

Multiple cranial nerve injury in Ramsay Hunt Syndrome: a case report

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Abstract

Herpes zoster oticus (Ramsay Hunt Syndrome) is characterized by facial nerve paralysis, ear pain and auricular skin rash. It occurs as a result of reactivation of latent varicella zoster virus infection in the geniculate ganglion of the facial nerve. Major clinical symptoms include 7th nerve paralysis or cranial nerve paralysis and vesicles along the nerve with cocomitant ear pain. Other cranial nerve involvement although uncommon, can be found in some cases. In this study, a 74-year-old female patient had ipsilateral 8th, 9th and 10th cranial nerves injury. Cranial nerve paralysis accompanied with injury has been reported in Ramsay Hunt Syndrome.

Keywords: Herpes zoster, cranial nerve, ramsay hunt.

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Introduction

Herpes zoster oticus (HZO) or Ramsay Hunt Syndrome (RHS) is one of the rare causes of facial paralysis and accounts for 3% to 12% of patients with facial paralysis.^{1,2} Diagnosis of Ramsay Hunt Syndrome includes severe pain in the external ear, facial paralysis and vesicles in the ear canal. Other cranial nerves are also frequently involved, especially, eighth nerve involvement may cause hearing loss and vertigo.^{3,4}

In the literature, the number of patients with RHS who have multiple cranial nerve involvement are rare. It was known that facial paralysis in RHS improved later on as compared to the sequelae of Bell's palsy.⁵ We aim to present a case due to its distinctive presentation with multiple cranial nerve involvement following several differential diagnosis and treatments.

Case Report

A 75-year-old female was admitted to the intensive care unit with a diagnosis of ischaemic attack during her initial presentation at a medical center with complaints of deformations on her face, difficulty in breathing, vomiting and dysphagia. Patient was then referred to Sanliurfa

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Figure-1: Bullous rashes around the right auricle, and some of them burst.

Mehmet Akif Inan Research Hospital's emergency department on 13th of September 2012 with complaints of hoarseness, difficulty in swallowing and facial paralysis. Patient was first evaluated by neurology and pulmonology department; brain MRI performed by neurology did not reveal any pathology. Patient was then evaluated by ENT clinic and routine examination showed bullous rash around the right auricle with some burst lesions. (Figure-1) Since, the patient wore a head scarf, it was assumed that these rashes were not identified before, this implies the importance of physical examination and observation. Patient had peripheral facial paralysis (Figure-2) (Hause Brackman Grade 5 on the right side of the face) with the absence of gag reflex due to paralysis of right vocal cord. Medical history revealed hypertension. Audiological examination was performed the following day due to complaints of hearing loss which showed 32 dB sensorineural hearing loss in the left ear and 68 dB



Figure-1: Peripheral facial paralysis (Hause Brackman Grade 5 on the right).

sensorineural hearing loss in the right ear. Examination on the same day further identified dizziness, nausea and spontaneous nystagmus. Cranial nerve involvement of nerve 7th, 8th, 9th and 10th was observed in this patient. Diphenhydramine 3x50 mg was given intravenously for three days. Valaciclovir 3x1000 mg was started and continued for 1 week. Oral methylprednisolone (1 mg / kg / day) was started and reduced by 3 weeks. Improvement in dizziness and nystagmus was seen in the subsequent week. At the end of the 6-months follow-up, her auditory and vocal cord paralysis improved and facial paralysis improved to Hause Brackman Grade 3.

Discussion

Ramsay Hunt syndrome is a cranial polyneuropathy caused by reactivation of varicella zoster viruses.¹⁻⁴ Ramsay Hunt syndrome, which has a prevalence of 1% in herpes zoster infections, was first described by Ramsay Hunt in 1907.² There are 3 different clinical forms according to the severity of the disease. In addition to herpes zoster infection in any region of the head or neck, the development of facial

paralysis is the second most common cause of the disease and the addition of the 8th nerve involvement symptoms is the third form of the disease.⁶ This is followed by the development of herpetic vesicles in the outer ear, tympanic membrane or anterior 2/3 part of the tongue within 3-4 days. Facial paralysis usually occurs within 1-2 weeks after the rash. This picture is often accompanied by symptoms of nausea, vomiting, vertigo, nystagmus, tinnitus and hearing loss. In addition, in some patients, 9th, 5th, 10th, 6th cranial and cervical nerve involvement can be seen in the order of frequency.⁷⁻⁹ In the literature, the number of cases with Ramsay Hunt syndrome with multiple nerve involvement is very rare.¹⁰

The most effective method for the treatment of Ramsay Hunt syndrome, which is usually diagnosed with medical history and clinical findings is the combination of antivirals to prevent replication of varicella-zoster viruses and systemic steroids due to its antiinflammatory and anti-oedematous effects. The importance of early diagnosis in terms of the success of the treatment should be emphasized.

Disclaimer: Consent was obtained from the patient and relatives of the patient for the publication of the case.

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