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Ramsay-Hunt syndrome

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Abstract

Ramsay-Hunt syndrome (RHS) is a disease caused by reactivation of varicella-zoster virus in the geniculate ganglion. It may be one of the common reasons of the facial palsy. Symptoms of the RHS include vesicular rash on the auricle, external auditory canal, soft palate, pharynx, facial nerve paralysis, ear pain, tinnitus, vertigo and hearing loss. There may be other symptoms connected to cranial nerves (CN) V, VIII, IX or X, when they are involved. Men are more likely to develop lesions than women. Risk of developing Ramsay-Hunt syndrome increases with age. Authors searched Google Scholar and PubMed, using searchterms Ramsay-Hunt syndrome, varicella-zoster virus, VZV, facial palsy. We manually searched the references of selected articles for additional relevant articles. We selected articles relevant to a general medicine readership and prioritised systematic reviews, clinical practice guidelines and cases. The literature contains the latest reports on Ramsay-Hunt syndrome. Clinicians should not ignore patients' symptoms and always pay attention to possibility of VZV infection and the development of RHS, as treatment should be started

within 72 hours after the onset of the symptoms. Main treatment includes antiviral drugs and corticosteroids. The results are promising.

Keywords: ramsay-hunt syndrome, herpes, varicella-zoster

Introduction

Ramsay-Hunt Syndrome (RHS) is defined as inflammation and degeneration of the 7th cranial nerve (facial nerve) as a reactivation of VZV-varicella-zoster virus in the geniculate ganglion. It appears with painful vesicles in the external auditory meatus or auricle and facial paralysis in the innervation of the facial nerve.[1,2] RHS was first described in 1907 by J. Ramsay Hunt as a facial palsy, dysfunction of the inner ear and vesicular rash.[3] Nerves connected to the facial nerve, such as CN V (trigeminal nerve), CN VIII (vestibulocochlear nerve), CN IX (glossopharyngeal nerve), CN X (vagus nerve), cervical nerves C2, C3, C4 may be affected.[4] Ramsay Hunt syndrome is a rare cause of otalgia and the second reason of non traumatic peripheral facial palsy. This syndrome is also claimed as a complication of herpes zoster.[2] Early diagnosis is crucial and should be based on the clinical examination.[5]

Definition and aetiology

There are many definitions of RHS in the literature which vary depending on the author and clinical center.[6] Over a hundred years ago, James Ramsay Hunt, an American neurologist, described cases of patients with herpetic lesions on oral mucosa and auricle, accompanied by neurological disorders of the cranial nerves.[7] Depending on the affected cranial nerves and their connections to each other and to the cervical nerves, patients' symptoms may vary and the authors redefine RHS.[8] Robilliard et al. described Ramsay Hunt syndrome as a peripheral facial nerve insufficiency, ear pain and herpes zoster lesions on auricle, external auditory canal or other cervical dermatome.[9] Sweeney and Gliden believe that RHS may be diagnosed when, in addition to paralysis of the peripheral facial nerve, vesicular lesions are visible on the oral mucosa or on the ear. However, they do not exclude other symptoms, such as acoustic disturbances, tinnitus or nausea and dizziness.[7] Jane Woo et al. reported a case of a 37-year-old patient (previously healthy) with Ramsay Hunt syndrome two days after he got vaccinated with the Pfizer-BioNTech vaccine. He showed RHS-related symptoms (ear pain, vesicles on the right ear and external auditory canal, facial paralysis). A PCR test was performed which confirmed the presence of varicella-zoster

virus DNA. The SARS-CoV-2 result was negative. Scientists claimed that the vaccine triggered varicella-zoster virus.[10]

Epidemiology

The infection with varicella-zoster virus most often manifests itself in lesions in the thoracic region and concerns about 59,2% of cases. 35% of infections are related to lesions in the neck and head region.[11,12] Other studies show that the thoracic area is involved in 49% of patients and the head and neck area in 13%.[6] Ramsay-Hunt syndrome seems to be one of the common causes of superficial facial nerve paralysis. Robillard et al. conducted a study of 1507 patients with facial palsy and found out that 185 of them (12%) were diagnosed with RHS.[9,13] Hato et al. performed a study of 2076 patients with unilateral facial palsy. He showed that there is no significant difference between the incidence of RHS in children over six years of age (16,7%) and in adults (18,1%). It is noteworthy that the risk of Ramsay-Hunt syndrome was significantly increased in children over six years old.[14] There is a predilection for gender and age. Men (18%) are more likely to develop lesions in cranial sections than women (8%).[11] Schniffer and Rohe showed that the incidence of RHS increases with age.[6] Hearing loss and vertigo were detected in 73-85% of patients with RHS.[15]

Clinical manifestation and symptoms

Ramsay-Hunt syndrome is characterised by vesicular lesions filled with serous fluid in the auricle, external auditory canal and in the mucosa of pharynx or soft palate, accompanied by paralysis of the superficial facial nerve.[1] Beside the main symptoms, depending on the cranial nerves involved, neurological symptoms may occur, such as voice alteration, trigeminal neuralgia, swallowing difficulty, tongue deviation or difficulty in moving the eyeballs. They may be caused by cranial polyneuropathy, changes in the cerebrospinal fluids of meningitidis.[4,16–19] Other neurological symptoms concern the vestibulocochlear nerve and include tinnitus, dizziness and even hearing loss. Dizziness has been found to worsen the prognosis of facial palsy.[20–22] Patients with dizziness are more likely to develop more severe hearing loss than patients without dizziness.[23] Montague et al. described in their case speaking difficulty and increasing confusion in their patient with Ramsay-Hunt syndrome.[24] Nishizawa et al. presented a case in which a patient also suffered from bilateral throbbing headache fever and fatigue. Four days later he developed dysgeusia.[25] Saniasiaya mentioned that patients may also occur with hyperacusis and facial

paralysis may be present without vesicles but with presence of increased number of VZV antibody and detected DNA of VZV from the skin, middle ear fluid or blood.[7,15]

Treatment

Diagnosis based on history and clinical symptoms is highly important since empirical treatment in the form of antiviral drugs should be administered within 72 hours of symptoms onset. [5] Intravenous or oral acyclovir and corticosteroids (prednisone) are recommended for patients with Ramsay-Hunt syndrome.[13,26,27] It is said that oral or intravenous ways of treatment are not statistically significant. [30] Acyclovir or famciclovir, antiviral drugs, are said to relieve pain and help heal wounds after vesicles. They may also prevent neuralgia. [29,31] On the other hand, corticosteroids have a strong anti-inflammatory effect, helping to reduce swelling of the nerves involved in RHS, which may accelerate their regeneration.[7] Quickly implemented treatment will not only alleviate the suffering of the patients, but can also prevent hearing loss.[13,28] Kinishi et al. checked the effectiveness of the therapy of combined aciclovir with corticosteroids and the therapy with corticosteroids alone. In the group of patients treated with antiviral drugs and steroids, improvement in nerve function was observed in 75% of patients. In the group treated with corticosteroids alone, improvement was seen in only 53% of patients, making this difference statistically significant.[29] Sampedro et al. proposed a treatment protocol with prednisolone 30 mg, ibuprofen 1800 mg and moxifloxacin 400 mg but without any improvement after 2 days, patient also got aciclovir and flucloxacillin. After 3 weeks with other symptoms improving, there was no improvement in facial paralysis. They decided to start acupuncture. They performed 18 sessions in 15 weeks. The punctures were done with a depth about 1-1,5 cm and the needles stayed in place for about 30 minutes with manual stimulation every 5 minutes. The clinicians observed significant improvement and after 15 weeks the patient regained the efficiency of the nerve.[30] Gupta et al. proposed a treatment with 60 mg/day of prednisone and 1000 mg/3 times a day of famciclovir for 1 week.[31] Khan et al. presented a protocol with 1000 mg/3 times a day of valacyclovir for 7 days and 60 mg/day of prednisone for 5 days.[32] Pradhan suggested oral prednisolone 40 mg/day and acyclovir 800 mg/5 times a day and physiotherapy.[33]

Conclusion

Ramsay-Hunt syndrome is a clinical manifestation of varicella-zoster virus replication. Patients show symptoms, such as vesicular rash with serous fluid on the ear, external auditory

canal, mucosa of pharynx or soft palate and facial palsy. Other symptoms, such as tinnitus, dizziness, hearing loss, tongue deviation, voice change, swallowing difficulty or difficulty in moving eyeballs, depend on included other cranial nerves (CN V, CN, XIII, CN IX, CN X) and cervical nerves (C2, C3, C4). The definition of Ramsay-Hunt syndrome may vary depending on affected structures. RHS is early diagnosed on the history and clinical picture. The PCR should be performed to confirm presence of varicella-zoster virus DNA. The treatment includes antiviral drugs (aciclovir, famciclovir, valaciclovir) and corticosteroids (prednison, prednisolon). It was proposed to perform acupuncture on patients with facial paralysis. Clinicians should pay attention to Ramsay-Hunt syndrome symptoms to may recognize it early and start to treat it. Ramsay-Hunt syndrome is a painful disease and may worsen the quality of the life of the patients.

References:

1. Lee J, Choi BY, Noh H, Jeong H, Shin JE, Kim CH. Nystagmus in Ramsay Hunt syndrome with or without dizziness. *Neurol Sci.* 2021;42(1):193–8.
2. Gaudencio M, Bertão MI, Carvalho A, Pena G, Bessa I, Gonçalves A. Ramsay Hunt Syndrome in a Patient with Ulcerative Colitis Treated with Infliximab. *Eur J Case Reports Intern Med.* 2021;3–6.
3. Zainine R, Sellami M, Charfeddine A, Beltaief N, Sahtout S, Besbes G. Ramsay Hunt syndrome. *Eur Ann Otorhinolaryngol Head Neck Dis.* 2012;129(1):22–5.
4. Shim HJ, Jung H, Park DC, Lee JH, Yeo SG. Ramsay Hunt syndrome with multicranial nerve involvement. *Acta Otolaryngol.* 2011;131(2):210–5.
5. Uscategui T, Doree C, Chamberlain I, Burton M. Antiviral therapy for Ramsay Hunt syndrome (herpes zoster oticus with facial palsy) in adults (Review). *Cochrane Database Syst Rev.* 2008;(4):1–11.
6. Wagner G, Klinge H, Sachse MM. Ramsay Hunt syndrome. *JDDG - J Ger Soc Dermatology.* 2012;10(4):238–44.
7. Sweeney C, Gilden D. Ramsay Hunt syndrome. *J Neurol Neurosurg Psychiatry.* 2001;71:149–54.
8. Walther LE, Prosowsky K, Walther A, Gudziol H. Herpes Zoster Oticus: Symptom Constellation and Serological Diagnosis. *Laryngo-Rhino-Otologie.* 2004;83:355–62.
9. Robillard RB, Hilsinger RL, Adour KK. Ramsay Hunt facial paralysis: Clinical analyses of 185 patients. *Otolaryngol - Head Neck Surg.* 1986;95(3 I):292–7.
10. Woo CJ, Chou OHI, Cheung BMY. Ramsay Hunt syndrome following COVID-19

- vaccination. *Postgrad Med J*. 2022;postgradmedj-2021-141022.
11. Eberhard P, Tillmann T. Zur Epidemiologie der Varizella-Zoster-Infektion. *Der Hautarzt*. 1996;47(8):604–9.
 12. Nahm FS, Kim SH, Kim HS, Shin JW, Yoo SH, Yoon MH, Lee DI, Lee YW, Lee JH, Jeon YH, Jo DH. Survey on the treatment of postherpetic neuralgia in Korea; multicenter study of 1,414 patients. *Korean J Pain*. 2013;26(1):21–6.
 13. Kunitomo K. Ramsay Hunt syndrome. *Postgrad Med J*. 2020;96(1133):168.
 14. Hato N, Kisaki H, Honda N, Gyo K, Murakami S, Yanagihara N. Ramsay Hunt syndrome in children. *Ann Neurol*. 2000;48(2):254–6.
 15. Saniasiaya J. Hypoacusis in Ramsay hunt syndrome: A hidden culprit. *Postgrad Med J*. 2020;0(0):2020.
 16. Choi JW, Nahm H, Shin JE, Kim CH. Atypical clinical manifestations of herpes zoster oticus: diagnostic usefulness of magnetic resonance imaging. *J Neurovirol*. 2019;25(6):874–82.
 17. Elliott KJ. Other neurological complications of herpes zoster and their management. *Ann Neurol*. 1994;35(1 S):S57–61.
 18. Kim YH, Chang MY, Jung HH, Park YS, Lee SH, Lee JH, Oh SH, Chang SO, Koo JW. Prognosis of ramsay hunt syndrome presenting as cranial polyneuropathy. *Laryngoscope*. 2010;120(11):2270–6.
 19. Yi HA, Lee H. Simultaneous occurrence of trigeminal herpes zoster and Ramsay Hunt syndrome presenting sudden prolonged vertigo. *Neurol Sci*. 2017;38(11):2049–51.
 20. Byun H, Cho YS, Jang JY, Chung KW, Hwang S, Chung WH, Hong SH. Value of electroneurography as a prognostic indicator for recovery in acute severe inflammatory facial paralysis: A prospective study of bell's palsy and ramsay hunt syndrome. *Laryngoscope*. 2013;123(10):2526–32.
 21. Shin DH, Kim BR, Shin JE, Kim CH. Clinical manifestations in patients with herpes zoster oticus. *Eur Arch Oto-Rhino-Laryngology*. 2016;273(7):1739–43.
 22. Yeo SW, Lee DH, Jun BC, Chang KH, Park YS. Analysis of prognostic factors in Bell's palsy and Ramsay Hunt syndrome. *Auris Nasus Larynx*. 2007;34(2):159–64.
 23. Kim CH, Choi H, Shin JE. Characteristics of hearing loss in patients with herpes zoster oticus. *Med (United States)*. 2016;95(46):1–5.
 24. Montague SJ, Morton AR. Ramsay Hunt syndrome. *Cmaj*. 2017;189(8):E320.
 25. Nishizawa T, Ishikawa K, Matsuo T, Higuchi N, Ishiguro K, Mori N. Atypical Ramsay Hunt syndrome (zoster sine herpette) with otitis media. *J Gen Fam Med*.

- 2021;22(6):344–6.
26. De Ru JA, Van Benthem PPG. Combination therapy is preferable for patients with ramsay hunt syndrome. *Otol Neurotol*. 2011;32(5):852–5.
 27. Kang HM, Jung SY, Byun JY, Park MS, Yeo SG. Steroid plus antiviral treatment for bell's palsy. *J Intern Med*. 2015;277(5):532–9.
 28. Furuta Y, Ohtani F, Mesuda Y, Fukuda S, Inuyama Y. Early diagnosis of zoster sine herpete and antiviral therapy for the treatment of facial palsy. *Neurology*. 2000;55(5):708–10.
 29. Kinishi M, Amatsu M, Mohri M, Saito M, Hasegawa T, Hasegawa S. Acyclovir improves recovery rate of facial nerve palsy in Ramsay Hunt syndrome. *Auris Nasus Larynx*. 2001;28(3):223–6.
 30. Giralt Sampedro I, Carvajal G, García-Janeras A, Fabà A, Nishishinya Aquino MB. A severe case of Ramsay Hunt Syndrome treated with acupuncture and related techniques. *Complement Ther Clin Pract*. 2020;39(October 2019).
 31. Gupta NM, Parikh MP, Panginikkod S, Gopalakrishnan V. Ramsay hunt syndrome. *QJM An Int J Med*. 2016;109(10):693.
 32. Khan YMT, Fatema N. Ramsay hunt syndrome. *Pan Afr Med J*. 2019;34:8688.
 33. Pradhan RR. Ramsay Hunt syndrome. *IDCases*. 2020;21:e00850.