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RAMSAY HUNT SYNDROME WITH MULTIPLE CRANIAL NEUROPATHIES.

Chok Tong KHAW[1], Rosdan SALIM[2], Julius Liang Chye GOH[1], Jothi SHANMUGANATHAN[1].

[1]Department of Otorhinolaryngology, Hospital Sultanah Aminah, 80100 Johor Bahru, Johor, Malaysia.
[2]Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan & Hospital Universiti Sains Malaysia.

ABSTRACT

Ramsay Hunt Syndrome is a rare type of shingles caused by reactivation of Varicella Zoster Virus in the geniculate ganglion hence causing painful vesicular rash at the area of distribution of geniculate ganglion and peripheral cranial nerve (CN) VII palsy. In this syndrome, the involvement of CN VIII is also relatively common causing hearing and balance problems. Nonetheless, despite rare, other cranial nerves such as CN V, IX, X, and XII might also be involved, making the diagnosis of RHS more complicated. The involvement of these nerves may cause physiological impairment leading to serious complications if not managed appropriately. Treatment which consists of steroid and antiviral combination should be commenced as early as possible to achieve the best outcome. Here, we report a rare case of Ramsay Hunt syndrome with multiple cranial nerves involvement and share a brief literature review of similar cases.

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Keywords: Ramsay Hunt syndrome (RHS), Cranial nerves, Facial nerve, Vagus nerve, Varicella zoster virus (VZV)

INTRODUCTION

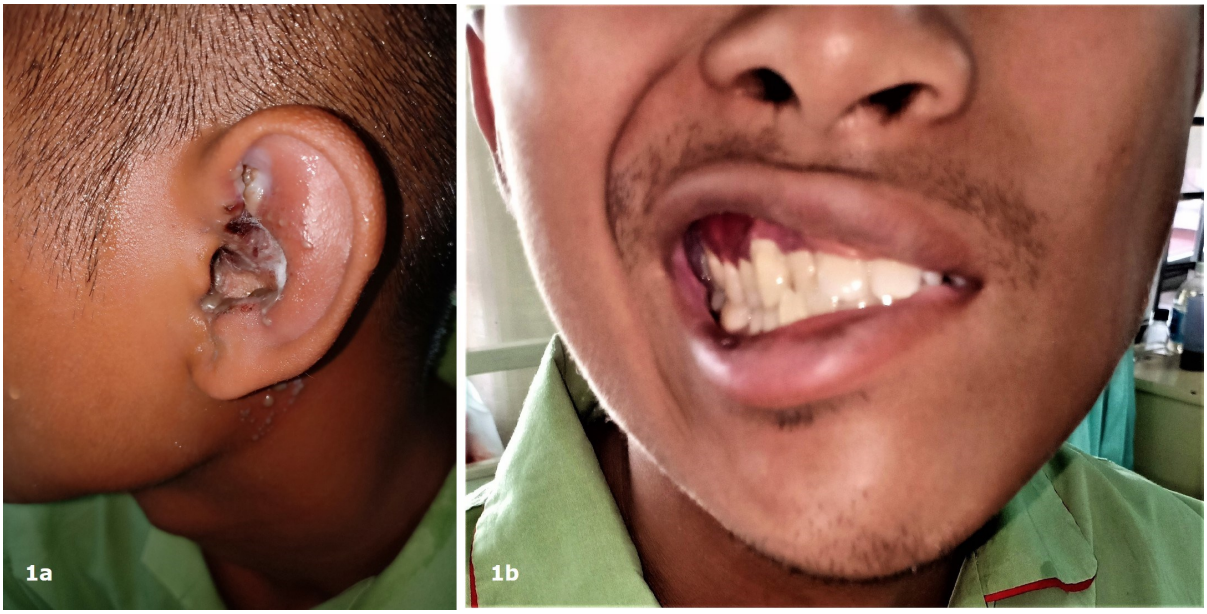
Ramsay Hunt syndrome(RHS) is a rare type of shingles caused by reactivation of Varicella Zoster virus (VZV) in the geniculate ganglion.[1] It is also known as herpes zoster oticus and has been classically described as a triad of lower motor neuron facial nerve palsy, neuralgia and erythematous vesicular rash along the sensory distribution of facial nerve.[2] Besides the cranial nerve (CN) VII, it can also affect CN VIII, IX, V, and VI in descending order of frequency.[3] It is very rare to encounter the involvement of CN X in RHS. Here we present a case of left ear RHS with the involvement of CN V, VII, VIII, IX and X and its clinical progression and treatment.

Corresponding author: Khaw Chok Tong (MBBS), Department of Otorhinolaryngology, Hospital Sultanah Aminah, 80100 Johor Bahru, Johor, Malaysia. Email: choktong87@gmail.com. Telephone: +60138181510

CASE REPORT

A 23-year-old man presented with hoarseness of voice and sore throat for three days, left ear swelling and painful vesicular rash and fever for two days. He also complained of slightly reduced hearing over the left ear with tinnitus. Since his change in voice, he would cough whenever he drank water too quickly.

On examination, vesicular lesions were noted over the left pinna and left external auditory canal (Figure 1a). The left tympanic membrane was partially seen due to the lesions and edema of the external auditory canal. There was also left facial paresis grade V House-Brackmann (Figure 1b). Oral and oropharyngeal examination revealed multiple vesicles seen over the soft palate and both anterior pillars and tongue. There were also multiple ulcers at the posterior oropharyngeal



Figures 1: a) Left ear vesicular lesion with pus exudate, b) left facial VII palsy .

al wall. The uvula was pulled over to the right side. Flexible laryngoscopy showed ulcerative lesion at left arytenoid and left vocal cord palsy in paramedian position which was partially compensated by the right vocal cord with minimal phonatory gap (Figure 2a).

His total white blood cell was only 6.2 $\times 10^9$ cells/L. Retroviral, hepatitis type B and C screenings were negative. A diagnosis of RHS with multiple cranial nerves involvement was made.

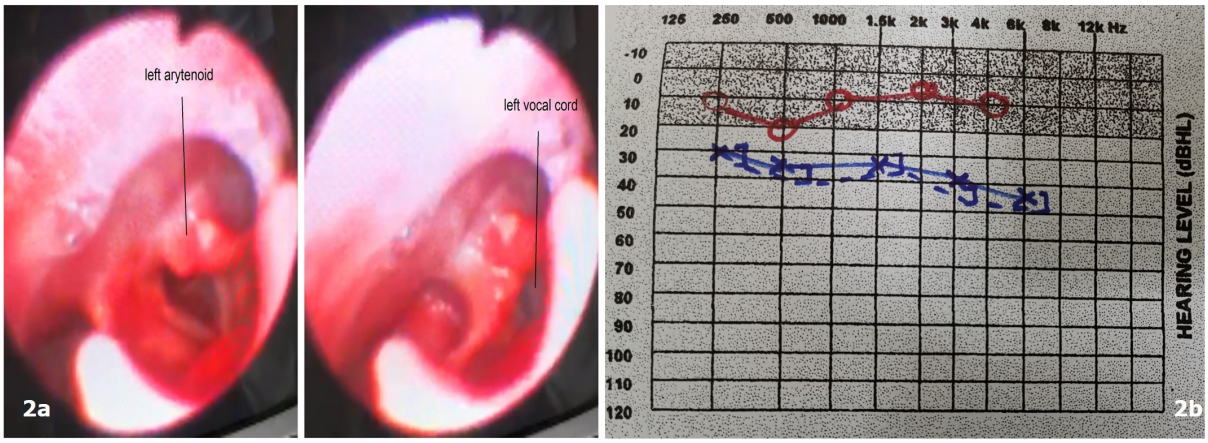
Patient was admitted to ward for intravenous dexamethasone 8mg TDS for three days followed by tapering dosage of oral prednisolone over two weeks. He was also given intravenous acyclovir for four days followed by oral acyclovir 800mg five times a day for two weeks. For prophylaxis of bacterial perichondritis, he was also given intravenous ciprofloxacin 400mg BD for four days. Eye cares were also prescribed. He was advised for nasogastric tube feeding but he refused and managed to eat and drink slowly without any problem. Upon discharge four days after admission, his facial nerve paresis improved to grade III House-Brackmann.

Upon follow-up in clinic six days later, his voice had improved and his left pinna lesions had dried up and left external auditory canal lesions had resolved. Otoscopy showed an intact left tympanic membrane. Therefore, a pure tone audiogram was performed and it showed left mild to moderate sensorineural hearing loss (Figure 2b). Oropharyngeal examination showed no vesicle or ulcer and uvula was central. Flexible laryngoscopy still showed left vocal cord palsy with minimal phonatory gap.

Three months later, his voice had improved significantly and left facial paresis had resolved completely. Flexible laryngoscopy showed bilateral vocal cords moving normally.

DISCUSSION

James Ramsay Hunt first introduced this syndrome in 1907.[3] VZV causes chickenpox in the primary infection and then it remains dormant in the geniculate ganglion of CN VII. Reactivation of this virus in the geniculate ganglion of CN VII later causes RHS. RHS commonly affects patients over the age of 60 or whose immunity has been compromised or weakened.[2,9] However, it can also affect



Figures 2: a) Flexible laryngoscopy showing ulcerative lesion at left arytenoid and left vocal cord palsy in para-median position, b) Pure tone audiogram showing left mild to moderate sensorineural hearing loss.

seemingly healthy persons as seen in our patient who is a fit young man with no prior illness.

Besides CN VII, other CN may also be involved probably due to close proximities of the CN ganglia or by vascular spread.[1,4] CN VIII has been reported to be involved in up to 50% of cases but the involvement of other CN is rare.[2] The various clinical presentations of previously published cases of RHS with CN X involvement have been summarised in Table I.

In our patient, there was a classical picture of RHS together with left-sided reduced hearing and tinnitus indicating the involvement of CN VIII while the dysphagia and left vocal cord palsy indicate left CN X involvement. His uvula was deviated to the right possibly due to CN V involvement as well. Moreover, multiple ulcers on the oropharyngeal wall might indicate CN IX involvement. Based on the above cases, an observation can be made that RHS can involve multiple CNs besides CN VII and the involvement of one CN can have variable presentations, some being life-threatening if undetected.

Diagnosis of RHS can usually be established based on history and examinations, especially for uncomplicated ones. For complicated cases with multiple CNs involvement, diagnosis can be confirmed by VZV polymerase chain reaction (PCR) of the ear exudate, blood, and cerebrospinal fluid (CSF).[2] In terms of imaging, gadolinium enhancement of geniculate ganglion and affected nerves in magnetic resonance imaging (MRI) can be appreciated in a proportion of patients.[3,5] In

Table I: Published cases of RHS involving CN X.

Publications	Clinical history and presentations
Steele et al., 2019.[2]	An immunocompromised 67-year-old woman with supraglotitis features and left vocal cord palsy complicated with aspiration pneumonia requiring ICU support. The woman also had reduced hearing and tinnitus.
Shim et al., 2019.[4]	A 48-year-old patient with right RHS with dysphagia, aspiration, and hoarseness secondary to right vocal cord palsy but with no documented hearing problem
Sahoo et al., 2019.[5]	A 54-year-old man had left RHS with dysphagia, hoarseness, ipsilateral uvula paralysis, tongue deviation to the ipsilateral side, loss of taste sensation at left anterior 2/3rd of the tongue and endoscopic finding of left vocal cord palsy in left paramedian position.
Shinha et al., 2015.[6]	A 66-year-old woman who had right RHS with laryngitis depicted by ulcerative lesions on the right side of the epiglottis and arytenoid but with no documented vocal cord palsy.
Ayoub et al., 2019.[7]	A case of RHS with atrial fibrillation postulated to be due to the involvement of the autonomic part of CN X

some patients, however, the MRI can be normal as seen in the case reported by Sahoo et al.[5] However, computed tomography (CT) scan and MRI, whichever appropriate, can be ordered in complicated cases to exclude tumors, abscesses or malignant otitis externa. Electroneurography and electromyography tests may estimate the severity of facial nerve involvement and assist to prognosticate.[3]

A combination of steroid and antiviral remains as the main treatment for RHS and best started within three days of onset of symptoms for the best prognosis.[1] There is no clear guideline on the dosage of antiviral and steroid for RHS. Antiviral has been shown to reduce the duration of rash and pain associated with herpes zoster while corticosteroids are postulated to reduce neuritis.[8] Stankus et al. recommended antiviral therapy with acyclovir 800mg five times a day for a duration of seven to ten days.[8] Famciclovir and Valacyclovir are alternative antivirals of choice.[8] A three-week course of tapering dosage of prednisolone is recommended in which 30mg BD is given for the first week, 15 mg BD is given for the second week and 7.5mg BD is given for the third week.[8] Some authors prefer a shorter course of prednisolone (1mg/kg body weight OD for five days).[9] Analgesia and eye care treatments should also be included. Last but not least, speech therapy and facial rehabilitation exercises may also hasten the recovery of the affected CNs. In our patient, complete recovery of cranial polyneuropathy was achieved as treatment was commenced early within 72 hours of the onset of symptoms. Otherwise, the prognosis of RHS remains grim with less than 50% of cases achieving completed resolution of facial palsy.[3]

CONCLUSION

Even though it is rare, clinicians should always bear in mind that RHS can result in cranial polyneuropathy. Hence, a complete ear, nose and throat examination together with full CN examination shall be performed in every case of RHS as the involvement of these nerves can lead to dire complications if not managed accordingly. A clinical diagnosis can usually be made based on history and clinical examination but complicated cases can be confirmed by laboratory tests such as VZV PCR. Imaging can assist to rule out other conditions. In terms of treatment, a good prognosis is dependent on early commencement steroid and antiviral therapy.

CONFLICT OF INTEREST DISCLOSURES

There is no conflict of interest to be disclosed by all authors.

CONSENT

We have acquired consent for the above images to be used for publication purpose.

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