Bell’s Palsy: Aetiology, Classification, Differential Diagnosis and Treatment Consideration: A Review


Abstract

Bell’s palsy (spontaneous idiopathic facial paralysis), which is the most common facial nerve disease, has a sudden onset. Bell’s palsy is the most common cause of facial paralysis, accounting for 70 per cent of facial palsies, when other causes have been eliminated. It has an incidence of 11–40 per 100,000 per year, and most commonly occurs in females in their teens and twenties. The distribution is almost equal in the thirties, with a slight predominance in males over 40. The annual incidence of Bell’s palsy in the Western world is approximately 20/1 00 000. Untreated Bell’s palsy leaves some patients with major facial dysfunction and a reduced quality of life. It is essential to rule out other causes of facial paralysis before making definitive diagnosis, which implies the intervention. Bell’s palsy has been termed a diagnosis of exclusion. This review emphasizes the etiology, diagnosis and management of patients with Bell’s palsy.


Key words: Bell’s palsy, facial paralysis, facial dysfunction.

Introduction

Facial nerve palsy results in the loss of facial expression and is most commonly caused by a benign self-limiting inflammatory condition known as Bell’s palsy (BP). BP is characterized by an acute onset of facial nerve palsy with no known cause. The incidence is about 20/year/100,000 population, and leads to a considerable disturbance in social activities among patients. Although the actual cause of BP is unknown, the widely accepted mechanism is inflammation of the facial nerve during its course through the bony labyrinthine part of the facial canal, which leads to compression and demyelination of the axons, and disruption of blood supply to the nerve itself.

BP is defined as a lower motor neuron palsy of acute onset and idiopathic origin. BP is regarded as a benign common neurological disorder of unknown cause. It has an acute onset and is almost always a mononeuritis. The facial nerve is

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a mixed cranial nerve with a predominant motor component which supplies all muscles concerned with unilateral facial expression. Knowledge of its course is vital for anatomic localization and clinical correlation. BP accounts for around 72% of facial palsies. Scottish surgeon anatomist Sir Charles Bell (1774-1842) described this as a syndrome of complete facial paralysis in a lecture ‘On the nerves: giving an account of some experiments on their structure and functions, which lead to a new arrangement of the system’ to the Royal Society of London in 1821. Almost a century later, the management and aetiology of BP is still a subject of controversy.

History

It was first reported by Nicolas A Friedrich two century ago in 1798. Sir Charles Bell originally described this condition in 1821. The term BP is used to describe an acute-onset, idiopathic facial paralysis resulting from a dysfunction anywhere along the peripheral part of the facial nerve from the level of the pons distally.

Epidemiology

The disease is common, with an annual incidence is reported in Table 1.

<table>
<thead>
<tr>
<th>Year</th>
<th>Annual incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1974-82</td>
<td>13-34/100,000</td>
</tr>
<tr>
<td>1998-2006</td>
<td>20-30/100,000</td>
</tr>
<tr>
<td>2001</td>
<td>20/100,000</td>
</tr>
</tbody>
</table>

Etiology

Since BP is a facial paralysis of unknown origin, it is essential to rule out other causes of facial paralysis before making the definitive diagnosis, which implies the intervention. BP has been termed a diagnosis of exclusion.

The causes include microcirculatory failure of the vasonervorum, viral infection, ischemic neuropathy, autoimmune reactions, surgical procedure such as local anesthesia, tooth extraction, infections, osteotomies, preprosthetic procedures, excision of tumors or cysts, surgery of TMJ, and surgical treatment of facial fractures and cleft lip/palate. A viral cause has been widely accepted, but no virus has been consistently isolated in patients with BP. The evidence for the viral hypothesis has been based primarily on clinical observation and changes in viral antibody titers. The pathogenesis of the paralysis may be a viral neuropathy alone or ischemic neuropathy caused by a viral infection. Although acute facial paralysis can occur during many viral illnesses such as mumps, rubella, herpes simplex, and Epstein-Barr virus infection or as a result of the reactivation of the human herpes virus in the geniculate ganglia.

Some patients may be more easily predisposed to facial nerve inflammation by exposure to a preceding pathogen, such as Herpes simplex virus, Epstein-Barr virus and cytomegalovirus. There have been an increasing number of reports on the Herpes simplex virus particle found on facial nerve biopsy in patients with BP. Facial nerve paralysis may be central or peripheral in origin, complete or incomplete. Its cause is varied and included trauma, tumor formation, iatrogenic problems, idiopathic conditions, cerebral infarct, pseudobulbar palsy and viruses. It results in a characteristic facial distortion that is determined in part by the nerves branches involvement.

The literature also reports three mechanisms, in which a dental procedure could damage a nervous structure: direct trauma to nerve from a needle, intraneural hematoma formation or compression and local anesthetic toxicity. Direct trauma seems unlikely since many patients report experiencing trauma to the nerve when they feel the electric shock sensation on injection of the needle. However, virtually all these symptoms resolve completely with no residual nerve damage. Besides common conditions like hypertension and diabetes mellitus, which may predispose to single or multiple attacks.

The familial cases of recurrent ipsilateral and alternating contralateral facial nerve palsy have both autosomal dominant and recessive inheritance. This genetic predisposition may also include variations in the immune response of each individual towards the inciting antigen.

Clinical features

There are three symptoms commonly noted by the patient in addition to the facial palsy. Epiphora
due to lack of tone in the lower eyelid and consequent failure of the punctum to make contact with the globe of the eye is often present. Pain is a frequent complaint, and may be in the ear, spread more widely over the head, down the neck or into the eye. It is usually present for a few days and may precede the palsy for up to 72 hours; but occasionally it comes on several days after the palsy and may be severe and persistent. Tenderness over the stylomastoid foramen may be present.\textsuperscript{38}

The other symptoms of BP include pain and numbness on the affected side of the face, especially in the temple, mastoid area, and along the angle of the mandible.\textsuperscript{39} The mouth may be dry due to decreased salivary secretion and altered taste sensation over the anterior two-thirds of the tongue and incomplete hyperaesthesia over the trigeminal nerve distribution as well as hyperacusis on the affected side.\textsuperscript{6, 40}

**Classification**

BP has been classified into the following 5 categories according to the clinical course of disease: unilateral non-recurrent, unilateral recurrent, simultaneous bilateral, alternating bilateral or recurrent bilateral.\textsuperscript{41}

**Differential Diagnosis**

The gradual onset and duration of the facial paralysis with associated facial pain are also consistent with a space-occupying lesion.\textsuperscript{42} In addition to cases with BP, minimal facial nerve thickening with contrast attenuation are possible to be observed also in cases of Guillain Bare Syndrome, postoperatively, traumatic facial paralysis and following radiotherapy. Unilateral central facial weakness (lower face muscles) may be due to a lesion of the contralateral cortex, subcortical white matter, or internal capsule. In addition to facial weakness, symptoms may include hemiparesis, hemisensory loss, or hemineglect (severe impairment of spatial perception).

Lyne neuroborreliosis- The spirochete *Borrelia burgdorferi* can affect central nervous system tissues. Lyme neuroborreliosis should be suspected in a patient who presents with isolated facial weakness and who has a history of tick bite with rash or who lives in an area where Lyme disease is endemic. Tumors involving the facial nerve account for fewer than 5% of all cases of facial nerve paralysis. A tumor should be suspected if weakness progress over weeks, if a mass is present in the ear, neck, or parotid gland, and if no functional improvement is seen within 4 to 6 weeks.\textsuperscript{42}

**BP associated with other disease**

Literature reviewed that “peripheral facial palsy (PFP)”, “chickenpox” and “varicella-zoster virus” to identify case reports of PFP associated with varicella published from 1980 to January 2008. From 1989 to date, 10 patients with bilateral Bell palsy associated with acute HIV-1 infection have been described in the literature (Table II).\textsuperscript{43-52}

<table>
<thead>
<tr>
<th>Year</th>
<th>Case patient (Ref.)</th>
<th>Age/ Sex</th>
<th>CD4+ cell count cells/mm(^3)</th>
<th>CD8+ cell count cells/mm(^3)</th>
<th>Outcome, weeks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1989</td>
<td>1 (43)</td>
<td>45 (M)</td>
<td>770</td>
<td>1550</td>
<td>Recovery (23)</td>
</tr>
<tr>
<td>1989</td>
<td>2 (44)</td>
<td>19 (F)</td>
<td>776</td>
<td>958</td>
<td>Recovery (3)</td>
</tr>
<tr>
<td>1990</td>
<td>3 (45)</td>
<td>40 (F)</td>
<td>N/R</td>
<td>1542</td>
<td>(3)</td>
</tr>
<tr>
<td>1993</td>
<td>4 (46)</td>
<td>32 (M)</td>
<td>718</td>
<td>2393</td>
<td>N/R</td>
</tr>
<tr>
<td>1995</td>
<td>5 (47)</td>
<td>21 (M)</td>
<td>N/R</td>
<td>N/R</td>
<td>(8)</td>
</tr>
<tr>
<td>2000</td>
<td>6 (48)</td>
<td>43 (M)</td>
<td>404</td>
<td>N/R</td>
<td>N/R</td>
</tr>
<tr>
<td>2002</td>
<td>7 (49)</td>
<td>37 (M)</td>
<td>533</td>
<td>1134</td>
<td>N/R</td>
</tr>
<tr>
<td>2002</td>
<td>8 (50)</td>
<td>73 (M)</td>
<td>513</td>
<td>2563</td>
<td>Persistent Paresia</td>
</tr>
<tr>
<td>2003</td>
<td>9 (51)</td>
<td>29 (F)</td>
<td>N/D</td>
<td>N/D</td>
<td>N/R</td>
</tr>
<tr>
<td>2006</td>
<td>10 (52)</td>
<td>26 (M)</td>
<td>327</td>
<td>N/R</td>
<td>Recovery (N/R)</td>
</tr>
</tbody>
</table>

N/D- Not done  N/R- Not reported

**Diagnosis**

The proper history and physical examination provide the key to the diagnosis of BP. Most patients do not require any laboratory testing. However, patients who have persistent weakness
without significant improvement requires further investigations.\textsuperscript{53}

1. Imaging: Computed tomography (CT) or MRI is indicated in cases of, no improvement in facial paresis even after 1 month, hearing loss, multiple cranial nerve deficits and signs of limb paresis or sensory loss.

2. Hearing testing - If hearing loss is suspected, then audiologic testing can be to rule out acoustic neuroma.

3. Laboratory testing is necessary if the patient has signs of systemic involvement without significant improvement over more than 4 weeks. A number of tests may be helpful.

Complete blood count with differential helps to rule out lymphoreticular malignancy, the first manifestation of which may be peripheral facial palsy\textsuperscript{54}. Blood glucose should be measured if diabetes mellitus is suspected. Serum antibodies against herpes zoster and \textit{B burgdorferi} (the agent of Lyme disease) can be checked if the patient has signs such as vesicular lesions on the external ear or lives in an area where Lyme disease is endemic. Serum calcium and angiotensin-converting enzyme levels should be tested if sarcoidosis is suspected; these levels are high in sarcoidosis.

Cerebrospinal fluid testing is helpful if infection or malignancy is suspected; however, in case of BP cerebrospinal fluid tends to show mild and inconsistently elevated cell counts and protein levels.

Electrodiagnostic testing is not routinely done in BP. It is not very reliable when BP is in the initial stages; however, after 2 weeks, it may detect denervation and demonstrate nerve regeneration.\textsuperscript{55}

**Management**

Treatment of BP is still controversial.\textsuperscript{55} Clinical and electrophysiological assessment should be done. Clinical evaluation for both the severity of paralysis and the presence of complication is the first step before the start of treatment or rehabilitation. The most popular method for assessing the severity or paralysis is the facial nerve grading system according to House and Brackmann (Table III).\textsuperscript{56}

For persons with persistent paralysis of the facial nerve, treatment modalities such as steroid therapy and surgical nerve decompression have been prescribed.\textsuperscript{57} Most authors agree that 75% of BP cases regress spontaneously with complete recovery. Approximately 15% of the cases have satisfactory recovery with a slightly detectable neurological deficit and 10% of the cases have permanent paralysis.\textsuperscript{58} A good prognosis is associated with BP seen in children.\textsuperscript{59} Corticosteroids are the most commonly used agents for reducing inflammation and edema in the nerve sheath.\textsuperscript{60}

**Table III: Facial nerve grading system**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Percent of Function</th>
<th>Degree of Function</th>
<th>Measurement</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>100</td>
<td>-</td>
<td>8/8</td>
</tr>
<tr>
<td>II</td>
<td>76-99</td>
<td>Slight</td>
<td>7/8</td>
</tr>
<tr>
<td>III</td>
<td>51-75</td>
<td>Moderate</td>
<td>5/8-6/8</td>
</tr>
<tr>
<td>IV</td>
<td>26-50</td>
<td>Moderately Severe</td>
<td>3/8-4/8</td>
</tr>
<tr>
<td>V</td>
<td>1-25</td>
<td>Severe</td>
<td>1/8-2/8</td>
</tr>
<tr>
<td>VI</td>
<td>0</td>
<td>Total Paralysis</td>
<td>0/8</td>
</tr>
</tbody>
</table>

*A centimeter is divided into four equal parts. On the affected side of the face, maximal voluntary lateral movement of the corner of the mouth is measured 0-4, and elevation of the eyebrow is measured 0-4, resulting in a sum from 0/8 to 8/8.

The intake of corticosteroids alone or in combination with anti viral drugs, improve the prognosis of BP (i.e. induces rapid and complete recovery in most of the patients) through preventing (or minimizing) axonal degeneration of nerve fibres. CS prevent or lessen nerve oedema and swelling in the facial bony canal; antiviral drug suppress viral replication in the neural tissue, thus they may protect the facial nerve from severe damage.\textsuperscript{61}

The route and extent of decompression for recurrent facial palsy is also controversial. Both transmastoid subtotal decompression and combined transmastoid-middle cranial fossa total decompression approach have been advocated.\textsuperscript{62} Early surgery including surgical decompression of the facial nerve may be recommended within 2 weeks following the onset of BP, if electroneurography revealed >90% degeneration.
of facial nerve fibres. On other hand, some may not recommended such surgical intervention in BP.63

The commonly used tests include: the maximum nerve excitability test, the maximum stimulation test (MST), electroneurography (ENoG). The MST and ENoG are the most reliable in predicting prognosis (and assessing the extent of facial nerve degeneration) if done 7-10 days after the onset of paralysis. Transcranial magnetic resolution is still inferior to the above mentioned techniques. The facial nerve conduction velocity may be done when side to side comparison is not possible as in bilateral facial palsy. The blink reflex is done mainly to exclude the lesion at the pons or medulla. Electromyography (EMG) of the facial muscles determines sign of denervation and/or reinnervation as well as the degree of recruitment of motor units.63

Physical treatment: It has also been recommended to use local superficial heat therapy (i.e hot packs or infrared rays) for 15min/session for the facial muscles prior to electrical stimulation. Massage which has been frequently been prescribes for facial palsy, improves circulation and may prevent contracture.64 Short wave diathermy (SWD) has been suggested as treatment of BP however, some may not recommend its use in BP because there is acute viral inflammation of the facial nerve in its early stage67 and heating of inflamed nerve may be contraindicated.65 Acupuncture66 and magnets67 have been used in combination with physiotherapy in the management of facial palsy, but their specific efficacy needs further investigation.

Treatment of hyperbaric oxygen, through the inhalation of 100% oxygen under high pressure (at pressure 2.8 times greater than normal atmosphere), should possibly be considered as one of the physical modalities. It was reported that this modality has induced better recovery than prednisone treatment in BP patients.68

Apart from above treatment eye and facial muscles protection is necessary. For eye protection, early treatment including use of artifical tears, night time eye patch ophthalmic ointment before sleep and eyeglasses are usually used to protect the light, dust and wind while in long term treatment, ophthalmic consultation is necessary for possible surgical interference, if there is a failure of spontaneous eye closure.44,57,58

Facial muscle protection from injury may be achieved by use of porous adhesive tape to prevent deviation of mouth to the healthy side during smiling. In case of management of facial hyperkinesis, when surgery is not indicated, local injection with botulinum toxin-A seems to be the most appropriate therapy.63 They should be performed while standing in front of a mirror and include trying to raise the eyebrows, opening and closing the eyes, blowing, and whistling. These exercises can be performed a few times daily. The efficacy of exercise has not been formally evaluated.69

Prognosis:

The prognosis depends to a great extent on the time at which recovery begins. If recovery begins within one week, 88% obtain full recovery, within one to two weeks 83% and within two to three weeks 61%. Early recovery gives a good prognosis and late recovery a bad prognosis.70

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