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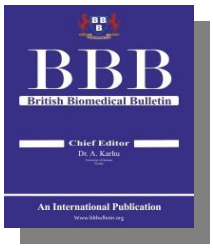

Case Report

Lower Motor Neuron Lesion Secondary to Ramsay Hunt Syndrome: A Case Report with Review of Literature

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A R T I C L E I N F O

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A B S T R A C T

Ramsay Hunt syndrome is a disease caused due to varicella zoster virus and is characterised by acute facial palsy and inner ear dysfunction with a herpetic eruption on the auricle and external ear canal. The clinical picture remains the cornerstone of diagnosis. It is hard to differentiate it from Bell's palsy and other conditions causing facial paralysis when vesicles are not present in patients. Otoscopic and audiometric examinations must be performed since Ramsay Hunt Syndrome can cause hearing loss. The standard treatment remains the combination of corticosteroid and antiviral therapy.

Significance: Early diagnosis of Ramsay Hunt syndrome is extremely important, as antiviral treatment within the first 72 hours of the onset of symptoms is generally considered to be crucial for better prognosis.

Introduction

Ramsay Hunt Syndrome (RHS) was first defined by James Ramsay Hunt in 1907 in a patient who had otalgia associated with cutaneous and mucosal rashes, which he ascribed to infection of the geniculate ganglion by varicella-zoster virus.[1,2]Hunt's syndrome, Zoster Oticus are the synonyms of this syndrome.[3,4] RHS develops in 0.2% of persons who had primary varicella zoster infections.[5]Characteristic features of RHS are painful herpetic vesicles on tympanic membrane and/or external auditory meatus with facial paralysis on the same side. Presence of these vesicles facilitates differential diagnosis from Bell's Palsy and other causes of facial paralysis. RHS

vesicular rashes might also be seen on ear auricles, external auditory canal, tympanic membrane, 2/3 front piece of tongue, face, neck, buccal mucosa, larynx. In addition to the facial nerve involvement, other cranial nerves are also reported to be involved in RHS.[4] Vestibulocochlear nerve is the most affected among them, causing tinnitus, nausea, vomiting, hearing loss, vertigo and nystagmus.[6] RHS is the second most common cause of atraumatic peripheral facial paralysis first being Bells palsy.[7] Compared with Bell's palsy patients with this syndrome often have more severe paralysis and are less likely to recover completely.[8] Here we present a case report of Ramsay hunt syndrome causing lower motor neuron lesion.

Case report

A 74 year old male patient reported to the Department of Oral medicine and radiology complaining of missing teeth in both upper and lower jaw since 1 year. History revealed that patient had developed fever accompanied by continuous dull ear ache present on left side of face 5 months back. Fluid filled eruptions were observed in his left ear region in next 48 hours. He consulted physician and was diagnosed with herpes zoster and antiviral was prescribed. On same day patient reported inability to hold the water on left side of mouth, followed by tinnitus in his left ear. He was taken to a hospital and was advised Acyclovir (Tab Ocuvir 800 mg x 5 times for 7 days),Eruptions got healed in 72 hours but facial paralysis, difficulty in closing the left eyelid and otalgia persisted. Patient also reported loss of hearing from left ear and mild taste alteration without any history of hyperacusis. Past medical history revealed a positive history of varicella zoster infection in childhood, cardiovascular accident1 year back and on medication for diabetics and hypertension for the past 5 years. On extraoral examination, slight facial asymmetry was observed without any vesicular eruptions on face or ears. Patient was unable to close his left eyelid with positive bell's phenomenon observed in left eye, he was unable to furrow on left side of his forehead, slight obliteration of left nasolabial fold along with triangular shaped smile was observed (Figure a,b,c). Neurological examination revealed weakness in marginal mandibular branch of facial nerve (Figure c). No sensory loss on face or any lymphadenopathy was noted. On Intraoral examination, completely edentulous both maxillary and mandibular ridges. Loss of taste sensation was noted on anterior 2/3rd of tongue. Based on history and clinical findings, diagnosis of Lower motor neuron lesion on left side of face secondarily to Ramsay hunt syndrome was given. Radiological and Hematological investigations were found to be normal.Pure tone audiometry showed sensorineural moderate hearing loss in patient's left ear and mild to moderate hearing loss in his right ear (Figure d). Patient was advised Neurobium forte o.d x 7 days, Artificial tears, galvanic physiotherapy with padding of eye, Patient was given referral for the fabrication of complete denture.

Discussion

Ramsay Hunt syndrome is an infectious disease and is a rare complication of the varicella zoster virus infection (VZV).[9] Activated many years after inoculation, VZV is followed by a latency period in the geniculate ganglion and spreads along the sensory tract of the facial nerve.[10] A positive history of varicella zoster infection had been reported in our case in his childhood. The incidence of the disease demonstrated an increase after fifth decade and peaked at eighth decade, parallel to the decreasing cellular immunity with the aging

process. It is 20% more common in females compared to males.[10,11]

The manifestations of Ramsey Hunt syndrome include ipsilateral peripheral 7thnerve palsy with or without herpetiform rash and vestibulocochlear symptoms.[12] In RHS; first symptom is mostly facial paralysis, at the same time vesicular rashes, hearing loss, sense of imbalance may also be seen.[4] In our patient fever developed first, followed by vesicular rashes and facial paralysis developed thereafter. RHS prognosis is reported to be better if vesicles appear before facial paralysis which may be due to earlier diagnosis and onset of treatment with observation of vesicles.[13] Symptoms and signs of vestibulocochlear dysfunction are not always present, if the nerve is involved it can cause tinnitus, vertigo, nystagmus, nausea and vomiting.[14] Our case also had tinnitus and then eventually total hearing loss developed with the progression of the disease that show that our case had facial as well as vestibulocochlear nerve involvement. The RHS diagnosis is mainly clinical, Bell's palsy can be commonly mistaken for the early stages of RHS. In Bell's palsy there is rapid onset facial paralysis which develops over hours to a day or two. Additional symptoms such as decreased tearing, hyperacusis, loss of taste sensation over the anterior two-thirds of the tongue and ear pain are variable. Bell palsy does not involve the presence of vesicles in the external meatus.[15-17]

Many conditions can produce isolated facial nerve palsy. Structural lesions in the ear or parotid gland tumors can produce facial nerve compression and paralysis. Tumors will present with a more insidious onset of symptoms over weeks or months. Other causes of peripheral nerve palsies include Lyme disease, otitis media, Guillain-Barre syndrome, sarcoidosis, and some influenza vaccines. Although these conditions can present as isolated facial nerve palsies, they usually have additional features. Patients with Lyme disease often have a history of tick exposure, rash, or arthralgias. Facial nerve palsies from acute and chronic otitis media have a more gradual onset, with accompanying ear pain and fever. Polyneuropathies (e.g., Guillain-Barre syndrome, sarcoidosis) will more often cause bilateral facial nerve involvement. Central nervous system lesions (e.g., multiple sclerosis, cerebro- vascular accident, tumor) can also cause facial nerve palsy. Multiple sclerosis is a chronic disorder characterized by periods of remission and exacerbation. This condition is characterized by several symptoms such as parasthesias of the trunk, the face or the extremities, weakness or clumsiness in a hand or leg, visual problems locomotor problems, muscular hypertonicity, lack of bladder control and dizziness. Multiple sclerosis can cause bilateral neuralgic type of pain.

Our patient has history of Cerebro vascular accident(CVA),but there was no facial paralysis during that time. Although Bell's palsy, RHS and CVA causes acute onset of facial paralysis CVA will cause more faster onset of facial paralysis and will usually bring paralysis or weakness to the lower face, while Bell's palsy, RHS usually brings paralysis or weakness to an entire side of the face. Also, Bell's palsy only affects only facial area, while a CVA can cause weakness to the extremities on the affected side of the body. Bell's palsy and RHS are not generally life-threatening, while CVA has often life threatening consequencies. It is hard to differentiate between RHS and Bell's palsy when vesicles are not present in patients. In such cases otoscopic and audiometric examinations must be performed as had been done in our case revealing loss of hearing in left ear. In some cases a blood test

for VZV antibodies levels are useful only when comparing the acute and convalescent stages of the condition.[1] Complement fixation tests and increased antibody levels may support the diagnosis. The detection of the virus by polymerase chain reaction in mononuclear cells of external auditory canal fluid, tear, CSF, and blood is accepted as the gold-standard in diagnosis of VZV.[18] It helps to distinguish between patients with Bell's palsy and patients with Ramsay Hunt syndrome in the very early stages.[19] When other cranial nerves are affected, Magnetic resonance imaging may be necessary to exclude intracerebral pathology. The House Brachmann scale should be used to assess the facial nerve function initially and at every subsequent follow-up. Facial nerve functions can be measured with electrodiagnostic methods. Edema and inflammation of the facial nerve are detected with Gadolinium-contrast-MRI, which is accepted as worst prognosis.[12,20,21]

In RHS the more severe the damage to the nerve, the longer it will take to recover, and the lower the chance to completely regain normal function. The chances of recovery are better if the treatment is started within 3 days after the symptoms begin. When treatment is started within this time, 70% of patients make a full recovery. However, when the treatment is delayed for more than 3 days, the chances of a complete recovery drop to about 50%. Children are more likely to have a complete recovery than adults. Untreated, the prognosis for Ramsay Hunt syndrome is much worse than for Bell's palsy with well over 50% suffering from permanent residual weakness and some having a permanently complete paralysis on one side of the face. Aggressive, early treatment lowers the bad-outcome risk.

The standard treatment of RSH syndrome is corticosteroid and antiviral therapy. The aim is to decrease the degeneration of the nerve. Corticosteroid therapy relieves pain, reduces vertigo, decreases the incidence of postherpetic neuralgia. It also reduces facial nerve inflammation and edema. Acyclovir, a synthetic acyclic purine nucleoside analog, is a selective inhibitor of herpes virus DNA polymerase.[22]Although there are no evidence-based dosing recommendations, published trials typically administered acyclovir at 800 mg by mouth 5 times/day for 7–10 days and prednisone at 1 mg/kg/day by mouth for 5 days followed by a taper.[23] The combination therapy with antivirals and steroids must be initiated as soon as possible in order to minimize the risk of permanent neuronal damage.[10] Patients with comorbid diseases (diabetes mellitus and hypertension) had been found to have poorer prognosis than those without comorbid diseases. This can be attributable to the presence of diabetic neuropathy.[24]Our patient also had diabetics and hypertension.

Intractable RHS cases resistant to medical therapy usually require surgical decompression of facial nerve.[4] RHS generally causes more severe dysfunction and has a poorer prognosis for facial nerve recovery than Bell's palsy.[25] RHS usually may not be responsive to therapy, prognosis will be better if the treatment is started within 72 hours of onset of disease.[14,23,26] In cases where treatment has been started within this time period, facial weakness recovers in up to 75% of patients

Conclusion

Ramsay Hunt syndrome is a rare complication of herpes zoster. This syndrome can be misdiagnosed if symptoms are not present at the onset of the disease. RHS is a diverse and challenging disease, since it can be associated with neurological symptoms mimicking other diseases. Prompt diagnosis and treatment (ideally within 72 hours of the onset of symptoms) are crucial

to secure the best outcomes. The ideal approach for treatment is still controversial and a multidisciplinary approach is essential for the follow up and recovery of these patients.

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Figure a: Bell's phenomenon observed in left eye

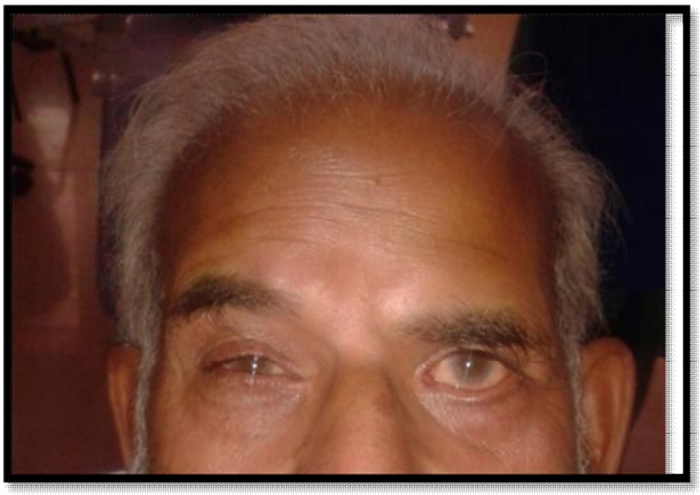

Figure b: Patient's inablility to furrow on left forehead

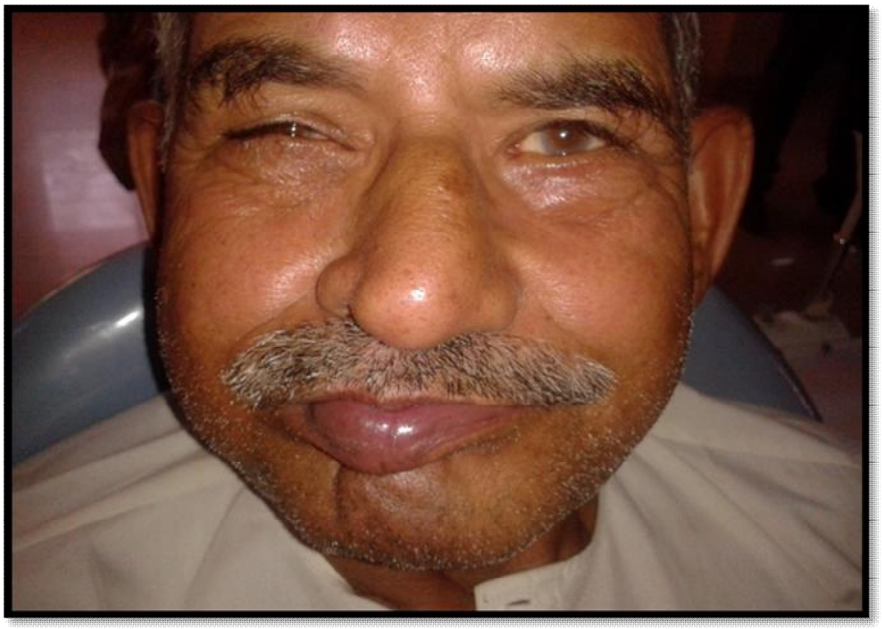

Figure c: Weakness of marginal mandibular branch of facial nerve

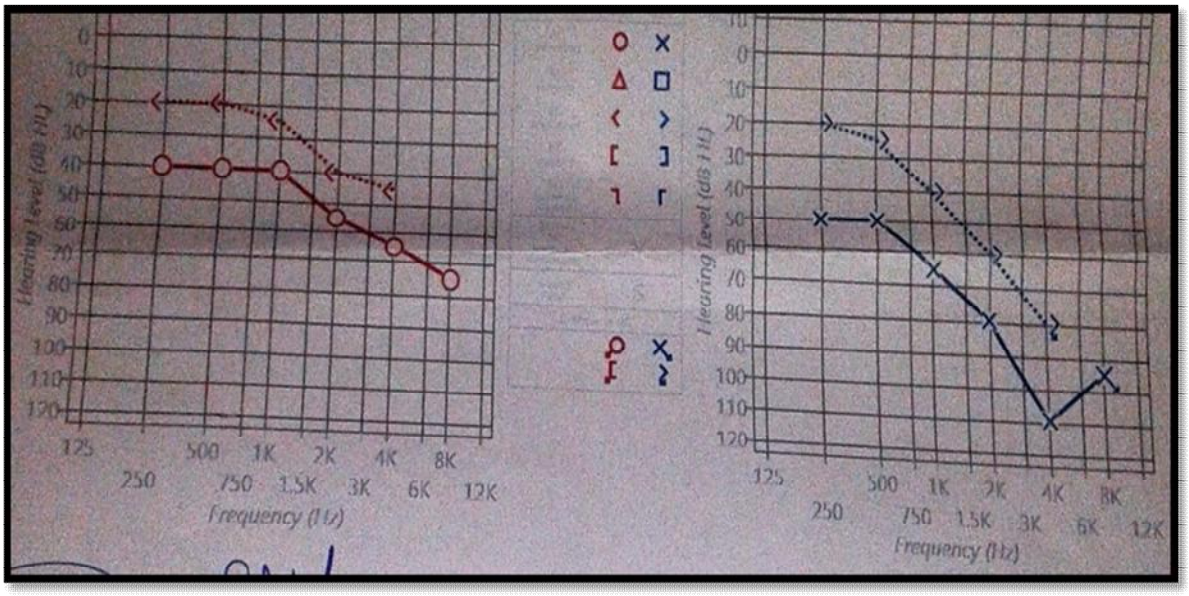

Figure d: Pure tone audiometry showing moderate hearing loss on the patient's left ear and mild to moderate hearing loss in his right ear