# ATYPICAL PRESENTATION OF RAMSAY HUNT SYNDROME WITHOUT FACIAL PALSY IN AN IMMUNOCOMPETENT YOUNG MALE

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***ABSTRACT***

*We report the case of a 28 year old man complaining otalgia, mild odynophagia, small vesicles on the neck and the swelling of regional lymph nodes. A dermatologist promptly diagnoses Herpes Zoster and prescribes acyclovir-steroid (prednisone) therapy (AS), that the patient buys and takes immediately. He does not complain fever, hearing loss, nausea, vomiting or vertigo. There was no reduction of taste sensation, no ataxia or nystagmus, Romberg sign was negative. Muscle strength was normal. The strict definition of Ramsay Hunt syndrome is peripheral facial nerve palsy accompanied by erythematous vesicular rash on the ear or in the mouth. Our patient targets two of the three criteria needed for the diagnosis of Ramsay Hunt syndrome. We explain that because he was immunocompetent, he was young and begun to take AS therapy very early. This suggests that prompt diagnosis and management improves outcome in Ramsay Hunt syndrome.*

***Key words****: Ramsay Hunt syndrome, Varicella Zoster Virus, immunocompetent, young, antiviral-steroid therapy.*

***Received*** *July 05, 2013;* ***Accepted*** *August 28, 2013*

## Introduction

In 1907, James Ramsay Hunt described a syndrome characterized by otalgia, auricular vesicles and peripheral facial paralysis. At that time, it was well accepted that infection of ganglia and skin by a herpes virus produced a characteristic dermatomal distribution of pain and vesicular rash. Hunt hypothesized that this syndrome was a result of herpetic infection of the geniculate (seventh nerve) ganglion. He also described other accompanying signs and symptoms including tinnitus, hearing loss, nausea, vomiting, vertigo and nystagmus[(1-2)].

Hunt described three different syndromes, the best known of which is zoster oticus with peripheral facial palsy. The second Ramsay Hunt syndrome encompasses the clinical features produced by carotid artery occlusion[(3)]. A third Ramsay Hunt syndrome is also known as dyssynergia cerebellaris progressiva[(4)].

Although Ramsay Hunt syndrome is traditionally defined as zoster oticus and lower motor neuron facial palsy, Hunt noted other regular symptoms and signs such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus (Tab.1).

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| **Main clinical features** |
| ➢ A painful red rash with fluid-filled vesicles occurring anywhere along the sensory distribution of the facial nerve, including the anterior two-thirds of the tongue, the soft palate,the pinna or the external auditory canal |
| ➢ Acute peripheral facial paralysis |
| ➢ Otalgia |
| **Other clinical features** |
| ➢ Tinnitus |
| ➢ Vertigo and ipsilateral hearing loss |
| ➢ Nystagmus |
| ➢ Nausea |
| ➢ Vomiting |
| ➢ Increased/Decreased lacrimation |
| ➢ Change in taste perception or loss of taste |

**Table 1**: Presenting clinical features of Ramsay Hunt syndrome.

He explained these eighth nerve features by the close proximity of the geniculate ganglion to the vestibulocochlear nerve within the bony facial

canal. Based on clinical presentations that indicated involvement of more than one ganglion, Hunt surmised that the gasserian, geniculate, petrous, accessory, jugular, plexiform, and second and third cervical dorsal root ganglia comprised a chain in which inflammation of a single ganglion could extend to nearby ganglia. This hypothesis explained cases of unilateral facial palsy accompanied by contiguous cranial neuropathies associated with vesicles in the mouth-usually on the tongue or hard palate-or ear.

Although this hypothesis remains valid, contiguous cranial neuropathies can also be explained based on the selective vulnerability of blood vessels to varicella zoster virus (VZV). Transaxonal spread of VZV from one or more cranial ganglionic nerves afferent fibres to the vasa vasorum of cranial nerves could produce infarction with resultant zoster polyneuritis cranialis[5].

Ramsay Hunt syndrome is the second most common cause of atraumatic peripheral facial paralysis and involves approximately 5 cases per 100.000 people. Compared with Bell palsy, Ramsay Hunt syndrome generally has more severe paralysis at onset and patients are less likely to recover completely. Before 1986, the frequency of zoster in patients with peripheral facial paralysis was estimated to be 4.5%-8.9%. In 1986, a retrospective review of 1507 consecutive patients presenting with unilateral facial palsy identified Ramsay Hunt syndrome in 185 (12% patients) based on the triad of facial paralysis, ear pain, and herpetic eruptions in any cranial dermatome[6]. Viral aetiology was confirmed in 46 patients by a fourfold rise in antibody to VZV. There were 20% more women (101) with Ramsay Hunt syndrome than men (84), and 56 (30%) were younger than 24 years, indicating a sampling bias due to the younger age of the patients. A retrospective review of 2076 patients presenting with unilateral facial palsy, with or without vesicles, from 1976 to 1996 in Japan disclosed a similar incidence of Ramsay Hunt syndrome in adults and children over age of 6 years[5].

In 1944, Denny-Brown et al[7] described a 62 year old man who developed a vesicular, painful rash in the right occiput, neck, and external ear canal. Eleven days later, he developed right peripheral facial palsy, right tongue deviation, and decreased right C2-3 distribution pain and temperature sensation. Two CSF examinations showed a lymphocytic pleocytosis with a normal protein and glucose concentration. Four weeks later, he died from a massive gastrointestinal haemorrhage. Pathological changes at post-mortem examination showed a necrotic second cervical dorsal ganglion and increased microglial proliferation in the grey matter of the second and third cervical posterior and anterior horns. Mild lymphocytic infiltrates were seen in the meninges and in patchy areas of the seventh cranial nerve. The geniculate ganglion was normal. The hypoglossal, trigeminal, and glossopharyngeal nerves were not examined.

Two case studies of pathological changes 6 and 9 weeks after the onset of Ramsay Hunt syndrome reported similar findings. In both patients, perivascular lymphocytic inflammation, demyelination, and axonal loss along the seventh nerve, and mild lymphocytic infiltration of the geniculate ganglion were seen. One patient showed extensive haemorrhage in the auditory nerve at the internal auditory meatus with complete destruction of the apex of the organ of Corti. In both of these cases, most histopathological changes occurred outside the geniculate ganglion[8-9]. In another study of 152 patients with Ramsay Hunt syndrome, 19% had an abnormal audiogram, but no correlation between severity of facial weakness and hearing loss was found[10].

Many authors have suggested that the data do not confirm Hunt's original concept of geniculate ganglionitis but rather illustrate viral attack of the facial nerve itself. By contrast with VZV induced dorsal root ganglionitis, the temporal range of histopathological findings in Ramsay Hunt syndrome is unknown[5]. The geniculate ganglia and the facial nerve are not commonly dissected at necropsy, and there are few reported cases. Thus the debate surrounding the existence of geniculate ganglionitis awaits further information.

## Case report

We report the case of a 28 year old man complaining otalgia and mild odynophagia. The day after he notes erythema of the auricle and the swelling of regional lymph nodes. While taking antibiotics for the suspect of impetiginis or erisipela, he decides to consult a dermatologist. Thanks to the presence of small vesicles on the neck, he promptly diagnoses Herpes Zoster and prescribes acyclovir-steroid (prednisone) therapy (AS), that the patient starts immediately. During the week before, he complained also tinnitus at least twice a day but he thought to link that to the swimming, he was practicing since the previous week. At

the light of the diagnosis, he decides to consult also a ear, nose and throat doctor who visits him concluding with Zoster oticus without rash in the external auditory canal. Further, the patient complains mild hyperesthesia of a thin slice of scalp between the forehead and the occiput, maybe due to the anatomic continuity of cranial nerves and C1 to C3 spinal nerves, without vesicular rash. He does not complain fever, hearing loss, nausea, vomiting or vertigo.

Physical examination doesn't show facial palsy but only a vesicular eruption in the right ear lobe and neck on that side (Fig. 1-2). There was no reduction of taste sensation, no ataxia in walking or nystagmus, Romberg sign was negative. Muscle strength was normal.

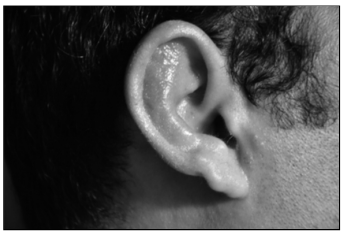

**Fig. 1**: Vesicular eruption in the right ear lobe.

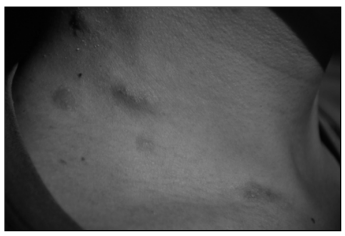

**Fig. 2**: On the same side, vesicular eruption in the neck.

## Discussion

The case of our patient targets two of the three criteria needed for the diagnosis of Ramsay Hunt syndrome. We have not measured VZV antibodies in the blood because the diagnosis is fundamentally clinical[11] and the lesions were without doubts vesicles. Viral cultures are often negative[12] and serologic testing is of limited use[11]. Also the irradiation of pain was typical for a dermatomeric distribution of the virus. Although swimming was not by itself the reason of the otalgia like suspected by the patient, maybe it was the reason of a reduction in immunologic defences that allowed the virus to reactivate. Our patient was immunocompetent, that is why, in our opinion, he promptly returned to his normal life and functionality in only four days of antiviral therapy, persisting for some other days a mild erythema of the points of the neck before involved and a mild regional lymphadenopathy. In fact, age greater than 60 years, diabetes mellitus, essential hypertension and associated vertigo were identified as prognostic factors for worse outcome and decreased chance of recovery, all conditions lacking in our patient[13]. But most important, he begun to take acyclovir-steroid (prednisone) therapy (AS) very early, we think that it is the reason of the absence of neural facial palsy as suggested by several studies in which there is evidence that in normal patients without immunosuppression, early administration of acyclovir-prednisone would improve the rate of healing of the skin lesions, diminish the duration of pain of herpes zoster, furthermore was proved to prevent nerve degeneration by nerve excitability test[14-15].

## Conclusion

The strict definition of Ramsay Hunt syndrome is peripheral facial nerve palsy accompanied by erythematous vesicular rash on the ear or in the mouth. Our patient targets two of the three criteria needed for the diagnosis of Ramsay Hunt syndrome. We explain that because he was immunocompetent, he was young and begun to take AS therapy very early. This suggests that prompt diagnosis and management improves outcome in Ramsay Hunt syndrome. We use corticosteroids, indicated in association to antiviral agents in the RHS, although our patient did not have the most worrying sign of peripheral palsy to prevent nerve degeneration and preserve its function, as suggested in literature. To improve his defences the ENT consultant suggested the patient to take immunostimulators, in particular he took pidotimod since the third day of the illness.

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