

# Ramsay Hunt syndrome with multicranial nerve involvement - our experience

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## Introduction

Ramsay Hunt syndrome is defined as an acute peripheral facial neuropathy associated with pathognomonic symptoms such as otalgia, erythematous vesicular rash of the skin of the ear canal, auricle (also termed herpes zoster oticus), and/or mucous membrane of the oropharynx. VZV (varicella zoster virus) infection/reactivation involving the geniculate ganglion of CN VII within the temporal bone is the main pathophysiological mechanism of Ramsay Hunt syndrome. The primary infection is usually acquired early in life and remains latent until it is reactivated later, when symptoms emerge, usually in association with an immune deficiency or suppression[1,2] Ramsay Hunt syndrome presenting with multiple cranial neuropathies is rare [8].

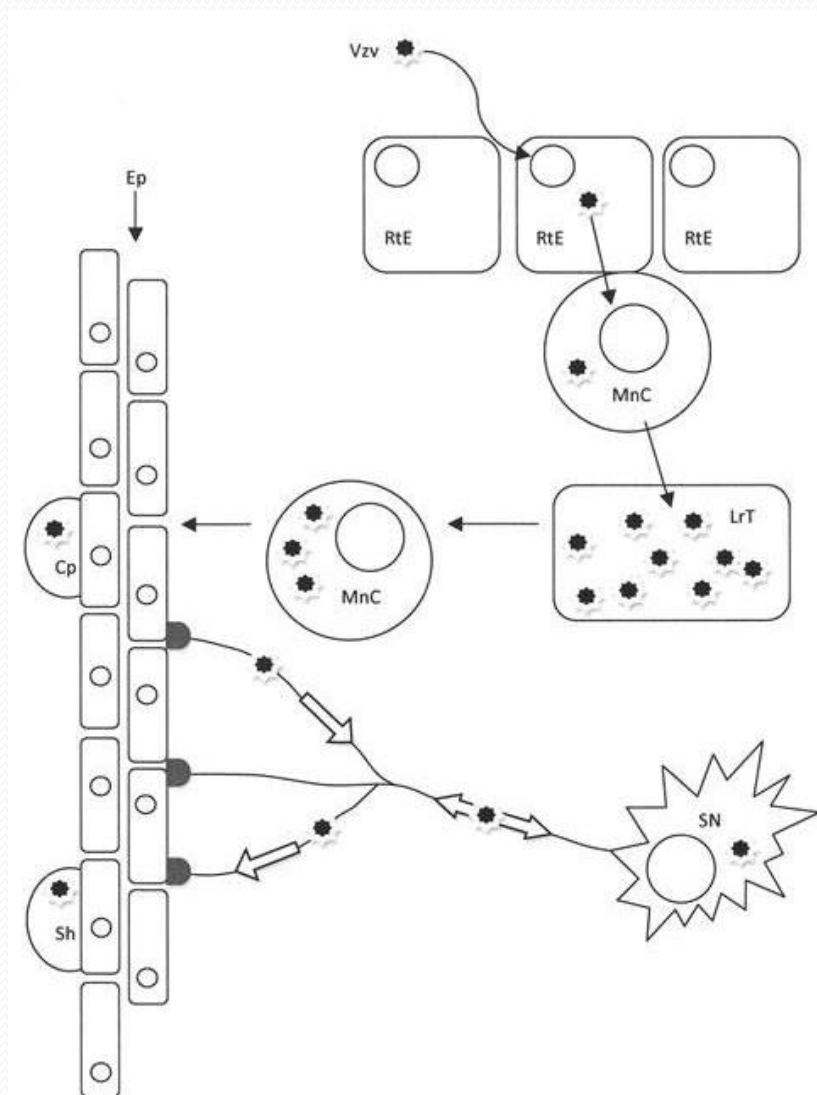


Figure 1. The Varicella Zoster Virus life cycle.

## Aim

To present two clinical cases of Ramsay Hunt syndrome with atypical unilateral multicranial nerve involvement.

**Methods:** a full neurological examination, ENT examination, PTA (pure-tone audiometry), CBC, basic metabolic panel, CSF analysis, serological tests, EMG, EEG, CT and MRI brain imaging.

**Case 1:** A 67-year-old female patient. PMH included arterial hypertension and thalassemia minor. Her illness started with flu-like symptoms, in the following week she developed pain, swelling and vesicular rash of the right auricle, chills and fever of 37.8°C, dysphonia, dysphagia (inability to swallow even water), drooping of the right side of the face, inability to close the right eye, vertigo, nausea, hearing loss, inability to lift her right arm above the shoulder, neck pain. The ENT, PTA and neurological examination revealed involvement of V, VII, VIII, IX, X and XI CN.



Fig.2 Case1 - hyperemic and edematous right aryepiglottic fold, paretic right vocal fold in the midline (X CN)



Fig.3 Case 1 - vesicles on the right side of the soft palate, uvula deviated to the left (IX CN)



Fig.4 Case 1 - herpetiform right ear rash, right Bell's syndrome (VII CN), weakness of right m.SCM and m.trapezius (XI CN)

CSF testing showed WBC 155x10<sup>6</sup> – mononuclear pleocytosis, total protein 0.49 g/l; glucose 3.9 mmol/l. Serological tests - VZV IgM Ab - negative; VZV IgG Ab >4000.00mIU/ml /positive if >165 /. CT and MRI – nothing abnormal detected.

Treated with a 14-day course of acyclovir 10mg/kg i.v., ceftriaxone 2.0 i.v., methylprednisolone 40mg i.v. A significant improvement was noted at day 14 with some residual right vocal fold paresis, full recovery was achieved at day 30 excluding the hearing loss.

**Case 2:** A 61-year-old woman with PMH of arterial hypertension, diabetes type II, asthma, osteoporosis. Presenting with needle-like pain in her right ear, swollen ear, right neck pain, slight dysphagia and dysphonia, drooping of the right side of the face. ENT, PTA and neurological examination revealed involvement of VII, VIII, IX and X CN: erythematous and edematous right auricle and external ear canal with vesicular herpetiform rash; right peripheral facial paresis; right tympanic membrane with decreased mobility and a single haemorrhagic bulla; assymetrical hypopharynx with retention of saliva on the right side, hyperemic and oedematous right aryepiglottic fold with a few confluent herpetiform lesions, right vocal cord paresis. PTA – mixed hearing loss. Head CT scan – NAD.

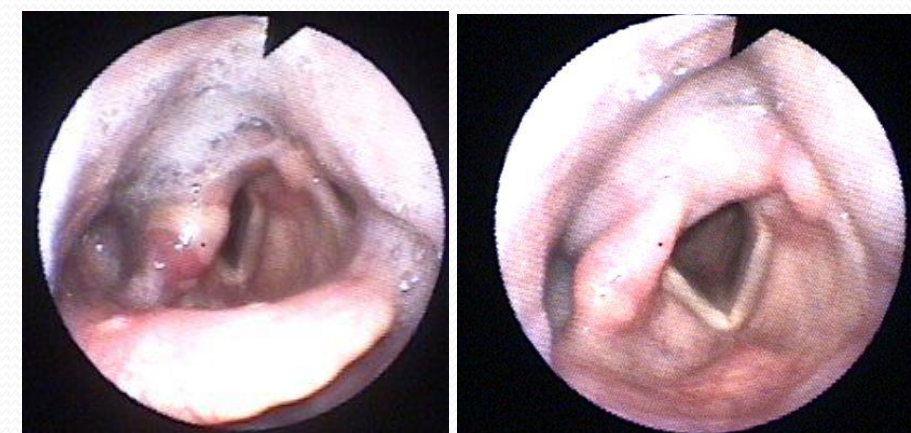


Fig.5 Case 2 – right vocal cord involvement

Fig.6 Case 2 – day 10 of treatment

The diagnose was based on the pathognomonic symptoms and confirmed by the effective treatment *ex juvantibus*. Treated with acyclovir 5x800 mg, dexamethasone 8mg for 4 days, ceftriaxone 2.0 i.v., Milgamma N<sup>TM</sup> 1amp i.v. In 10 days a significant improvement was noted regarding CN VII along with full recovery of CN VIII, IX and X functions.

## Discussion and conclusion:

Different theories have been proposed as to where exactly VZV and associated inflammation are situated during a flare up of herpes zoster, but generally it is acknowledged that the cause is ganglionitis[7]. Theories involving viral spread due to vasculitis or perineuritis have also been considered [3,4] and VZV has been electron microscopically observed in sensory nerves during HZ [5]. VZV is also found in the zoster elements during the vesicular phase [6].

RHS should be considered in patients with unilateral multiple cranial nerve palsies, as early antiviral and steroid treatment significantly improves the prognosis [9,10], as this is a difficult and severe diagnosis with low full recovery rate, especially with multicranial involvement [8]. Diagnosis is purely clinical, but in some cases a blood test for VZV antibodies can be considered[7]. RHS can be deceiving since the herpetic lesions are not always present (zoster sine herpette) and it might mimic other severe neurological intracerebral illnesses.

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