetant individuals. Concomitant diseases such as diabetes and chronic renal failure may lead to an aggressive course of infection and can predispose to encephalitis and meningitis.\(^{[8,9]}\) Nagano K et al reported a case of RHS with local meningitis, 5\(^{th}\), 6\(^{th}\), 7\(^{th}\), 8\(^{th}\) cranial nerve palsies and second cervical nerve involvement.\(^{[4]}\) The possible mechanism of VZV spreading to CNS is reactivated viruses, which establish latency in geniculate ganglia, upward spread along with the facial canal, and eventually enter intracranially.

In the treatment of RHS, the combination of acyclovir and prednisone is the most recommended therapy.\(^{[10]}\) Although the optimal dosage of acyclovir and prednisone has not been established and has varied in previous studies. We used IV acyclovir (30mg/kg/day) for first 10 days followed by oral acyclovir (800mg five times a day) for additional for days with oral steroids with tapering doses over 3 weeks.

### 4. Conclusion

RHS commonly presents with painful vesicular eruption over pinna and in external auditory meatus, vertigo and hearing impairment with ipsilateral LMN facial palsy. Other lower cranial nerves involvement and meningitis can be seen rarely, as in this case. Diagnosis of suspected VZV CNS infection should be based on CSF analysis for the detection of VZV DNA by PCR and/or intrathecal antibody production. Here we highlight that early and prompt initiation of therapy in such cases improves the prognosis.

**Figures**

**Figure 1:** Vesicular eruptions over left pinna

**Figure 2:** left LMN facial palsy: deviation of angle of mouth to right
Figure 3: Bell’s phenomenon on left side on closing the eyes.

Figure 4: T2 weighted axial MRI image shows partial loss of pneumatisation of left temporal bone (shown by arrow)

References


