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Case report

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Ramsay hunt syndrome: case report

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Abstract

Ramsay-Hunt Syndrome is an uncommon disease caused by the reactivation of latent Varicella zoster virus in the geniculate ganglion. It is characterized with a herpes zoster oticus associated with an acute peripheral facial nerve paralysis, and possible other cranial nerve lesions. We describe the case of a 27-year-old man who presented with severe ear pain for two days, with vesicular eruptions on the external ear and ipsilateral peripheric facial paralysis. He was treated with the association of intravenous steroids and acyclovir for 7 days. The evolution showed gratifying results. The one-year follow-up showed no relapse of the syndrome. The purpose of this report is to highlight the importance of a thorough clinical examination and early treatment of the Ramsay-Hunt syndrome.

Introduction

The Ramsay Hunt syndrome (RHS) was described for the first time by the neurologist James Ramsay Hunt in 1907. It is an unusual disease that includes a herpes zoster oticus and an ipsilateral peripheral facial paralysis. It is caused by reactivation of the Varicella-Zoster virus (VZV) at the geniculate ganglion. This report highlights the importance of a complete clinical examination and precocious treatment in management of the Ramsay-Hunt syndrome.

Patient and observation

A 27-year-old man presented with intense right-sided ear pain for two days. The patient did not complain of headache, dizziness, tinnitus, or hearing impairment. There was no history of trauma. Physical examination revealed right facial muscles weakness and incomplete eye closure at effort, consistent with a grade IV at House-Brackmann (HB) scale. We also noted a right-sided Charles Bell´s phenomenon. (Figure 1). The otoscopy showed a vesicular rash at the right concha extended to the external auditory meatus, with a slight swelling of the auricular area (Figure 2). Audiometric evaluation showed an abolished right stapedial reflex and a normal pure tone audiometry. Neurological examination showed no other cranial nerve lesion. Ophthalmological examination showed no signs on corneal lesion. The patient was treated with intravenous steroids (prednisolone 1mg/kg per day) with antivirals (acyclovir 15mg/kg per day) and antalgics. The follow-up showed gratifying results with the disappearance of the earache and vesicular rash, as well as the improvement of facial paralysis, becoming a grade II HB. The one-year follow-up showed no relapse of the syndrome.

Discussion

Ramsay-Hunt syndrome is an uncommon disease, with incidence ranged from 0.3 to 18% of acute atraumatic facial palsies, the most common cause being Bell´s palsy [1]. RHS is characterized by vesicular eruptions on the external ear and severe ear pain (herpes zoster oticus) with ipsilateral peripheric facial paralysis. It is caused by reactivation of latent Varicella Zoster Virus in the geniculate ganglion. The neuropathogenesis of RHS is, according to Grose *et al*. the contamination of the ganglion by the virus during chickenpox, via the sensory branches of the facial nerve located on the ear and tongue. Upon reactivation, VZV travels back along the sensory branches of the facial nerve, causing, by a bystander effect, the inflammation of the adjacent motor branches of the facial nerve, leading to facial palsy [2]. Histopathological studies revealed perivascular, perineural, and intraneural aggregation of round cells in the facial nerve in patients with RHS. The VIII cranial nerve can be affected by VZV as well, causing vestibulocochlear symptoms [3]. There are complicated anastomoses between cranial nerves including facial, trigeminal, glossopharyngeal, vagal, or cervical nerves, which enable herpetic lesions to progress further. While skin lesions lead to the diagnosis of herpes zoster oticus, various neurological disturbances can occur such as tinnitus, hearing loss, nausea and vomiting, vertigo, and nystagmus [4].

Early diagnosis of RHS is a crucial factor to improve nerve damage in Ramsay Hunt syndrome. Diagnosis is mainly based on the history, clinical findings, and neurological examination. Polymerase Chain Reaction assays may be useful to detect herpes zoster virus DNA in biological fluid [5]. Magnetic Resonance Imaging after injection with gadolinium, although usually unnecessary, can also contribute to the diagnosis by showing intense enhancement of the geniculate ganglion [6]. Combination treatment with anti-viral agents and steroids is recommended for the treatment of RHS. The parenteral dose of acyclovir is 15 mg/kg per day and the oral dose is 800 mg 4 times a day. Murakami *et al.* [7] compared 48 patients with oral acyclovir versus 32 patients with intravenous acyclovir and observed that there was no significant difference between the two routes, except for the advantage of reducing hospital time for the oral acyclovir. When compared with idiopathic facial palsy (Bell´s

palsy), RHS seems to have a more severe paralysis at onset and is less likely to recover completely [8]. Ryu´s study compared 202 patients with RHS and 155 patients with Bell´s palsy. Both groups were treated with oral steroids and antiviral agents. Ryu concluded that RHS showed poorer prognosis for recovery from facial weakness compared with Bell´s palsy [9]. Murakami´s study [7] investigated the importance of an early treatment and established that the sooner treatment was initiated, the better was the recovery. Early diagnosis and treatment of Ramsay Hunt syndrome is therefore crucial as prognosis of cranial nerve damage depends on the time at which acyclovir-corticosteroid therapy is started [10].

Conclusion

Ramsay-Hunt syndrome is a rare but easily diagnosed disease, based on clinical symptoms and examination. Treatment recommended is the combination of anti-viral agents and steroids. Prognosis is good, but any delay of the care could lead to sequelae. This report emphasizes how an early on treatment can offer remarkable results with a prompt and complete recovery.

Competing interests

The authors declare no competing interests.

Authors' contributions

IO has written the article. All the authors have read and agreed to the final manuscript.

Figures

Figure 1: vesicle rash on the right concha with a slight swelling of the pavilion

Figure 2: facial palsy of the right side with incomplete eye closure and Charles-Bell phenomenon

References

1. Gondivkar S, Parikh V, Parikh R. Herpes zoster oticus: a rare clinical entity. Contemporary clinical dentistry. 2010; 1(2): 127-129. **PubMed**| **Google Scholar**
2. Grose C, Bonthius D, Afifi AK. Chickenpox and the geniculate ganglion: facial nerve palsy, Ramsay Hunt syndrome and acyclovir treatment. The Pediatric Infectious Disease Journal. 2002;21(7): 615-617. **PubMed**| **Google Scholar**
3. Shin DH, Kim BR, Shin JE, Kim CH. Clinical manifestations in patients with herpes zoster oticus. European archives of oto-rhino-laryngology: official journal of the European Federation of Oto-Rhino-Laryngological Societies (EUFOS): affiliated with the German Society for Oto-Rhino-Laryngology - Head and Neck Surgery. 2016;273(7): 1739-1743. **PubMed**| **Google Scholar**
4. Jeon Y, Lee H. Ramsay Hunt syndrome. J Dent Anesth Pain Med. 2018;18(6): 333-337. **PubMed**| **Google Scholar**
5. Sauerbrei A, Eichhorn U, Schacke M, Wutzler P. Laboratory diagnosis of herpes zoster. Journal of clinical virology: the official publication of the Pan American Society for Clinical Virology. 1999;14(1): 31-36. **PubMed**| **Google Scholar**
6. Sartoretti-Schefer S, Wichmann W, Valavanis A. Idiopathic, herpetic, and HIV-associated facial nerve palsies: abnormal MR enhancement patterns. AJNR American journal of neuroradiology. 1994;15(3): 479-485. **PubMed**| **Google Scholar**
7. Murakami S, Hato N, Horiuchi J, Honda N, Gyo K, Yanagihara N. Treatment of Ramsay Hunt syndrome with acyclovir-prednisone: significance of early diagnosis and treatment. Annals of neurology. 1997;41(3): 353-357. **PubMed**| **Google Scholar**

8. Gupta NM, Parikh MP, Panginikkod S, Gopalakrishnan V. Ramsay Hunt syndrome. QJM: monthly journal of the Association of Physicians. 2016;109(10): 693. **PubMed**| **Google Scholar**
9. Ryu EW, Lee HY, Lee SY, Park MS, Yeo SG. Clinical manifestations and prognosis of patients with Ramsay Hunt syndrome. American Journal of Otolaryngology. 2012;33(3): 313-318. **PubMed**| **Google Scholar**
10. Wagner G, Klinge H, Sachse MM. Ramsay Hunt syndrome. Journal of the German Society of Dermatology: JDDG. 2012;10(4): 238-244. **PubMed**| **Google Scholar**

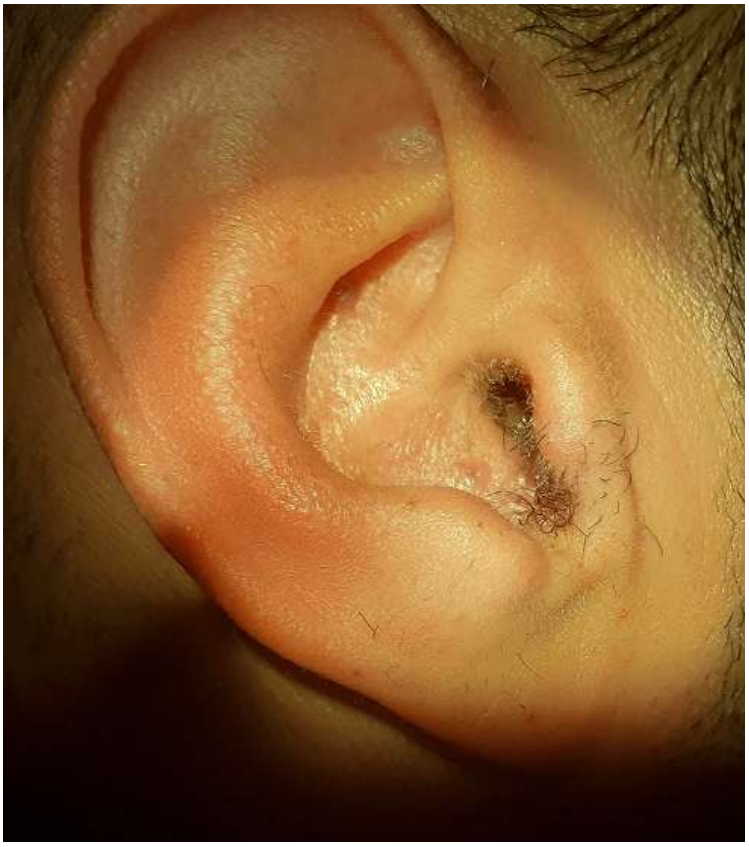

Figure 1: vesicle rash on the right concha with a slight swelling of the pavilion

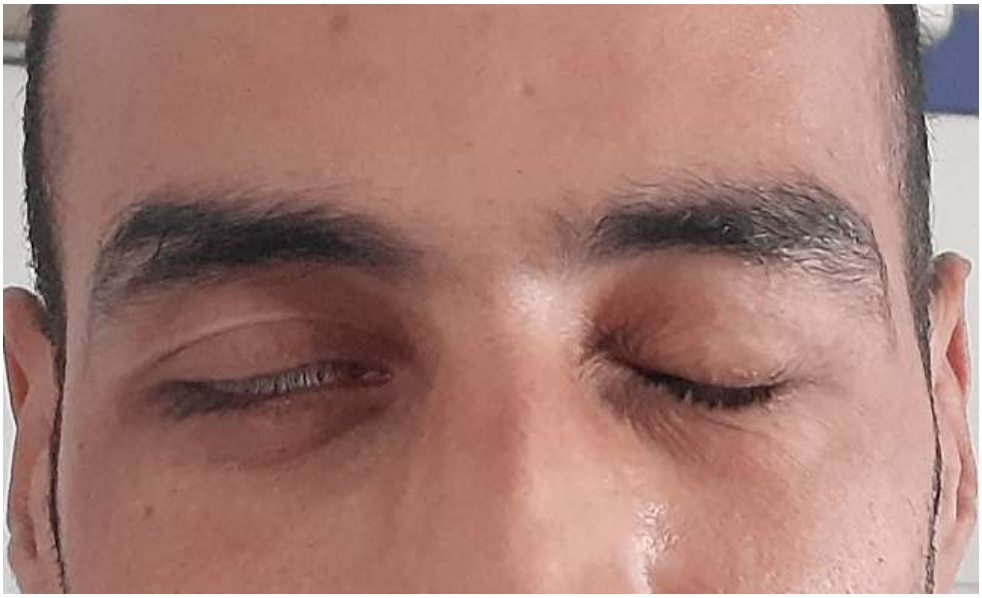

Figure 2: facial palsy of the right side with incomplete eye closure and Charles-Bell phenomenon