The Journal of Laryngology & Otology (2012), **126**, *833–836*.

doi:10.1017/S002221511200120X

CLINICAL RECORD

Differential diagnosis of recurrent or bilateral peripheral facial palsy

D M OOSTERVEER[1], C P BÉNIT[2], E L L M DE SCHRYVER[3]

[1]*Sophia Rehabilitation Centre, Delft,* [2]*Department of Neurology, Westeinde Hospital, The Hague,* [3]*Department of Neurology, Rijnland Hospital, Leiderdorp, The Netherlands*

Abstract

Objective: To describe the differential diagnosis of recurrent or bilateral peripheral facial palsy.

Method: Case report and literature review.

Results: Two patients with recurrent, alternating, peripheral facial palsy are described. In both patients, additional investigation was performed to search for a specific diagnosis. In the first patient, only a positive family history was found, indicating a possible familial susceptibility. In the other patient, diabetes mellitus and hypertension were identified as risk factors.

Conclusion: There is an important and extensive differential diagnosis of recurrent or bilateral facial palsy. However, in a large proportion of patients the cause remains unknown.

Key words: Facial Palsy; Lower Motor Neuron Disease; Peripheral Neuropathy; Genetics

Introduction

Bell's palsy is peripheral facial palsy of unknown cause. It is relatively common, with an estimated incidence of 13–34 cases per 100 000.[1,2]

Patients with Bell's palsy typically present with a unilateral facial paresis which develops within hours. Examination shows eyebrow sagging, inability to close one eye, disappearance of one of the nasolabial folds, and mouth asymmetry. The onset is generally acute, with maximum facial palsy within three or four days. In 85 per cent of patients, facial nerve function returns within three weeks.[3] Recurrence rates of 7–11 per cent have been reported.[4–7]

Simultaneous, bilateral facial palsy is much rarer than unilateral palsy, with an incidence of 0.3–2 per cent.[8]

When a peripheral facial palsy recurs or affects both sides of the face, an idiopathic cause is less likely.

We present two cases of recurrent and (at times) bilateral facial palsy, and we summarise the differential diagnoses of a recurrent or bilateral peripheral facial palsy.

To prepare this summary, we searched PubMed for 'Bell's palsy', 'peripheral' OR 'bilateral' OR 'recurrent' AND ('facial palsy' OR 'facial palsy', 'facial paresis').

Case reports

Case one

A 76-year-old woman suddenly developed a left facial palsy, with inability to close her left eye firmly, asymmetry of the mouth and disappearance of the nasolabial fold. Her medical history included hypothyroidism, for which she took levothyroxine 0.05 mg daily. The patient's mother had previously suffered Bell's palsy.

We diagnosed Bell's palsy and prescribed the patient prednisone 25 mg twice daily for 10 consecutive days.[9]

However, two months later she returned to our out-patient clinic because of deterioration of the Bell's palsy: her facial palsy was now complete and she could not move her left facial musculature.

Blood and cerebrospinal fluid (CSF) examination results (including angiotensin-converting enzyme and lysozyme testing) were normal. Tests for *Treponema pallidum* and *Borrelia burgdorferi* in both serum and CSF were negative, as was viral polymerase chain reaction testing of the CSF. Magnetic resonance imaging (MRI) of the brain showed several white matter lesions but gave no explanation for the facial palsy. A chest X-ray was normal.

A few weeks later, the patient could close her left eye again and use her mouth adequately, but a mild paresis was still seen.

Less than three months later, she returned to our out-patient clinic because of salvia loss from the right corner of her mouth. Examination identified loss of the right nasolabial fold without evident weakness of the upper facial muscles. Another MRI scan was performed due to suspicion of a central facial paresis, but this scan was identical to the previous one suggesting a partial peripheral facial palsy. Blood and CSF examinations were again normal.

Thus, despite extensive additional investigation, we could find no underlying cause of this patient's alternating, recurrent facial palsy, other than a possible familial susceptibility.

Case two

The second patient was a 44-year-old man who had previously suffered a left-sided Bell's palsy at the age of 35 years, with complete recovery. He visited our out-patient

Accepted for publication 3 November 2011

clinic with a four-week history of right-sided peripheral facial palsy, which had been preceded by an upper airway infection. Recently, he had been diagnosed with diabetes mellitus and hypertension, and had started a diet to control his diabetes.

A brain MRI scan showed hyper-intensity of the labyrinthine segment of the right facial nerve and of the right geniculate ganglion, typical of Bell's palsy. A chest X-ray was normal.

Three weeks later, the patient also developed a left facial palsy, with no other neurological deficits.

At this stage, blood examination demonstrated an elevated erythrocyte sedimentation rate (29 mm/hour) and a slightly elevated γ globulin test (59 U/l), without any other abnormalities (the angiotensin-converting enzyme concentration was normal). The CSF had a high protein content (839 mg/l) and a slightly elevated glucose concentration (3.9 mmol/l), with a normal number of leukocytes. Microscopic examination of the CSF showed a cell-rich liquor with many lymphocytes and some monocytes. However, viral polymerase chain reaction and *T pallidum* and *B burgdorferi* testing of both CSF and serum were all negative.

Four months later, the patient's left-sided Bell's palsy was observed to have almost completely recovered. On the right side, there was some improvement: he could close his right eye again, but there was still diminished blinking and marginal movement of the mouth.

The underlying cause of this patient's bilateral facial palsy was never found, although diabetes mellitus and hypertension were identified as risk factors.

Discussion

The two patients described above illustrate the difficulty of identifying an underlying cause for recurrent and bilateral facial palsies. An idiopathic aetiology is still possible: Navarrette *et al.*[10] reported that 84 per cent (77/92) of their recurrent facial palsy cases remained idiopathic, while Keane[11] reported that 23 per cent (10/43) of their bilateral facial palsy cases remained idiopathic. However, efforts should be undertaken to identify a (treatable) cause in each patient with recurrent or bilateral peripheral facial palsy. Table I shows the differential diagnosis of recurrent or bilateral facial palsy.

As shown in Table I, there are several viral and bacterial agents which can cause a bilateral or recurrent facial palsy. It is hypothesised that Bell's palsy is caused by infection with herpes simplex virus (HSV). The best proof of this is given by Murakami *et al.*,[56] who found HSV DNA in the endoneural fluid surrounding the facial nerve, obtained during surgical decompression in patients with Bell's palsy. Endoneural fluid cannot be obtained routinely, but we did screen both our patients for several viral agents (including HSV in the CSF) and also for *T pallidum* and *B burgdorferi* in both serum and CSF. However, in both patients all such tests were negative.

Guillain–Barré syndrome is an important post-infectious cause of facial palsy. However, patients with this syndrome typically present with bilateral facial palsy after an infection, generally accompanied by weakness of the extremities and absence of tendon reflexes.

Another post-infectious cause of bilateral facial palsy is malaria infection, although we found only one report of this.[57]

TABLE I
CAUSES OF RECURRENT OR BILATERAL PERIPHERAL FACIAL PARESIS

Category	Cause
Trauma	Bilateral temporal bone fracture[12]
	Mandibular fracture[13]
	Forceps delivery[14]
	High voltage electrical injury[15]
Infectious	Bacterial[†]
	Viral[‡]
	Parasitic (*Trichinella spiralis*)[16]
Systemic, autoimmune or unknown	Guillain–Barré syndrome[11,17]
	Sarcoidosis (Heerefordt syndrome)[11,17]
	Amyloidosis[17,18]
	Wegener's granulomatosis[19]
	Kawasaki disease[20]
	Systemic lupus erythematosus[11]
	Sjögren syndrome[21]
	Scleroderma[22]
	Periarteritis nodosa[23]
	Myasthenia gravis[24]
	Multiple sclerosis, acute disseminated encephalomyelitis[25]
	Idiopathic benign intracranial hypertension[11]
	Behçet's disease[26]
Idiopathic	Bell's palsy[11,27]
Neoplastic	Tumours or (leptomeningeal) metastasis[11,17,28]
	Middle-ear adenoma[29]
	Facial cyst[30] or neurinoma[31]
	Fibrous dysplasia of temporal bone[32]
	Temporal bone haemangioma[33]
Toxic	Vincristine[34]
	Ethylene glycol[35]
	Paclitaxel[36]
	Linezolid[37]
Iatrogenic	External carotid artery embolisation[38]
Metabolic	Diabetes mellitus[11,39]
	Wernicke–Korsakoff syndrome[40]
	Acute porphyria[41]
Genetic or congenital	Familial[42,43]
	Osteopetrosis[44]
	Möbius syndrome[11,17]
	Melkersson–Rosenthal syndrome[17]
	Kennedy's disease[17]
	Amyotrophic lateral sclerosis[45]
Other	Hypertension[12]
	Pregnancy[17,46]
	Hymenoptera sting[47]

Note that muscle disorders and causes located in the central nervous system (e.g. pontine haemorrhage or infarction)[11,48] can mimic bilateral peripheral facial palsy. [†]*Borrelia burgdorferi*,[17] *Treponema pallidum*,[11,17,49] *Mycobacterium tuberculosis*,[11] *Mycobacterium leprae*,[11] *Mycoplasma pneumoniae*,[50] *Leptospira interrogans*,[51] *Clostridium tetani*[52] and bacterial otitis caused by other bacteria.[11,26] [‡]Epstein– Barré virus,[17,28] herpes simplex virus,[53] human immunodeficiency virus,[11,17] varicella zoster virus,[28] rubella virus[54] and poliovirus.[55]

In the Introduction, we describe the typical course of Bell's palsy. An atypical course, as seen in our first patient, could indicate a less another cause. This patient's facial palsy was slowly progressive, which raises suspicion of a tumour. In addition, the involvement of only one of the distal branches of the facial nerve suggested local compression.[58–61] However, the brain MRI did not show any signs of tumour or leptomeningeal metastasis. In case two, an MRI scan was performed three weeks before development of bilateral facial palsy. This MRI demonstrated enhancement

https://doi.org/10.1017/S002221511200120X Published online by Cambridge University Press

of the labyrinthine segment of the facial nerve, typical of Bell's palsy.[62]

- **Recurrent or bilateral facial palsy has an extensive differential diagnosis**
- **Searching for causes reduces the number of idiopathic (Bell's palsy) diagnoses**
- **No cause is found for 84 per cent of recurrent facial palsy cases**
- **No cause is found for 23 per cent of bilateral facial palsy cases**

A notable aspect of case one was the presence of a positive family history of Bell's palsy. Previous reports have mentioned the possibility of genetic susceptibility, which has been demonstrated in 2.4–28.6 per cent of cases.[63] However, no consistent pattern of inheritance has been found.[42,64] Human leukocyte antigens were hypothesised to be involved; however, studies have shown ambiguous results.[43,65–68] Cawthorne and Haynes[5] suggested that this familial tendency was due to extremely cellular mastoids; they noted that all patients with recurrent Bell's palsy had extensive, cellular mastoids.

Diabetes mellitus and hypertension have been mentioned as risk factors for Bell's palsy, and were also found in our second case.[68] Pitts *et al.*[4] have stated that patients with recurrent facial palsy are 2.5 times more likely to have diabetes, probably because diabetic patients are more prone to nerve degeneration. Hypertension also increases the risk, but the association is less strong. It is hypothesised that hypertension causes facial paresis through direct pressure from a dilated vessel, oedema or haemorrhage within the facial canal.[69]

Conclusion

When a patient has a recurrent or bilateral facial palsy, diagnoses other than Bell's palsy should be considered. In addition to extensive anamnestic data collection, investigation should include blood examinations and contrast-enhanced MRI of the brain. If the diagnosis still remains unclear after such investigation, a chest X-ray should be performed to search for systemic disease, and the CSF should be examined to diagnose infection and exclude leptomeningeal metastasis. Anamnestic data may prompt further investigation.

Unfortunately however, no specific cause can be found for a large number of recurrent or bilateral facial palsy cases, as exemplified by our presented patients.[10,11] In the future, expanding clinical knowledge and diagnostic possibilities may enable the identification of a specific cause in more of these patients.

References

1 Peitersen E. The natural history of Bell's palsy. *Am J Otol* 1982; **4**:107–11
2 Hauser WA, Karnes WE, Annis J, Kurland LT. Incidence and prognosis of Bell's palsy in the population of Rochester, Minnesota. *Mayo Clin Proc* 1971;**46**:258–64
3 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
4 Pitts DB, Adour KK, Hilsinger RL Jr. Recurrent Bell's palsy: analysis of 140 patients. *Laryngoscope* 1988;**98**:535–40
5 Cawthorne T, Haynes DR. Facial palsy. *Br Med J* 1956;**2**: 1197–2000
6 Hallmo P, Elverland HH, Mair IWS. Recurrent facial palsy. *Arch Otorhinolaryngol* 1983;**237**:97–102
7 van Amstel AD, Devriese PP. Clinical experiences with recurrences of Bell's palsy. *Arch Otorhinolaryngol* 1988;**245**:302–6
8 Sherwen PJ. Bilateral facial nerve palsy: a case study and literature review. *J Otolaryngol* 1987;**16**:28–33
9 Salinas RA, Alvarez G, Daly F, Ferreira J. Corticosteroids for Bell's palsy (idiopathic facial paralysis). *Cochrane Database Syst Rev* 2010;**3**:CD001942
10 Navarrete ML, Cespedes R, Mesa M, Grasa J, Perez M, Raguer N *et al*. Recurrent Bell's facial palsy: our experience [in Spanish]. *Acta Otorrinolaringol Esp* 2001;**52**:682–6
11 Keane JR. Bilateral seventh nerve palsy: analysis of 43 cases and review of the literature. *Neurology* 1994;**44**:1198–202
12 Li J, Goldberg G, Munin MC, Wagner A, Zafonte R. Post-traumatic bilateral facial palsy: a case report and literature review. *Brain Inj* 2004;**18**:315–20
13 Kumar S, Gupta R. Simulaneous bilateral facial palsy as a result of isolated mandibular fractures. *Int J Oral Maxillofac Surg* 2006;**35**:1156–9
14 Edgeworth FH. Case of bilateral facial paralysis, due to injury by forceps at birth. *Br Med J* 1894;**1**:11
15 Vasquez JC, Shusterman EM, Hansbrough JF. Bilateral facial nerve paralysis after high voltage electrical injury. *J Burn Care Rehabil* 1999;**20**:307–8
16 Lopez-Lozano JJ, Merino JAG, Liano H. Bilateral facial paralysis secondary to trichinosis. *Acta Neurol Scand* 1988;**78**:194–7
17 Ropper AH, Samuels MA. *Adams and Victor's Principles of Neurology*, 9th edn. Boston: McGraw-Hill, 2009
18 Massey EW, Massey JM. Facial diplegia due to amyloidosis. *South Med J* 1986;**79**:1458–9
19 Nikolaou AC, Vlachtsis KC, Daniilidis MA, Petridis DG, Daniilidis IC. Wegener's granulomatosis presenting with bilateral facial nerve palsy. *Eur Arch Otorhinolaryngol* 2001;**258**: 198–202
20 Lim TC, Yeo WS, Loke KY, Quek SC. Bilateral facial nerve palsy in Kawasaki disease. *Ann Acad Med Singapore* 2009;**38**: 737–8
21 Uchigara T, Yoshida S, Tsukagoshi H. Bilateral facial paresis with Sjögren's syndrome. *J Neurol* 1989;**236**:186
22 Taesdall RD, Frayha RA, Shulman LE. Cranial nerve involvement in systemic sclerosis (scleroderma): a report of 10 cases. *Medicine (Baltimore)* 1980;**59**:149–59
23 Dudley JP, Goodman M. Periarteritis nodosa and bilateral facial paralysis. *Arch Orolaryngol* 1969;**90**:139–46
24 Cucurachi L, Cattaneo L, Gemignani F, Pavesi G. Late onset generalized myasthenia gravis presenting with facial weakness and bulbar signs without extraocular muscle involvement. *Neurol Sci* 2009;Epub ahead of print (PMID:19484183)
25 Smith V, Traquina DN. Pediatric bilateral facial paralysis. *Laryngoscope* 1998;**108**:519–23
26 Menassa J, Sawaya R, Masri AF, Arayssi T. Recurrent peripheral facial paresis may constitute the sole clinical manifestation in neuro-Behcet disease. *Neurologist* 2008;**14**:77
27 Kim YH, Choi IJ, Kim HM, Ban JH, Cho CH, Ahn JH. Bilateral simultaneous facial nerve palsy: clinical analysis in seven cases. *Otol Neurotol* 2008;**29**:397–400
28 Ramsey KL, Kaseff LG. Role of magnetic resonance imaging in the diagnosis of bilateral facial paralysis. *Am J Otol* 1993;**14**: 605–9
29 Mori E, Kojima H, Wada K, Moriyama H. Middle ear adenoma diagnosed by recurrent facial paralysis. *Auris Nasus Larynx* 2009;**36**:75–8
30 Michalopoulos K, Bajaj Y, Strachan DR. Recurrent facial nerve palsy caused by a facial cyst. *Br J Hosp Med (Lond)* 2008;**69**:475
31 Scholz E, Langer J, Begall K. Recurrent facial paresis with facial neurinoma [in German]. *Laryngorhinootologie* 2007;**86**:443–7
32 Zaytoun GM, Dagher WI, Rameh CE. Recurrent facial nerve paralysis: an unusual presentation of fibrous dysplasia of the temporal bone. *Eur Arch Otorhinolaryngol* 2008;**265**:255–9
33 Eby TL, Fisch U, Makek MS. Facial nerve management in temporal bone hemangiomas. *Am J Otol* 1992;**13**:223–32
34 Sarkar S, Deb AR, Saha K, Das CS. Simultaneous isolated bilateral facial palsy: a rare vincristine-associated toxicity. *Indian J Med Sci* 2009;**63**:355–8
35 Mallya KB, Mendis T, Guberman A. Bilateral facial paralysis following ethylene glycol ingestion. *Can J Neurol Sci* 1986; **13**:340–1

https://doi.org/10.1017/S002221511200120X Published online by Cambridge University Press

36 Lee RT, Oster MW, Balmaceda C, Hasdorffer CS, Vahdat LT, Papadopoulos KP. Bilateral facial nerve palsy secondary to the administration of high-dose paclitaxel. *Ann Oncol* 1999;**10**: 1245–7
37 Thai XC, Bruno-Murtha LA. Bell's palsy associated with linezolid therapy: case report and review of neuropathic adverse events. *Pharmacotherapy* 2006;**26**:1183–9
38 Metson R, Hanson DG. Bilateral facial nerve paralysis following arterial embolization for epistaxis. *Otolaryngol Head Neck Surg* 1983;**91**:299–303
39 Hattori T, Schlagenhauff RE. Bilateral facial palsy: occurrence with diabetes mellitus. *N Y State J Med* 1977;**77**:1492–3
40 Rice JP, Horowitz M, Chin D. Wernicke-Korsakoff syndrome with bilateral facial nerve palsies. *J Neurol Neurosurg Psychiatry* 1984;**47**:1356–7
41 Lewis M, Kallenbach J, Hockman M, Zaltzman M, Zwi S. Otolaryngological complications of acute porphyria. *Laryngoscope* 1983;**93**:483–4
42 Clement WA, White A. Idiopathic familial facial nerve paralysis. *J Laryngol Otol* 2000;**114**:132–4
43 Döner F, Kutluhan S. Familial idiopathic facial palsy. *Eur Arch Otorhinolaryngol* 2000;**257**:117–19
44 Kulkarni GB, Pal PK, Shyambabu C, Kovoor JM, Senthilkumar E. Osteopetrosis manifesting as recurrent bilateral facial palsy in childhood: a case report. *Clin Neurol Neurosurg* 2011;**113**: 230–4
45 Salameh JS, Atassi N, David WS. SOD1 (A4V)-mediated ALS presenting with lower motor neuron facial diplegia and unilateral vocal cord paralysis. *Muscle Nerve* 2009;**40**:880–2
46 Walling AD. Bell's palsy in pregnancy and the puerperium. *J Fam Pract* 1993;**36**:559–63
47 Raucq E, Dupuis MJ. Facial diplegia after hymenoptera sting. *Acta Neurol Belg* 1998;**98**:215–20
48 Roh JK, Kim BK, Chung JM. Combined peripheral facial and abducens nerve palsy caused by caudal tegmental pontine infarction. *Eur Neurol* 1999;**41**:99–102
49 Strauss MJ. Diplegia facialis in early syphilis: a report of a case. *Arch Dermatol Syph* 1929;**20**:306–14
50 Ernster JA. Bilateral facial nerve paralysis associated with *Mycoplasma pneumoniae* infection. *Ear Nose Throat J* 1984; **63**:585–9
51 Silva AA, Ducroquet M, Pedrozo JC Jr. Bilateral facial palsy associated with leptospirosis. *Braz J Infect Dis* 2009;**13**: 319–21
52 Brown AJ. Cephalic tetanus with report of a case. *Ann Surg* 1912;**55**:473–84
53 Santos DQ, Adour KK. Bilateral facial paralysis related to sexually transmitted herpes simplex: clinical course and MRI findings. *Otolaryngol Head Neck Surg* 1993;**108**:298–303
54 Jamal GA, Al-Husaini A. Bell's palsy and infection with rubella virus. *J Neurol Neurosurg Psychiatry* 1983;**46**:678–80
55 Sherman IC, Kimelblot SJ. Facial paralysis in poliomyelitis: report of 3 patients with unusual delayed paralysis. *Neurology* 1959;**9**:282–7
56 Murakami S, Mizobuchi M, Nakashiro Y, Doi T, Hato N, Yanagihara N. Bell palsy and herpes simplex virus: identification of viral DNA in endoneurial fluid and muscle. *Ann Intern Med* 1996;**124**:27–30
57 Kochar DK, Sirohi P, Kochar SK, Bindal D, Kochar A, Jhajharia A *et al*. Post-malaria neurological syndrome – a case of bilateral facial palsy after *Plasmodium vivax* malaria. *J Vector Borne Disease* 2007;**44**:227–9
58 Boahene DO, Olsen KD, Driscoll C, Lewis JE, McDonald TJ. Facial nerve paralysis secondary to occult malignant neoplasms. *Otolaryngol Head Neck Surg* 2004;**130**:459–65
59 Catalano PJ, Sen C, Biller HF. Cranial neuropathy secondary to perineural spread of cutaneous malignancies. *Am J Otol* 1995; **16**:772–7
60 Breadon GE, Cody DT, Weiland LH. Facial palsy: unusual etiology. *Laryngoscope* 1977;**87**:26–34
61 May M, Hughes GB. Facial nerve disorders: update 1987. *Am J Otol* 1987;**8**:167–80
62 Schwaber MK, Larson TC, Zealear DL, Creasy J. Gadolinium-enhanced magnetic resonance imaging in Bell's palsy. *Laryngoscope* 1990;**100**:1264–9
63 Auerbach SH, Depiero TJ, Mejlszenkier J. Familial recurrent peripheral facial palsy: observations of the pediatric population. *Arch Neurol* 1981;**38**:463–4
64 Qin D, Ouyang Z, Luo W. Familial recurrent Bell's palsy. *Neurology India* 2009;**57**:783–4
65 Smith CI, Hammarström L, Sidén A. No significant association between HLA and Bell's palsy. *Tissue Antigen* 1978;**12**:404–6
66 Shibahara T, Okamura H, Yanagihara N. Human leukocyte antigens in Bell's palsy. *Ann Otol Rhinol Laryngol Suppl* 1988;**137**: 11–13
67 Schwartz MS, Tiwari JL, Rice DH. Bell's palsy and HLA-DR: a possible association. *Arch Otolaryngol Head Neck Surg* 1986; **112**:753–4
68 Savadi-Oskouei D, Abedi A, Sadeghi-Bazargani H. Independent role of hypertension in Bell's palsy: a case-control study. *Eur Neurol* 2008;**605**:253–7
69 Harms MM, Rotteveel JJ, Kar NC, Gabreëls FJ. Recurrent alternating facial paralysis and malignant hypertension. *Neuropediatrics* 2000;**31**:318–20

Address for correspondence:
Dr D M Oosterveer,
Sophia Rehabilitation Centre,
Reinier de Graafweg 1,
2625 AD Delft, The Netherlands

Fax: +31 15 27 80 219
E-mail: dmoosterveer@hotmail.com

Dr D M Oosterveer takes responsibility for the integrity of the content of the paper
Competing interests: None declared