The Journal of Laryngology & Otology
March 2005, Vol. **119,** pp. 184–188

An analysis of diagnostic delay in unilateral facial paralysis

A Alaani, FRCSEd, FRCSI, R Hogg, FRCS (ORL-HNS), N Saravanappa, FRCS,
R M Irving, MD FRCS (ORL-HNS)

Abstract

Bell's palsy or idiopathic facial palsy is the commonest cause of unilateral lower motor neuron facial palsy. Misdiagnosis of facial nerve palsy as Bell's palsy is still seen in clinical practice. The clinician should always consider the possibility of a potentially serious underlying pathology before making the diagnosis of Bell's palsy.

We present a series of 13 patients referred to our ENT department with an initial diagnosis of Bell's palsy. Further clinical examination and investigation revealed the underlying cause. Many had additional symptoms and signs related to the ear.

In all patients with unilateral facial palsy a detailed history should be taken and thorough clinical examination carried out. Where no recovery occurs within the expected time period further radiological investigations such as computerized axial tomography (CT) and magnetic resonance imaging (MRI) should be performed. Current scanning techniques provide good quality images, which can show occult lesions of the temporal bone, internal acoustic canal and/or cerebellopontine angle. Radiologists with a special interest and experience in otoneurological radiology should ideally report these images, and a close co-operation between ENT surgeon and radiologist is essential in arriving at a proper diagnosis.

Key Words: Facial Paralysis; Tomography, X-ray Computed; Magnetic Resonance Imaging

Introduction

The aetiology of unilateral facial palsy is diverse. Bell's palsy is the commonest cause of unilateral lower motor neuron facial palsy, accounting for up to 80 per cent of all cases of peripheral facial palsy.[1] The incidence of Bell's palsy in the general population is estimated to be around 10-30 cases per 100 000 per annum.[2,3] The other main causes of facial palsy include trauma, herpes zoster, neoplasm, infection and congenital anomalies.[4] Since the majority of cases of unilateral facial palsy are idiopathic, it is not uncommon to miss other underlying lesions. Therefore it is essential to exclude other causes of facial palsy before making a definitive diagnosis.[2,4,5] The accurate evaluation of facial palsy, with early recognition of signs and symptoms inconsistent with Bell's palsy, is crucial to avoid misdiagnosis of facial palsy.[5] Every patient with facial palsy should have a complete neurological examination,[6] and repeated assessments are required to detect signs that may not present initially.[3,6] Furthermore, a progressive facial palsy should always be considered to be of neoplastic origin until proven otherwise.[6] Before the advent of high-resolution imaging, tiny lesions of the facial nerve were difficult to detect. With magnetic resonance imaging (MRI) and high-resolution computerized tomography (CT), virtually all cases of facial nerve lesions can be identified. These are the most sensitive and complementary diagnostic tests.[7] While MRI can visualize the whole course of the facial nerve from the brainstem to the parotid gland, T1-weighted MRI with gadolinum is considered the imaging method of choice to diagnose neoplastic and inflammatory lesions of the facial nerve in the cerebellopontine angle and internal acoustic canal.[8] When temporal bone disease is associated with facial palsy, a high-resolution CT scan can provide the diagnosis.[9] This is considered the imaging method of choice for the intra-temporal portion of the facial nerve.[10]

Patients presenting with unilateral facial paralysis, erroneously labelled as Bell's palsy, frequently experience considerable delay between initial presentation and definitive diagnosis and treatment. Such patients could benefit from early surgical intervention that could improve the prognostic outcome of facial nerve function. We present our experience of treating 13 patients with unilateral facial palsy diagnosed initially as Bell's palsy. We have tried to analyse the reasons for diagnostic delay

From the Department of Otorhinolaryngology, University Hospital Birmingham/Queen Elizabeth Hospital, Birmingham, UK.
Presented at the 7th International Congress on Cholesteatoma and Ear Surgery, The Hague, June 2004.
Accepted for publication: 13 December 2005.

TABLE I

ADDITIONAL SYMPTOMS AT INITIAL PRESENTATION OF PATIENTS WITH FACIAL NERVE TUMOURS AND THEIR FINAL HISTOPATHOLOGICAL DIAGNOSIS

Patient	Initial presentation	Type of tumour
Case 1	None	Schwannoma of facial nerve
Case 2	Unilateral deafness	Schwannoma of facial nerve
Case 3	Unilateral tinnitus, facial spasm	Geniculate ganglion meningioma
Case 4	Vertigo	Geniculate ganglion haemangioma
Case 5	Unilateral tinnitus, facial twitching and spasm	Parotid adenocarcinoma

and suggest how this process can be improved. We would also like to emphasize the important role of careful clinical examination and imaging in such patients.

Materials and methods

This is a retrospective case-note review of patients with unilateral facial palsy who were initially erroneously diagnosed as having Bell's palsy and later found to have a different aetiology. Thirteen patients with lower motor neuron facial palsy referred to the Queen Elizabeth Medical Centre in Birmingham, during the period 1999–2003, with an initial diagnosis of Bell's palsy were studied. There were seven females and six males. The age of these patients on initial presentation with unilateral facial palsy ranged from 17 to 70 years. The majority of the patients underwent MRI and high-resolution CT scanning of the temporal bone.

Results

The duration of the delay between the initial presentation of the facial palsy and referral to our department was between six months and 25 years with a mean of nine years and median of seven years. At the time of first consultation at our department, on clinical examination, eight patients were found to have House-Brackman Grade VI facial palsy, and the remainder had Grade III facial palsy.

Analysis of the final diagnosis showed that five patients had a neoplastic lesion, four patients had a petrous apex cholesteatoma, three patients had a middle-ear cholesteatoma and one patient had an arachnoid cyst. The neoplastic lesions in this series were predominantly primary benign facial nerve tumours (Figure 1a and b). The associated symptoms are summarized in Tables I–III. Twelve patients had associated symptoms at initial presentation

TABLE II

ADDITIONAL SYMPTOMS AT INITIAL PRESENTATION OF PATIENTS WITH PETROUS APEX CHOLESTEATOMA

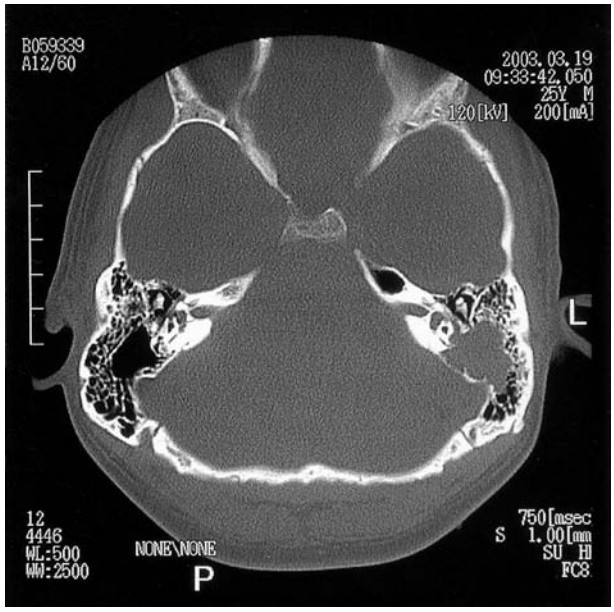
Patients	Additional symptoms
Case 6	Unilateral deafness, facial spasm
Case 7	Unilateral deafness, otorrhoea
Case 8	Unilateral deafness, otorrhoea
Case 9	Sudden unilateral hearing loss, three episodes of bacterial meningitis

TABLE III

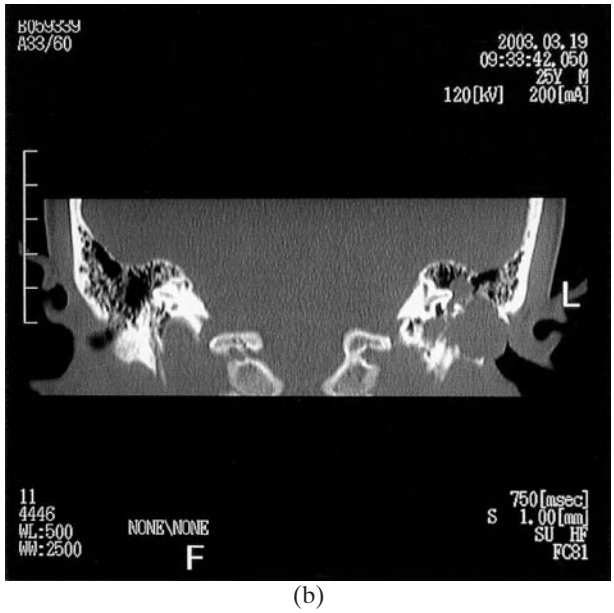
ADDITIONAL SYMPTOMS AT INITIAL PRESENTATION OF PATIENTS WITH MIDDLE-EAR CHOLESTEATOMA

Patients	Additional symptoms
Case 10	Unilateral deafness
Case 11	Unilateral deafness
Case 12	Bilateral hearing loss, history of bilateral chronic otitis media

suggesting a possible underlying cause for the facial palsy. Interestingly, a thorough history revealed that 12 patients had ear symptoms at the time of referral and two patients had a clear history of chronic suppurative otitis media. Ten patients had hearing loss at the time of presentation. Four patients had a history of facial spasm and twitching associated with the facial palsy. Two patients had previously



(a)

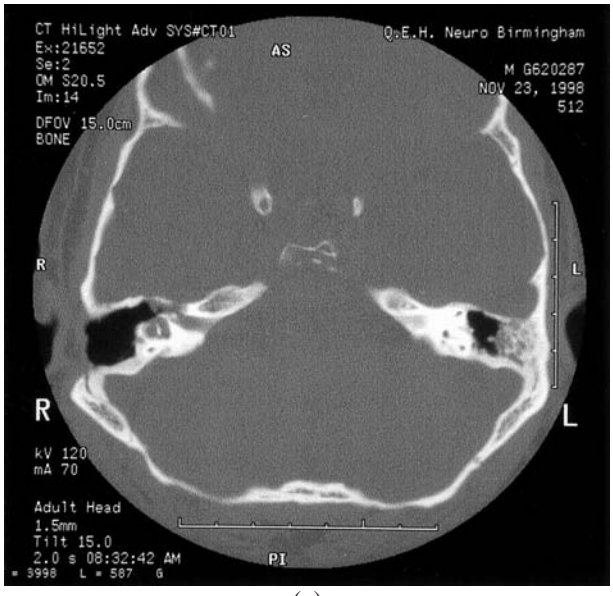


(b)

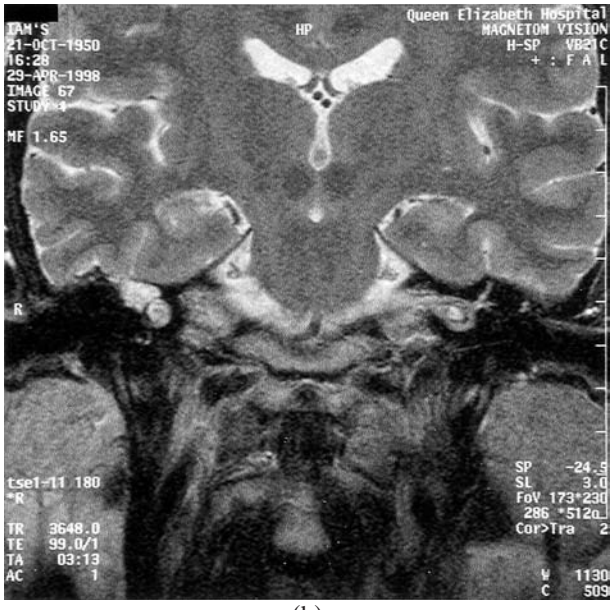
FIG. 1

Axial (a) and coronal (b) CT scans of the temporal bones showing a schwannoma of the left facial nerve involving the inner ear and the mastoid cavity.

https://doi.org/10.1258/0022215053561477 Published online by Cambridge University Press



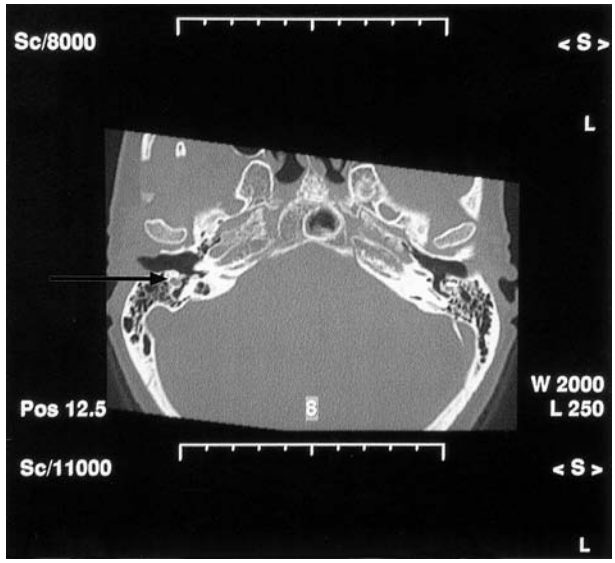
(a)



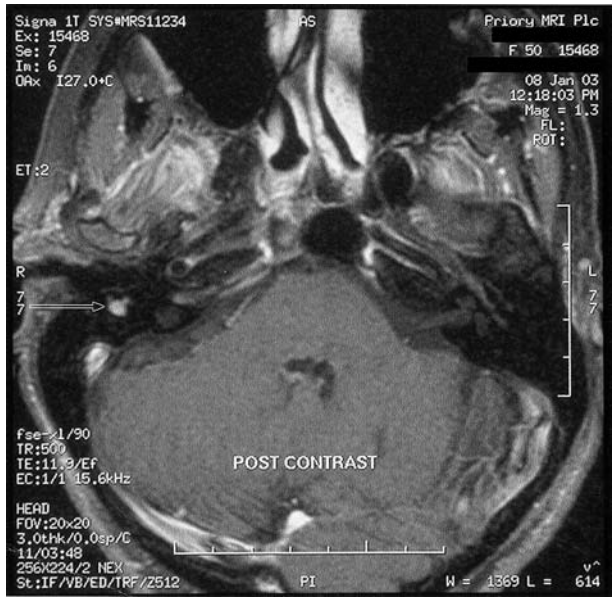
(b)

FIG. 2

(a) Axial CT scan of the petrous bone showing a right-sided residual cholesteatoma involving the inner ear and the petrous apex. (b) Coronal T2-weighted MRI scan of brain showing right-sided petrous apex cholesteatoma as a hyperintense lesion involving the right cochlea.



(a)



(b)

FIG. 3

(a) Axial CT scan of the temporal bone showing enlargement of the vertical part of the right facial nerve (arrow) due to perineural spread of a parotid adenocarcinoma, (b) Axial T1-weighted MRI scan of the brain demonstrating enlarged vertical part of the right facial nerve (arrow) secondary to parotid adenocarcinoma.

undergone mastoid surgery for cholesteatoma before the onset of facial palsy. CT (Figure 2a) and MRI (Figure 2b) scans showed residual cholesteatoma with extension to the inner ear several years after the mastoidectomy. Five patients had presented with a sudden onset of facial palsy and eight developed a progressive palsy over a period of a few weeks. Two patients gave a history of recurrent facial palsy, of which one developed the first episode at the age of nine. Three patients developed a VIth cranial nerve palsy. One patient had a history of multiple cranial nerve involvement including VI, X, XI and XII and was finally diagnosed with cholesteatoma involving the jugular foramen. One of the patients suffered with three episodes of meningitis and facial palsy before the diagnosis of petrous apex cholesteatoma was made. In two patients the aetiology of the facial nerve palsy was missed due to incorrect reporting of their MRI and CT scans. One patient had a tumour of the parotid gland which was missed on the CT (Figure 3a) and MRI (Figure 3b) scans. Another patient had cholesteatoma in the middle ear and mastoid that was not reported on the CT scan of the temporal bone. Three patients had incorrect imaging with CT or MRI which did not include the cerebellopontine angle or the parotid region.

Detailed analysis of our patients showed that these patients could be categorized into three groups. The first group comprised those in which the

https://doi.org/10.1258/0022215053561477 Published online by Cambridge University Press

initial consultation had failed to detect the additional clinical features inconsistent with Bell's palsy, and the diagnosis of idiopathic palsy was not questioned. In the second group the diagnosis of Bell's palsy was questioned but the underlying pathology was missed. In the third group the diagnosis was only questioned with the occurrence of further neurologic sequale.

Nine patients were investigated using both MRI and CT, one patient had only MRI, and the remaining patients had only CT. In our series, tumour of the facial nerve was the commonest pathology as it was found in five patients, followed by petrous apex cholesteatoma in four patients. Middle-ear cholesteatoma was diagnosed in three patients. Eleven patients had surgical treatment with or without facial nerve grafting. Two patients, one with a cerebellopontine angle arachnoid cyst and the other with a facial nerve schwannoma, were treated conservatively. The facial nerve function improved from grade VI to grade III in only three patients, all of whom had a middle-ear cholesteatoma. The grade of facial nerve palsy remained unchanged in all the others. In some of these cases, principally the apical cholesteatomas, we believe that earlier diagnosis would have resulted in earlier surgical intervention and a better ultimate outcome for facial nerve function.

- **Bell's palsy is the commonest cause of unilateral lower motor neuron injury to the facial nerve**
- **This paper presents 13 patients who were referred with an initial diagnosis of idiopathic palsy who were subsequently found to have alternative pathologies**
- **The paper highlights the need for CT and MRI, especially in patients presenting with hearing loss or other neurological signs or if facial nerve function has not recovered within three months**
- **In addition the authors recommend that close cooperation between neuro-otologist and radiologist is imperative if inaccurate reporting of the radiology is to be avoided**

Discussion

Many aetiological factors can cause unilateral facial palsy of which idiopathic facial palsy or Bell's palsy is the commonest. However, other possible causes of facial nerve palsy should be meticulously searched for before the diagnosis of Bell's palsy is made, which is essentially a diagnosis of exclusion.[3,4] Bell's palsy affects people of all ages, but most commonly individuals of 15–45 years old.[11]

Thirty years after the famous phrase of Sir Terence Cawthorne "All that Bell's is not Bell's", cases of misdiagnosis of facial palsy are still seen in clinical practice.[12] Our results show that over a period of four years 13 patients had been misdiagnosed as having Bell's palsy. Three main reasons were identified: the lack of clinical awareness of this problem, incorrect imaging and poor reporting. The experience of acute facial palsy is frightening for patients. In clinical practice a patient with acute facial palsy seeks urgent medical advice from their general practitioner or is seen at the accident and emergency department. The patient is usually labelled as Bell's even though there could be other underlying causes of facial palsy.[4] In a series of 2856 cases of acute facial palsy, only 51 per cent of the cases were due to Bell's palsy.[4] The reported incidence of misdiagnosis of unilateral facial palsy as Bell's palsy ranges between 13 and 20 per cent in all referred cases of peripheral palsy.[4,13] In one study of 1675 patients referred for acute facial palsy, 224 cases had a different aetiology.[4]

Careful history taking and examination are crucial. Early recognition of the signs and symptoms inconsistent with a diagnosis of Bell's palsy is important for prompt and appropriate referral.[4] Bell's palsy is acute unilateral facial palsy[4] which usually recovers in three months.[3] A history of recurrent facial palsy, other cranial nerve involvement, facial twitching, no recovery after three to six weeks or only partial recovery after three to six months, and slowly progressive palsy over more than three weeks indicate more serious aetiologies[6,9] and require referral to a specialist with expertise in managing such conditions. In this series eight out of 13 patients presented with progressive palsy and 12 had additional ear symptoms. Eight patients were missed because either they did not receive an appropriate specialist opinion or the clinical examinations were incomplete. Ear examination is important in all patients presenting with facial palsy.[9] Furthermore, incorrect imaging and poor reporting were the reasons behind the misdiagnosis in five cases.

Investigations may be needed to diagnose the aetiology of the facial palsy, usually pure-tone audiometry and imaging. The development of radiological investigations revolutionized the diagnostic process of facial nerve palsy. Patients with unilateral facial palsy may have an underlying pathology that is clinically silent and can only be detected by further MRI or CT scanning. Petrous apex cholesteatoma is a classical example, which accounted for one third of our series. There is no consensus opinion available with regard to the clinical stage of facial palsy at which CT and MRI scanning should be performed. It has previously been suggested that six months is a reasonable period to wait before performing any further investigations in patients with Bell's palsy.[6] An opportunity to detect and treat facial nerve lesions might be missed if there is a delay in performing the MRI scan for six months knowing that most cases of Bell's palsy recover in three months. In particular, the longer the nerve is paralysed, the less likely is its chance of full recovery. T1-weighted gadolinium images are the most useful in diagnosing neoplasia, inflammation and oedema of the facial nerve, as they are associated with an increased extracellular extravasation of the contrast.[8] The facial nerve within the parotid gland is best visualized by MRI.[11] High-resolution CT scanning of temporal bone

https://doi.org/10.1258/0022215053561477 Published online by Cambridge University Press

should be performed first in patients with a history suggesting ear disease and may rule out the need for an MRI scan in the presence of obvious ear pathology. High-resolution CT scanning has proven to be the most valuable tool in the diagnosis of intratemporal facial nerve tumour, where it indicates the anatomical correlation of the tumour with the facial canal,[14] and permits a detailed and accurate evaluation of the temporal bone and related bony structures.[10] High-resolution CT scanning and enhanced MRI are the two most sensitive methods available for the diagnosis of facial nerve tumours including haemangiomas.[7,15,16] Thin-sectioned CT and MRI scans of the facial nerve from the brain stem to the terminal branches may be required to diagnose small facial nerve tumours.[7] The advantage of MRI is its high sensitivity for detecting small lesions. MRI is the better method for evaluating the brain stem/cerebellopontine angle segment of the facial nerve, whereas high-resolution CT is particularly useful in the evaluation of the intratemporal segment of the facial nerve and assessment of the anatomic perspectives of a lesion within the temporal bone.[10,14] MRI should be performed before a CT scan in the evaluation of facial nerve dysfunction as it demonstrates all tumours in the internal acoustic canal and some in the geniculate ganglion.[17] However, when there are symptoms suggesting ear pathology CT is the radiological investigation of first choice. A radiologist with a special interest and experience in otoneurological imaging would help in identifying the abnormality and reduce the incidence of false reporting. A coordinated approach between the otolaryngologist and the radiologist is needed to perform a tailored radiographic study to localize the pathology.[18] All these measures are essential for making an early, accurate diagnosis and for appropriate, prompt surgical management.

Conclusion

In all cases of unilateral facial palsy other aetiologies must be meticulously searched for before labelling it as idiopathic facial palsy. All patients with unilateral facial palsy should be referred to an otorhinolaryngology department for evaluation and further follow up. Detailed clinical history and a thorough clinical examination including ENT evaluation are essential to differentiate Bell's palsy from other causes of facial palsy.

We believe that radiological investigations should be performed in all patients presenting with facial palsy in association with hearing loss or other neurological signs. If the cause of facial palsy is not suspected as idiopathic, or if there is no recovery of facial nerve function within three months, appropriate imaging studies should be carried out, including an MRI scan with gadolinium injection covering the whole course of the facial nerve from the brain stem to parotid gland. In some cases, it may be necessary to perform a high-resolution CT scan of the temporal bone. It is important to ensure that an expert radiologist with a special interest in ENT/neuroradiology reports such scans. This line of approach will help in making an appropriate diagnosis and a prompt surgical intervention that, in turn, will improve the outcome for facial nerve function in some cases.

References

1 Bleicher JN, Hamiel BS, Gengler JS. A survey of facial paralysis: etiology and incidence. *Ear Nose Throat J* 1996;**75**:355-8
2 Petruzzelli G, Hirsch B. Bell's palsy: a diagnosis of exclusion. *Postgrad Med* 1991;**90**:115-23
3 Peitersen E. Bell's palsy: the spontaneous course of 2,500 peripheral nerve palsies of different etiologies. *Acta Otolaryngol* 2002;**549**:4-30
4 May M, Klein S. Differential diagnosis of facial nerve palsy. *Otolaryngol Clin North Am* 1991;**24**:613-45
5 Brach JS, Van Swearingen JM. Not all the facial paralysis is Bell's palsy: case report. *Arch Phys Med Rehabil* 1999;**80**:857-9
6 Jackson CG, Glassocock ME, Hughes G, Sismanis A. Facial paralysis of neoplastic origin: diagnosis and management. *Laryngoscope* 1980;**90**:1581-95
7 Salib RJ, Tziambazis E, McDermott A, Chavda SV, Irving RM. The crucial role of imaging in detection of facial nerve haemangiomas. *J Laryngol Otol* 2001;**115**:510-3
8 Yetiser S, Kazkayas M, Altinok D, Karadeniz Y. Magnetic resonance imaging of the intratemporal facial nerve in idiopathic peripheral facial palsy. *Clin Imaging* 2003;**27**:77-81
9 Ruckenstein MJ. Evaluating facial paralysis, expensive diagnostic tests are often unnecessary. *Postgrad Med* 1998;**103**:187-92
10 Schwaber MK, Zealear D, Netterville JL, Seshul M, Ossoff RH. The use of magnetic resonance imaging with high-resolution CT in the evaluation of facial paralysis. *Otolaryngol Head Neck Surg* 1989;**101**:449-58
11 Peitersen E. Natural history of Bell's palsy. *Am J Otol* 1982;**4**:107-11
12 Cawthorne T. Intratemporal facial palsy. *Arch Otolaryngol* 1969;**90**:789-99
13 Jungehuelsing M, Sittel C, Fischbach R, Wagner M, Stennert E. Limitations of magnetic resonance imaging in the evaluation of perineural tumour spread causing facial nerve paralysis. *Arch Otolaryngol Head Neck Surg* 2000;**126**:506-10
14 Ito H, Hoshino T, Okawa Y, Asai Y, Nozue M, Yokoyama T, *et al*. Pre-operative diagnosis of intratemporal facial nerve tumour: high-resolution CT and otoscopic findings in four cases. *Auris Nasus Larynx* 1996;**23**:75-82
15 Weber AL, McKenna MJ. Radiological evaluation of the facial nerve. *Isr J Med Sci* 1992;**28**:186-92
16 Shelton C, Brackmann DE, Lo WW, Carberry JN. Intratemporal facial nerve haemangiomas. *Otolaryngol Head Neck Surg* 1991;**104**:116-21
17 Lo WW, Shelton C, Waluch V, Solti-Bohman LG, Carberry JN, Brackmann DE, *et al*. Intratemporal vascular tumours:detection with CT and MR imaging. *Radiology* 1989;**171**:445-8
18 Leslie PA, Zinreich SJ. Facial nerve imaging. *Otolaryngol Clin North Am* 1991;**24**:571-85

Address for correspondence:
Mr Richard M Irving,
Consultant ENT surgeon,
Department of Otolaryngology,
University Hospital Birmingham/Queen Elizabeth Hospital,
Birmingham B15 2TH, UK.

Fax: +44 121 6272299
E-mail: rmi@globalnet.co.uk

Mr R.M. Irving takes responsibility for the integrity of the content of the paper.
Competing interests: None declared