



Ramsay Hunt Syndrome

H B NONGRUM, P S NAGPURE

INTRODUCTION

In 1906 James Ramsay Hunt gave a classic description of the syndrome consisting of blisters, facial paralysis, and inner ear disturbances due to herpes zoster. Since his description, the name Ramsay Hunt and the designation Herpes Zoster Oticus have been synonymous. He felt the problem to be geniculate ganglionitis due to the herpes virus. Subsequent investigators with the benefit of histopathologic studies of autopsy cases of patients with herpes zoster oticus demonstrated little, if any, ganglion involvement. They did find heavy lymphocytic infiltration in the substance of the facial nerve. These findings were present in facial nerve in several patients with paralysis who recovered^[1].

CASE 1

A 20yrs male had presented with pain and swelling in the post and pre auricular area since 9 days. 4 days later he developed eruptions over the right ear (pinna) which rapidly progressed to involve the whole pinna and the pre and post auricular area. The following day he had difficulty in closing the right eye (but able to do so forcefully) and deviation of angle of mouth to the left. Over the next 2 days the weakness progressed and he was not able to close the right eye completely. Examination revealed multiple vesicular eruptions in the right Pinna and the pre and post auricular area with congestion in the posterior wall of the external auditory canal and tympanic membrane. Cranial nerve examination showed a grade II (House – Brackmann classification) paresis of right facial nerve which progressed to grade IV over the next 2 days of admission. On conducting the localization test clinically the lesion of the facial nerve was infrageniculate. Nose and throat examination were within normal limits. Routine blood and radiological investigations were within normal limits. Audiometry showed normal hearing threshold with absence of stapedial reflex on impedance audiometry. He was diagnosed as Ramsay hunt syndrome grade II. He was started on prednisolone (2mg/kg body weight) and the dose was tapered over the next 15 days. He was also started on Acyclovir orally (800mg 5 times a day) and local application for 10 days. Following treatment the pain and eruptions over the ear subsided but facial nerve paralysis remained static. Patient was discharged on the 10th day of treatment and came for follow up after 2 weeks and after a month; the facial nerve was still in grade I V paresis.

CASE 2

A 17-year-old female presented with complaints of painful vesicular eruptions over the left pinna of 7 days duration and



FIG Ramsay Hunt syndrome with vesicular eruption (*arrow*) right pinna

left facial weakness, inability to close the left eye completely and deviation of angle of mouth to the right of 6 days duration. On examination we found vesicular eruptions which were present over the left outer surface of the pinna (concha and helix portion of the pinna). Cranial nerve examination showed left infranuclear facial nerve paresis, grade IV. On performing the localization test, the lesion of the VIIIth nerve was found to be infrageniculate. Her hearing threshold, routine blood and radiological investigations, were within normal limits. She was diagnosed as Ramsay Hunt syndrome Grade II. She was started on the same regime as the previous case. She was discharged on the 10th day. On discharge status of facial nerve was Grade III paresis. She was again seen in the out patient department 2 weeks and a month later. She had a full recovery of the nerve function with no evidence of any residual paresis.



DISCUSSION

Herpes zoster oticus or Ramsay Hunt syndrome includes facial paralysis associated with hearing loss, dizziness, and herpetic eruption around the auricle (commonest site being the concha of the auricle). Herpes zoster oticus is the cause of 2 to 10% of all cases of facial paralysis, including 3 to 12% of adults and approximately 5% of children.

The facial nerve is the commonest to be involved followed by the ocular nerve. This is due to the fact that these nerves pass through bony canal in the skull. This course in the bony canal increases the chances of entrapment.

Pathologically, the theory of inflammatory changes as a cause of Bell's palsy or facial paralysis has been proposed by many authors. Sade described cases of facial paralysis secondary to external otitis with the inflammation traveling along the chorda tympani or sensory anastomoses to the facial nerve. Denny - Brown showed that pressure on a nerve causes ischemic paralysis. Even a small amount of inflammation will suffice to cause pressure; thus, a relatively small amount of edema or inflammatory exudates could cause strangulation of the nerve. Fisch proved these changes with photography of the inflammatory changes in the labyrinthine portion of the facial nerve^[1].

Hunt classified the disease into 4 grades as follow^[7]:

- (1) Disease affecting the sensory portion of the CN VII.
- (2) Disease affecting the sensory and motor divisions of the CN VII
- (3) Disease affecting the sensory and motor divisions of the CN VII with auditory symptoms
- (4) Disease affecting the sensory and motor divisions of the CN VII with both auditory and vestibular symptoms.

Many authors have shown that there is no real difference between herpes zoster and Bell's palsy. Complement fixation test carried out by Tomita et al.^[3] in 1973 found that 25% of patients with Bell's palsy had positive complement fixation tests.

Paralysis of herpes zoster may more likely be a complete one but predictability from electrical tests, and the time of recovery seem similar to those of Bell's palsy. However the natural history between the two differs in several ways^[2].

1. Bell's palsy recurs in 12% of cases, but Herpes Zoster rarely recurs.
2. With Bell's palsy the decrease in response to electrical testing peaks in 5-10 days but in Herpes zoster the peak is later (10-14 days).

3. 84% of those suffering from Bell's palsy have satisfactory recovery, but 60% of those with Herpes zoster oticus recover to a satisfactory degree.

The medical management for facial paralysis of herpes zoster is aimed at eliminating inflammation and ischemia of the nerve, thereby restoring facial function as quickly as possible. Steroid dose as recommended for adults is a daily total of 1mg/kg body weight in divided dose. If the palsy is incomplete by the fifth day the dosage can be tapered to zero during the next 5 days. If there is a question about the severity or the progression of severity, full dosage is recommended for 10 days and then tapered over the next 5 days^[4, 5].

Acyclovir, a virostatic drug developed for the use in the treatment of herpes simplex has been found to prevent replication of varicella-zoster virus (VZV). Acyclovir in the host cell is converted to acyclovir triphosphate and gets incorporated in the newly formed viral DNA, resulting in termination of the DNA molecular chain. Because VZV is generally less sensitive to acyclovir than is HSV, higher dose must be used to treat VZV infection. Oral Acyclovir is available; however, absorption from the gastrointestinal tract is only 15% - 25% of the ingested dose. A dose of 800mg five times a day has a modest beneficial effect to localize the lesion. For these reasons intravenous dose of 10 mg/kg every 8 hours over a 7 days hospitalization has been recommended. This intravenous route has more inherent expenses than an oral route of administration. Prognosis depends primarily on immediate initiation of therapy^[6]. Alternate antiviral agents such as valacyclovir (1g orally three times a day for 10 to 14 days) or famciclovir (500mg orally three times a day for 10 days), which achieve adequate levels by an oral route, are now available as an alternative to intravenous therapy.

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AUTHORS

CORRESPONDENCE

1. H B Nongrum,
H B Nongrum, MS
Consultant Surgeon, Department of ENT
Nazareth Hospital, Shillong

2. P S Nagpure
Professor and Head,
Department of ENT-HNS, MGIMS, Sewagram

H B Nongrum, MS
Consultant Surgeon
Department of ENT
Nazareth Hospital
Shillong
+91-94363 09304
henry.nongrum@gmail.com



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1. WHO Collaborating Centre for Drug Information, WHO, Geneva, 1998.
2. WHO Collaborating Centre for Drug Information, WHO, Geneva, 1998.



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