CASE STUDY

Ramsay-Hunt Syndrome presenting laryngeal paralysis

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KEYWORDS

Ramsay-Hunt; Varicella-zoster virus; Laryngeal palsy

Abstract The Ramsay-Hunt syndrome is the association of facial palsy and varicella-zoster virus infection with involvement of the ear canal and eardrum. It may be associated with deafness, tinnitus and dizziness. It can sometimes affect the lower cranial nerves. A case of an immunocompetent patient with affection of the VII, VIII and X cranial nerves is presented.

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PALABRAS CLAVE

Ramsay-Hunt; Herpes zoster; Parálisis laringea

Resumen El síndrome de Ramsay-Hunt consiste en la asociación de parálisis facial periférica (PFP) e infección por virus varicela zoster (VVZ) con afectación del conducto auditivo externo y membrana timpánica. Se puede acompañar de sordera, acúfenos y vértigos. En ocasiones puede afectar los pares craneales bajos. Se presenta el caso de un paciente inmunocompetente con afectación de los pares craneales VII, VIII y X.

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Introduction

Varicella-zoster virus (VZV) can cause otological manifestations such as peripheral facial palsy (PFP), vesicular skin rash, tinnitus, hearing loss and vertigo. We report the case of a patient suffering classic Ramsay-Hunt syndrome with involvement of the seventh and eighth cranial nerves. The subject was immunocompetent, but with the particularity of presenting involvement of the tenth cranial nerve. The diagnosis was confirmed clinically, as well as by laboratory tests and imaging techniques. Our objective is to inform of this extremely rare neuritis, in order to facilitate the early diagnosis of other, similar patients.

Case Report

The patient was a 70-year-old male who was admitted to the Emergency Unit due to right otalgia, tinnitus, hearing loss, vertigo, hiccups and dysphonia. Personal history included hypertension, diabetes mellitus and dyslipidemia. An otoscopy showed vesicles and scabs in the right ear canal. Vestibular examination revealed a harmonic vestibular syndrome with strong right lateropulsion and grade II spontaneous nystagmus towards the left. Clinical
examination of the facial nerve showed grade IV PFP in the House-Brackmann scale. Nasofibroscopy observed paralysis of the right vocal cord (RVC). The patient was admitted to the ENT Unit with a diagnosis of Ramsay-Hunt syndrome, and was treated intravenously with 800 mg/4 h of acyclovir and 60 mg/8 h of methylprednisolone, followed by a decreasing dosage of 50 mg/12 h of sulpiride for 5 days and analgesia with paracetamol and metamizole. Laboratory tests were normal. A computed tomography (CT) scan of the head, neck and thorax showed no relevant findings, whilst a magnetic resonance imaging (MRI) scan of the skull showed neuritis of the facial nerve affecting its entire path from the fundus of the internal auditory canal to the mastoid portion (Fig. 1).

The patient was discharged after 10 days. Outpatient monitoring conducted after 1 month detected a persistence of grade II PFP and RVC paralysis, whilst hearing loss, vertigo and hiccups had improved considerably. In the review at 6 months, the patient still suffered RVC paralysis in a para- median position, with good compensation by the left vocal cord (LVC). There were no facial asymmetries and both vertigo and hiccups had disappeared, but a mild sensorineural hearing loss of 40 dB still remained (Fig. 2).

Discussion

We present a case of Ramsay-Hunt syndrome characterised by the classic triad of facial palsy, otic herpes and hearing loss, as described by Ramsay-Hunt in 1907, but with the peculiarity of involvement of the vagus or pneumogastric nerve, causing hiccups and right recurrent paralysis. Following the description by Ramsay-Hunt, the involvement of other cranial nerves was reported. In reality, it is a multiple peripheral neuritis which may affect cranial nerves 7, 8, 9 and 10 due to their close proximity, common embryonic origin in the same branchial arch or by blood involvement through their vessels.

Ramsay-Hunt syndrome presents very broad clinical variants, with the common feature of affecting various cranial nerves. Vestibular disorders are more common than audiological (72%–85% of patients), but tend to evolve towards total compensation. Cochleovestibular manifestations become more common with increasing facial involvement. Otic zoster is the cause of 10.7% of PFP.

The aetiology of laryngeal paralysis has been studied by several authors, who have obtained mixed results. These include laryngeal and extralaryngeal tumours, thyroid surgery, prolonged intubation, trauma, neurological and idiopathic causes and others.

Flexible laryngoscopy is the gold standard for the diagnosis of vocal cord paralysis; direct observation of abnormal movement confirms the diagnosis. In asymptomatic patients, sensitivity can be increased with phonation, deep breathing or gasping. The differential diagnosis between laryngeal paralysis and fixation as such, can be complex. A detailed medical history may help, although some cases may require an electromyographic study.

Regarding the clinical evolution at 6 months, the recovery of PFP is related to the initial level of involvement. Vestibular disorders often show a good clinical response.

We can conclude that classic Ramsay-Hunt syndrome may occur in various clinical presentations, with the common feature of cranial nerve neuritis in varied forms. Ramsay-Hunt syndrome is a rare complication of geniculate ganglion involvement by VZV. We must be aware that the main clinical features are not always present at the same moment in time. In such cases, serological testing will provide confirmation of the suspected diagnosis.

Early diagnosis is important, so that treatment with acyclovir and corticosteroids may be initiated as soon as possible, in order to maximise the recovery rate of nerve function.
Conflict of Interest

The authors have no conflict of interests to declare.

References