CASE REPORT

Vocal cord paralysis associated with Ramsay Hunt syndrome: looking back 50 years

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SUMMARY

Ramsay Hunt syndrome is defined by herpes zoster oticus and peripheral facial nerve palsy which is often associated with otalgia. The syndrome is, in rare cases, associated with other cranial nerve paralyses including the vagal nerve causing unilateral vocal cord paralysis. Vocal cord paralysis is more often seen as a symptom of various other diseases, that is, malignant tumours, neurodegenerative illness, cerebrovascular assaults, inflammatory processes or as a result of intubation or surgical procedures. The symptoms of unilateral vocal cord paralysis are mainly hoarseness, dyspnoea and dysphagia. We present a case of Ramsay Hunt syndrome combined with unilateral hearing loss and left vocal cord paralysis. The patient underwent MRI, CT and a lumbar puncture causing anxiety in the patient and delaying the initiation of antiviral and anti-inflammatory treatment, which is only efficient when initiated within 72 h. We hope to raise the awareness of this disease.

BACKGROUND

Ramsay Hunt Syndrome (RHS) is defined by herpes zoster oticus and peripheral facial nerve palsy often associated with otalgia.¹ The syndrome accounts for about 12% of all facial nerve palsies and in rare cases the disease is associated with other cranial nerve paralyses including the vagal nerve causing unilateral vocal cord paralysis.^{2–3} Vocal cord paralysis is more often seen as a symptom of various other diseases, that is, malignant tumours, neurodegenerative illnesses, cerebrovascular assaults, inflammatory processes, other infections or as a result of intubation or surgical procedures. However, affection of the recurrent nerve is often found to be idiopathic. The symptoms of unilateral vocal cord paralysis are mainly hoarseness, dyspnoea and dysphagia often accompanied by aspiration.^{4–5} The diagnosis of RHS with other cranial nerve affections is often mistaken for more serious illnesses resulting in extensive clinical diagnostic workup, including neuroimaging and lumbar puncture, which in turn may delay initiation of the appropriate treatment. We present a rare case of RHS combined with unilateral hearing loss and left vocal cord paralysis. In this case, the patient sought medical attention due to a swollen, painful ear (patient's own description) and peripheral facial nerve palsy. This should have led the doctor to the correct diagnosis, but instead the patient went through MRI, CT and a lumbar puncture, causing much anxiety in the patient. This also delayed the initiation of acyclovir treatment, which is only efficient when initiated within 72 h from

the onset of herpetic rash.⁶ We would like to raise the awareness of this disease, which might be underdiagnosed and possibly mistreated when diagnosis is delayed.

CASE PRESENTATION

A 43-year-old Caucasian man was referred to our ear, nose and throat department 1 week after initial symptoms consisting of severe otalgia and a swollen external auditory canal. A general practitioner had diagnosed the swelling as external otitis and treated with hydrocortisone–terramycin–polymyxin B ear drops and oral penicillin. This had no effect, and 1–2 days before presentation at our department, he had developed a peripheral facial nerve palsy of the left side and significant hoarseness, but no dyspnoea or loss of hearing was noted. At the first objective physical examination, the patient was alert and vital signs were normal. The left facial nerve scored 5 at the House-Brackmann scale. The external auditory canal was red and swollen but vesicles were not visible. A diagnosis of external otitis and idiopathic facial nerve palsy was made and hydrocortisone 50 mg/day was prescribed in addition to the anti-inflammatory and antibiotic ear drops. The patient consulted our outpatient department again 5 days later. The physical examination was extended to an audiogram showing a 15 dB asymmetric loss of hearing in the high frequencies of the left ear. The fiberoptic assessment and rigid video laryngoscopy showed total paralysis of the left vocal cord, but otherwise no pathology was found (video 1). A diagnosis of herpes zoster oticus was proposed and acyclovir 800 mg×5/day was prescribed. The patient was admitted to the department of neurology to have a lumbar puncture carried out in order to examine the spinal fluid for Lyme disease, herpes simplex and herpes zoster. The spinal fluid showed no trace of active infection. MRI of the cerebrum and a CT scan of the neck showed no pathology. A diagnosis of RHS associated with unilateral hearing loss and vocal cord paralysis was made. The patient was discharged with oral treatment of acyclovir and hydrocortisone.

INVESTIGATIONS

The patient had a lumbar puncture performed because encephalitis was suspected. MRI of the cerebrum and a CT scan of the neck were performed to exclude cerebral or local pathology. An audiogram, Multi-Dimensional Voice Programme (MDVP), fiberoptic and rigid video laryngoscopic assessment were also performed. The

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Video 1 Decreased movement of the left vocal cord and cartilago arythnoidea. The vocal fold is fixated in a paramedian position.

House-Brackmann scale was used to assess the facial nerve function.

It is very important to perform a cerebrospinal fluid analysis in order to exclude encephalitis before starting antiviral treatment in cases like these, since this serious diagnosis must not be overlooked.

DIFFERENTIAL DIAGNOSIS

- ▶ Cerebral assault/stroke
- ▶ Encephalitis
- ▶ Malignant disease (including temporal bone carcinoma)
- ▶ Autoimmune disorders (ie, myasthenia gravis and sarcoidosis)
- ▶ Necrotising otitis externa
- ▶ Otitis media
- ▶ Otitis externa
- ▶ Idiopathic facial nerve palsy
- ▶ Idiopathic vocal cord paralysis

Video 2 Normal function of both vocal cords after remission.

- ▶ Lyme disease
- ▶ Neurodegenerative disorders

A high-resolution CT scan of the temporal bone could have been relevant in this patient to exclude temporal bone carcinoma and would also have been helpful in excluding necrotising otitis externa.

TREATMENT

Initial treatment: hydrocortisone–terramycin–polymyxin B ear drops and oral penicillin.

One week from first symptom: oral corticosteroids 50 mg daily for 10 days.

One week and 5 days from first symptom: acyclovir 800 mg five times daily.

Optimal treatment for the disease would be acyclovir 400–800 mg five times daily combined with corticosteroids initiated within 72 h.⁶

Figure 1 Multi-Dimensional Voice Programme radial graphic display of a patient with recurrent nerve palsy showing breathy function and instability in the voice, with increased perturbation values and elevated amplitude values (above threshold level in the lower right quarter of the radial graph) as well as a small increase in tremor values (SPI/ATRI).

Table 1 Review of the previous reported cases with Ramsay Hunt syndrome (RHS) and vocal cord paralysis (English literature in the past 50 years)

Author	Number of patients	Year of Publication	Age/sex	Cranial nerve involvement	Clinical symptoms (in relation to pharynx and larynx)	Endoscopic features	Treatment	Follow-up
Coleman	1	2012	81/F	V,VII,VIII,X	Breathy voice, dysphagia, Odynophagia	Left VCP+ Vesicles aryepiglottal fold	Initial VAL: 3 days ACY:10 days Solumedrol: 10 days	FNP wanes on steroids No or little remission on swallowing and voice
Shim	1	2011	48/M	VII,VIII,IX,X	Odynophagia, hoarseness, Aspiration	Right VC palsy	IV ACY: 14 days Prednisolone: 28 days	Improved swallowing, but no improvement to VC palsy or FNP
Morelli	1	2008	82/M	V,VII,VIII,IX,X,XI,XII	Dysphonia, dysphagia	Left VC paresis+ vesicles in left hemi larynx, palatal paresis (left)	IV ACY: 14 days Prednisolone: 3 days	Swallowing improved, FNP and dysphonia remained after 42 days
Kim	1	2007	27/M	VII,VIII,IX,X	Dysphagia, hoarseness	Left soft palate paralysis+ left VCP	ACY: 14 days Methylprednisolone: 10 days	Only vertigo improved HL, FNP and laryngopharyngeal atrophy persisted
Nishioka	1	2006	78/M	III,IV,V,VI, VII, VIII, IX,X,XI,XII	Odynophagia, dysphagia, hoarseness and tongue weakness	Left tongue deviation, left soft palate paralysis and left VCP	ACY: 7 days Hydrocortisone: 3 days	After 1 year: tongue weakness resolved Can swallow soft meals, but FNP, VCP still affected
Espay	1	2005	83/F	VII,VIII,IX,X,XII	Dysphonia, dysphagia	–	ACY: 10 days	Dysphagia improved, other cranial nerves remained paralysed
Saito	1	2003	66/F	VII,VIII,X	Hoarseness	Left VCP	–	FNP recovered, VCP persisted
Radhakrishna	2	2000	49/F 40/F	(1) VII,IX,X (2) VII,IX,X,XI	(1) Hoarseness, dysphagia (2) Nasal regurgitation, hoarse	(1)– (2)–	(1)+(2) Steroids, ACY + sup. t	(1) (2) Improvement during 13 day
De	1	1999	67/F	V,VII,VIII,IX,X,XI,XII	Weak/husky voice, dysphagia	Ulcers in pharynx and larynx, right VCP	IV ACY Sup.t	Initial recovery of FNP on ACY, [–] on recovery of VCP
Rothschild	1	1994	68/M	VII,VIII,X	Hoarseness, dysphagia	Left VCP	ACY: 28 days	VCP resolved after 42 days
Golden	2	1990	51/M 69/M	V,VII,VIII,IX, X, XII VII, VIII, X	Dysphagia and voice change	Tongue weakness, diminished gag-reflex, right VCP VCP	IV cortisone+ sup.t.	Dysphagia, tongue paralysis and VCP resolved over months VCP partially resolved
Bhattacharya	1	1986	50/M	VII,VIII,IX,X	Hoarse/husky voice Nasal regurgitation	Left VC paresis Absence of gag-reflex	Prednisolone Sup.t., nasogastric feeding tube	Remission on skin-lesions, but only some improvement on voice and regurgitation

All cases have the features compatible with the RHS definition.

(–) is indicated when no information was available.

ACY, acyclovir; Asp, aspiration; FNP, facial nerve palsy; HL, hearing loss; IV, intravenous; Supt, supportive treatment; VAL, valacyclovir; VC, vocal cord; VCP, vocal cord paralysis.

OUTCOME AND FOLLOW-UP

At 2-months follow-up, the facial and laryngeal recurrent nerve had regained function (video 2). A MDVP radial graphic display of a patient with recurrent nerve paralysis showed breathy function and instability in the voice, with increased perturbation values and elevated amplitude values (above threshold level in the lower right quarter of the radial graph) as well as a small increase in tremor values (SPI/ATRI; figure 1). No audiogram was carried out at the follow-up, but the patient reported normal hearing function. At 4-months follow-up, the facial nerve had regained function to a House-Brackmann score of 2 and the vocal cord paralysis had resolved. The patient reported (self-assessed) normal function of the voice.

DISCUSSION

RHS associated with paralysis of the recurrent nerve and affection of the voice is an extremely rare condition. With an extensive search of the literature dating back to 1960, only a small number of cases have been reported (table 1).^{7–18} The rate of full recovery is found to be 67.7–82.9% in isolated RHS. However, when multiple cranial nerves are involved, the rate is as low as 27.3%.³ The data on recovery rate for individual cranial nerves are scarce in the literature. The rate of full recovery from facial paralysis alone was found to be in 45.5–54.5% and that of hearing loss was 11–80%.^{3 19} The recovery rate, however, seems to be better for vocal cord paralysis. In a study of RHS with multiple cranial nerve involvement, vocal cord paralysis was reported in eight patients; in five of these patients, a full recovery of 62.5% was observed.³ Kim *et al*²⁰ report a 60% full recovery rate of vagal nerve paralysis, but do not report on the vocal cord paralysis separately. It is generally acknowledged that ganglionitis is the cause of RHS and two theories explaining the viral spread are discussed in the literature. Either the virus spread along the nerves or by vascular anastomosis from the infected carotid artery.^{20 21} Conclusive evidence favouring one of these theories is still lacking. In patients suffering from varicella zoster infection of the ear, it has been shown that treatment with acyclovir and corticosteroids should be initiated within 72 h from the first symptom to be effective.⁶ In this patient, the treatment was unfortunately delayed due to initial failure to acknowledge the viral infection. Awareness of the disease should be raised and we hope to help doing so by reporting this case.

An interesting fact is the declining incidence of herpes zoster in the USA subsequent to the introduction of the varicella zoster vaccine into the children's vaccination programme.²² RHS, with and without other cranial nerve paralyse, might thus become even rarer in the future if the vaccination programme is extended to more countries table 1.

Learning points

- ▶ The Ramsay Hunt syndrome is defined by herpes zoster oticus and peripheral facial nerve palsy often associated with otalgia.
- ▶ The syndrome can mimic more serious conditions such as stroke, encephalitis and malignancy by causing multiple cranial nerve paralyse.
- ▶ Lumbar puncture and imaging modalities are essential when considering more debilitating differential diagnoses.
- ▶ Correct treatment with antivirals and corticosteroids should be initiated within 72 h.

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REFERENCES

- 1 Hunt J. On herpetic inflammations of the geniculate ganglion: a new syndrome and its complications. *J Nerv Ment Dis* 1907;34:73–96.
- 2 Peitersen E. Bell's palsy: the spontaneous course of 2,500 peripheral facial nerve palsies of different etiologies. *Acta Otolaryngol Suppl* 2002;549:4–30.
- 3 Shim HJ, Jung H, Park DC, *et al*. Ramsay Hunt syndrome with multicranial nerve involvement. *Acta Otolaryngol* 2011;131:210–15.
- 4 Brunner E, Friedrich G, Kiesler K, *et al*. Subjective breathing impairment in unilateral vocal fold paralysis. *Folia Phoniatr Logop* 2011;63:142–6.
- 5 Leder SB, Ross DA. Incidence of vocal fold immobility in patients with dysphagia. *Dysphagia* 2005;20:163–7; discussion 168–9.
- 6 Murakami S, Hato N, Horiuchi J, *et al*. Treatment of Ramsay Hunt syndrome with acyclovir-prednisone: significance of early diagnosis and treatment. *Ann Neurol* 1997;41:353–7.
- 7 Coleman C, Fozo M, Rubin A. Ramsay Hunt syndrome with severe dysphagia. *J Voice* 2012;26:e27–8.
- 8 Shim JH, Park J, Kwon BS, *et al*. Dysphagia in Ramsay Hunt's syndrome. *Ann Rehabil Med* 2011;35:738–41.
- 9 Morelli N, Mancuso M, Cafforio G, *et al*. Ramsay-Hunt syndrome complicated by unilateral multiple cranial nerve palsies. *Neurol Sci* 2008;29:497–8.
- 10 Kim JH, Chung PW, Oh S, *et al*. Ramsay Hunt syndrome complicated by a brainstem lesion. *J Clin Virol* 2007;39:322–5.
- 11 Nishioka K, Fujishima K, Kobayashi H, *et al*. An extremely unusual presentation of varicella zoster viral infection of cranial nerves mimicking Garcin syndrome. *Clin Neurol Neurosurg* 2006;108:772–4.
- 12 Espay AJ, Bull RL. Petrositis in Ramsay Hunt syndrome with multiple cranial neuropathies. *Arch Neurol* 2005;62:1774–5.
- 13 Saito S, Ochi K, Kobayashi T, *et al*. Vestibular-evoked myogenic potentials in two patients with Ramsay Hunt syndrome. *Auris Nasus Larynx* 2003;30(Suppl):S89–92.
- 14 Radhakrishna H, Malakondaiah T, Reddy IC, *et al*. Polyneuritis cranialis following herpes zoster. *Indian J Dermatol Venereol Leprol* 2000;66:264–5.
- 15 De S, Pfeleiderer AG. An extreme and unusual variant of Ramsay Hunt syndrome. *J Laryngol Otol* 1999;113:670–1.
- 16 Rothschild MA, Drake W, Scherl M. Cephalic zoster with laryngeal paralysis. *Ear Nose Throat J* 1994;73:850–2.
- 17 Golden LI, Deeb ZE, DeFries H. Atypical findings in cephalic herpes zoster polyneuritis: case reports and radiographic findings. *Laryngoscope* 1990;100:494–7.
- 18 Bhattacharya SK, Sunder S. Unusual cases of herpes zoster involving cranial nerves. *J Assoc Physicians India* 1984;32:535.
- 19 Wayman DM, Pham HN, Byl FM, *et al*. Audiological manifestations of Ramsay Hunt syndrome. *J Laryngol Otol* 1990;104:104–8.
- 20 Kim YH, Chang MY, Jung HH, *et al*. Prognosis of Ramsay Hunt syndrome presenting as cranial polyneuropathy. *Laryngoscope* 2010;120:2270–6.
- 21 Hunt J. The symptom-complex of the acute posterior poliomyelitis of the geniculate, auditory, glossopharyngeal and pneumogastric ganglia. *Arch Intern Med* 1910;5:631–75.
- 22 Baxter R, Ray P, Tran TN, *et al*. Long-term effectiveness of varicella vaccine: a 14-year, prospective cohort study. *Pediatrics* 2013;131:e1389–96.

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