Case Report

Ramsay Hunt Syndrome: A Rare Disease of Multiple Cranial Nerve Involvement

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ABSTRACT

Ramsay Hunt Syndrome (RHS) also known as herpes zoster oticus is a viral disease, a member of the human herpes virus family, is a late complication of varicella-zoster virus infection that results in inflammation of the geniculate ganglion of cranial nerve VII. Ramsay Hunt is a clinical diagnosis. The hallmark of the condition is multiple unilateral erythematous vesicles, which are distributed over the auricle and preceded by severe otalgia. If these symptoms are associated with facial nerve palsy, the condition is called RHS which is usually accompanied by vestibulocochlear abnormalities. Diagnosis is often missed or delayed, which can lead to an increased incidence of long-term complications. The condition is self-limiting, but treatment is targeted at decreasing the total duration of the illness as well as providing analgesia and preventing the complications that can occur. This activity reviews the role of the inter professional team in the diagnosis and treatment of RHS.

Keywords: *Ramsay Hunt syndrome, herpes zoster oticus, varicella-zoster virus, erythematous vesicles.*

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INTRODUCTION

Ramsay Hunt Syndrome (RHS) also known as herpes zoster oticus or geniculate ganglion herpes zoster, is a late complication of varicella-zoster virus (VZV) infection, resulting in inflammation of the geniculate ganglion of cranial nerve VII.[1] The syndrome is named after James Ramsay Hunt (1872-1937), an American neurologist and Army officer in first world war who described three different syndromes, the most famous of which is the second, which is discussed herein as "Ramsay Hunt syndrome."[2] Early stages of VZV infection cause fever and diffuse vesicular rash, a condition that is commonly referred to as chickenpox. After the initial infection, the virus will often remain dormant in the body. Subsequent reactivation of the virus causes a "zoster" or "herpes zoster" phenomenon. This syndrome consists of pain and a vesicular rash along the involved nerves distribution, typically corresponding to a single dermatome. The distribution and associated symptoms depend on the nerve involved. Less than 1% of zoster cases involve the facial nerve and result in Ramsay Hunt syndrome.[3]

Although the classic triad of RHS is ipsilateral facial paralysis, otalgia and a vesicular rash, there is significant variability in clinical presentation, with some patients demonstrating facial paralysis before the rash or sometimes, no rash at all.[4,5] In the latter,

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the patient's chief complaints are severe ear pain and facial weakness; this variant is known as zoster sine herpete and can be very difficult to clinically distinguish from Bell's palsy. Zoster sine herpete has been reported to comprise up to 30% of Ramsay Hunt cases.[6] If a rash is present, it may be frankly vesicular or maculopapular and can involve the affected side of the face, scalp, palate, and tongue. Additional symptoms that may be reported include a change in taste sensation, dry eye, tearing, hyperacusis, nasal obstruction, and dysarthria. Hearing loss, tinnitus, and vertigo can be seen with involvement of the vestibulocochlear nerve, and hoarseness or aspiration may indicate involvement of the vagus nerve.

CASE SUMMARY

A 42-year-old diabetic normo tensive woman 9 days back presented at the Otolaryngology clinic with a sudden onset of dysphagia, odynophagia and hoarseness of voice,after 4-5days of these symptoms she complained of left-sided mild deafness, tinnitus, severe otalgia and erythematous vesicular eruptions over the left pinna and face. After that 2 day she developed complete left lower motor neuron facial nerve palsy, House Brackmann grade III, with dry eye.(Fig-1)

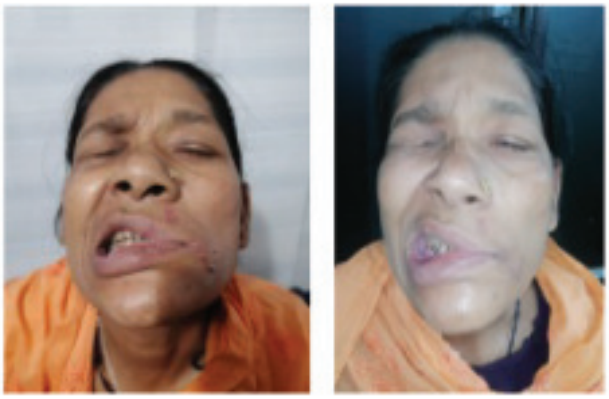

Fig-1: *VII N palsy at p resentation & days later*

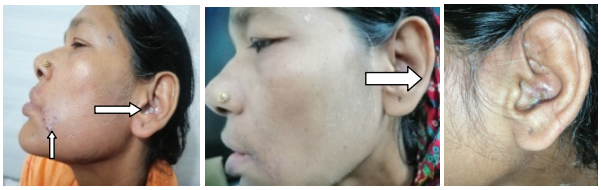

Fig-2: *Vesicle over face & pinna, at presentation-a & 15 day later-b*

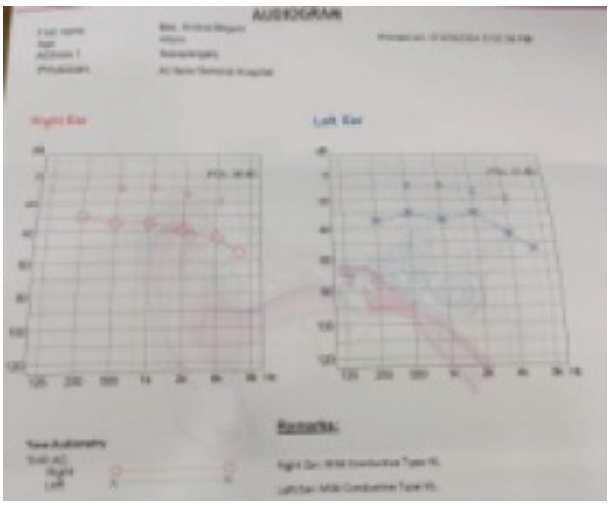

Fig-3:*Pure tone audiogram, mild sensory hearing loss, left ear*

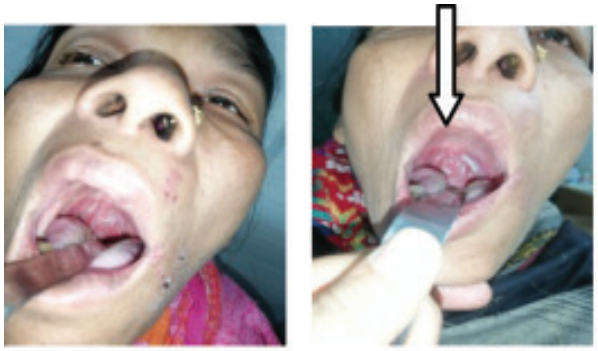

Fig-4 :*Palatal palsy(IX, X Nerve palsy), at presentation & 15 day later*

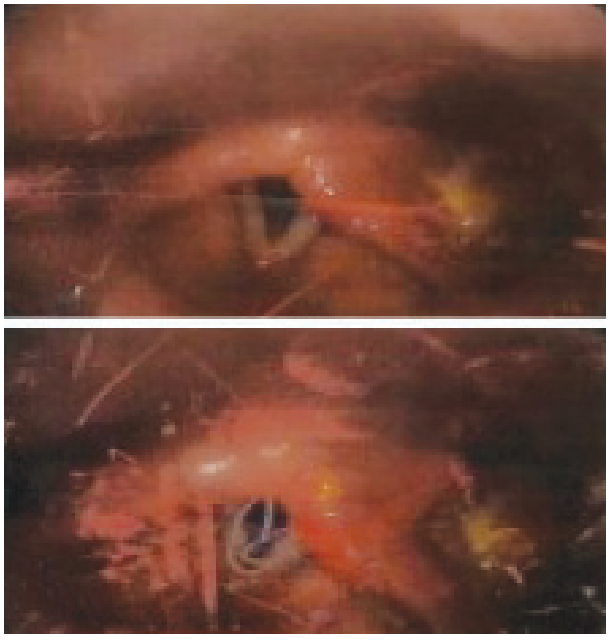

Fig-5:Videolaryngoscopy showed left vocal cord palsy & aytenoid oedema(X Nerve palsy)

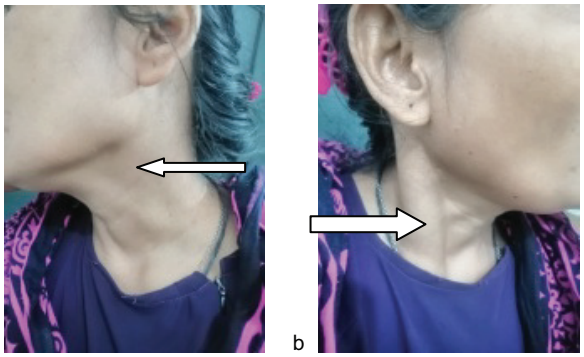

Fig-6:*Showing weakness-a and sterngth-b of sternoclaidomastoid muscle(CN XI)*

She was apparently healthy before presentation of these symptoms. The patient's past medical history was unremarkable. Neither the patient nor her family had similar problem. Physical examination revealed(after 9 days of presentation) erythematous vesicular eruptions over the left auricle, angle of mouth, temple (Figure-2).Tuning fork tests showed that the Rinne test was positive on both sides, the Weber test lateralized to the right ear and the absolute bone conduction test reduced on the left side. In pure tone audiometry there was also mild sensoryneural hearing loss in the left ear(Fig-3). Oropharyngeal examination revealed that the uvula –soft palate deviated to the right side on phonation and reduced gag reflex on the left side(Fig-4).Telelaryngoscopic examination revealed erythema and white exudate covering the mucosa over the left arytenoid, accompanied by vocal cord palsy(Figure-5).Blood tests revealed that the erythrocyte sedimentation rate (ESR) was 25 mm in 1st hour (reference range 0-15 mm/hour) and C-reactive protein was 12 mg/dL(Normal range). On serological test, the IgM and IgG antibody against VZV was not done due to unavailability.MRI with gadolinium enhancement of the brain revealed no abnormal lesions.No definite abnormal findings were found on neck and chest CT scan. The patient was diagnosed with RHS with polyneuropathy. She was hospitalized and a loading dose of 1 mg/kg/day of prednisolone orally were started. Subsequently, gradual tapering was done over three weeks. In addition, oral acyclovir was given at a dose of 25 mg/kg/day for 14 days and acyclovir cream over the skin lesion also applied. Dressing of the ear on a daily basis was performed and a pain killer was given with pregabalin 50mg bid 7days, carboxymethylcellulose sodium eye drop applied in left eye. Physiotherapy of the patient was initiated in the physiotherapy & rehabilitation unit. No side effects were detected during or following the completion of the treatment regimen. After two weeks of follow-up, the vesicles dried up changed to scabs and dropped off. Her hearing impairment partially recovered with partial improvement of facial movement but palatal palsy and vocal cord palsy persisted.She is now under observation, continuing treatment and follow up.

DISCUSSION

RHS affects both immunocompetent and immunocompromised patients and has an incidence of about 5 per 100,000 people per year; in contrast, the incidence of Bell palsy is much higher, at about 15-30 per 100,000 people per year.[7,8] RHS accounts for roughly 7% of acute facial paralysis cases, with zoster sine herpete comprising up to 30% of those. RHS mostly presents in individuals aged 60-80 years, any age from 19 to 89 years can be involved [2].However, the more severe disease form and less favorable outcomes occur in patients with hypo immunity. There are many predisposing factors that can increase the incidence of RHS, such as stress, infection, malnutrition, cytotoxic drugs, diabetes mellitus and malignant tumors. The classical presentation of RHS is a triad of unilateral severe otalgia which is preceded by the appearance of erythematous vesicular lesions over the auricle, external ear canal ,eardrum and lower motor neuron facial nerve paralysis. The vestibulocochlear nerve is the most common cranial nerve associated with the syndrome [2]. The oculomotor, trigeminal, abducent, glossopharyngeal, and vagus nerves are also associated with RHS also reported in the literature as RHS with polyneuropathy [4,6,7]. Moreover, non-classical presentations such as RHS without the involvement of the facial nerve have also been reported [8]. There are five possible mechanisms of simultaneous cranial polyneuropathy, namely, VZV induces occlusive vasculitis which results in ischemic neuropathy, the spread of the infection through the synapse, a neuroinvasive feature of the virus, inflammation of contiguous ganglia, and anastomoses between the affected nerves and other cranial nerves [9,10]. The reason for the slowly developed facial paralysis was probably the result of reactivation of the VZV that remained dormant in the nerve root ganglion. Trigeminal and vestibulocochlear

nerves and spinal ganglia C2-C4 are known to be the commonly affected nerves. Severe inflammation in one ganglion can spread to another ad jacent ganglion. It can also cause infarction by invading microvascular structures that have a common distribution in other brain nerves. VZV can directly invade the brainstem parenchyma by way of nerve axons or via synapses. Recently, a hypothesis that, VZV causes synaptic conduction along the reflex pathways of the brainstem, causing multiple neuropathies, has also been raised[11]

On serologic test, the positive rate of serum VZV IgM in patients with RHS was reported to be significantly higher (30.8%) than that in patients with Bell's palsy (9.8%).[12] Serologic screening for IgG against VZV will aid in identifying non immune individuals. The presence of IgM against VZV is suggestive of an acute or recent infection. However, results should be correlated with the clinical presentation because the patient's symptoms are the most important criteria in RHS diagnosis. RHS with multiple CN palsy has rarely been reported 11 patients with 10 years of RHS with polyneuritis, and the frequency of CN involvement was VII, VIII, IX, X, and V in that order reported, 11 cases for 15 years, and the following CN VII, VIII nerves, the frequency of other CN was CN X, IX, and V in that order[13,14] In a paper analyzing RHS with polyneuritis combined with vocal cord palsy (CNX) reported in the literature for 50 years; of the 14 patients, the incidence was CN VII,VIII,IX,V and XII.[15]There are few prospective controlled studies for RHS with multiple CN involvement in the medical literature.

The diagnosis of RHS is usually based on the classical presentation of the syndrome. Confirmation of the diagnosis is carried out with serological tests of IgG and IgM antibodies against VZV. Brain MRI is mandatory for the exclusion of tumors or demyelinated lesions or when there is a suspicion of infective brain tissue complications of the VZV. Although RHS is considered to resolve without treatment, early initiation of treatment is advised to reduce long-term complications such as postherpetic neuralgia and spastic facial nerve palsy. Several investigations have reported a significant reduction in late complications with the combination of steroid and antiviral therapy[12,14]. Our patient showed partial improvement of all presentation with these two treatment modalities till date. The prognosis of RHS is less favorable than Bell's palsy [14]. The full recovery rate was reported to be 63.63% in patients with an RHS with cranial polyneuropathy [7]. Furthermore, the prognostic factor that seems as reported in the literature is the severity of the presenting features [13]. Even though there was slow recovery of the involved cranial nerves in the presenting case, complete recovery was achieved at the four-month follow-up.

CONCLUSION

RHS affects patients in a myriad of ways and behavioral health concerns all occurring commonly in the acute period. While most patients do recover the majority of their pre morbid function when managed appropriately, long-term pain, facial dysfunction, scarring, and behavioral health concerns may all persist. For this reason, optimal patient outcomes occur when healthcare teams include members with expertise across a broad range of specialties. In the short term, primary care, otolaryngology, neurology, ophthalmology, and psychology/psychiatry are often required. In the long-term, facial plastic surgery or otolaryngology, pain management, ophthalmology, speech or physical therapy, and psychology/psychiatry may be needed. It is critical to surround these patients with an experienced inter professional team early on in the treatment process in order to provide the care and support they need to maximize their quality of life outcomes.

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