American Medical Journal 3 (2): 237-239, 2012 ISSN 1949-0070 © 2012 Science Publications

Ramsay Hunt Syndrome: Case Report

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Abstract: Problem statement: Ramsay Hunt syndrome is characterized by peripheral facial palsy and eruptions in external ear reportedly due to the reactivation of latent varicela zoster virus in the sensory ganglia of facial nerve. Our purpose is to describe a case of this syndrome accompanied by the Neurology Service of Nova Iguacu General Hospital. Approach: A 60-year old female patient sought the emergency room due to a complaint because she was not able to close her left eye, followed by a drift of the labial fold to the right as well as a sensation of "burning eyes" for the last three days. She also referred vertigo and bilateral hypoacusis, more intensely felt on the left side. Results: Physical examination showed a left facial palsy with a vesicular eruption in the left external auditory canal, ear lobe and neck on that side. Her taste sensation was decreased on the anterior 2/3 of the tongue, a negative rinne test, a positive weber test indicating a neurossensorial hearing loss, ataxia in walking, a Romberg sign and an abnormal fukuda pace test, however the CT scan was normal. Conclusion: The diagnosis is basically clinical, in turn, treatment is controversial. In addition to clinical findings, the diagnosis is confirmed by the presence of viral DNA in the involved tissue and vesicular exudate, as assessed by polymerase chain reaction. Ramsay Hunt syndrome involves severe dysfunction, with poorer facial nerve prognosis than in Bell's palsy. Some studies suggest that treatment with prednisone and acyclovir may improve outcome, although a prospective randomised treatment trial remains to be undertaken.

Key words: Facial paralysis, herpes zoster, deafness, ramsay hunt syndrome

INTRODUCTION

In 1907, James Ramsay Hunt described a clinical syndrome-Ramsay Hunt Syndrome (RHS) that carries his name, in which peripheral facial paralysis, pain and ipsilateral vesicular lesions are caused by involvement of the geniculate ganglion by the Varicella Zoster Virus (VZV) (Goldani *et al.*, 2009; Gilden *et al.*, 2010; Santos *et al.*, 2010; Demir and Basut, 2012). Highlighting that vesicles can also appear on the eardrum (or tympanic membrane), in the external audiotry canal, the outer ear as well as along the mastoid process. Occasionally, the

anterior facial pillar of the palate or neck may also be involved (Uscategui *et al.*, 2008).

Hunt described two kinds of syndrome: an otalgic one (characterized by ear pains) and a prosopalgic one (facial pains) (Uscategui *et al.*, 2008). Facial palsy is accompanied by disorders in taste, hyperacusis and a decrease in salivary as well as tear production (Uscategui *et al.*, 2008). Variable degrees of involvement of the Eighth Cranial nerve occurs in about 20% of cases (Esteves *et al.*, 2010). It is believed that RHS is responsible for 7-16% of unilateral facial palsies of nontraumatic origin (Demir and Basut, 2012;

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Lima and Junior, 2011). The VZV posseses a tropism for ganglionary nerve tissue, thus causing an intense inflammatory reaction, especially in the elderly, diabetics and the immunosupressed ones (Goldani et al., 2009; Esteves et al., 2010; Oviedo et al., 2007). Diagnosis is basically clinical (Esteves et al., 2010). Once that, clinical course is more severe than the facial palsy itself, the complete clinical recovery occurs in only 30% (Demir and Basut, 2012). Treatment is controversial, some authors reccomending analgesics and eye lubricants while others suggest corticosteroids in order to lessen the inflammatory process (Secchi et al., 2000; Alcantara et al., 2000). Studies show that aciclovyr decreases the total duration of disease as well as eye complications (Wagner et al., 2012; Esteves et al., 2010; Secchi et al., 2000).



Fig. 1: Left facial palsy



Fig. 2: Vesicular eruption in left external auditory canal, ear lobe and neck

Case report: A 60-year old female patient sought the emergency room of Nova Iguaçu General Hospital due to a complaint of not being able to close her left eye accompanied by a drift of the labial fold to the right as well as a sensation of "burning eyes" for the last three days, but, there was no ear pain. She also referred vertigo and bilateral hypoacusis, more intensely felt on the left side. Physical examination showed a left facial palsy with a vesicular eruption in the left external auditory canal, ear lobe and neck on that side (Fig. 1 and 2). Taste sensation was decreased over the anterior 2/3 of her tongue, a negative rinne test, a positive weber test indicating a neurossensorial hearing loss, ataxia in walking, a Romberg sign and an abnormal fukuda pace test. There was no nystagmus, muscle strength and superficial sensibility were normal. Deep tendon reflexes were both normoactive and symmetrical. At all, there was no babinksi sign, no dysmetrias or dysdiadococinesias and her CT scan was normal. The treatment with 40 mg of prednisone along with 800 mg of aciclovyr was begun over a five-day period. Eye lubricant was applied as well as occlusion of the left eye (eyepatch, gauze) the patient was eventually discharged and is currently being evaluated on an outpatient basis by a multi disciplinary approach (neurologist, physical therapist, ear/nose and throat specialist and ophtamology).

MATERIALS AND METHODS

For this clinical case we decided address a brief review about Ramsay Hunt Syndrome. A research was made at Scielo and Pubmed using terms facial palsy, herpes zoster, deafness and Ramsay Hunt Syndrome. Articles published since 1998 were included in this study.

RESULTS

The research resulted in 30 articles with free full text available. From these, were selected those who had close relation with the patient's case. The selected articles allowed better fundamentation to the treatment and follow of this patient.

DISCUSSION

Facial nerve is a mixed nerve, having a motor, sensory and visceral root; the latter called the intermedius nerve. Seventh Cranial nerve is in close association with the VIII CN, the latter an exclusively sensorial one (Morrow, 2000; Terada *et al.*, 1998). A clinical presentation of peripheral facial nerve palsy, vesicular ear lobe and auditory canal eruptions,

neurossensorial deafness and vertigo is classic for Ramsay Hunt Syndrome (Esteves *et al.*, 2010).

The case report explained is a typical one, except for the absence of ear pain, an extremely common complaint in these patients. Noteworthy, the physical exam showed an involvement of the seventh and eighth cranial nerves, as well as the intermedius. Similarly, studies suggest that age over 50 and the presence of a complete facial paralysis are unfavorable prognostic signs. Complications include post-herpetic neuralgia and corneal inflammation (Oviedo *et al.*, 2007). An early diagnosis of RHS is important as prognosis of cranial nerve damage (Wagner *et al.*, 2012). Antiviral agents and steroids as a combination therapy improve recovery of facial nerve palsy and other cranial nerve palsies (Kim *et al.*, 2010).

CONCLUSION

Ramsay Hunt syndrome is more likely to presenting variable clinical picture, depending on the number of cranial nerve pairs involved. It is caused by a reactivation of a previously acquired and heretofore latent VZV infection in the geniculate ganglion. The ideal approach for treatment is still controversial and a multidisciplinary approach is essential for the follow up and recovery of these patients.

REFERENCES

- Alcantara, L.J.L., F. Gavaszoni, A.L. Ataíde and E.A.P. Carvalho, 2000. Síndrome de ramsay-hunt: descrição de caso. Rev. Bras. Otorrinolaringologia, 66: 536-538.
- Demir, U.L. and O. Basut, 2012. Ramsay Hunt syndrome and current approaches. Kulak Burun Bogaz Ihtis Derg., 22: 59-63. DOI: 10.5606/kbbihtisas.2012.012
- Esteves, M.C.B.N., L.A.F. Brandao, K. Kobari, J.C. Nardi, *et al.*, 2010. Síndrome de ramsay hunt: Relato de caso e revisão de literatura. ACTA ORL/técnicas em Otorrinolaringologia, 28: 37-39.
- Gilden, D., R.J. Cohrs, R. Mahalingam and A.M. Nagel, 2010. Neurological disease produced by varicella zoster virus reactivation without rash. Curr. Top Microbiol. Immunol., 342: 243-53. DOI: 10.1007/82_2009_3

- Goldani, L.Z., L.F.D. Silva and J.M. Dora, 2009. Ramsay Hunt syndrome in patients infected with human immunodeficiency virus. Clin. Exp. Dermatol., 34: e552-e554. DOI: 10.1111/j.1365-2230.2009.03228.x
- Kim, Y.H., M.Y. Chang, H.H. Jung, Y.S. Park and S.H. Lee *et al.*, 2010. Prognosis of Ramsay Hunt syndrome presenting as cranial polyneuropathy. Laryngoscope, 120: 2270-2276. PMID: 20824789
- Lima, M.A. and J.D.N. Junior, 2011. Ramsay Hunt syndrome following otoplasty. Braz. J. Otorhinolaryngol., 77: 808-808. PMID: 22183292
- Morrow, M.J., 2000. Bell's palsy and herpes zoster oticus. Curr. Treat. Options Neurol., 2: 407-416. PMID: 11096766
- Oviedo, A.M. and M.T.L. Zamarro and J.J.U.Del Hoyo, 2007. Sindrome de Ramsay-Hunt. Med. Interna, 24: 31-34.
- Santos, M.A., H.H. Caiafa-Filho, M.F. Vianna, A.G. Almeida and P.R. Lazarini, 2010. Varicella zoster virus in Bell's palsy: A prospective study. Braz. J. Otorhinolaryngol., 76: 370-373. PMID: 20658018
- Secchi, M.M.D., R.G.D.S. Bastos, C.M.J. Finamori, L.B. Coelho and M.C.L. Chayamiti, 2000. Síndrome de Ramsay Hunt: relato de caso. Acta Medica Misericordiae, 3: 65-67.
- Terada, K., T. Niizuma, S. Kawano, N. Kataoka and T. Akisada *et al.*, 1998. Detection of varicella-zoster virus DNA in peripheral mononuclear cells from patients with Ramsay Hunt syndrome or zoster sine herpete. J. Med. Virol., 56: 359-363. PMID: 9829642
- Uscategui, T., C. Dorée, I.J. Chamberlain and M.J. Burton, 2008. Antiviral therapy for Ramsay Hunt syndrome (herpes zoster oticus with facial palsy) in adults. Cochrane Database Syst. Rev., 8: CD006851-CD006851. PMID: 18843734
- Wagner, G., H. Klinge and M.M. Sachse, 2012.
 Ramsay Hunt syndrome. J. Dtsch. Dermatol. Ges., 10: 238-244. DOI: 10.1111/j.1610-0387.2012.07894.x