

# Ramsay-Hunt syndrome: a case report

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## INTRODUCTION:

Ramsay-Hunt syndrome is a disease caused by reactivation of latent varicella zoster virus in the geniculate ganglia. It involves facial nerve paralysis and it is usually associated with a rash on the auricle and/or external ear [1]. Sometimes the blisters are not very noticeable and can be overlooked by the inspector. Other accompanying symptoms may be ache and fullness of the ear (78%), vertigo/dizziness (51%), hearing loss (45%), nausea, vomiting, tinnitus and hyperacusia [2].

The diagnosis is mainly based on the history, clinical findings and neurological examination. Polymerase chain reaction assays may be useful by detecting DNA of the virus in exudates from ear scraping or cerebrospinal fluid (CSF) [3].

It represents 7% of all peripheral facial paralysis and recovery rates are lower in this group [4,5]. Early treatment with a combination of acyclovir and prednisone is reported to be effective [6-8].

## CASE DESCRIPTION:

A 46-year-old previously healthy woman was admitted to the emergency department complaining of mouth drooping and vertigo since that morning. She also had a headache and pain on the right ear since the night before. She denied fever, hearing loss, hyperacusia, tinnitus, nausea or vomiting.

On physical examination, she was alert and oriented, with a Glasgow Coma Scale score of 15, without focal neurologic deficits except for right peripheral facial palsy [Figure 1]. Gait was not tested because of her vertigo but she had no dysmetria on finger-to-nose and heel-to-knee tests. She was hemodynamically stable and afebrile with no meningeal signs. She had no visible vesicular eruptions nor any abnormalities on otoscopy.

Lab work [Table 1] showed a total leukocyte count of 12.270/uL (normal range 4.000-12.000/uL). Other than that, blood routine examination was normal, including a low level of C-reactive protein (4mg/L, normal range <5mg/L).

Contrast-enhanced head CT scan [Figure 2] showed no signs of acute vascular, hemorrhagic or edematous events and documented normal permeability of deep and superficial venous system.

Because of persistent headache, she underwent a lumbar puncture with drainage of clear CSF. CSF analysis showed normal glycorrhachia, no pleocytosis and slight proteinorrhachia (84 mg/dL, normal range 15-60mg/dL). Screening for varicella zoster DNA on CSF by polymerase chain reaction was positive.

A clinical diagnosis of Ramsay-Hunt syndrome was established. The patient was started on acyclovir 10 mg/kg every 8 hours for 14 days, prednisolone 1mg/kg for 5 days, humidifying eye drops and right eye

protection. Physiotherapy for the face was initiated and the patient slowly improved, maintaining at the time of discharge a class III palsy on the House-Brackmann Facial Nerve Grading System.

## DISCUSSION:

Ramsay-Hunt syndrome is perhaps an underdiagnosed form of peripheral facial palsy [5], since the diagnosis is mainly based on clinical findings of facial weakness and a vesicular rash [1]. Regarding the timing of these manifestations, the clinical picture includes three possibilities. When the vesicular rash occurs before or simultaneously with the facial palsy, the diagnosis is usually easy. Sometimes the blisters develop only after the onset of facial weakness and occasionally they are not very noticeable and can be overlooked. A minority of patients, like ours, do not develop them at all. One retrospective study showed that in 6% of patients, only the ear canal or the ear drum were affected, without visible blisters outside. This can make the diagnosis more difficult and indistinguishable from Bell's palsy [2].

Our patient also complained of ear pain, headache and vertigo. This syndrome may also present with acute herpetic neuralgia that can radiate to the face, ear, head and neck [9]. Approximately 50% of patients are expected to have an injury of the vestibulocochlear nerve. This can cause nausea, vomiting, gait insecurity with a tendency to fall to the affected side, vertigo and nystagmus [10]. Damage of the cochlear component leads to tinnitus and hearing loss [11,12].

Analysis of cerebrospinal fluid and brain magnetic resonance imaging has limited diagnostic and prognostic value. However, polymerase chain reaction assays may be useful to detect herpes zoster virus DNA in exudates from ear scrapings or CSF and MRI can visualize inflammation of the cranial nerves [3].

It is important to differentiate Ramsay-Hunt syndrome from other forms of facial palsy, since the former often has more severe paralysis and has less chances of recovery [13]. Treatment of this condition involves both glucocorticoids and antiviral therapy, while for Bell's palsy it remains uncertain whether antiviral therapy adds benefit [5,8,10]. Antiviral agents improve acute pain, recovery rate and prevent from the occurrence of postherpetic neuralgia [14]. Glucocorticoids have a strong anti-inflammatory action, reducing edema in the nerves involved, which can lead to a hasten recovery [10]. There does not seem to be a difference between the use of oral versus intravenous antivirals [11]. Starting treatment in the first 48 hours of symptoms, as we did in this case, is crucial as nerve damage and therefore prognosis depends on the timing at which the combination therapy is started [5,8,10].

## CONCLUSIONS

A high index of suspicion is necessary to diagnose a Ramsay-Hunt syndrome in a patient presenting with peripheral facial palsy, especially in the absence of a typical vesicular rash. Analysis of CSF has limited value but the detection of varicella zoster DNA may help to prove the diagnosis. Furthermore, early diagnosis and adequate treatment is crucial to improve damaged nerves and maximize the chances of full recovery.

## AUTHOR CONTRIBUTIONS:

Author 1: Involved in patient care, data and information collection and main author of the manuscript.

Author 2: Involved in patient care and manuscript review.

Author 3: Involved in patient care and manuscript review.

Author 4: Involved in patient care and manuscript review.

Author 5: Involved in manuscript review.

Author 6: Involved in manuscript review.

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