Ramsay Hunt Syndrome

Nyilo Purnami¹, Cita Rosita Sigit Prakoeswa², Damayanti Tinduh³

Keywords: Ramsay Hunt Syndrome, Varicella Zoster Virus, Audiogram Examination, Audiology.

Abstract:

Background: Ramsay Hunt syndrome (RHS) is a rare disease that caused by a varicella zoster virus (VZV) infection who common in the geniculate ganglion. RHS causes severe facial nerve damage compared to Bell's palsy but there is an important condition to be evaluated i.e., damage to the vestibulocochlear nerve. Objectives: To reports the cases that include diagnostic, therapeutic, and prognosis of RHS patients. Methods: To observed the case reports from patients admitted to the hospital until patient returns. Case: A 29-year-old man with vertigo symptoms, a mouth congealed, perceived skin efflorescence of the right arm, and hearing loss. Audiogram examination was obtained sensory neural hearing loss desktra with masking bone conduction of 60 dB. Electromyography amplitude results in 70% of frontal muscles, 57% of orbicularis oculi muscle, 40% of nasalis muscle, and 43% of the orbicularis oris. The patient received corticosteroid therapy and performed facial massage. The patient improved after treatment for 2 months with mild facial paralysis and hearing function within normal limits. Conclusions: The RHS patient that observed by audiological examination needs to performed in order to prevent the progression of a threshold reduction.

1 INTRODUCTION

Varicella-zoster virus (VZV) causes chickenpox is a characteristic disease that characterized by skin lesions distributed and mostly occurs in childhood (John and Canaday, 2017). Several cases of VZV infection were found in the ganglion geniculate. The condition is commonly known as Ramsay Hunt syndrome (RHS). Typical manifestations of RHS consist of triad herpes eruption symptoms in the outer ear, facial nerve paralysis, and ear pain (Chen et al., 2016). The condition is caused by the reactivation of VZV that had previously been infected because of the decrease in immune system (Park et al., 2015; John and Canaday, 2017).

RHS is a relatively rare case, this disease was lasts for a short time and self-limited. Furthermore, RHS causes a disturbance in the facial nerve (John and Canaday, 2017; Kuya et al., 2017) and vestibulocochlear nerves (Psillas et al., 2012). RHS has a poor prognosis with 10-30% of patients have

residual symptoms (Kayakurt et al., 2014; Kuya et al., 2017). Thus, a fixed diagnosis is required through some tests. Audiological examination is important to determine the condition of the vestibulocochlearis nerves (Psillas et al., 2012) and electromyography (EMG) examination was to evaluate the facial nerve conditions (Xing et al., 2013). The goal is to determine therapy and evaluate the prognosis of RHS patients.

2 CASE

The subject of this study was a man (29 years) that experienced right ear pain since a week ago, vertigo, and decreased hearing. In the external aureus was obtained in bulls and crusts, while in the external acoustic meatus that has no secretions were found, but the facial palsy was found. The patient had a history of otorea at age 5 and recovered a year later. Patient working in the fish storage area with room

¹ Department of Otolaryngology, Faculty of Medicine, Universitas Airlangga, Dr. Soetomo General Hospital, Surabaya, Indonesia

² Department of Dermatology Venereology, Faculty of Medicine, Universitas Airlangga- Dr. Soetomo General Hospital, Surabaya, Indonesia

³ Department of Medical Rehabilitation, Faculty of Medicine, Universitas Airlangga Dr.Soetomo General Hospital, Surabaya, Indonesia

temperature below 20^oC since 3 months and undenied of having chickenpox.

In the initial symptoms was appear in 10 days ago before the patient had to go to a general practitioner and was diagnosed with strep throat. The patient also had control twice to the general practitioner however it did not improve. Then the patient went to Otolaryngology unit at Dr. Soetomo General Hospital Surabaya, Indonesia. The initial condition of the patient comes are shown figure 1.



Figure 1: The external auric conditions.

Examination of the right auricle was obtained a picture of vesicles that partially dried and covered in crust around the front concha. Meatus acoustic externus was obtained the hyperemia and cicatrik. While, Weber test was obtained by conduction of sinusra and examination of audiogram obtained sensory neural hearing loss desktra with masking bone conduction of 60 dB. On examination of tympanometry was obtained a results within normal limits. Then, patients were diagnosed with ramsay hunt syndrome and received dexamethasone therapy 1 mg per 8 hours and dicloxacillin 250mg per 8 hours.

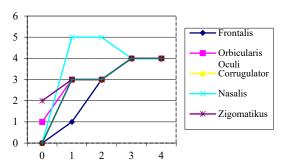


Figure 2: Graph examination of facial muscles.

The results of EMG examination were obtained a ratio of 70% frontalis amplitude, 57% orbicularis

oculi, nasalis 40%, and oris 43% orbicularis. Electrodiagnosis was showed a reversible prognosis and patients were given the facial massage. The results of examination of the facial muscles are shown in Figure 2.

The second week of the patient was eximined by weber test 3 times that leads to the left and 2 in the middle. Audiogram examination of the light-conductive dexterous deafness was dexogram with the masking of the left bone conduction of desktra 70 dB at a frequency of 4000 Hz. Tympanometry within normal limits and positive left and left stapedius reflexes. The patients also complained uncomfortably in the eyes and performed Schirmer tests (Table 1). Then, the patients also given the dexamethasone therapy 1 mg per 12 hours and added neurotrophic up to week six that showed good prognosis.

3 DISCUSSION

The VZV infection of the geniculate ganglion is usually called RHS. The condition is a rare case, so it is often overlooked in diagnosis and therapy. Detailed clinical examination was obtained a typical set of symptoms of herpes eruption in the ear, ear pain, and facial nerve paralysis that easily found in early stage to prevent complications (Chen et al., 2016; Kuya et al., 2017).

Herpes Zoster infection could occur due to viral reactivation after previous varicella infections (Kuya et al., 2017). However, in this case was denied that the patient ever getting infected by Herpes Zoster before. One possible factor triggering reactivation of viral infections was extreme room temperature because the patient works in an air-conditioned room with temperatures below 200 C. These conditions possibility lower the individual's immune system if it was exposed continuously (Park et al., 2015; John and Canaday, 2017).

The condition of oblique mouth and stiffness on the right face is caused by an infection that attacks the nerves vii (facial nerves) and decreased hearing (The vestibulocochlear nerve). The condition was complained by RHS patients by 84.6% (Zainine et al., 2012) and if not managed properly it possibility causes a persistent symptoms as much as 12% (Kayakurt et al., 2014). The identified herpes eruption is the usual stage of crustation that occurring at 1-2 weeks after infection (Avci and Ertam, 2014). Thus, the Tzanck smear cytological examination was not carried out with such considerations (Riahi et al., 2014).

Table 1: The results of Schirmer test.

-	Week	II	III	IV
	Schirmer	Different >	Different >	Different
	test	50%	50%	< 25%

The results of the audiogram examination was obtained that sensory neural hearing loss desktra in the first week and re-examination on the second week that obtained a deaf conduction dexter sinistra. This neurological disorder was caused by inflammation of the geniculate ganglion thus suppressing the cranial nerve of the nerves VIII (Godani et al., 2013; Kuya et al., 2017). The improvement was caused by a decrease in compression of nerves VIII. The EMG examination of the amplitude illness/healthy were ranged from 40-70%, that estimated the lower the amplitude ratio than the muscle contractures then more visible and the prognosis was getting worse (Xing et al., 2013).

Antiviral (acyclovir) was not given with the consideration of patients who coming a week after not infection and did show immunocompromised symptoms. Also, thus given dexamethasone at a dose of 3 times 1 mg (Avci and Ertam, 2014; Chen et al., 2016; Kuya et al., 2017). The treatment was given for 2 weeks and gradually decreased the dose and the patient also gets face massage. Massage proved capable of blood circulation, so the cells more rapid regeneration and expected facial paralysis can be minimized (Field, 2014). The second week the patient were complained of blurred vision and examination, eye condition within normal limits.

Postherpetic neuralgia complications were a common complication by 10-30% (Kayakurt et al., 2014; Kuya et al., 2017). It is necessary to take preventive steps in the case of RHS which is the determination of accurate diagnosis and follow prognosis by using EMG. The prognosis in this case was good which shows an amplitude ratio above 25% (Xing et al., 2013).

4 CONCLUSION

RHS is a rare occurrence with a characteristic triad of symptoms. In this case, the patients come to health services a week after the appearance of paralysis facial and hearing loss. Patients receive corticosteroid, neurotrophic and facial massage therapy without antiviral therapy. A few weeks after the therapy the patient has improves their facial muscles and audiology functions. As a preventive measure to prevent the occurrence of RHS

complications required audiological diagnosis and facial paralysis.

REFERENCES

- Avci, O., Ertam, I., 2014. Viral infections of the face. Clinics in Dermatology. doi:10.1016/j.clindermatol.2014.02.010
- Chen, I., Fohtung, R.B., Oughli, H.A., Bauer, R., Mattar, C., Powderly, W.G., Thoelke, M.S., 2016. Concurrent Ramsay Hunt syndrome and disseminated herpes zoster in a patient with relapsed chronic lymphocytic leukemia. *IDCases* 6, 79–82. doi:10.1016/j.idcr.2016.10.003
- Field, T., 2014. Massage therapy research review. Complementary Therapies in Clinical Practice. doi:10.1016/j.ctcp.2014.07.002
- Godani, M., Giorli, E., Traverso, E., Beronio, A., Capellini, C., Del Sette, M., 2013. Ramsay Hunt syndrome with trochlear nerve involvement and EEG abnormalities: Multicranial neuritis or encephalitis?

 Journal of Clinical Virology. doi:10.1016/j.jev.2012.08.018
- John, A.R., Canaday, D.H., 2017. Herpes Zoster in the Older Adult. *Infectious Disease Clinics of North America*. doi:10.1016/j.idc.2017.07.016
- Kayayurt, K., Yavasi, O., Bilir, O., Ersunan, G., Giakoup, B., 2014. A case of Ramsay Hunt syndrome with atypical presentation. *Turkiye Acil Tip Dergis*i 14, 142–145. doi:10.5505/1304.7361.2014.82788
- Kuya, J., Kuya, K., Shinohara, Y., Kunimoto, Y., Yazama, H., Ogawa, T., Takeuchi, H., 2017. Usefulness of high-resolution 3D multi-sequences for peripheral facial palsy: Differentiation between Bell's Palsy and Ramsay Hunt syndrome. *Otology and Neurotology*. doi:10.1097/MAO.000000000001605
- Park, Y.M., Kim, D.R., Park, J.Y., Kim, S.K., Kim, S.Y., Kim, J.S., Lee, Y.H., Kim, Y.G., Jeong, K.H., Moon, J.Y., Lee, S.H., Ihm, C.G., Lee, T.W., 2015. A case of Ramsay Hunt syndrome diagnosed after kidney transplantation. *Kidney Research and Clinical Practice* 34, 241–244. doi:10.1016/j.krcp.2014.11.004
- Psillas, G., Arnaoutoglou, M., Gatsios, T., Rizos, D., Koutsouraki, E., Vital, V., 2012. Autoimmune recurrent facial palsy and bilateral sudden sensorineural hearing loss following Ramsay Huntlike syndrome. *Auris Nasus Larynx* 39, 229–232. doi:10.1016/j.anl.2011.03.007
- Riahi, R.R., Yamazaki, M.L., Kelly, B.C. 2014. Tzanck smear utilized in the diagnosis of Hailey-Hailey disease mimicking genital herpes. *International journal of dermatology* 53(1), 85-7.
- Xing, Y., Chen, L., Li, S., 2013. Evoked electromyography to rocuronium in orbicularis oris and gastrocnemius in facial nerve injury in rabbits. *Journal of Surgical Research* 185, 198–205. doi:10.1016/j.jss.2013.05.087

Zainine, R., Sellami, M., Charfeddine, A., Beltaief, N., Sahtout, S., Besbes, G., 2012. Ramsay Hunt syndrome. *European Annals of Otorhinolaryngology, Head and Neck Diseases* 129. doi:10.1016/j.anorl.2011.08.003

