Ramsay Hunt syndrome with ipsilateral involvement of the glossopharyngeus nerve

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Background

Ramsay Hunt syndrome is caused by varicella zoster virus (VZV). It's clinical manifestation is facial palsy, otic pain, herpetic vesicles around the auricle and external auditory canal. [1]

The initial infection, also known as varicella, is a common exanthemous disease characterized by a highly contagious generalized vesicular rash. [2] VZV remains latent in the neurons of cranial nerves and dorsal root ganglia.

Many causes for the reactivation of VZV infection are supposed: stress (although not objectively measured); immunodeficiency (HIV; immune depressing medications, cancer, radio or chemotherapy; having varicella before the age of 1).

Ramsay-Hunt syndrome is a rare complication of latent VZV infection [3]. In some cases it might occur in the absence of pathological cutaneous alterations.

Some authors report that up to 20% of the clinically diagnosed cases of Bell's palsy are caused by VZV. [4]

In studies of patients with Ramsay-Hunt syndrome 27% were observed with additional cranial polyneuropathy, especially in immune suppressed cases. [5]

Case Report

A 19 years old patient with sudden hearing loss, asymmetry of the face and sore throat came in the

emergency. The ENT examination showed an erythematous vesicular rash of the skin fig. 1 of the right ear canal and auricle, sensorineural hearing loss on the same side fig. 2, ipsilateral peripheral facial nerve palsy fig. 3 and aphthous lesions on the right side of the soft palate fig. 4. The patient had a classical anamnesis of stress (loss of a parent) and two days later the herpes zoster infection.

Fig. 1

Fig. 2

Fig. 3

Fig. 4

However the patient was consulted with infectious diseases specialist (ID specialist) to determine treatment and future behavior. The localization and distribution of a vesicular rash make the diagnosis of herpes zoster highly likely; however, other viral exanthemas can occasionally be confused with

this disease (more often enteroviral infections and mostly group A coxsackieviruses). [6]

Serology showed high titers of anti VZV IgG and anti VZV IgA (ELISA) which correlated with the diagnosis.

The patient was treated with Acyclovir 3 x 500 mg i.v. for five days, and Methylprednisolon i.v. 60 mg (40 + 20) two days, 40 mg the next two days and 20 mg before dischargement . [7].

Three days later the patient had significant improvement of the facial nerve palsy (the patient could fully close his eye), no pain in the auricular region and average 30 dB improvement in the hearing. 20 days later the patient was recovered from the Ramsay-Hunt Syndrome with ipsilateral involvement of the glossopharyngeus nerve.

After five day-treatment the patient was discharged with home treatment – Acyclovir 5 x 800 mg p.o. for 5 days, Nimesulide 2 x 100 mg p.o. for 7-10 days and Pregabalin 2 x 100 mg p.o. for 7 days. [8]

Ten days later he was hospitalized in Physiotherapy clinic for aftercare. The patient was again consulted with ID specialist because of the persistent rash and low-grade fever. His status was not as better as it had to be according to the therapy. New serology was set, that time for Human Immunodeficiency Virus (HIV). After two positive tests (ELISA) and one confirmative test (Western blot) the patient was diagnosed as HIV-positive, so he was pointed at Clinic for Immunodeficiency patients.

Discussion

Ramsay-Hunt syndrome is a rare complication of the latent VZV infection. It is topic for ENT neurology and dermatology department. Numerous articles have reported various neuropathies additionally to the VII and the VIII CNV.

The following case presents additional to the Ramsay-Hunt's syndrome involvement of cranial nerve (glossopharyngeus). We would like to focus on the fact that although we have very typical anamnesis of stress (the patient has lost a close family member 2 days earlier) in cases of additional involvement of cranial nerves an immune suppressing therapy or disease has to be excluded.

The patient was only 19 years old but HIV positive. The close collaboration between the ENT specialist and the ID specialist had positive result for the quick diagnosis and precise therapy of the patient.

The early diagnosis is very important for seropositive patients, because the treatment must be started as soon as possible. The patient has better chances to keep a normal quality of life for many years if he is in early or latent stage of the illness. At present, patients have been maintained on combination therapy for several years, with a low rate of relapse due to development of resistance and low

viral burdens. Immune restoration and long-term biologic control or complete eradication of the viral burden with minimal toxicity remain elusive goals, while intense efforts continue to focus on new anti-HIV therapies. [9] However the early treatment nowadays is the only opportunity for a better and extended life.

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