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Dr Hardeep K. Bhupal
MBBS, MRCGP, MRCP (UK), DRCOG
GP, Hillingdon, Middlesex

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AUTHOR
Dr Hardeep K. Bhupal
MBBS, MRCGP, MRCP (UK), DRCOG
GP, Hillingdon, Middlesex

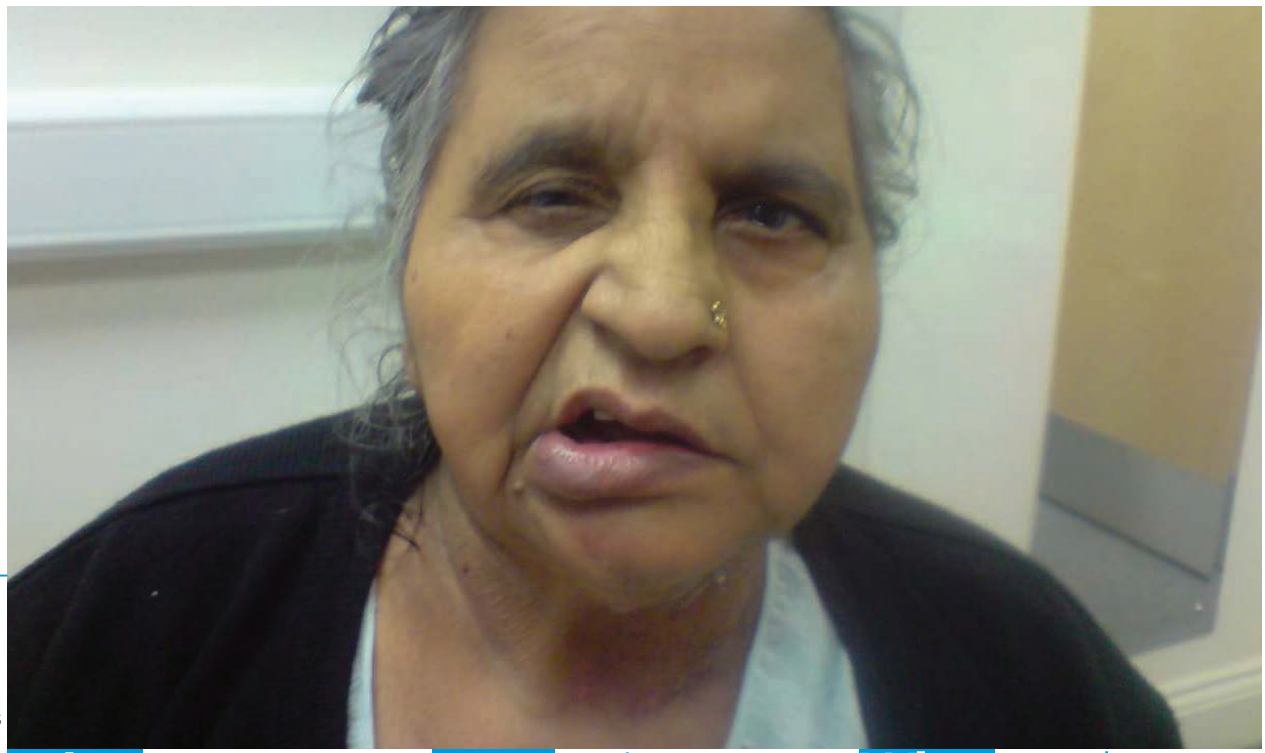


FIGURE 1
Ramsay Hunt syndrome. On asking the patient to smile the left-sided facial weakness becomes more apparent

What causes Ramsay Hunt syndrome?

RAMSAY HUNT SYNDROME IS A LOWER MOTOR NEURONE WEAKNESS OF THE SEVENTH (FACIAL) cranial nerve caused by reactivation of the herpes zoster virus.

It was first described by James Ramsay Hunt (1874 – 1937), an American neurologist. The virus infects the geniculate ganglion of the nerve causing facial weakness. The onset of a motor neuropathy thereby makes it inherently different from the more typical presentation of shingles, which predominantly causes a sensory neuropathy. A vesicular erythematous rash on, or around, the pinna (zoster oticus) may be present.

EPIDEMIOLOGY

Herpes zoster, or shingles, occurs through reactivation of the varicella zoster virus, which also causes chickenpox. After the initial infection the virus remains within the sensory ganglia and with increasing age has a propensity to reactivate and present as herpes zoster. Hence herpes zoster and Ramsay Hunt syndrome are seen as diseases of older people.

It is believed that approximately 20-30% of individuals will be affected by herpes zoster during their lives.[1,2] However, Ramsay Hunt syndrome is a rare disease, affecting fewer than 1 in 1,500 people in the United States. Vulnerability to reactivation occurs as a result of immunosenescence, and a gradual decline in cellular and possibly humoral immunity.[3]

Patients who are immunocompromised are at greater risk of developing herpes zoster and reactivation can result in a more severe widespread presentation.

How is the condition diagnosed?

CASE REPORT

A 70-year-old woman presented to her GP with a two-day history of left ear pain and no further symptoms.

She was immunocompetent and apart from lisinopril for her hypertension was not on any other medication.

On examination, there was mild erythema around the left ear canal, however, there were no other obvious abnormalities. Otitis externa was suspected so she was prescribed a course of oral antibiotics and advised to come in should she develop any further symptoms.

Three days later she was seen by another GP. She had developed a vesicular, erythematous rash extending from the external meatus, to involve »

What are the treatment options?



FIGURE 2 (right) Vesicular erythematous lesions exuding a serous discharge and covered in calamine lotion



FIGURE 3 (far right) Left-sided facial weakness, however, the ability to protect the eye is preserved

the post-auricular area, and also down the mandible, see figure 2, above. The diagnosis at this stage appeared clear, the patient had developed varicella zoster. She was started on aciclovir with a plan to review over the next few days.

However, two days later the patient presented with left-sided facial weakness and was seen by yet another GP. A diagnosis of Ramsay Hunt syndrome was made.

The patient was advised to continue her aciclovir. At review three days later, most of the active lesions had regressed. She was prescribed oral prednisolone and amitriptyline along with paracetamol and codeine phosphate for pain relief.

Her corneal reflex and ability to close her left eye fully were preserved, see figure 3, above. She was followed up regularly over the next two weeks. The lower motor neurone facial weakness of the seventh cranial nerve resolved and her skin lesions regressed.

SYMPTOMS AND SIGNS

Pain

The pain can be subjective in nature and severity but has commonly been described as a dull background ache with acute exacerbations. Post-herpetic neuralgia is a well known complication and can result in typical features associated with neuropathic pain.

The pain around the ear may also be associated with headaches and a general malaise along with allodynia (pain at the site provoked by innocuous stimuli). Patients may also report a numbness or tingling sensation around the area.

Ipsilateral auditory manifestations can occur and these include: tinnitus, deafness, hyperacusis (due to paralysis of the branch of the facial nerve supplying the stapedius) and a sensation of aural fullness.

Additional symptoms include nausea, vomiting, vertigo and a change in taste perception.

Rash

Early presentations may mimic otitis externa, as was the case in this patient. The locality of the rash is variable, and common sites include: the auditory canal, in and around the pinna; the palate; and occasionally the anterior two thirds of the tongue. It commonly presents as an erythematous vesicular rash, with the vesicles coalescing to form bullous lesions, resolution usually takes 10 to 14 days. However, complete resolution can take 2-4 weeks and may be complicated by superimposed bacterial infection.

The presence of pain and allodynia, without the onset of a rash, is indicative of the condition zoster sine herpete.

Ipsilateral facial weakness

This can cause significant alarm and is often a major factor in the patient seeking immediate medical attention. It characteristically involves the lower motor neurone fibres of the seventh (facial) cranial nerve.

Preservation of the motor function of the forehead muscles is characteristic and can be assessed by asking the patient to raise his or her eyebrows. If the patient is able to furrow the forehead muscles then the upper motor neurones have been spared and the lesion is exclusive to the lower motor neurone fibres.

In 12% of patients presenting with lower motor neurone facial weakness the cause is Ramsay Hunt syndrome.[4] Other causes include Bell's palsy, trauma to the facial nerve, parotid gland tumours and cerebrovascular events.

Table 1

Differential diagnosis

Bell's palsy	One-sided facial weakness also occurs in Bell's palsy. However, the characteristic feature of Ramsay Hunt syndrome remains the presence of the typical erythematous vesicular rash. The facial palsy associated with Ramsay Hunt syndrome tends to be more severe and the prognosis less favourable.[4]
Otitis externa	Early presentation in Ramsay Hunt syndrome can be mistaken for otitis externa, as happened in this case. The natural disease progression and onset of the rash remains a key differentiating factor, as does the development of facial palsy.[6]
Referred pain	This can be from a variety of sources including the temporomandibular joint, teeth, ear and nasopharynx.
Herpes simplex virus (HSV) infection	The rash may be morphologically similar, however there is no associated facial palsy. HSV infections have a tendency to recur whereas only 4% of patients will experience a second episode of shingles, and third episodes are even rarer.[7]
Atypical headaches	Trigeminal neuralgia, cluster headaches and occasionally temporal arteritis may have a similar presentation.
Acute vestibular neuronitis and Ménières disease	Both these conditions can present with symptoms similar to those associated with Ramsay Hunt syndrome.

Severe eruptions can result in significant ophthalmological complications. Thus the cornea, along with the corneal reflex, and the patient's ability to close the eye completely should be fully assessed. Severe eruptions are an indication for an urgent ophthalmological opinion as there is higher risk of ocular complications.[5]

Differential diagnoses are listed in table 1, opposite.

DIAGNOSIS

Within the general practice setting the diagnosis remains clinical. The presence of severe eruptions or significant complications such as ocular involvement or severe cellulitis may warrant acute admission.

Diagnosis can be confirmed by obtaining scrapings from the lesions and direct immunofluorescence staining of the varicella zoster virus. Polymerase chain reaction is also an effective way of confirming the diagnosis and may play a role in those individuals who present early without typical features, or in those who present with zoster sine herpete or in whom the diagnosis remains ambiguous.[8,9]

MANAGEMENT

Antivirals

A recent review concluded that management of Ramsay Hunt syndrome remains controversial with a lack of evidence to support the use of antivirals.[10] However, a retrospective study, published in 1997, showed a significant improvement in outcomes in patients treated with antiviral medication within 72 hours of the onset of symptoms. Complete recovery occurred in 75% of patients treated within 72 hours compared with 30% of patients treated within 7 days of the onset of symptoms.[11]

A prospective study, in 2001, concluded that aciclovir improved treatment outcome.[12] Several randomised, placebo-controlled trials have also found this.[13-16]

Although there is no clear guidance on recommended dosing regimens, trials commonly used 800 mg aciclovir orally 5 times a day for 7-10 days. This appears to be the recommended dosing regimen in the UK with the preferred duration of treatment being 7 days.[17] There is no evidence to suggest increased benefit of using intravenous aciclovir over oral aciclovir.[11] Although most benefit is obtained if antivirals are prescribed within the first 72 hours, all patients should be assessed individually and those at high risk of developing post-herpetic neuralgia (over 50 years of age, severe eruptions and prodromal symptoms) should be prescribed antiviral medication.

Corticosteroids

Large controlled trials have concluded that adjuvant corticosteroid therapy resulted in improved and quicker healing of cutaneous lesions and improved relief of neuralgic symptoms.[17,18] Trials have used a regimen of oral prednisolone 1 mg/kg daily for 5 days followed by tapering the dose.[12] An alternative regimen of 60 mg of oral prednisolone daily for 5 days followed by tapering has been proposed.[19]

Care should be taken in prescribing steroids in patients with comorbidity such as diabetes mellitus, hypertension, osteoporosis and associated metabolic disorders.

Adjuvant therapy

Oral analgesics, lubricating eye drops, eye patches and taping down of the eyelid can also play an important role in management.

Post-herpetic neuralgia: Pain persisting for three months after the onset of symptoms is termed post-herpetic neuralgia. It is more commonly seen in older patients and treatment may include amitriptyline and neuroleptics such as gabapentin. Topical capsaicin and lidocaine patches have also been shown to be of benefit.[20,21]

PROGNOSIS

Advanced age, diabetes mellitus and hypertension appear to be significant poor prognostic features with patients more likely to suffer complications.[22] With regards to facial paralysis, 66% of patients with untreated partial loss of nerve function at onset regain complete function, in contrast to 10% of patients recovering completely when presenting with complete loss of nerve function.[23]

CONCLUSION

Ramsay Hunt syndrome can present with vague and non-specific symptoms, especially during early onset of the disease. A high index of suspicion and frequent follow up are imperative for early diagnosis. There is clear evidence to support the use of early antiviral therapy along with oral corticosteroid use. Early and appropriate intervention has resulted in significant improvement in treatment and complication outcomes.

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Useful information

www.ramsayhuntsyndrome.org