RAMSAY HUNT SYNDROME-CASE REPORT

Marcelo José da Silva de Magalhães Medical Assistant Professor from FUNORTE University and FipMoc University Department of Neurosurgery, Aroldo Tourinho Hospital, Montes Claros-MG, Brazil.

Maeds Soares Cardoso Acadêmicos do curso médico das Faculdades Unidas do Norte de Minas- FUNORTE.

Isabelle Lopes Gontijo Acadêmicos do curso médico das Faculdades Unidas do Norte de Minas- FUNORTE.

Correspondence: Marcelo José da Silva de Magalhães, Neurocirurgia - marcelo7779@yahoo.com.br

ABSTRACT

Purpose: The Ramsay Hunt Syndrome (RHS), also called by Herpes Zoster Oticus, is caused by reactivation of Varicella Zoster Virus (VZV) in the geniculate ganglia. This syndrome is consisting of vesicles in auditory pavilion, ipsilateral otalgia and peripheral facial paralysis. This case report demonstrates classic symptoms of the disease, but the clinician must look carefully to the several presentations, like vague symptoms or atypical scenario. Case description: A 57 years old man presented with edema in the right external auditory pavilion, otalgia and, posteriorly, ipsilateral peripheral facial paralysis accompanied by vesicles in the right external auditory canal. After treatment with Acyclovir, there was parcial improvement of the peripheral facial paralysis with disappearance of lesions.Conclusions: The first-line treatment includes an association of antivirals e corticosteroids agents, which must be instituted within first 72 hours of syndrome evolution, with better results.

Keywords: Herpes Zoster Oticus; Facial paralysis; Otalgia; Ramsay Hunt Syndrome.

SÍNDROME DE RAMSAY HUNT-RELATO DE CASO

RESUMO

Objetivo: A Síndrome de Ramsay Hunt (SRH), também chamada Herpes Zoster ótico (HZO), é causada pela reativação do Vírus Varicela Zoster (VVZ) que ocorre no gânglio geniculado. Sua síndrome é composta por lesões vasculares em pavilhão auricular, otalgia ipsilateral e paralisia facial periférica. Este caso demonstra um quadro clássico da doença, mas o clínico deve estar atento à diversidade de sua apresentação, como sintomas vagos ou atípicos.Descrição do caso: Paciente do sexo masculino, 57 anos, apresentou edema em pavilhão auditivo externo direito, dor auricular e, posteriormente, paralisia facial periférica ipsilateral associado ao aparecimento de vesículas em pavilhão auditivo externo direito. Após tratamento com Aciclovir, houve melhora parcial da paralisia facial periférica com desaparecimento das lesões.Conclusão: O tratamento de primeira linha inclui uma associação de agentes antivirais e corticoterapia, o qual deve ser instituído dentro das primeiras 72 horas de evolução da síndrome, por mostrar melhores resultados.

Palavras-chave: Herpes Zoster Ótico; Paralisia facial; Otalgia; Síndrome de Ramsay Hunt

1 INTRODUCTION

The Ramsay Hunt Syndrome (RHS), also called by Herpes Zoster Oticus (HZO), is a disease caused by reactivation of varicella zoster virus (VZV) in the geniculate ganglia. This illness represents the second cause of peripheral facial paralysis (PFP) atraumatic and occurs in 7-16% of cases.[1,2] Although it could manifest in any age, the incidence of RHS is higher in the second and third decades of life, and it is uncommon in childhood. It is also higher in immunosuppressed patients, like HIV positives, in use of cytotoxic and steroid medications.[1] The presence of PFP and ipsilateral otalgia accompanied by vesicles in the auditory canal are classic syndrome of HZO.[1-3] Vertigo, nausea, vomiting, nystagmus and sensorineural hearing loss are additional clinical features.[1,2] Involvement of cranial nerves trigeminal, glosopharyngeal, vagus and hypoglossal occurs less frequently.[1]

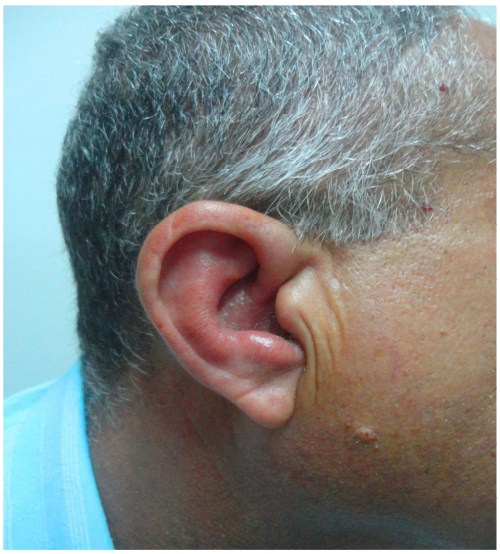
The diagnosis is basically clinic and, in doubtful cases, by laboratorial confirmation. The recommended treatment for RHS is corticosteroid and antiviral therapy.[2-4] Studies demonstrate that the combined therapy reveals a better prognostic of the patient.[4] The presented case demonstrates the classic triad of the disease, but it is essential that the clinician look carefully to the several presentations, since RHS can present only vague symptoms or atypical scenario.

2 CASE REPORT

A 57 years old man with previous healthy condition of osteomyelitis, gout and hypertension presented with edema in the right external auditory pavilion and otalgia. He presented with edema in the right external auditory pavilion and earache. On his first consultation by general practitioner it was prescribed Amoxicilin after suggestion of a bacterial infection process, but without symptoms improvement. In the third day of evolution, the patient presented ipsilateral peripheral facial paralysis accompanied by vesicles in the right external auditory canal, no fever, confirming the diagnosis of Ramsay Hunt Syndrome.(Figura 01)

He started his treatment with Lacrima plus® eye drops, Epitezam® and orientations about eye protection. He started oral Acyclovir 800mg, every 5 hours for 7 days besides physical therapy. In control appointment after 30 day, it was observed disappearance of lesions in external auditory pavilion with partial improvement of the peripheral facial paralysis.

Figura 1 - vesicles in the right external auditory canal, confirming the diagnosis of Ramsay Hunt Syndrome.



3 DISCUSSION

The RHS is a rare presentation of Herpes Zoster characterized by peripheral facial paralysis, erythematous vesicles in auditory pavilion and cochlear and vestibular symptoms.[2] The incidence is 5/100.00 habitants/year and it increase by age: about 3/100/year in 20-50 years old people and 10/1000/year in age of 80.[5] It shows a worst prognostic than Bell palsy, having total recover in 30% of cases.[3,6]

The HZO appears by reactivation of varicella zoster virus, resulting from immunosuppression factors - like stress, fever, radiotherapy, tissue damage - in geniculate ganglia.[2,6-8] The VZV, during RHS evolution, replicates in dorsal spinal nerve roots and cause intense inflammatory activity and tumor necrosis responsible for disease symptoms.[2,6] As differential diagnosis we have Bell palsy, Lyme disease, trauma, metabolic diseases and tumors.[3]

The PFP can precede vesicular eruptions.[2] In prospective study, 14% of diagnosed patients with RHS presented vesicular eruptions after facial involvement.[1] Although the classic triad of HZO is PFP and ipsilateral earache accompanied by vesicular lesions in auditory pavilion, some experts do not considerate the presence of vesicles necessary to diagnosis of this pathology - RHS sine herpete.[3] Several clinical presentations are possible in this scenario, having an essential characteristic the unilateral involvement of facial nerve.[1] The literature

brings as most prominent symptoms the presence of PFP, earache, erythematous lesions in auditory pavilion, nausea, vertigo, sensorineural hearing loss.[1,2,6,9,10] Buzz and nistagmus may also be present in HZO.[11] Other related symptoms are due to lesion of other cranial nerves like trigeminal, glosopharyngeal, vagus and hypoglossal.[1,6,9,11]

Although the RHS diagnosis is predominantly clinical, virological and serological tests to identification of VZV are indicated in atypical cases, like HZO *sine herpet*.[1,9,10] It is important to establish the topographic diagnosis of facial nerve lesion through tearing evaluation by Schirmer test, stapes reflex by impedance audiometry and, finally, gustometry of anterior portion of the tongue.[3] The early diagnosis is essential, since the early institution of therapy minimize the risk of disease's sequelae.[9]

Retrospectives studies and case reports demonstrate that the better therapy option to RHS is the association between antiviral and corticosteroids agents.[1,6] In a study, the institution of combined therapy in the first 72 hours of the symptoms resulted in better prognostic and sequelae rate of patients, having a percentage of total healing in 75% of cases.[1,3] An antiviral medication that is most widely used is Acyclovir 800mg/dia, orally, 5 times per day for 7 days.[3] Immunosuppression patients, children or in case of serious disease complications, it is recommended a therapy with Acyclovir, endovenously, at a dose of 10mg/kg/day, every 8 hours, for a period of 7 to 10 days.[7] Other therapy options are Valacyclovir, that present efficiency slightly higher than Acyclovir and Fancyclovir.[11] This medication reduces the acute symptoms duration of HZO and prevents nerve damages in long term.[3] The corticotherapy is indicated in cases of polyneuritis, moderate to severe pain, since corticosteroid decreases the pain degree associated with HZO and nerve inflammation and can be helpful in the management of the facial paralysis of RHS.[3,6] However, many authors caution against implementing steroid therapy, especially with periocular lesions, as they fear dissemination of the VZV infection.[6] Other option is surgical decompression of facial nerve in case of persistent neuralgia and severe PFP.[2,4]

It is noteworthy that despite the institution of appropriate therapeutic, patients with sequelae correspond to 24 to 90% of cases, being possible complications the post herpetic neuralgia, syncinesis, ophthalmopathies, segmental myelitis and encephalitis.[2] The most common and important sequelae of this syndrome is facial palsy, that can attain maximum

intensity within the first week of the symptomatology beginning. Factors of bad prognostic to recovery are the age over 50 years and complete facial palsy.[1]4 **Conclusion**

The classic triad of RHS is PFP, ipsilateral earache and vesicular lesions in auditory pavilion, generally unilateral, and the diagnosis is basically clinic. The first-line treatment includes an association of antivirals e corticosteroids agents, which must be instituted within first 72 hours of syndrome evolution. The RHS can determinate sequelae in patient generally if not treated in a timely manner, being essential the early diagnosis of this pathology.

REFERENCES

1. Oviedo AM, Zamarro MTL, Hoyo JJU. Síndrome de Ramsay Hunt. An Med Interna 2007;24 (1):31-34.

2. Rafael LB, María Luiza N, Ana María GA, Susana LC, Domingo G, Enrique PS: Síndrome de Ramsay Hunt: nuestra experiência. Acta Otorrinolaringol Esp 2010;61(6):418-421.

3. Worme M, Chada R**,** Lavallee L. An unexpected case of Ramsay hunt syndrome: case report and literature review. BMC Res Notes 2013;6:337-40.

4. Sweeney CJ, Gilden DH. Ramsay Hunt syndrome. J Neurol Neurosurg Psychiatry. 2001;71:149–54.

5. Anna G: Varicella-zoster vírus infections of the central nervous system. [tese]. Gothenburg: University of Gothenburg; p. 11-20;2013.

6. Ahmed MJ, Taylor PM, Cameron MLC, George KBS: Unilateral Facial Swelling Caused by Ramsay Hunt Syndrome Resembles Odontogenic Infection. J Can Dent Assoc 2006;72(9):829-832.

7. Ganesh PD: A rare case of herpes zoster oticus in an immunocompetent patient. Dermatol Online J 2012;3(4):349-351.

8. Veit G, Rüdiger VK, Eva R, Bernd HB, Johannes L: Immunocompetent children account for the majority of complications in childhood Herpes Zoster. J Infect Dis 2007;196:1455-8.

9. Antonio GT, Antonio MV, Antonio AJ, Francisco EO: Ramsay-Hunt Syndrome presenting laryngeal paralysis. Acta Otorrinolaringol Esp 2013;64(1):72-74.

10. Barreira JACS. Paralisia Facial Periférica: Impacto na qualidade de vida. 2010. Disponível em: https://www.dropbox.com/s/bsu14ox7w0cu3as/tesedemestradojo%C3%A3o.pdf . Acessado em: 2014 (Abril 7).

11. Augustine JS, Patrick AT: Ramsay Hunt Syndrome in a patient with human Immunodeficiency Virus Infection. JABFP 2001;14(5):392-394.

12. Schulza P, Arbusowa V, Struppa M, Dietericha M, Sautierb W, Brandta T. Sympathetic contralateral vestibulopathy after unilateral zoster oticus. J Neurol Neurosurg Psychiatry 1999; 66:672–676.