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An extreme and unusual variant of Ramsay Hunt syndrome

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Abstract
Ramsay Hunt syndrome is characterized by facial nerve paralysis, herpetic vesicles in or around the ear and pain often associated with vestibulocochlear nerve involvement. It is thought to be a cranial polyneuropathy caused by the herpes zoster virus. We present an extreme and unusual variant of this disease with involvement of VIIth, VIIIth, Xth, XIth and XIIth cranial nerves as well as C2–4 sensory dermatomes and profound systemic upset which caused some diagnostic uncertainty.

Key words: Herpes zoster oticus; Facial paralysis; Complications

Introduction

Ramsay Hunt syndrome was first described by Ramsay Hunt in 1907 as a syndrome of facial paralysis, ear pain and a herpetic auricular rash. It was originally classified into four groups (Aviel and Marshak, 1982):

(1) herpes oticus without neurological signs;
(2) herpes oticus with facial palsy;
(3) herpes oticus with facial palsy and auditory symptoms;
(4) herpes oticus with auditory and labyrinthine symptoms.

It has long been known that it is caused by reactivation of varicella zoster virus (VZV). Ramsay Hunt suggested that the syndrome was due to geniculate ganglionitis but it is now thought to represent a neuritis or cranial polyneuropathy (Wackym, 1997).

We report a case with an extreme and unusual presentation characterized by widespread and progressive neurological involvement that caused some diagnostic concern.

Case report

A 67-year-old retired radiographer presented to the outpatient clinic with a one-week history of right earache, severe sore throat and dysphagia for solids associated with general malaise and fever. Her voice had also become weak and husky two days previously. However, there were no specific complaints of hearing loss or balance disturbance. She had been diagnosed by her general practitioner as having 'quinsy' and was treated with erythromycin and ciprofloxacin with no improvement in her symptoms.

On examination, she was generally unwell, while inspection of her ears, nose and throat showed the presence of haemorrhagic bullae on the right tympanic membrane and a number of large aphthous ulcers in the pharynx and larynx involving the right faucial pillar, epiglottis and the aryepiglottic fold on the right side. Laryngoscopy also revealed paralysis of the right vocal fold.

Initially these findings were thought to be due to a non-specific viral illness for which supportive and symptomatic treatment was recommended. However, within 48 hours her general malaise and presenting symptoms had worsened. She had also developed a right partial facial weakness and was therefore admitted to hospital for further investigation and treatment.

This revealed a normal haemoglobin and white cell count, and an ESR of 8 mm. Her chest was clear and throat swabs revealed no bacterial growth. Pure tone audiometry showed a sensorineural high tone hearing loss in the right ear of 40 dB. Blood samples were sent for virology studies.

At this stage, the possibility of Ramsay Hunt syndrome was considered. She was given intravenous acyclovir as well as supportive therapy with intravenous fluids, analgesics and anti-emetics.

This resulted in a fairly rapid resolution of her constitutional symptoms with some improvement in her facial strength and she was discharged from hospital on oral acyclovir a week after admission.

However, one week later she developed further neurological symptoms and was readmitted for thorough neurological assessment. She was noted to have pain involving the C2–4 sensory dermatomes on the right and found to have weakness of the right trapezius and sternocleidomastoid muscles as well as slight weakness involving the right side of her tongue.

In view of the widespread neurological involvement and in the absence of serological confirmation the diagnosis of Ramsay Hunt syndrome was reconsidered. However magnetic resonance image (MRI) brain scan ruled out an alternative underlying neurological lesion while the results of virology studies subsequently showed a varicella zoster IgG titre of >100 U/ml. Antibodies to varicella zoster virus (VZV) were positive and VZV IgM was positive. Viral studies of CSF were negative for herpes zoster DNA.

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These findings were consistent with recent infection with VZV which showed that this was in fact a very unusual variant of the Ramsay Hunt syndrome.

Discussion

There have been reports of Ramsay Hunt syndrome presenting with multiple cranial nerve involvement in the past. Aviel and Marshak (1982) reviewed the literature and found that the following nerves are involved in order of decreasing frequency: VIIth, VIIIth, IXth, Vth, Xth and VIth with involvement of Ist, IInd, IIIrd, IVth, XIth and XIIth being rare. Other central or peripheral phenomena described in the literature include cervical dermatome involvement (Steffen and Selby, 1972), encephalitis (Bhattacharyya and Kakati, 1993), Horner's syndrome and herpes zoster uveitis (Aviel and Marshak, 1982) and the syndrome of inappropriate secretion of antidiuretic hormone (SIADH) (Kagayama *et al.*, 1989).

Our case led to diagnostic uncertainty because of the unusual presentation and widespread involvement. The profound systemic upset experienced by our patient is also very uncommon. In addition to the late appearance (10 days after the onset of symptoms) of facial palsy there was eventually evidence of involvement of the VIIIth, Xth, XIth and XIIth cranial nerves as well as cervical dermatomes C2–4 on the ipsilateral side. This specific pattern of nerve involvement has not previously been described.

Diagnosis of Ramsay Hunt syndrome is usually based on a small recognizable group of clinical features. However, in atypical cases, it can be confirmed by isolation of viral antigen from the vesicles or by viral serology studies which should always be part of the acute investigation even though the results are only obtained after empirical treatment with acyclovir has been commenced on grounds of reasonable suspicion. VZV IgM and IgA are specific markers of recent infection or reactivation (Cicala *et al.*, 1977). In this case the diagnosis was confirmed by high titres found on viral serological studies.

The role of MRI in the diagnosis of Ramsay Hunt syndrome is controversial. Abnormal enhancement of the VIIth and VIIIth nerves in the internal acoustic meatus has been demonstrated in some cases of Ramsay Hunt syndrome although it is usually impossible to differentiate between Ramsay Hunt syndrome and Bell's palsy on MRI findings alone (Tada *et al.*, 1994). In this case however, MRI was carried out to exclude other pathology in view of the progressive widespread neurological signs.

In conclusion, although Ramsay Hunt syndrome usually presents with the classical triad of pain, vesicles and facial nerve paralysis, it must be kept in mind that it is a cranial polyneuropathy. It may therefore present with involvement of one or more cranial nerves, cervical dermatome involvement or as a systemic illness. It is important that other central and peripheral causes of multiple nerve involvement are excluded in such cases. However, the benefit of early recognition of Ramsay Hunt syndrome is that early commencement of antiviral treatment improves the prognosis significantly (Murukami *et al.*, 1997) as the clinical course tends to be more severe and the recovery profile less favourable than in Bell's palsy (Robillard *et al.*, 1986).

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