



CASUISTIC PAPER

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Ramsay Hunt syndrome with deep hearing loss and meningitis

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ABSTRACT

Introduction. Ramsay Hunt syndrome is a clinical manifestation of varicella zoster virus reactivation. It is characterized by an erythematous vesicular rash in the external auditory canal and pinna with otalgia, vertigo and ipsilesional facial palsy. Symptoms develop over a few days with prodromal signs of facial weakness, tingling, facial numbness. Usually, cranial nerves VII and VIII are involved in the inflammatory process. Possible consequences of Ramsay Hunt syndrome are hearing loss, encephalitis and meningitis.

Description of the case report. The authors present the case of a 63-year-old woman with a vesicular rash, earache, vertigo and left-sided facial paralysis who was treated with antiviral drugs and analgesics. These symptoms were complicated by conductive hearing loss in the left ear and meningitis. After treatment facial paralysis decreased. Unfortunately, hearing loss was permanent.

Discussion. Rapid administration of antivirals and corticosteroids limited facial paralysis and improved facial expression. The prognosis for facial palsy is poorer in Ramsay Hunt syndrome than in idiopathic forms.

Conclusions. A past history of vertigo and hypertension could be a predisposing factor for the severe manifestation of Ramsay Hunt syndrome and subsequent complications.

Key words. facial palsy, hearing loss, meningitis, Ramsay Hunt syndrome

Introduction

Ramsay Hunt syndrome (RHS) is characterized by an erythematous vesicular rash in the external auditory canal and pinna with severe otalgia and ipsilesional facial palsy. It is a clinical manifestation of varicella zoster virus (VZV) reactivation. Neurological complications include changes in cerebrospinal fluids, peripheral motor neuropathy, aseptic meningitis, and cranial polyneuropathy. Both cranial nerves VII and VIII are commonly involved in the inflammatory process, producing vestibulocochlear symptoms such as vertigo, hearing loss, and

tinnitus and symptoms of peripheral facial paralysis. Among these, vertigo, hearing loss, and tinnitus most commonly occur in patients with RHS.¹⁻⁴

Description of the case report

A 63-year-old woman with a past medical history of vertigo and hypertension first presented a left-sided headache and burning for the past few days. One day later she had developed multiple vesicles on the left side of her face. She complained of earache and vertigo. ENT examination did not reveal any inflammatory process

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Figure 1. Facial paralysis on the left side (3rd degree according to the House-Brackmann grading scale) after six month

of the left ear. Computed tomography (CT) of the head revealed no changes. An ENT examination revealed a vesicular rash in the pinna of the left ear, peripheral paralysis of nerve VII on the left side (V degree according to the House-Brackmann grading scale) and deep conductive hearing loss (60-70 dB in audiometric tests). Brain magnetic resonance imaging (MRI) detected a high signal intensity lesion in both a T₂-weighted image and fluid-attenuated inversion recovery (FLAIR) apparent in white in both parietal patches, most likely vascular-related. Intraventricular fluid spaces were normal. In addition, the brain structure was without focal changes and with the correct signals. After intravenous administration of the paramagnetic agent, there was no focus of pathological contrast enhancement. On the other hand, there was a slightly more intense contrast enhancement of the mid-range and the front of the skull, which could be attributed to the inflammatory process of the meninges. Air entrapment of the pneumatic pyramidal cells of the left temporal bone was significantly reduced- probably by inflammatory lesions. Laboratory tests detected WBC 7.2 HGB 12.6 g/dl, CRP 2.35 mg/l. She was treated with antiviral drugs: acyclovir 800 mg 5 times daily every 4 hours with a night break. Tramadol hydrochloride (37.5 mg) and acetaminophen (325 mg) were administered 4 times daily to alleviate pain, carbamazepine 200 mg daily and ketoprofen 100 mg and metamizol 0.5 mg intravenously once ad hoc in the case of severe pain. Six months later, facial weakness on the left side (3rd degree according to the House-Brackmann grading scale) was seen. Conductive hearing loss of the left ear (60-80 dB) was without improvement and required the use of equipment.

Discussion

The incidence of hearing loss in patients with RHS has been shown to range from 6.5% to 85%. According to Chang et al., the incidence of hearing loss in patients

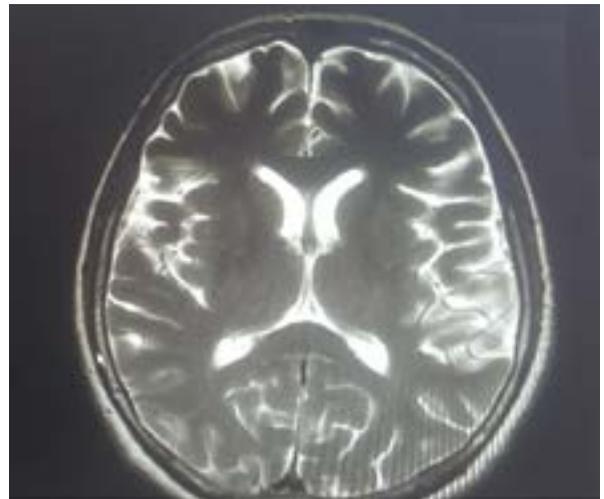


Figure 2. Changes in MRI examination

with RHS is 76% and is more severe in the high frequency range than in the low frequency range.¹ Transmission of VZV infection from a dehiscence facial canal to the inner ear and organs through the oval or round window has been suggested as a potential route for inner ear involvement. For interneural transmission, the spread of VZV across the perineural tissues inside the internal auditory canal has been proposed as a possible route of infection. Cerebrospinal fluid infection is more likely to be a source of cochlear deficit rather than nerve anastomosis and direct connection between cranial nerve VII and VIII. Hearing loss and facial nerve paralysis are the predominant permanent consequences of VZV infection. The permanent character of these complications are caused by deep tissue processes. Histopathological examinations reveal pronounced segmental or partial atrophy and degeneration of cranial nerve VII and/or VIII.² Vertigo and facial nerve palsy depend on each other, because severe vestibular symptoms can be related to the severity of facial paralysis after the onset of herpetic symptoms.⁵ The prognosis for facial palsy is poorer in Ramsay Hunt syndrome than in idiopathic forms.⁶ Only 10% of patients with complete facial palsy are totally cured, with full recovery in as few as 20% of cases.⁷ Facial nerve involvement is initially due to inflammation caused by the viral neuronitis and secondarily to the facial nerve edema.^{8,9} Advanced age, elevated arterial blood pressure, vertigo and diabetes can be factors of poor recovery in Ramsay Hunt syndrome.¹⁰ Hypertension and vertigo, which might have been the causes of poorer recovery from facial paralysis. Early administration within 72 hours of antivirals and corticosteroids improves the prognosis.^{8,9,10,11} Recovery rates in patients with RHS are higher following treatment with steroid plus famciclovir than with steroid plus acyclovir, especially in patients without hypertension and diabetes mellitus.¹² A past history of vertigo is

a predisposing factor for hearing impairment. Hearing impairment is more severe in patients with vertigo than in those without vertigo in both the high and low frequency ranges, even though the degree of hearing impairment is not significantly different between patients with and without facial palsy. These findings indicate that the mechanisms of viral spread from CN VII to CN VIII may differ between vestibular and audiologic deficits.¹ The coexistence of Ramsay Hunt syndrome and varicella zoster encephalitis and meningitis is rare. Concomitant diseases such as diabetes and chronic renal failure may lead to an aggressive course of infection and can predispose to encephalitis and meningitis.^{13,14} Organ transplant recipients also had herpes oticus more frequently. The incidence of VZV infection is reported as 11.2% at 4 years after kidney transplantation, an incidence that is approximately nine times greater than that in the general population.⁸ HZ develops in 12% of liver transplant recipients.¹⁵ For a transplant surgeon, it is imperative to remember that viral prophylaxis is essential in the follow-up of the transplant patients.¹⁶ The possible mechanism of VZV spreading to CNS is reactivated viruses, which establish latency in geniculate ganglia, upward through the acoustic internus along with the facial canal, and eventually enter intracranially and first invade the basis pontis. Meanwhile, VZV may also spread downwards along with general somatosensory fibers to the skin of external auditory canal, resulting in herpes zoster formation.¹⁷ Some data indicate that vasculitis might also be involved in other VZV CNS manifestations, such as herpes zoster-associated encephalitis. For this reason, VZV CNS infection must be suspected in several CNS syndromes and diagnostics should be based on CSF analysis for the detection of VZV DNA by PCR and/or intrathecal antibody production.^{18,19,20}

Conclusions

Meningitis is possible but rare complication of herpes oticus. Hearing loss is quite often consequence of Ramsay Hunt syndrome. Favourable facial nerve paralysis recovery depends on rapid administration of antivirals and corticosteroids. The prognosis for facial palsy is poorer in Ramsay Hunt syndrome than in idiopathic forms. A past history of vertigo can be a predisposing factor of hearing impairment.

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