PERIPHERAL SEVENTH CRANIAL NERVE PALSY, IS IT STROKE?

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INTRODUCTION

Patients' presentation of lower motor seventh nerve palsy to the Family Physician is not an uncommon encounter. The majority of peripheral seventh cranial nerve palsy cases remain without an identified etiology and will eventually be diagnosed as idiopathic or Bell's palsy. Some features of this condition may be characteristic of a viral infection. Indeed, several herpes viruses have been implicated as potential causative pathogens. Besides varicella-zoster virus, shown to cause Bell's palsy under the Ramsay-Hunt syndrome, recent years have seen an increased interest and focus on the possible herpes simplex virus type 1 (HSV-1) etiology in idiopathic facial paralysis. Many other pathogens are also implicated as a causative agent for Bell's Palsy. The Department of Otorhinolaryngology or Neurology of the University of Wuerzburg tested serologically patients with a Bell's Palsy who were admitted between 2000 and 2002 for the presence of antibodies against Borrelia burgdorferi, herpes viruses (HSV-1/2, VZV) and M. pneumoniae. Fifteen patients showed a reactivation of a VZV (n=12) or of a HSV-1 (n=3) infection. In six cases the immunoblot revealed specific antibody bands for B. burgdorferi. In 24 patients (26.4%) a seroconversion of M. pneumoniae could be detected[1].

These however are not important in the patients' mind. The question in these patients that need the family physician to answer is "Doctor, am I having a stroke?"

Ramsay Hunt Syndrome was first described in 1907 by J. Ramsay Hunt (1872-1937), an American neurologist, in patients who had otalgia associated with cutaneous and mucosal rashes, which he ascribed to infection of the geniculate ganglion by human herpesvirus 3 (ie., varicella-zoster virus). Synonyms for this syndrome include geniculate neuralgia, herpes zoster oticus and nervus intermedius neuralgia.

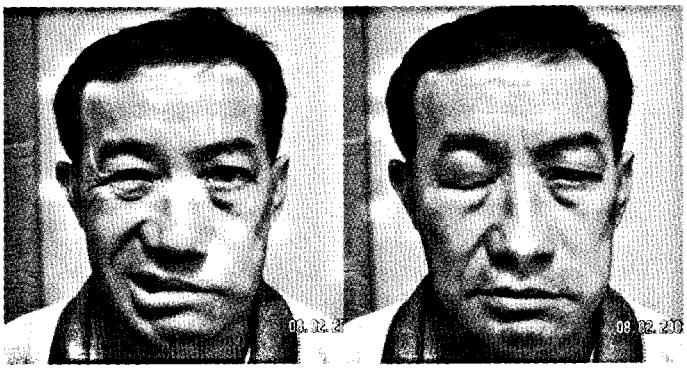
A CASE STUDY

History

Mr A, a 50-year-old taxi driver walks into your clinic one evening and complains that there is something wrong with his face. Of particular concern to him is a 1-day history of weakness of the left side of his face. He does not have any limb weakness or any diplopia. Speech is still fairly clear and swallowing is normal. He has no chronic medical problems such as hypertension and diabetes mellitus. In fact he has never been seen at any clinic before, except an occasional visit to the Chinese Physician for body aches and pain.

Physical findings

The physical findings that catch your attention are shown in Photograph 1 and Photograph 2.



Photograph 1 Photograph 2

Looking at Mr A at Photographs 1 and 2 there is loss of left forehead wrinkling compared to the right. He has weakness of the periorbital muscle and perioral muscle groups. In Photograph 2, we can see drooping of the left nasolabial fold and also Bell's sign.

Anatomical diagnosis

Mr A has a left-sided lower motor neuron Facial Nerve (CN VII) palsy.

Etiological causes

Possible causes of a lower motor facial nerve palsy to be considered are:

1. Bell's Palsy (idiopathic) – commonest cause.
2. Middle ear pathologies eg. surgery or otitis media.
3. Associated tumours eg. parotid gland, skull base.
4. Facial trauma, including petrous temporal bone fractures.
5. Osteomyelitis of skull base.
6. Guillain-Barre Syndrome.
7. Ramsay Hunt Syndrome.
8. Others: Sarcoidosis; Poliomyelitis.

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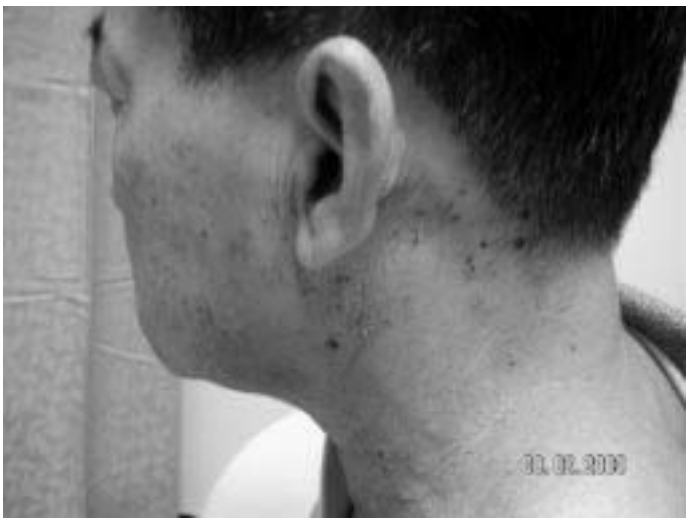
Further findings

Further examination revealed another significant finding, as shown in Photograph 3.



Photograph 3: Crops of papulovesicles with varying degrees of crusting

In Photograph 3 there are crops of erythematous papulovesicles with varying crusting on his neck and face on the left side.

Upon further questioning, Mr A says that the painful rash over the left side of his neck and face appeared 3 days ago and has become more extensive.

What is the etiological diagnosis?

Mr A has Ramsay Hunt Syndrome. This is an acute facial paralysis associated with an ipsilateral herpes zoster infection manifesting classically as blisters within the ear canal or auricle. It is caused by reactivation of a latent Varicella Zoster infection which has affected the geniculate nucleus of CNVII (Facial nerve).

What are the other important areas to examine and for what?

The other important areas to examine are:

1. The ear on the affected side for similar rash with the external canal, and for any superimposed bacterial infection.
2. The eye on the affected side for exposure keratoconjunctivitis.
3. A general neurological examination to ensure sole involvement of the unilateral CNVII distributation.
4. Ipsilateral hearing loss or balance problem.

It should be remembered that the nervus intermedius supply the following areas and hence, the characteristic blisters may seen any of these areas:

1. Anterior two thirds of the tongue.
2. Soft palate.
3. External auditory canal.
4. Pinna.

What does the patient usually complain of in this syndrome?

The common main worry for the patient is of a stroke. Indeed, this was Mr A's main concern in this consult. They will complain of weakness of the ipsilateral facial muscles, and varying degree of paraesthesia and pain of the affected dermatome, associated with the characteristic blistering rash.

He may also complain of ipsilateral eye dryness or pain, vertigo or balance problem, hearing loss or tinnitus. The possible differentials in a patient with ipsilateral facial pain includes the following:

1. Atypical facial pain (before the eruption of a typical rash.
2. Bells palsy (still the most common world wide cause of a lower motor neuron facial paralysis.
3. Post-herpetic neuralgia.
4. Teperomandibular Joint Syndrome.
5. Trgeminal Neuralgia.

Diagnosis

The diagnosis of Ramsey Hunt Syndrome is clinical. Further investigations that may be considered are:

1. Viral studies
   - Serologic tests also can reveal VZV, although prior history of chickenpox may lead to a positive result
   - The diagnosis of VZV usually is made without difficulty when the characteristic rash is present as well as vesicular eruption. If necessary, VZV may be isolated from vesicle fluid and inoculated into susceptible human or monkey cells for identification by serologic means
   - Antibody determinations on paired sera may be helpful in establishing the diagnosis by comparing titers at time of presentation and a few weeks later
   - VZV can be detected by PCR on samples of tear fluid from affected individuals.
2. Imaging Studies
   - Structural lesions can be ruled out by CT scan, MRI, or magnetic resonance (MR) angiography
   - Gadolinium enhancement of the vestibular and facial nerves on MRI has been described in Ramsay Hunt syndrome.
3. Other Tests
   - Audiometry usually demonstrates sensorineural hearing loss.

TREATMENT

Treatment usually comprises a combination of corticosteroids and oral acyclovir. It is thought that the corticosteroids (high dose for a week and taper dose subsequently) reduced inflammation within the geniculate ganglion while acyclovir (7-10 days duration) is an established antiviral effective against varicella zoster infections. This should ideally be started within 3 days of

onset of the syndrome (see below). Vestibular suppressants may be helpful if vestibular symptoms are severe. Analgesics are prescribed as required for pain-relief. Carbamazepine and neurotin may be helpful as an anti-neuralgic, especially in cases of idiopathic geniculate neuralgia.

Complications such as superimposed bacterial infection may occur. The unprotected cornea should be well lubricated with regular eye drops and protected during sleep either by medical tape or a gauze covering or eye patch.

After initiation of medical therapy, the patient should be seen in follow-up at 2 weeks, 6 weeks, and 3 months. The pain experienced is usually transient and the prognosis for this is good although patients are still at risk of post-herpetic neuralgia. Regarding the facial paralysis, it is said that starting treatment within 3 days of onset of symptoms is associated with a better recovery rate of up to 70%. If delayed beyond 3 days, recovery may only occur in up to 50% of patients. Children tend to have a better prognosis. Hearing loss and tinnitus may be prolonged.

HOW WOULD YOU ADVISE A PATIENT WITH RAMSEY HUNT SYNDROME OR IDIOPATHIC BELLS PALSY?

A few points are useful:

1. Enquire about the patient's main concern. Often the patient will be worried whether he or she is having a stroke. We should seek to allay their anxiety regarding this, and explain the cause and prognosis of this disorder to them clearly.
2. Teach the patient cornea care as mentioned above.

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