Unusual presentation of Ramsay–Hunt syndrome without facial nerve palsy

Introduction

Ramsay–Hunt syndrome is usually diagnosed on history and examination as zoster oticus and lower motor neuron facial palsy. However, atypical cases may delay diagnosis and appropriate treatment. This article presents a case of delayed appearance of the auricular rash without facial palsy, in which there was cranial polyneuropathy associated with severe systemic illness.

Discussion

Ramsay–Hunt syndrome was first described in 1907 as facial paralysis, earache and a herpetic auricular rash. Other common presentations noted by Hunt included tinnitus, hearing loss, nausea, vomiting, vertigo and nystagmus. This syndrome was initially thought to be a result of geniculate ganglionitis by varicella zoster virus (VZV) reactivation (Sweeney and Gilden, 2001). A review of case reports suggests that this syndrome is also a cranial polyneuropathy, with VIIth, VIIIth, IXth, Vth, Xth and VIth nerves being involved in decreasing frequency (Aviel and Marshak, 1982). Vocal cord paralysis, facial pain, trapezius and sternocleidomastoid muscle weakness have been described (De and Pfleiderer, 1999; Kuhweide et al, 2002) in patients who have Ramsay–Hunt syndrome.

Case Report

A 65-year-old woman presented to the outpatient clinic with severe left earache associated with nausea, dizziness and left-sided hearing loss. Her symptoms, which started 5 days previously, had got progressively worse. There was no history of ear discharge or trauma. She was otherwise fit and healthy with no remarkable past medical history.

On examination, she was generally unwell. The left pinna was swollen and tender. The auditory canal was tight and inflamed with a small amount of discharge. These findings were initially thought to be otitis externa associated with perichrondritis of the pinna. She was admitted for symptomatic relief, aural toileting and intravenous antibiotics.

Her full blood count was normal and swabs taken revealed only normal skin flora. Her symptoms, however, did not improve as expected. Her dizziness and nausea had worsened although she did not have true rotatory vertigo or ataxia. She also complained of dysphagia to solids. There was no significant change to her speech but a fibreoptic endoscopy revealed sluggish movement of the left vocal cord and reduced sensation of the supraglottis. The rest of her neurological examination was otherwise normal.

At this stage, the possibility of Ramsay–Hunt syndrome was considered but there was still no evidence of herpetic lesions or facial palsy. Pure tone audiometry showed a sensorineural hearing loss of 30 dB in the left ear. Computed tomography scan of the temporal bones and magnetic resonance imaging of the brain were normal. Her symptoms gradually improved over the course of 1 week. On discharge, all her presenting symptoms had completely subsided except for mild intermittent nausea.

She was readmitted a week later with recurrence of left earache. However, both the nausea and the dizziness that presented themselves on initial admission were absent. The pinna was not swollen but there were now herpetiform vesicles in the left auditory meatus and tympanic membrane. Neurological examination was normal. Antibodies to varicella zoster virus (VZV) and VZV immunoglobulin M were positive.

These findings were consistent with VZV infection, which suggest that this may have been an unusual variant of Ramsay–Hunt syndrome. She was treated with acyclovir and gabapentin for post-herpetic neuralgia. The patient responded to treatment and was discharged 3 days later. Her follow-up examination revealed no evidence of residual symptoms.

This case led to diagnostic uncertainty because of the unusual presentation. Patients with viral labyrinthitis or vestibular neuronitis may present similarly. The profound systemic illness experienced by this patient did not match initial clinical findings. The late appearance of herpetiform lesion and the absence of facial palsy are unlike typical Ramsay–Hunt syndrome. In addition the IXth and Xth cranial nerve manifested as dysphagia and vocal cord palsy.

The diagnosis of Ramsay–Hunt syndrome is usually based on history and neurological examination. In unusual cases, it can be confirmed by the isolation of viral antigen from the vesicles or by polymerase chain reaction (PCR) (Gnann, 2002). A negative VZV immunoglobulin G or immunoglobulin M does not rule out Ramsay–Hunt syndrome. Examination of CSF and magnetic resonance imaging (MRI) has no diagnostic or prognostic value (Jonsson et al, 1995).

Patients with Ramsay–Hunt syndrome should receive antiviral treatment during the acute phase of the illness. Recovery of the facial nerve has shown the greatest rate of improvement with acyclovir and steroids rather than steroids alone (Wood et al, 1994). However, the use of steroids is not without danger and thus patients have to be properly selected.

The benefits of using steroids is unclear. There was no significant benefit gained with the addition of steroids in the healing of the herpetic lesions (Adour et al, 1996) or recovery of the facial nerve (Stankiewicz, 1987). Despite the paucity of large randomized, controlled prospective treatment trials, data from

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the collective case reports and retrospective reviews suggest that both prednisolone and acyclovir, if given early, improve the overall prognosis. There was statistically significant improvement in patients treated with prednisolone and acyclovir within 3 days of onset (Murakami et al, 1997). Dual therapy has not shown any influence on the incidence or severity of post-herpetic neuralgia (Wood et al, 1994).

Conclusions

Ramsay Hunt–syndrome is usually recognized as zoster oticus and lower motor neuron facial palsy. However, it may present as a polyneuropathy associated with systemic illness. In cases like this, it is important to rule out other causes of central and peripheral neuropathy with appropriate investigations. An urgent MRI of the brain and temporal bones with a neurology opinion may be warranted. A high index of suspicion is required to diagnose cases where presentation is atypical. **BJHM**

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For getting your numbers wrong

As a columnist, I need a firm view about Professor Sir Roy Meadow. But I keep vacillating.

Medical expert witnesses and child protection have had a bad press this year. First the General Medical Council (GMC) admonished Professor David Southall for watching television and jumping to conclusions; now they've struck Meadow off for getting his numbers wrong over Sally Clark. As he is well over retirement age, that has something of the farce about it, but this is no laughing matter: not for Meadow, nor for the wrongly convicted, and nor for the babies murdered by their parents. How much of it, I wonder, is the result of the adversarial British court system, which encourages experts to be certain?

The newspapers gave wide coverage. Editorial comment was mixed; there was little triumphalism. There were interviews with the wronged parents: some of them thought Meadow deserved everything he got, others thought the verdict harsh. More than one said an apology would have been appreciated. The *Lancet* came out strongly against the rigour of the penalty, both warning the GMC before and criticizing them afterwards. The *British Medical Journal* (BMJ) reported the facts but its opinion was muted. Not so the BMJ's rapid responders: doctors bewailed the decision, certain that it would wreck child protection forever; lay responders applauded: an arrogant wrecker of lives given his comeuppance. On the more informal internet doctor forums, language was more intemperate.

Almost everything, if the media reports are to be believed, hinged on Meadow's misunderstanding of probability: his statement to the court of a 1 in 70 million chance of deaths occurring in the same family. As many correspondents have pointed out, if this is such a simple matter, why didn't someone else in the court correct his error at the time?

David Southall took covert video film of parents harming their own children. Some people will never forgive him. As a paediatric colleague ruefully said, 'They defend the right to kill their children.' The courts don't always get things right, but there is a difference between a verdict being overturned because of fresh evidence or a confession, and being overturned on a technicality. The guilty sometimes go free.

We have already had the Wolff report on court procedure and expert witnesses; another review, or at least some more advice, seems likely. And I wonder about the role of the GMC in all this. If an expert chemist, say, misinterpreted their evidence, what would happen to them? Would their professional body be able to censure them? Does an opinion given in court bear on one's right to practise clinical medicine?

So I think I'm in the 'deserved censure but not striking off' camp. But the camp that I know I'm in is the 'glad I'm not there' camp. These columns are about as far above the parapet as I want my head to be. A colleague who couldn't be counted as anything other than calm and collected acted as expert witness in a criminal case with nowhere near the implications of child protection and found it so stressful that he had to take ranitidine to get him though the week. I'm either a coward, or recognize that pinko liberal tendencies is not what barristers want. **BJHM**

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