Case Study

Ramsay Hunt Syndrome: A Case Report

Syahmi Amar [1], Rustam Siregar [1], Husnia Auliyatul U [1]

Author's Affiliation:
1- Department of Child Health, Universitas Sebelas Maret Medical School / Dr. Moewardi Hospital

Correspondence:
Syahmi Amar, Email: syahmiamar@gmail.com

Received on: 21-Nov-2020 Accepted for Publication: 01-Aug-2021

ABSTRACT

Background: One of the types of cranial Herpes Zoster is herpes zoster oticus. It is rarely found in children with possible clinical manifestations including vesicular lesions according to dermatome and peripheral facial nerve paralysis. The combination of all these symptoms is known as the Ramsay Hunt Syndrome (RHS).

Aims: To describe the diagnosis, treatment, and prognosis of a RHS case.

Case Description: A 14-year-old boy complained of pain and vesicular lesion in the right ear, facial asymmetry. From ear examination found a crusted vesicular lesion in the right ear. Cranial nerve examination found weakness in the peripheral facial nerve House-Brackman score of 4, reduced tear secretion, and impaired taste on the right side. The examination found no abnormality of vestibulocochlear nerve. The patient was offered physiotherapy, oral acyclovir and oral prednisone. After day five, the patient showed an improvement in his facial motor with a House-Brackman score of 3.

Conclusion: Providing comprehensive early therapy including physiotherapy, corticosteroids, and antiviral therapy can improve the recovery rate.

Keywords: Ramsay Hunt Syndrome, Herpes Zoster Virus

INTRODUCTION

Herpes zoster is a long-term complication of the reactivation of the varicella virus. The virus stays in the ganglion cells and reacts when our immunity decreases.[1,2] One of the types of cranial Herpes Zoster is herpes zoster oticus. It is rarely found in children; with an incidence of 2.7/100,000.[3] Possible clinical manifestations include vesicular lesions in the auricula, or the ear canal, ear pain, and peripheral facial nerve paralysis. The combination of all these symptoms is known as the Ramsay Hunt Syndrome (RHS).[4,5] The treatment of RHS is the administration of antivirus and corticosteroid. However, there is no standard recommendation on the treatment of RHS.[5]

This case report aims to describe the diagnosis and treatment of an RHS case.

CASE DESCRIPTION

A 14-year-old boy complained of pain in his right ear and the occurrence of a vesicular lesion in the right ear, with facial asymmetry. There was no complaint of reduced hearing, tinnitus, or spinning dizziness. The patient had a history of varicella infection when he was 8 years old and had a fever a week before the occurrence of the lesion. He had no history of immunodeficiency disease, use of corticosteroid, cancer, hypertension, varicella vaccination, or head trauma.

The patient was conscious, and his vital signs were within normal limits. Ear examination found a crusted vesicular lesion in the right ear and an intact tympanic membrane (Figure 1). Neurological examination found that the motor strength, physiological reflex, and pathological reflex were within normal limits. Cranial nerve examination found a weakness in the peripheral facial nerve motor with House-Brackman 4, reduced tear

secretion, and impaired taste on the right side. The examination found no abnormality on nerve vestibulocochlear. Tzanck test was conducted and there was no multinucleated giant cell. The patient was given physiotherapy, oral acyclovir therapy (80 mg/kg body weight/day) for 7 days, oral prednisone (1 mg/kg/day) for 5 days, and tapered off. He was also given artificial tears during the day and lubricating ointment during the night. After the fifth day, the patient showed an improvement in his facial motor with a House-Brackman score of 3.

DISCUSSION

One of the chronic complications of varicella is varicella zoster. The incidence of herpes zoster increases along with age and rarely affects children. Studies in the United States reported the incidence of 20, 30, 59, and 63 per 100,000 children per year within the age group of 0-4, 4-9, 10-14, and 15-19, respectively. RHS is rarely found in children. However, it placed second for non-traumatic peripheral facial nerve paralysis after Bell's palsy with the incidence of 2.7/100,000 population.[2,3,6,7]

After the incubation period of 4-20 days, if there is an involvement of the facial nerve, then vesicles may appear according to its dermatome, accompanied by pain surrounding the area, ear pain, facial paralysis, reduced tear secretion, and impaired taste. The involvement of the vestibulocochlear nerve can be characterized by signs and symptoms such as nausea, vomiting, vertigo, nystagmus, tinnitus, and reduced hearing.[8,9,10]

When encountering a facial nerve paralysis, it is important to assess the location of the lesion and other nerves involved. This can be accomplished through history taking, physical examination, and adjunctive examination. In the central lesion, there is a unilateral weakness of the lower part of facial muscles and is usually accompanied by contralateral hemiparesis/hemiplegia, albeit without autonomous impairment such as impaired taste or salivation. Peripheral lesions provide descriptions of unilateral facial weakness on all upper and lower facial muscles.[11,12]

The diagnosis of RHS is established based on the occurrence of herpetic vesicle along its dermatome and facial nerve paralysis. Examination of the facial nerve function is needed to determine lesion location, the severity of paralysis, and treatment evaluation. The examination includes facial muscle motor function, facial muscle tone, and the existence of synkinesis or hemispasm, gustatometry, and the Schimer test.[9,11] To determine the degree of severity and predict the probability of improvement of the facial nerve weakness, a modified House-Brackmann scale can be used.[5] Possible adjunctive examinations include Tzanck test before the lesion is crusted. MRI and IgG and IgM serum level anti-VZV with ELISA are often not needed because of the clear clinical appearance of RHS.[7,13]

The standard treatment of RHS is an antivirus and high-dose corticosteroid. The use of artificial tears during the day and lubricant ointment during the night is needed in RHS with ocular complications because the eyes could not close completely. Acyclovir, famciclovir, or valacyclovir can be given for antiviral therapy. However, the use of famciclovir and valacyclovir is not recommended for children. Delayed administration of medicine for more than 72 hours reduced its effectiveness. Patients with motoric nerve involvement can be given corticosteroids to eliminate pain, reduce the incidence of postherpetic neuralgia, reduce inflammation, and facial nerve edema.[14,15]

There is no standard recommendation on RHS therapy. Several case reports and studies stated different administration duration, route of administration, and tapering off duration. Adour only used prednisone with 1 mg/kg/day for 14 days followed by dose tapering off for 7 days.[16] Wackym suggested the administration of intravenous acyclovir because the oral dose was not effective in treating RHS.[17] Contrary to Wackym, Murakami *et al.* suggested that intravenous and oral acyclovir had similar effectivity on the recovery of the facial nerve.[18] Kim and Bhimani used 800 mg of acyclovir 5 times a day and prednisone with 1 mg/kg/day for 5 days, followed by tapering off for 6-7 days for RHS therapy.[10]

Several factors are affecting the recovery of RHS patients, include patients with incomplete paralysis, the time of initial therapy, and younger age who have a better prognosis.[19,20] Early diagnosis and appropriate treatment can improve the recovery of the facial nerve. Several studies showed significant improvement from facial muscle

paralysis after acyclovir and corticosteroid therapy. The administration of therapy before 3 days, 4-7 days, and more than 7 days showed 75%, 48%, and 30% recovery, respectively.[18] Meanwhile, Kinisi *et al.* explained that the administration of corticosteroids before one week could lead to 62% of complete recovery.[21] If antiviral therapy is added before 1 week, then the recovery rate increased to 90%. The prognosis of the skin lesion will recover within several days to weeks. The use of electrophysiological examination such as electromyography or electroneurography is often used to assess the recovery prognosis. However, its effectiveness is yet to be determined.[22]

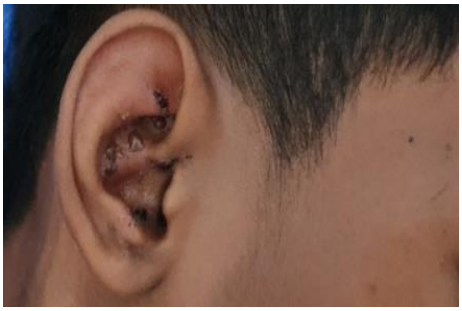

Figure 1. clinical features of Ramsay Hunt syndrome

Patient's consent: Written informed consent was obtained from the parents of the patient for publication of this case report and the accompanying images.

REFERENCES

1. Tom WL, Freidlander SF. Viral exanthem. In: Irvine AD, Hoeger PH, Yan AC. Harpers' Textbook of Pediatric Dermatology. 3rd ed. Oxford: Blackwell publishing; 2011; p. 491-4.
2. Basuki PS. Varisela. In: Hadinegoro SR, Moedjto I, Hapsari MMDEAH, Alam A, editors. Buku ajar infeksi dan penyakit tropis. Jakarta: Badan Penerbit Ikatan Dokter Indonesia; 2018. p. 389-96
3. S. Ogita, K. Terada, T. Niizuma, Y. Kosaka, and N. Kataoka. Characteristics of facial nerve palsy during childhood in Japan: Frequency of varicella-zoster virus association. Pediatrics International. 2006;48:245–9.
4. D.E. Mattox, Clinical disorders of the facial nerve. In: C.W. Cummings, J.M. Fredrickson, L.A. Harker, C.J. Krause, M.A. Richardson, D.E. Schuller, editors. Otolaryngology head and neck surgery. 3rd ed. St. Louis: Mosby-Year Book; 1998. p. 2773
5. L Kansu, I Yilmaz. Herpes zoster oticus (Ramsay Hunt syndrome) in children: Case report and literature review. International Journal of Pediatric Otorhinolaryngology. 2012;76: 772–6.
6. N. Hato, H. Kisaki, N. Honda, K. Gyo, S. Murakami, N. Yanagihara, Ramsay Hunt syndrome in children. Ann. Neurol. 2000;48:254–6.
7. Y. Furuta, F. Ohtani, H. Aizawa, S. Fukuda, H. Kawabata, T. Bergstro¨m, Varicella zoster virus reactivation is an important cause of acute peripheral facial paralysis in children. Pediatr. Infect. Dis. J. 2005;24:97–101.
8. Soetirto, Indro. Gangguan nervus fasialis. In: Soepardi EA, Iskandar HN, editors. Buku ajar ilmu kesehatan telinga hidung tenggorok kepala leher. 7th ed.. Jakarta : Balai Penerbit FK UI; 2012. p. 89-92
9. Harvey BS. Bell's palsy. In: Kliegman RM, et al., editors. Nelson Textbook Of Pediatric. 20th ed. Philadelphia: Elsevier, 2016. p. 3014-15.
10. D. Kim, M. Bhimani, Ramsay Hunt syndrome presenting as simple otitis externa. CJEM. 2008;10:247–50.
11. Tiemstra JD, Khatkhate N. Bell's palsy: diagnosis and management. Am Fam Phys. 2007; 76: 997-1004
12. Gilden DH. Bell's palsy. N Engl J Med. 2004;351:1323-31.
13. H. Nakatani, K. Yamakawa, M. Hamada, T. Takeda, A. Kakigi, M. Iwai, Initial lesions in Bell's palsy and Ramsay-Hunt syndrome. J. Otorhinolaryngol. 2009;71:105–11.
14. LaRussa PS, Marin N. Varicella-Zoster virus. In: Kliegman RM, et al., editors. Nelson Textbook Of Pediatri. 20th ed. Philadelphia: Elsevier; 2016. p. 1579-85.
15. Rafael da CM, Aline GB, Natal JBN, Silvia CA, Fabio TM, Raquel S. Treatment and prognosis of facial palsy on ramsay hunt syndrome: results based on a review of the literature. Int Arch Otorhinolaryngol. 2016;20:394–400.
16. K.K. Adour, Otological complications of herpes zoster. Ann. Neurol. 1994;35:S62–S64.

17. P.A. Wackym, Molecular temporal bone pathology. Ramsay Hunt syndrome (herpes zoster oticus). Laryngoscope. 1997;107:1165–75.
18. S. Murakami, N. Honda, M. Mizobuchi, Y. Nakashiro, N. Hato, K. Gyo, Rapid diagnosis of varicella zoster virus infection in acute facial palsy. Neurology. 1998;51:1202–05.
19. S.W. Yeo, D.H. Lee, B.C. Jun, K.H. Chang, Y.S. Park, Analysis of prognostic factors in Bell's palsy and Ramsay Hunt syndrome. Auris Nasus Larynx. 2007;34:159–64.
20. E. Peitersen, Bell's palsy: the spontaneous course of 2500 peripheral facial nerve palsies of different etiologies. Acta Otolaryngol. 2002;549:4–30.
21. M. Kinishi, M. Amatsu, M. Mohri, M. Saito, T. Hasegawa, S. Hasegawa, Acyclovir improves recovery rate of facial nerve palsy in Ramsay Hunt syndrome. Auris Nasus Larynx. 2001;28:223–26.
22. W.X. Chen, V. Wong, Prognosis of Bell's palsy in children—analysis of 29 cases. Brain Dev. 2005;27:504–8.