

Case Report

Ramsay Hunt Syndrome with multiple cranial nerve palsy

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Abstract:

Ramsay Hunt Syndrome classically presents with VII nerve palsy. Other cranial nerve involvement is rare. We describe a case here who presented with VII, IX and X cranial nerve palsies and review the literature.

Key words: Ramsay Hunt; syndrome; cranial nerve palsy

Introduction:

Ramsay Hunt Syndrome was first described by Johns Ramsay Hunt in 1907. It is defined as acute facial nerve palsy with vesicular eruption of the skin of pinna and external auditory canal caused by varicella zoster virus^{1, 2}. Reactivation of the dormant viral particles within the geniculate ganglion of facial nerve acquired by primary varicella infection or chicken pox is responsible for its clinical manifestation. Classically it presents with VII nerve palsy on one side. But because of its close relationship with VIII nerve and its ganglions as it travels through the internal auditory canal and inner ear; infection may spread to the VIII nerve and presentation with

sensory neural hearing loss or dizziness is not unusual. But other cranial nerve involvement is rare²⁻⁴.

Pathophysiologically other cranial nerve involvement can be explained to occur either through the numerous neurological anastomosis between V, VI, VII, VIII, IX, X, XI, XII, cervical cutaneous nerves C2, C3 and C4 at the base of the skull or through their vasa nervorum or through localized leptomeningitis or by way of the cerebrospinal fluid^{2, 4}.

Review of the literature shows V and VII cranial nerves are the nerves commonly infected by herpes zoster virus in ENT- Head and Neck region⁵. While in reports of Ramsay Hunt Syndrome with multiple cranial nerve palsy IX and X cranial nerves are found most frequently affected⁴.

Case report:

A 50-year-old gentleman attended to the outpatient department of ENT- Head and Neck surgery of Banga Bandhu Medical Hospital(BBMH), University of Science and Technology, Chittagong(USTC) with five days complaints of severe pain in left ear followed by inability to close the left eye, pain in throat,

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hoarseness of voice and difficulties in swallowing. On examination of oral cavity angle of the mouth was found to be deviated towards the right with two big irregular ulcers over the left edge of tongue. Examination of throat showed multiple intermingled ulcerations over the soft palate and pharyngeal wall mostly on the left side with flattening of the left palatal arch and deviation of the uvula towards the right. Gag reflex was absent. On Indirect laryngoscopy extensive ulcerations of the base of the tongue was noted along with left vocal cord palsy. Left ear revealed multiple hemorrhagic blisters covered with crust over the concha extending up to the deep meatus. Rinne was positive on the right and negative on the left side. Weber was lateralized towards the left indicating conductive type of hearing loss of left ear. Facial nerve function test showed complete left facial nerve palsy of lower motor neuron type. We thus reached our clinical diagnosis of "Ramsay Hunt Syndrome with VII, IX and X cranial nerve palsy". We made the patient hospitalized, established an intravenous channel, started antiviral and steroid injections.

Patient was advised to take a semisolid diet. Ulcerations of the oral cavity and throat showed very quick recovery and patient started eating and drinking in full swing after two days of start of treatment. Earache reduced too along with the healing of the blisters of the ear canal. After five days of start of treatment, antiviral and steroid injections were replaced by oral preparations. Patient was very happy and pleased as discharged ten days later.

Discussion:

Chicken pox caused by varicella zoster virus is an infection of childhood where as Herpes zoster infection is a disease of elderly population which is said to develop due to

decline of cell mediated immunity with age or disease⁶. Reactivation of the varicella zoster virus after many years of primary infection is popularly known as "Shingles" and is seen most commonly in the face and trunk region⁷. Classically it starts in a ganglion and spreads along the nerve routes producing excruciating pain and vesicular eruption in the regional territory of that nerve. This is true in case of Ramsay Hunt Syndrome too where the herpes virus is reactivated within the geniculate ganglion of the facial nerve. Symptoms and signs of involvement of sensory and motor division of facial nerve are the principal presentation of Ramsay Hunt Syndrome. In ENT - Head & Neck region since the cranial nerves are very closely related and interconnected to one another presentation of this syndrome with multiple cranial nerve involvement is not unlikely though not common.

Ramsay Hunt Syndrome is diagnosed clinically from paroxysmal deep otalgia on one side along with appearance of rash or herpetic blisters in the distribution of nervous intermedius and VII nerve palsy. Presence of virus can be proofed by the rise of serological titer of VZV in serum or CSF. Viral DNA in the blister fluid can also be confirmed polymerase chain reaction (PCR)¹⁻⁶.

Treatment consists of anti viral agents (Acyclovir, Valacyclovir and Famciclovir) for about 7-10 days, steroid and a strong pain reliever. Best results are reported when treatment is started within 3 days of appearance of symptoms⁸. The overall incidence of Shingles and Ramsay Hunt Syndrome can be reduced by bringing more children in the coverage of varicella vaccine. In case of those who have already had chickenpox in childhood can also be benefitted by giving another vaccine which prevents reactivation of the varicella virus.

The prognosis of Ramsay Hunt Syndrome is not as good as that of Bell's palsy. Unlike Bell's palsy only 50% victims are likely to get a complete return of facial nerve function². Though there is good clinical evidence that treatment with antiviral agent, steroid and pain killer can improve recovery and lessen extreme facial discomfort and earache. However, some residual facial deformity might be left behind due to incomplete return of facial nerve function

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