CASE REPORT

Facial palsy and atrial fibrillation: a special case of Ramsay Hunt syndrome

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SUMMARY

Ramsay Hunt syndrome (herpes zoster oticus) is a rare complication of latent Varicella Zoster virus infection. It can be complicated by permanent hearing loss, loss of taste and postherpetic neuralgia. Although Ramsay Hunt syndrome most prominently involves the facial nerve, a number of other cranial nerves can be involved such as the vestibulocochlear, glossopharyngeal and the vagus nerve. We report on a case of Ramsay Hunt syndrome with cranial polyneuritis complicated by atrial fibrillation. Vagal involvement as evidenced by physical examination and MRI findings was present in our patient. We hypothesise that viral vagal neuritis likely contributed to the development of atrial fibrillation in our patient through involvement of the autonomic components of the vagus nerve.

BACKGROUND

Ramsay Hunt syndrome (herpes zoster oticus) is a rare complication of latent Varicella zoster virus (VZV) infection. Although Ramsay Hunt syndrome (RHS) most prominently involves reactivation of VZV in the facial nerve, a number of other cranial nerves have been reportedly involved such as the vestibulocochlear nerve, glossopharyngeal nerve and the vagus nerve.¹ RHS can be complicated by permanent hearing loss, loss of taste and postherpetic neuralgia. We report on a case of Ramsay Hunt syndrome complicated by paroxysmal atrial fibrillation in a 60-year-old woman. Initially diagnosed as acute otitis media, our patient's presentation evolved from left ear fullness to fever, hoarseness of voice, left ear rash, facial palsy and paraesthesias. Varicella zoster DNA PCR from a blood sample was positive. Brain MRI revealed involvement of cranial nerves 5, 7, 8 and 10 with viral inflammation. The presentation was later complicated by development of atrial fibrillation with rapid ventricular response. We believe that the development of atrial fibrillation is a novel manifestation of Ramsay-Hunt syndrome and was likely autonomically mediated through viral vagal neuritis. We discuss our patient's case and the likely pathophysiology underlying her unique presentation.

CASE PRESENTATION

A 60-year-old woman with no significant cardiac history presented to her primary care physician with several weeks of left ear fullness and tinnitus and a week of left-sided soreness of the throat and sharp left-sided ear pain. On examination, she

appeared well with no visible external ear abnormalities. Her oropharynx was clear and moist, with no erythema or exudates. Her neck was non-tender and without lymphadenopathy. Examination of the left tympanic membrane revealed erythema, with no purulence or bulging. Examination of the right tympanic membrane was normal. Her presentation was thought to be explained by acute otitis media and the patient was prescribed 1 week of oral amoxicillin, as well as ciprofloxacin ear drops.

Five days after her initial presentation, she presented to the emergency department with facial asymmetry, a left ear rash, fatigue, fever, dysphagia, nausea and vomiting. Examination in the emergency department was significant for an elevated temperature of 39.5°C and an erythematous, oedematous external acoustic meatus with multiple vesicular lesions (figure 1). Oropharyngeal examination revealed small vesicular and friable mucosal lesions of the left oropharynx (figure 2). The left tympanic membrane was difficult to visualise. Cranial nerve examination demonstrated involvement of left-sided cranial nerves V (loss of sensation to light touch in the distribution of V1, V2, V3), VII (moderate lower motor neuron facial weakness, House-Brackmann grade III), VIII (left-sided hearing loss), IX and X (reduced gag reflex and dysphonia). The patient was admitted to the inpatient medicine service for management.

During her admission, the patient developed sudden onset tachycardia. On examination, she had an irregularly irregular rhythm. An ECG was obtained and revealed atrial fibrillation with a rapid ventricular response. The patient had no prior history of cardiac arrhythmias. Within hours, the patient returned to normal sinus rhythm with rate control treatment. After a 5-day inpatient stay, she was discharged in normal sinus rhythm. One day after discharge, she returned to the emergency department with palpitations and was found to have another episode of atrial fibrillation with rapid ventricular response. She was then admitted to the inpatient cardiology service for management.

INVESTIGATIONS

On her initial admission to the medicine service, a complete blood count was within normal limits. She was acidotic with a serum bicarbonate of 21 mmol/L, along with an anion gap of 21 and a lactic acid of 3.2 mmol/L. Her erythrocyte sedimentation rate was elevated at 63 mm/H. Varicella Zoster DNA PCR from a blood sample was positive. MRI of the brain was obtained. Axial

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Figure 1 External acoustic meatus with multiple vesicular lesions.

T1-weighted images postcontrast revealed inflammation of the intracanalicular portions of cranial nerves VII and VIII, dural inflammatory enhancement within the internal auditory canal (figure 3) as well as inflammation of cranial nerve X (figure 4). In context of the patient's clinical presentation, this was highly suggestive of viral cranial polyneuritis due to Ramsay Hunt syndrome. Speech and swallow evaluation revealed a poor labial seal, a markedly raspy voice, mild pocketing of fluid in the left buccal cavity and an immediate cough with thin liquid swallow trials.

After her initial episode of tachycardia, an ECG was obtained and revealed atrial fibrillation with a rapid ventricular response. Lab testing for thyroid function with thyroid-stimulating

Figure 2 Small vesicular lesion of the left oropharynx (circled).

Figure 3 Contrast-enhanced axial T1-weighted MRI image showing inflammation of cranial nerves VII and VIII (arrow).

hormone and free T4 was within normal limits. Three sets of troponin T were <0.03 ng/mL. A transthoracic echocardiogram evidenced normal biventricular systolic function and no significant valvular dysfunction.

Treatment

During her initial admission to the inpatient medicine service, the patient received intravenous acyclovir, as well as intravenous methylprednisolone. Her diet was adjusted to include only thickened fluids and she was educated on measures to avoid aspiration. Following her episode of atrial fibrillation with rapid ventricular response, bolus intravenous doses of diltiazem were administered twice, after which she reverted to normal sinus

Figure 4 Contrast-enhanced axial T1-weighted MRI image showing inflammation of cranial nerve X (arrow).

rhythm. She was later discharged in normal sinus rhythm and prescribed a 14-day course of oral acyclovir as well as oral diltiazem to be taken as needed for rate control in case of atrial fibrillation recurrence.

One day after discharge, the patient returned to the emergency department for symptomatic palpitations and was found to have atrial fibrillation with a rapid ventricular response at 152 bpm. She was started on intravenous continuous diltiazem infusion for rate control and reverted to normal sinus rhythm a few hours later. The patient was started on oral metoprolol and after 2 days of inpatient rhythm monitoring remained in sinus rhythm and was discharged.

OUTCOME AND FOLLOW-UP

After discharge, the patient was scheduled for a 2-week portable heart rhythm monitor to evaluate arrhythmia burden, and this did not reveal any recurrence of atrial fibrillation. Nine months after discharge, the patient had not had any recurrence of her atrial fibrillation.

Her external auditory canal and buccal vesicular lesions had mostly resolved. Her facial asymmetry, dysphagia, hoarseness of the voice and hearing loss had not improved. Her loss of sensation to light touch in the distribution of V1, V2 and V3 had evolved into facial paraesthesias, possibly indicating gradual neurological recovery. One month after discharge, laryngeal videostroboscopy demonstrated near immobility of the left vocal fold. An audiological evaluation demonstrated moderate to severe left sensorineural hearing loss.

DISCUSSION

VZV is a human herpesvirus that causes chickenpox in humans. Following primary infection, VZV persists in dorsal root and cranial nerve ganglia for life. Later in life, reactivation and replication of the virus can lead to various clinical manifestations depending on the site of reactivation. Classically, reactivation is in the form of herpes zoster, manifesting as cutaneous eruptions in a thoracic or lumbar dermatome. Reactivation has been associated with increasing age or immunosuppression, however, the biological mechanisms underlying reactivation remain unclear.²

First described by Hunt in 1907, RHS is classically defined as a triad of lower motor neuron facial paralysis, ear pain and herpetic eruptions on the ear or in the mouth. The classic manifestations of RHS are due to reactivation of VZV in the geniculate ganglion of the facial nerve. However, involvement of other cranial nerves such as the vagus, glossopharyngeal and vestibulocochlear nerve has been described. Involvement of the aforementioned cranial nerves can be explained by the fact that all three travel together and derive from the same brachial arch.¹ Such polyneural involvement is not common however; in a series of 325 patients with RHS, Murakami *et al* found glossopharyngeal or vagal nerve paralysis concomitant with facial nerve paralysis in only 2.5% of patients.³

Atrial fibrillation is the most common sustained cardiac arrhythmia, with an estimated overall prevalence of 1%. Increasing age is a primary risk factor, with more than 9% of persons over the age of 80 affected.⁴ Hypertensive, coronary and valvular heart disease are common underlying causes for atrial fibrillation. Non-cardiac causes such as hyperthyroidism and surgery are less commonly implicated. The autonomic nervous system, primarily through vagal afferents, may possibly be involved in the pathogenesis of atrial fibrillation. This is particularly important in paroxysmal atrial fibrillation,

as it has been shown that either increased vagal or sympathetic tone can promote atrial fibrillation.^{5,6}

We hypothesise that in our patient with RHS with cranial polyneuritis, involvement of the vagus nerve with inflammation likely induced autonomically mediated atrial fibrillation. Evidence of vagal involvement was through both physical examination showing vocal cord paralysis as well as brain MRI showing involvement of cranial nerve X with inflammation. Based on our review of the literature, we believe this manifestation of RHS is novel and worthy of report. While systemic inflammation in general has been implicated in the initiation and maintenance of atrial fibrillation,⁷ this is usually due to a prolonged inflammatory process rather than an acute onset of an inflammatory condition. Our patient also did not have any history of cardiac disease, had a normal echocardiogram and had no recurrence of her atrial fibrillation on a 2-week portable heart rhythm monitor and remains free of atrial fibrillation 9 months after her initial presentation. We believe this supports our hypothesis that involvement of the vagus nerve with inflammation is the more likely explanation for the onset of paroxysmal atrial fibrillation in our patient rather than the systemic inflammation associated with RHS.

Early treatment of RHS with antivirals and corticosteroids has been shown to improve pain, increase the chance of hearing recovery and reduce nerve degeneration.⁸ In treatment of vagally mediated atrial fibrillation that has not spontaneously reverted to sinus rhythm, antiarrhythmics with vagolytic properties such as flecainide and disopyramide have been shown to be more successful in maintaining sinus rhythm.⁹

Learning points

- ▶ Ramsay Hunt syndrome (RHS; herpes zoster oticus) is a rare complication of latent Varicella zoster virus (VZV) infection.
- ▶ Although Ramsay-Hunt syndrome (RHS) most prominently involves reactivation of VZV in the facial nerve, a number of other cranial nerves can be involved such as the vestibulocochlear nerve, glossopharyngeal nerve and the vagus nerve.
- ▶ Early treatment of RHS with antivirals and corticosteroids has been shown to improve pain, increase the chance of hearing recovery and reduce nerve degeneration.
- ▶ As demonstrated in our patient, involvement of the vagus nerve with inflammation may cause autonomic dysfunction increasing the likelihood of arrhythmias such as atrial fibrillation. In our patient, resolution of the acute inflammation prevented recurrence of atrial fibrillation.

Contributors FA, DM, R-AF drafted the case report and jointly drafted the discussion. FA obtained the images, patient consent and MRI images. MK was the attending physician and reviewed the case for accuracy and approved the final draft.

Competing interests None declared.

Patient consent Obtained.

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