Unilateral Cranial Polyneuropathy in Herpes Zoster Oticus: Infection teaching us Anatomy

Abdul Qavi DM1, Ashutosh Tiwari DM2, Pradeep Kumar Maurya DM3, Ajai Kumar Singh DM4, Pradeep Kumar MD5

1,2,3,4,5 Department of Neurology, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

Date of submission: 29th May 2020  Date of acceptance: 16th November 2020  Date of publication: 1st December 2020

Abstract
Herpes zoster oticus or Ramsay Hunt syndrome is an uncommon neurological manifestation of herpes virus infection causing external ear rash with otalgia and facial nerve palsy. Rarely herpetic infection may present with multiple cranial nerves palsies involving VII, VIII, IX and X cranial nerves. Here we report a case of herpes zoster oticus with multiple cranial nerve palsy. This case study will help in understanding the dermatomal distribution of cranial nerves with cranial polyneuropathy due to reactivation of neurotropic herpes virus.

Some interesting case reports regarding different cranial nerve involvement in herpetic infection are discussed in the table which helps in understanding the neurotropism of herpes virus.

Key words: Cranial Polyneuropathy, Ear rashes, Facial palsy, Herpes Zoster oticus

Introduction

Herpes zoster oticus or Ramsay Hunt syndrome is an uncommon neurological manifestation of herpes virus infection causing external ear rash with otalgia and facial nerve palsy.1 Sometimes this herpetic infection may present with multiple cranial nerves palsies involving VII, VIII, IX and X cranial nerves.2

Case report

A 55 years old man presented to us with right ear pain, deviation of angle of mouth to left side and unsteadiness while walking. First, he developed ear pain and on the next day he noticed rashes around right ear involving pinna. On the same day he developed a nasal twang of voice with nasal regurgitation. He was complaining of tinnitus in right ear with difficulty in walking and swaying on the third day of illness. On examination there was lower motor neuron palsy of right cranial nerve VII with facial deviation to the left side and involvement of right sided cranial nerve VIII including both vestibular and cochlear divisions. Gag reflex on the right side was absent with drooped soft palate depicting right sided cranial nerves IX and X palsies. Local examination revealed herpetic rashes and crust on the right pinna, external ear canal and on right sided soft palate (Figure 1).

There was no neck rigidity and signs of meningeal irritation were absent. On higher mental function examination, the patient was conscious and oriented to time place and persons, alert and following commands with intact memory. Cerebellar examination was normal but the patient had vestibular ataxia. The clinical features of meningitis and/or encephalitis were absent.

Patient was admitted and Ryle’s tube was inserted for feeding and oral medications. In routine blood investigation he was found to be recently diagnosed
diabetes mellitus. His CSF examination was showing lymphocytic pleocytosis (WBC count was 45 cells/mm$^3$, polymorphs 10%, lymphocytes 90%) with slightly raised proteins (95.0 mg/dl) and normal sugar level (82 mg/dl with corresponding blood sugar was 156 mg/dl). CSF was clear watery in appearance and ADA level was 1.1. CSF PCR for Herpes Simplex Virus and Varicella Zoster virus were negative. MRI brain with contrast was also normal without any evidence of meningeal enhancement or parenchymal intensity changes. The abnormality in the CSF examination can be explained by immunological reaction to reactivation of latent herpes virus infection presenting in the form of Herpes zoster oticus. Patient was treated with antiviral oral valacyclovir 1 gm three times a day for seven days, acyclovir ointment for local application, prednisolone 1 mg/kg with tapering and oral hypoglycemic agents monitoring sugar levels and physiotherapy. The patient improved symptomatically allowing removal of Ryle’s tube and reduction in pain, rashes and gait difficulty in two weeks of treatment. However residual facial palsy was present on the last follow up (three months after discharge).

Figure 1. (A) Rashes on right palate, (B) Resolving rashes and crusting over pinna and neck, (C) Right palatal palsy, on attempt to phonation flat and drooped right soft palate with median raphe deviated to left, (D) Right sided LMN facial palsy
<table>
<thead>
<tr>
<th>S.N.</th>
<th>Author</th>
<th>Age/ Sex</th>
<th>Risk Factors/ Co-morbidity</th>
<th>Clinical Features</th>
<th>Cranial Nerve Involved</th>
<th>Treatment given</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Sato K. et al. 1991</td>
<td>56y/ female</td>
<td>none</td>
<td>Right ear and facial pain, respiratory distress, fatigue</td>
<td>V, VII, VIII, IX, X, XI and XII</td>
<td>Ventilatory support, Antiviral, steroid</td>
<td>Improved with late recovery of hoarseness of voice</td>
</tr>
<tr>
<td>2</td>
<td>Kikuchi H. et al 1995</td>
<td>50y/ male</td>
<td>none</td>
<td>Left sided tinnitus, hoarseness, dysphagia, facial deviation</td>
<td>Left VII, IX, X, XI and right VII, IX, X</td>
<td>Antiviral, steroid</td>
<td>Improved</td>
</tr>
<tr>
<td>3</td>
<td>Xanthopoulos J. et al. 2002</td>
<td>63y/ female</td>
<td>Old age</td>
<td>features of multiple cranial nerve palsy</td>
<td>V, VII, VIII, IX, and XII</td>
<td>Acyclovir, steroid</td>
<td>improved</td>
</tr>
<tr>
<td>4</td>
<td>Sugita-Kitajima A et al. 2009</td>
<td>58y/ female</td>
<td>Rheumatic heart disease</td>
<td>Right ear ache, vertigo, swallowing difficulty, hoarse voice</td>
<td>VII, VIII, IX and X</td>
<td>Antiviral, steroid</td>
<td>Improved with late recovery of hoarseness of voice</td>
</tr>
<tr>
<td>5</td>
<td>Lauridsen AG et al. 2010</td>
<td>56y/ male</td>
<td>none</td>
<td>Ear rashes, features of multiple cranial nerve palsy</td>
<td>V, VII, VIII, IX, X and XII</td>
<td>Antiviral, steroid</td>
<td>Not available</td>
</tr>
<tr>
<td>6</td>
<td>Sun W et al. 2011</td>
<td>-</td>
<td>Diabetes Mellitus</td>
<td>features of multiple cranial nerve palsy</td>
<td>V, VII, VIII, IX, and XII</td>
<td>Insulin, antiviral, steroid</td>
<td>Improved</td>
</tr>
<tr>
<td>7</td>
<td>Kim CH. et al 2012</td>
<td>66y/ female</td>
<td>none</td>
<td>Left ear pain and rashes, diplopia, vertigo, facial palsy</td>
<td>VI, VII, VIII</td>
<td>Acyclovir, steroid</td>
<td>improved</td>
</tr>
<tr>
<td>8</td>
<td>Coleman C et al 2012</td>
<td>80y/ female</td>
<td></td>
<td>Left sided ear and facial pain, ear rashes, tinnitus, left SNHL, facial deviation</td>
<td>V, VII, VIII, X and XII</td>
<td>Valacyclovir, steroid</td>
<td>improved</td>
</tr>
<tr>
<td>9</td>
<td>Patil V. et al. 2014</td>
<td>70y/ male</td>
<td>Old age</td>
<td>Ear rash, headache, facial deviation, hiccup, swallowing difficulty</td>
<td>VII, VIII, IX and X</td>
<td>Valacyclovir, steroid</td>
<td>improved</td>
</tr>
<tr>
<td>10</td>
<td>Talukdar J. et al. 2016</td>
<td>66y/ male</td>
<td>hypothyroidism</td>
<td>Ear ache and rashes, swallowing difficulty, change in voice, giddiness</td>
<td>VII, VIII, IX and X</td>
<td>Valacyclovir, steroid</td>
<td>Improved with residual facial weakness</td>
</tr>
<tr>
<td>11</td>
<td>Arya D. et al. 2018</td>
<td>29y/ male</td>
<td>Retroviral positive</td>
<td>Headache, facial deviation, gait disturbance, abnormal tongue sensation</td>
<td>V, VII, VIII, IX, X</td>
<td>Valacyclovir, steroid along with continuation of antiretroviral therapy</td>
<td>Symptomatic improvement</td>
</tr>
</tbody>
</table>

*Table 1: Review of some interesting cases of Cranial Polyneuropathy in Herpes Zoster Oticus*
Discussion

Herpes is a neurotropic virus having the ability to remain dormant in dorsal root and cranial nerve ganglia. The reactivation can cause zoster in a dermatomal distribution usually in elderly, diabetes mellitus and immuno-compromised patients.3 Sometime due to atypical presentation or involvement of multiple cranial nerves, misdiagnosis occurs. In our case, cranial nerves having fibers of Nucleus Tractus Solitarius, namely VII, IX and X were involved. Small carotid artery branch supplying contiguous cranial nerves, peripheral anastomosis of V, VII, IX and X cranial nerves and cranial nerves neighborhood in cavernous sinus has been considered as the plausible explanations for such presentations.2, 4

After the Bell’s palsy, Ramsay Hunt Syndrome is described in literature as second most common cause of unilateral lower motor neuron type of facial nerve palsy with incidence of 12%.5 Rarely this Ramsay Hunt Syndrome can present with cranial polyneuropathy involving V, VII, IX and X cranial nerves as shown in some of interesting case reports in Table 1. 1, 2, 6

Sharing such cases help us identify simple infections with complex anatomical presentations and simultaneously provide an insight of anatomy and interconnections of cranial nerves.

Conclusion

Herpes zoster oticus is one of the common causes of unilateral facial palsy and should always be kept as a differential diagnosis of unilateral cranial polyneuropathy especially in elderly patients and associated with comorbidities. Early and prompt management with antiviral and steroid results a favorable outcome.

Conflicts of interest: None
Source(s) of support: None

References