Understanding Bell's Palsy – a review

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Abstract

Bell's palsy is a common neuropathic cranial condition causing acute unilateral lower motor neuron facial paralysis. Immune, ischaemic and infective mechanisms are the potential contributors for the development of Bell's palsy, but the exact cause remains unclear. It affects 11.5-53.3 per 100,000 individuals a year across different populations. Over 70-75% of patients make a full recovery. Recently published management modalities for the acute treatment of Bell's palsy advocate for steroid monotherapy. For those with longstanding episodes from incomplete recovery, aesthetic, functional (nasal patency, eye closure, speech and swallowing) and psychological considerations need to be addressed by the treating team. A patient centred holistic approach utilising physiotherapy, targeted botulinum toxin injection and selective surgical intervention has reduced the burden of long-term disability in facial palsy. Hence this review deals with aetiology signs and symptoms, diagnosis and treatment management for Bell's palsy.

INTRODUCTION

Bell's palsy (BP), named after the Scottish anatomist Sir Charles Bell, is the most commonly sought diagnosis linked to facial nerveparalysis as well as the most encountered acute mono-neuropathy. It affects individuals various age groups and both sexes, with an annual incidence ranging from 11.5 to 53.3 per 100,000 persons across multiple populations. Typically, BP results in partial or complete inability to automatically move the affected side of the facial muscles[1]. Although it usually resolves within few days or months, BP facial paralysis may lead to severe temporary oral insufficiency and incapability to close the eyelids, which may result in permanent eye injury[2]. In approximately 25% of patients with BP, facial asymmetry of moderate to severe intensity may persist, frequently impairing patients' quality of life. These are among BP's long-term adverse consequences, which can be a psychological and functional devastation for patients. Despite its severe effects, the exact aetiology of BP remains unclear. The Guideline Development Group (GDG) has put forth that the diagnosis of BP as one of exclusion, requiring careful clinical elimination of other potential aetiologies of facial paralysis, such as trauma, neoplasms, congenital or syndromic problems, postsurgical facial paralysis, or infection by agents including zoster and Lyme disease. This diagnosis also fails to address cases of recurrent facial paralysis. The GDG has also recognized the "acute" or "rapid onset" nature of BP and that the occurrence of paralysis usually reaches its maximum severity in less than 72 h of paralysis onset.There are five major theories regarding the aetiology of BP including anatomical, viral infection, ischemia, inflammation, and cold stimulation.[1]

Anatomy

The human facial nerve is the seventh cranial nerve (CNVII) and comprises of three components- motor, sensory and parasympathetic. Its function is responsible for voluntary and mimetic facial movement, taste to the anterior two-thirds of the tongue, and control of secretions of salivary gland and lacrimal gland. The facial nerve receives axons from the superior part of the solitary nucleus and superior salivary nucleus that form the nervus intermedius component (sensory and parasympathetic axons) and motor efferent fibres from the facial nucleus, which receives synaptic input from the contralateral motor cortex for all facial movements except the forehead, which has bicortical input. The path of the facial nerve has intracranial, infratemporal and extratemporal components. Its intracranial course runs from the pontomedullary angle to the internal acoustic meatus where it is accompanied by the vestibulocochlear nerve (CNVIII). The infratemporal course of the facial nerve is long and tortuous. During its infratemporal course, the nerve comprises the geniculate ganglion and gives rise to the superior petrosal nerve, the nerve to stapedius and chorda tympani nerve branches, and exits the skull base through the styloid foramen. The extratemporal facial nerve courses through the substance of parotid gland dividing it into deep and superficial lobes. It gives off the posterior auricular nerve and nerve to the posterior belly of digastric before dividing into its main terminal facial branches. There is significant variation in the branching pattern of the terminal facial branches, which are traditionally conceptualised into 5 branches-temporal, zygomatic, buccal, marginal mandibular and cervical branches. These terminal motor branches are responsible for all facial expression and functional tasks such as eye and mouth closure and nasal patency during inspiration. Throughout its course, the facial nerve forms multiple communications between its own branches and with adjacent cranial nerves.[2]

Aetiology

There are five major theories regarding the causes of BP including[1]

- Anatomical
- Viral infection
- Ischemia
- Inflammation
- Cold stimulation

Anatomical

The facial nerve (CN VII) is a unique motor nerve, emerging from the facial nerve nucleus in the pons. From this point, it is accompanied by the CN VIII along its cisternal pathway to the internal auditory meatus. Specifically, its petrous route includes a labyrinthine segment, a horizontal tympanic segment, and a vertical mastoid segment, which extends until it reaches the stylomastoid foramen and theparotid gland. CN VII then follows a long-drawn-out pathway through the temporal bone within the fallopian canal. Given this extended and convoluted pathway, it is more susceptible to palsy than other nerves in the body.[1]

Viral infections

Another possible aetiology of BP that has been suggested is infection by reactivated viruses, such as the varicella zoster virus (VZV), herpes simplex virus type 1 (HSV-1), human herpes virus [6], and the Usutu virus. Herpesviruses (HV) are large, enveloped viruses with doublestranded linear DNA. A-HV infections, which target peripheral neurons (e.g., HSV-1, HSV-2, and VZV), are among the most common viral infections worldwide. HSV and VZV infections can persist across the host's lifespan. A-HVs enter the human body via the mucosa and establish their latent presence in multiple ganglia of the neuroaxis by highly restricted gene transcription for the host's entire life, including in the autonomic and sensory ganglia of the head, neck, and cranium. When there is no active viral replication or assembly, this latent form in the ganglia is characteristic and widely distributed throughout diseased and normal populations. Both HSV and VZV can reactivate in the presence of circulating antibodies or in an immunocompetent host, while reactivation is more likely in cases of immunodeficiency.[1]

Ischemia

BP, an acute idiopathic lower motor neuron palsy, is commonly a unilateral and self-limiting inflammatory condition. Most cases of BP remit within 4-6 months and almost always resolve completely within 1 year. Among those cases that do not resolve, studies have implicated secondary ischemia, tertiary ischemia, or their sequelae, and this in turn, can result in thickening of the facial nerve sheath, forming one or more fibrous bands that cause nerve strangulation and compression, thereby hampering recovery.[1]

Inflammation

Numerous studies have suggested that BP results from acute, inflammation-caused demyelination. The evidence supporting this aetiology is supported by histologic changes in the facial nerve, first identified by Liston and Kleid, the characteristics of which are summarized as follows.[1]

1. The nerve, from the internal acoustic meatus to the stylomastoid foramen, is infiltrated by round, small inflammatory cells.
2. A breakdown of neuron myelin sheaths, which involves macrophagesoccurs.
3. Interneuronal space is increased.
4. The bony fallopian canal is normal, with no sign of facial nerve compression by the fallopian canal bone.

Cold stimulation

There are multifactorial possible risk factors for BP, including severe preeclampsia, psychological factors, glucose metabolism abnormalities, radiation exposure, hypertension, and migraine. Recently, epidemiological studies have revealed that the incidence of BP is also related to extreme temperature exposure. Campbell and Brundage investigated climate and season correlates of BP risk using a centralized surveillance system containing medical encounter and demographic data. The results revealed that both season and climate were independent predictors of BP risk. There is a clear correlation between the cold season and the number of cases observed. A previous study also found that BP occurs more frequently during the spring and summer, with its incidence peaking in September and a significantly higher incidence of BP in the summer.[1`]

Symptoms

Symptoms of Bell's palsy vary from individuals and range in severity from mild weakness to total paralysis. The symptoms include twitching, weakness, or paralysis on one or both sides of the face, drooping of the eyelid and corner of the mouth, headache, drooling, dryness of the eye or mouth, impairment of taste, and excessive tearing in one eye. However, these symptoms usually begin suddenly and reach their peak within 48 hours. Other symptoms may include pain or discomfort around the jaw and behind the ear, ringing in one or both ears, headache, loss of taste, an increased sensitivity to sound on the affected side, impaired speech, dizziness, and difficulty eating or drinking. Adour et al stated that the anoxia of the nerve, caused by a primary or secondary ischemia, followed by compensatory dilatation of the blood vessels supplying the nerve, is part of the process causing the occurrence of ear pain.[3]

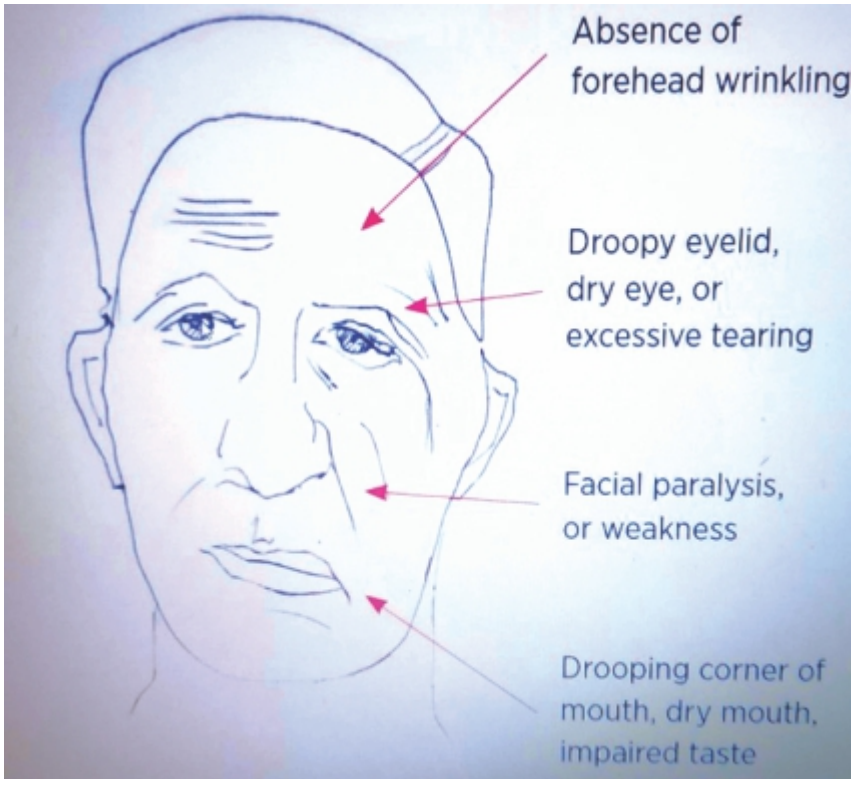
- upper motor neurone lesion – based on innervation, absence of forehead wrinkling is a reliable way of differentiating Bell's palsy from an upper motor neuron lesion
- herpes zoster oticus (Ramsay Hunt syndrome)
- rarer causes including otitis media, HIV infection, sarcoidosis, autoimmune disorders or tumours of the parotid gland.

In addition to ocular problems, complications of Bell's palsy include:

- motor synkinesis (involuntary movement of muscles occurring at the same time as deliberate movement, e.g. involuntary mouth movement during voluntary eye closure)
- crocodile tears (tears when eating due to misdirection of regenerating gustatory fibres destined for the salivary glands, so that they become secretory fibres to the lacrimal gland and cause ipsilateral tearing while the patient is eating)
- incomplete recovery
- contracture of facial muscles
- reduction or loss of taste sensation
- problems with dysarthria due to facial muscle weakness.[3]

Prognosis

The severity of symptoms of Bell's palsy varies from mild weakness to severe paralysis, but



the prognosis is generally good. The Copenhagen Facial Nerve Study found that around 71% of patients recover normal function without treatment. Around 13% are left with slight weakness and around 4% with severe weakness resulting in major facial dysfunction. Contracture of the facial muscles on the affected side was found in 17% and associated movements were found in 16%.[6] Scoring systems such as the House-Brackmann scale used in randomised controlled trials and systematic reviews may be helpful to monitor progress.[7] Although the study was underpowered to detect significant differences in recovery between patients with different degrees of severity, the recovery rate in one randomised controlled trial was significantly higher for those with moderate severity at onset compared to those with severe Bell's palsy. Recovery was 90% with those moderately affected and 78% in those severely affected.8 The frequency of review depends on the individual patient and the severity of their symptoms. If there is no improvement after a month the patient should be referred. A referral is also indicated if there is only partial recovery after 6-9 months. The palsy recurs in 7% of patients, with equal incidence of ipsilateral and contralateral recurrence. There are insufficient data on whether treatment affects the rate of recurrence.[3]

Differential diagnosis

The differential diagnosis for facial palsy is broad and misdiagnosis is not uncommon. Causes of facial palsy may be divided into congenital and acquired aetiologies. Congenital causes include genetic syndromes, birth-related trauma and isolated disorders of development (eg, developmental hypoplasia of facial muscles). Acquired causes include infective (VZV, Lyme disease, mycobacterium tuberculosis, HIV), traumatic (iatrogenic or head trauma), inflammatory (vasculitis, sarcoidosis, autoimmune disease), neoplastic (benign or malignant) and cerebrovascular causes, among others. In the experience of one of the authors (GC) in an expert referral setting, the rate of misdiagnosis of Bell's palsy by the initial consulting clinician is 10.8%. Missed diagnoses include tumours (eg, facial nerve schwannoma, parotid malignancy and, rarely, acoustic neuroma), herpes zoster oticus and granulomatous diseases such as sarcoidosis and granulomatosis with polyangiitis (Wegener's granulomatosis). A structured clinical approach that considers the pattern of facial palsy along with patient characteristics and a thorough physical examination will generally provide evidence for an alternative diagnosis, and prompt appropriate investigation. Particular patterns of facial palsy that require thoughtful consideration include:[4]

1. Fluctuant, step-wise or slowly progressive (beyond 72 h).
2. Bilateral palsy (GBS, carcinomatosis, lymphoma).
3. Recurrent facial palsy (facial nerve neuroma).
4. Prolonged complete palsy (>4 months).
5. Sudden complete facial palsy (haemorrhage into a tumour).

These patterns should prompt a detailed search for an underlying cause. Likewise, the presence of a mass in the parotid region, a history of cutaneous malignancy or segmental facial nerve weakness should raise suspicion for a tumour. A history of trauma, ear symptoms such as ipsilateral deafness, tinnitus, fullness or discharge, or systemic symptoms such as fever, are also red flags warranting further investigation and specialist otological consultation. The consideration of cerebrovascular disease as a cause of facial palsy is important with this being the main concern for many patients and clinicians, often prompting expert neurology consultation. The preservation of upper facial movement (frontalis contraction) is a discriminator between cortical (central) and peripheral facial nerve weakness. Rarely, ipsilateral pontine pathology may result in lower motor neuron pattern facial weakness due to direct compromise of the facial nucleus. This will be accompanied by other cranial nerve, and long tract symptoms and signs. Ipsilateral abducens nerve (CNVI) dysfunction (lateral gaze palsy) is a particularly useful sign.[4]

TREATMENT

The treatment of Bell's palsy aims to speed recovery and reduce long-term complications. An inability to close the eye on the affected side increases the risk of corneal complications. Eye protection is crucial so an eye patch and lubricants are used to prevent drying of the cornea. Eye drops, such as hypromellose drops, should be applied for lubrication during the day and ointment at night. In severe cases, the eye may have to be taped or partially sutured shut.[3]

Drug therapy

The treatments considered for Bell's palsy include oral corticosteroids (prednisolone) and antiviral drugs. Although the aetiology of Bell's palsy is uncertain, it is known that inflammation and oedema of the facial nerve are responsible for the symptoms. Corticosteroids have therefore been used for their anti-inflammatory effect.[3]

Corticosteroids

The maximum benefit is seen when steroids are commenced within 72 hours of the onset of symptoms. There is no optimum regimen, but in adults 50-60 mg prednisolone daily for 10 days has been commonly used. Prednisolone has been used at a dose of 1 mg/kg/day up to a maximum of 80 mg in some studies. Doses of more than 120 mg/day have been used safely in patients with diabetes. There was also a significant reduction in motor synkinesis in those who received prednisolone. There was no significant reduction in cosmetically disabling sequelae.[3]

Antiviral drugs

The antiviral drugs used in trials were acyclovir (400mg five times daily for five days) or valaciclovir (1000 mg/day for five days). There is currently no evidence to support the use of either antiviral drug on its own and there is uncertainty regarding the benefit of adding them to corticosteroids.[3]

Combination therapy

There have been several studies looking at the benefit of antiviral drugs with or without prednisolone. A randomised prospective study found that a combination of an antiviral and a steroid was more effective in treating severe to complete Bell's palsy than steroid alone. A GDG found that there was low-quality evidence of benefit from adding antivirals. Patients who are offered them in addition to corticosteroids should be counselled that the increase in recovery is less than 7%. A Cochrane review in 2015 found that antivirals combined with corticosteroids improved rates of incomplete recovery compared with corticosteroids alone, but this was not significant and the evidence was low quality. None of the treatments had significant differences in adverse effects, but the evidence was again of low quality. The best management of bells BP children with Bell's palsy is also unknown. A major trial (BellPIC) in Australia is addressing this question.[3]

Adverse effects of treatment

Treatment courses are short, but can cause adverse effects. Prednisolone should be used with caution in immuno suppression and sepsis. It may lead to:

- induction or worsening of peptic ulcer disease
- hyperglycaemia especially in diabetics, however higher doses may be required in diabetes
- malignant hypertension
- hepatic and renal dysfunction

Antiviral drugs may cause:

- nausea and vomiting
- abdominal pain
- diarrhoea
- neurological reactions - dizziness, convulsions (more common with higher doses)
- very rarely, hepatitis and jaundice[3].

Non-drug therapy

Physical therapies including tailored facial exercises, acupuncture to affected muscles, massage, thermotherapy and electrical stimulation have been used to hasten recovery. However, there is no evidence for any signifi-

cant benefit. A Cochrane review concluded from poor-quality evidence that tailored facial exercises can help improve facial function, mainly for moderate paralysis and chronic cases. Early facial exercise may reduce recovery time, long-term paralysis and number of chronic cases.18 Surgical treatment to free the facial nerve has been considered. However the evidence for this procedure is of very low quality.[3,4]

CONCLUSION

Bell's palsy is an acute benign cranial polyneuritis and is currently considered as the leading disorder affecting the cranial nerves. Most patients with Bell's palsy regain normal function with or without medical therapy, often within 3 weeks. In some cases, full recovery takes up to 9 months, but up to 30% are left with potentially disfiguring facial weakness, involuntary movements, and/or persistent lacrimation. A delay in the diagnosis and administration of medications could play a role in residual weakness of the face and mouth. Hence this review will help in understanding the aetiology, early detection methodologies, complications and treatment strategies of Bell's palsy so as to protect from recurrence.

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