



Ramsay Hunt syndrome a case report and review of literature

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Abstract:

This is a case report of a rather rare disorder i.e. Ramsay Hunt syndrome. This is caused by Varicella zoster infections involving geniculate ganglion of facial nerve. This syndrome is manifested by the presence of blebs in the external auditory canal, ear ache, and lower motor neurone type of facial paralysis. This patient had excellent recovery following administration of oral steroids and acyclovir.

Introduction:

This syndrome characterised by herpetic blisters in the external auditory canal, lower motor neurone type of facial paralysis and otalgia. This syndrome was first described by Ramsay Hunt¹ in 1910. This disease is caused by varicella zoster virus type 3 which has a predilection to involve sensory nerves. Ramsay Hunt in his detailed study demonstrated that Herpes virus involves cranial nerves and used the term cephalic herpes zoster to describe this condition. His studies demonstrated that peripheral facial nerve paralysis were common in patients with cephalic herpes. He attributed this to involvement of geniculate ganglion by Herpes virus.

He surmised that in patients with herpetic eruptions over concha and external auditory meatus the geniculate ganglion could be affected by inflammatory oedema causing pressure against the facial nerve trunk causing paralysis of 7th nerve. In patients with herpetic infections of trigeminal nerve then facial paralysis could be caused due to degenerative changes involving the geniculate ganglion.

Differences between Bell's Palsy and Ramsay Hunt syndrome:

Ramsay Hunt syndrome needs to be differentiated from Bell's palsy. Bell's palsy is actually idiopathic in nature and is a diagnosis of exclusion. Prognostically Bell's palsy² carries better prognosis than that of Ramsay Hunt syndrome. More than 90% of patients with Bell's palsy regain facial nerve function in comparison to 70% of those with Ramsay Hunt syndrome.

According to published literature etiology of Bell's palsy could be viral. Pubmed search did not throw up any literature where virus has been identified in patients with Bell's palsy even though reactivation of Herpes virus has been implicated as a probable etiological factor³. A dip in the cell mediated immunity could cause reactivation of dormant herpes virus in these patients.

Case Report:

50 Years old male patient came with complaints of:

1. Pain right ear – 6 days
2. Deviation of angle of mouth to left – 3 days
3. Inability to close right eye – 3 days

On examination:

Bleb seen in the right external auditory canal.

Tympanic membrane was normal on both sides.

Patient did not have any tenderness over mastoid process.

Lower motor neurone paralysis of facial nerve on the right side.



Photograph showing bleb in the external auditory canal



Photograph of patient showing right facial nerve palsy (LMN type)

House Brackmann ⁴ scoring system was used to grade the severity of facial nerve paralysis. Facial nerve paralysis was found to be of Grade V (Severe).

House Brackmann scoring system helps in grading severity of facial nerve paralysis. This scaling system is dependent on measuring upward movement of the middle of upper eyebrow and outwards movement of angle of the mouth. 1 point is awarded for each 0.25 cm movement up to a maximum of 1 cm, Scoring is made to a scale of 8.

| Grade | Description | Measurement | Function% | Estimated Function% |
|-------|-------------------|-------------|-----------|---------------------|
| I | Normal | 8/8 | 100 | 100 |
| II | Slight | 7/8 | 76-99 | 80 |
| III | Moderate | 5/8-6/8 | 51-75 | 60 |
| IV | Moderately Severe | 3/8-4/8 | 26-50 | 40 |
| V | Severe | 1/8-2/8 | 1-25 | 20 |
| VI | Total | 0/8 | 0 | 0 |

Nerve conduction study:

Showed poor muscle contraction amplitude on electrophysiological stimulation.

Results of Topognostic studies:

1. Patient had metallic taste in the anterior 2/3 of tongue
2. Impedance audiometry showed intact stapedial reflex
3. Normal nasolacrimal reflex

Topognostic tests are reliable in assessing the prognosis of the disease.

Persistence of taste

Intact stapedial reflex

Normal nasolacrimal reflex

Are all indicators of good prognosis ⁵.

Management:

Patient was started on steroids. T. Prednisolone ⁶ was administered in doses of 10 mg four times a day. The dose is tapered during the course of 6 weeks.

Oral acyclovir in doses of 4000 mg/day ⁷ was started from the 8th day of facial paralysis. Patient showed rapid improvement.

Facial paralysis which was Grade V of House Brackmann scale improved to Grade I within 1 week

of starting acyclovir. Oral Acyclovir was continued for a period of 9 days.

Conclusion:

This case report is presented to demonstrate the usefulness of a combination of steroid and acyclovir therapy in the management of Ramsay Hunts syndrome. In addition this patient had all the positive prognostic factors (taste, intact stapedial reflex and nasolacrimal reflex). It is advisable to start these patients with acyclovir as early as possible to prevent further damage to the nerve due to Herpes virus.

References:

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