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Acute unilateral facial nerve palsy

Keywords: cranial nerve diseases; diagnosis, differential

Case study

Mrs PS, 78 years of age, presented with acute left-sided otalgia, ear swelling and subsequent unilateral facial paralysis (Figure 1). She denied any otorrhoea or hearing loss. Past medical history relevant to the presenting complaint included:

- Bell palsy diagnosed 20 years ago with no residual effect
- biopsy confirmed benign parotid lump (diagnosed 3 years previously). Histopathology revealed a pleomorphic adenoma. Mrs PS declined surgical intervention at the time
- chicken pox as a child
- normal fasting blood glucose 1 month previously and no known immune compromise.



Figure 1. Patient shown with left sided unilateral facial nerve palsy

Examination revealed yellow crusts and small vesicles on the external acoustic meatus (Figure 2). A 10 mm well defined firm and nontender nodule was palpable at the ramus of the mandible.

Question 1

What are the possible differential diagnoses and which is the most likely?

Question 2

What is the recommended management of this condition?

Question 3

What is the likely prognosis of this condition?

Question 4

What preventive strategy could help reduce the incidence of herpes zoster and postherpetic neuralgia?

Answer 1

Possible differential diagnoses include:

- Ramsay Hunt syndrome (herpes zoster oticus). Ramsay Hunt syndrome typically presents with a triad of otalgia, cutaneous vesicles in a dermatomal distribution and unilateral facial nerve palsy. It results from the reactivation of latent varicella zoster virus within the geniculate ganglion.¹ Histopathology shows inflammation and neuritis of the facial nerve. Herpes zoster may also affect other cranial nerves, especially V, VI, VIII, IX and X
- recurrent Bell palsy. Bell palsy is the most common cause of acute unilateral facial nerve paralysis. It may also involve other cranial nerves. However, recurrent Bell palsy occurs in only 12% of patients with an initial diagnosis of Bell palsy.² Recurrences are more common

in patients with diabetes mellitus, immune compromise or a family history of Bell palsy. The rarity of recurrent Bell palsy however should warrant careful workup to exclude other aetiology prior to the diagnosis²

- malignant transformation of pleomorphic adenoma. Pleomorphic adenoma is the most common benign neoplasm of the salivary gland. Patients usually present with a slow growing painless mass in the parotid with an intact facial nerve. Pleomorphic adenoma has a 1% risk of malignant transformation after 10 years into carcinoma ex-pleomorphic adenoma and this can lead to facial nerve palsy³
- malignant otitis externa. This rare but severe form of otitis externa is caused by *Pseudomonas aeruginosa* and results in osteomyelitis of temporal bone. It is usually seen in the elderly, in those with diabetes and in immunosuppressed patients. Patients typically present with unremitting otalgia, otorrhoea, hearing loss and the pathognomic sign of granulation tissue at the bony osseous junction of the external auditory canal. Unilateral facial nerve palsy occurs when osteomyelitis involves the skull base.⁴

The most likely differential diagnosis is herpes zoster oticus. Mrs PS's symptoms are consistent with this diagnosis and the trademark vesicles were seen on the external auditory canal on examination (*Figure 2*).

Answer 2

The management of Ramsay Hunt syndrome can be considered under the headings 'facial nerve recovery', 'management of postherpetic neuralgia' (if present), and 'eye care'.

Facial nerve recovery

Current treatment involves the use of antivirals and corticosteroids within 72 hours of the onset of rash. However, a Cochrane review^{5,6} could not prove the benefit of this treatment regimen due to a lack of randomised control trials.

The recommended antiviral regimen is:

- acyclovir 800 mg five times per day, OR
- famciclovir 250 mg three times per day, OR
- valacyclovir 1 g three times per day for 7–10 days.⁷

The hypothesis is that antiviral treatment inhibits the viral replication process and prevents



Figure 2. Swollen left ear with crusting. A close look reveals small vesicles

further spread.⁸ High dose steroids are also recommended for 14 days for analgesia. Recent studies have found that steroids also reduce residual weakness of the facial nerve, synkinesis (involuntary muscle movement accompanying voluntary movement) and autonomic synkinesis (autonomic lacrimation postvoluntary muscle movement).⁹ Additional analgesic treatment may be required during the acute phase.

Postherpetic neuralgia

The incidence and duration of postherpetic neuralgia is found to be directly related to age. Patients can present with paresthesia, dysesthesias and allodynia (pain resulting from stimulus that does not usually evoke injury) in the dermatome affected. Gabapentin has been found to be effective and can be weaned 6 months after onset of the disease or when neuralgia resolves.¹⁰

Eye care

The eyes must be examined for the ability to close fully at rest to prevent abrasion and exposure keratopathy. Patients may require regular artificial eye drops during the day with eye ointment and taping of the paralysed lid at night. Ophthalmological opinion for consideration of temporary tarsorrhaphy may be necessary.

Answer 3

The natural history of Ramsay Hunt syndrome varies between patients. However, patients with Ramsay Hunt syndrome generally have a poorer outcome than patients with Bell palsy, and are more likely to have permanent effects.¹¹ There are limited studies examining the prognostic factors in Ramsay Hunt syndrome. Yeo et al¹² found that elderly patients were more likely to suffer severe initial facial nerve paralysis and had a lower probability of complete recovery. They also found that diabetes mellitus was a negative prognostic factor and therefore recommended tight glucose control in diabetic patients with Ramsay Hunt syndrome.¹²

Answer 4

The high morbidity associated with Ramsay Hunt syndrome has resulted in the consideration of vaccination to prevent herpes zoster. The Shingles Prevention Study evaluated the effectiveness of high titre, live attenuated herpes zoster vaccine in patients over the age of 60 years. They were able to conclude that the incidence was lowered by 50% and in the vaccinated group that still had shingles, the incidence of postherpetic neuralgia was reduced.¹³ In Australia, this vaccine is available on prescription for patients over the age 50 years. However the full cost must be met by the patient.¹⁴

Case follow up

Mrs PS was diagnosed with herpes zoster oticus and managed according to the treatment approach outlined in *Answer 2*. On subsequent follow up however, she had persistent facial weakness. The prolonged history of pleomorphic adenoma made it difficult to exclude ex-pleomorphic adenocarcinoma as the underlying pathology. She is currently awaiting elective parotidectomy.

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