

### Ramsay Hunt syndrome in focal segmental glomerulosclerosis

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Facial paralysis has been reported in the literature subsequent to multiple insults but its occurrence after herpes zoster infection is a drastic, disfiguring and painful event with poorer outcomes. Ramsay Hunt syndrome (RHS) is characterized by the presence of cochleovestibular symptoms in association with facial paralysis. Facial nerve palsy has long been considered to have mainly infectious etiology. Recent diagnostic analysis have provided convincing evidence that the virus most likely enters the geniculate ganglion during chicken pox via sensory branches of facial nerve and remains in dormancy before it is reactivated.

This case presents the lower motor neuron facial paralysis along with vesicular rashes in the external auditory canal and pinna, giddiness, otalgia and hearing impairment in a young patient diagnosed histopathologically as having focal segmental glomerulosclerosis subsequent to nephrotic syndrome, 3 months before.

Our patient is a 14-year-old Saudi girl who reported in the emergency room with the complaints of sore throat, cough, fever and some dyspnea for 4 days. A probe into past history revealed that the girl had nephrotic syndrome along with hypertension diagnosed based on clinicals and investigations 3 months before and was put on steroids and anti hypertensives. One month before renal biopsy at the King Faisal Specialist Hospital, Riyadh, Kingdom of Saudi Arabia was carried out and a diagnosis of focal segmental glomerulosclerosis was established and the patient was advised to use nifedipine, furosemide and to continue oral steroids. Patient's parents could not exclude the occurrence of chickenpox in her childhood. On general examination at the time of recent presentation, her temperature was 38.7°C, blood pressure 140/100 mm Hg, pulse rate of 120 per minute regularly and respiratory rate was 28 per minute. Her throat examination revealed hyperemic posterior pharyngeal wall and tonsils with jugulodigastric lymphadenopathy. The patient was admitted for the management of acute upper respiratory tract infection and was administered parenteral cefuroxime. Approximately 18 hours after admission, the patient developed skin eruptions on lateral surface of right auricle (**Figure 1**) along with severe otalgia. Eruptions were vesicular in nature, hyperemic and tender. There was a small rash on the right cheek. The pain was experienced in the right ear and para auricular areas.



**Figure 1** - Ramsay Hunt syndrome eruptions.

Approximately 24 hours after admission the patient complained of facial asymmetry of the angle of mouth. Patient had developed right sided lower motor neuron type facial paralysis.

The next day, the patient felt more pain and slight numbness on the right side of the face. The vesicles became prominent and extended to the external auditory canal although drumhead was spared but hyperemic. She also had off and on dizziness spells and some difficult in hearing on right side. She was examined vigilantly at regular intervals and a battery of investigation was carried out. Blood routine examination showed that hemoglobin was 12.5g/L, white blood cell was 12.6 and the differentials within the normal limits. Urine routine examination showed turbid appearance, red blood cells 40-45, albumin 2++ and increased concentration of urobilinogen. Urine culture grew *E coli*, resistant to cephradine, co-trimoxazole, genticycyn and ampicillin but sensitive to nitrofurantoin and nalidixic acid. Urea and creatinine were within normal. Uric acid was raised up to 325, while sodium and potassium were at lower normal limits 132 and 3.5. Liver and lipid profiles and other biochemical analysis had unremarkable changes. Skin scrapping from the vesicles was also subjected to culture but no bacterial growth was reported. Ultrasonic renal imaging revealed multiple echopenic spots suggestive of infarcts. Schermir test showed reduced lacrimation on the effected side. Pure tone audiometry results were in favor of mild sensorineural hearing loss. The facilities such as electrical stimulation tests, magnetic resonance imaging, viral studies, polymerase chain reaction (PCR) and other sophisticated audiological and serological tests and so forth, which have been used as diagnostic tools in such cases are not available at our center. Based on the clinical information and investigations, the diagnosis of RHS was established in this patient who has focal segmental glomerulosclerosis and has immunosuppressed status due to steroids intake. She

started with Acyclovir tablet 400 mg 5 times daily along with adequate hydration, Acyclovir topical application, analgesics, and she also continued taking steroids. Supportive measures such as physiotherapy and biofeedback exercises were also offered. Progress of patient was closely observed. On the second day of admission, she experienced numbness, pain hearing loss, dizziness and vesicles. On the third day vesicles did not increase in number but there was edema of external auditory canal. On the fourth day otalgia was moderate, there was no dizziness and vesicles started settling down and on the fifth day of admission, pure tone audiometry was repeated and hearing loss was minimal. Facial nerve also recovered up to Grade III (House and Brackmann Grading system). During her admission days, renal status was regularly assessed and it remained within normal limits. There were no major adverse reactions to Acyclovir. On the seventh day of admission patient had minimal complaints and rashes faded significantly. On the eighth day, she was discharged and advised to discontinue the Acyclovir but to continue the steroids, antihypertensives, physiotherapy, analgesics and adhere to follow up plans.

Herpes zoster oticus (HZO) is caused by a specific neurotropic virus - varicella zoster as first described by Letulle (1882) and Korner (1884) but particularly studied by Ramsay Hunt (1907) who reported it as a herpetic disease of geniculate ganglion and hence herpes zoster oticus associated with facial paralysis got the name of RHS.<sup>1</sup> The eighth nerve features such as hearing loss, tinnitus, nausea, vomiting, vertigo and nystagmus are due to proximity of the geniculate ganglion to the vestibulocochlear nerve within the bony facial canal.<sup>1</sup> In a prospective study of RHS patients, 14% developed vesicles after facial weakness; therefore, initially RHS may be indistinguishable from Bells paralysis.<sup>1</sup> In another study 23% of RHS cases were misdiagnosed as Bells palsy. Therefore, it is important to adopt an accurate diagnostic technique to distinguish between these 2 conditions. The presence of Varicella Zoster virus DNA in the tear fluid, sub mandibular gland saliva and parotid gland saliva have been confirmed quantitatively by PCR and saliva collected hybridization method. In a study of 102 specimens of tears and saliva collected from 15 patients, virus was detected in 40 specimens (39 %) from 12 patients (80%).<sup>2</sup>

Ramsay Hunt syndrome, on rare occasions can involve many nerves and in literature we found a case of a woman with involvement of V.VII, VIII, IX and XII cranial nerves; therefore, RHS should be recognized as a polycranial neuritis characterized by damage to sensory and motor nerves. Another case presentation was about a patient who presented with oral lesions on the anterior two thirds of the tongue and palate. A recent study investigated the audiological features of a group of 15 patients with this syndrome with a comprehensive audiological battery. This data

suggested cochlear or retrocochlear involvement or involvement at more than one site along the auditory pathway.<sup>3</sup>

Contrast enhanced magnetic resonance imaging (MRI) on 147 patients<sup>4</sup> with facial paralysis concluded with the impression that enhancement of the distal intrameatal and a labyrinthine segment is specific of such lesions. In a similar study authors present a clinical case to show the important role played by gadolinium MRI in the differential diagnosis of facial paralysis. Corneal epithelial keratitis in HZO was evaluated in a non contact photo micro graphic in-vivo study in the human cornea and findings strongly suggest that such lesions occurring in the absence of cutaneous rash are in fact recurrent episodes of virus shedding. The neurologic complications associated with HZO are infrequent except for post herpetic neuralgia. A study on 100 patients of RHS concluded that such complications other than post herpetic neuralgia occurred in 12 % of patients of this series with male predominance and peripheral neuropathies were the most frequent complications.<sup>5</sup> Several studies have implied that oral acyclovir improves the outcome of facial paralysis for patients with RHS. Varicella Zoster is less sensitive to acyclovir than herpes simplex hence, higher doses are recommended to treat RHS. There is a real evidence to suggest additional benefit of oral steroids in facial zoster and their use can be supported in immuno-competent individuals. However, steroids use is debatable and its use has met variable degree of success. Facial nerve decompression surgery for HZO has experienced varying levels of enthusiasm over the years. Recent work implies that early extensive decompression of the nerve through a middle fossa craniotomy may benefit patients at risk for persistent deficits. However, until this procedure is subjected to a rigorous, controlled trial comparing it with maximal medical therapy, it is difficult to justify the very high cost and risks.

Our patient developed RHS when she had already existing underlying disease-focal segmental glomerulosclerosis and had an immuno-suppressed status due to steroids intake. This patient had a typical onset and progression of disease. Evidence as regards RHS in glomerulosclerosis is lacking in literature. Patients diagnosis were based on clinical examination mainly because more sophisticated diagnostic work ups were beyond our approach as the patient was managed here, where such facilities are not available. A board of doctors including Internist, Nephrologist, Physiotherapist and Dermatologist participated in her management after joint discussions. Patient was put on oral Acyclovir 400 mg x 5 times daily for a period of 7 days. Her steroid intake (prednisolone 40 gm daily) and antihypertensive treatment continued, in addition to adequate hydration and supportive physiotherapy were practiced. Patient responded very well to this treatment protocol and showed significant improvement. She was discharged on the eighth day

## Clinical Note

with grade III recovered facial paralysis, but other complaints were minimal. She was followed up at regular intervals and there are no significant post herpetic complications. The patient was also experiencing psychological stress before this illness and we found in the literature that it has a role. In one of such studies, 55 patients with facial paralysis were asked if they had a physical or psychological stress one week before the attack and it was reported that 76.9% had experience of physically fatigued while approximately 52% were reported existence of psychological stress.<sup>6</sup> Ramsay Hunt syndrome is a rare disorder. Early diagnosis by adopting an accurate diagnostic work up and early institution of treatment with antiviral agents and steroids help hasten healing and is more likely to prevent irreversible complications affecting the facial and other cranial nerves involved.

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## References

1. Sweeney CJ, Gilden DH. Ramsay Hunt Syndrome. *J Clin Microbiol* 2001; 39: 2856-2859.
2. Hiroshige K, Ikedia M, Hondo R. Detection of varicella zoster virus DNA in tear fluid and saliva of patients with Ramsay Hunt syndrome. *Otol Neurotol* 2002; 23: 602-607.
3. Kaberos A, Balatsouras DG, Korres SG, Kandiloros D, Economou C. Audiological assessment in Ramsay Hunt Syndrome. *Ann Otol Rhinol Laryngol* 2002; 111: 68-76
4. Kinoshita T, Ishii K, Okitsu T, Okudera T, Ogawa T. Facial nerve palsy: evaluation by contrast - enhanced MR imaging. *Clin Radiol* 2001; 56: 926-932.
5. Sanchez-Guerra M, Infante J, Pascual J, Berciano J, Polo J. Complications of herpes Zoster. A retrospective study in 100 patients. *Laryngoscope* 2001; 111: 719-723.
6. Kuga M, Ikeda M, Kukimotor N, Abiko Y. An assessment of physical and psychological stress of patients with facial Paralysis. *Nippon Jibunkok Gukkai Kaino* 1998: 101; 1321-1327.