Familial and Functional Facial Palsy: Case Series and Clinical Approach

Ali Raheem Hashim
Professor of Medicine and Neurology – Department of Medicine
College of Medicine – University of Basrah, Basrah, Iraq
Ali.hashim@uobasrah.edu.iq

Haithem Jawad Kadhum
Lecturer of Physiology – Department of Physiology
College of Medicine – University of Basrah, Basrah, Iraq
Haithem.kadhum@uobasrah.edu.iq

Lana Ahmed Kadhim
Neurophysiology Specialist – Department of Neurophysiology
Al-Sadr Teaching Hospital – Basrah Health Directorate, Basrah, Iraq
Ibrahim10553@gmail.com

Hassan Ala Farid
Neurology Resident and PG Student – Department of Medicine
College of Medicine – University of Basrah, Basrah, Iraq
Hassan.farid@uobasrah.edu.iq

Nareen Haikaz Hasrat
Neurophysiology Resident and PG Student – Department of Physiology
College of Medicine – University of Basrah, Basrah, Iraq
Nareen.haikaz@bjes.edu.iq

Abstract

Bell's palsy is a common clinical syndrome of uncertain etiology. Familial clustering has long been noted in the literature. In addition, patients with conversion disorder may be presented with features that mimic facial play. This article reported two cases of familial bell' palsy from Basrah city in the south of Iraq in which more than one member of their families complains of the same condition, as well as one bizarre functional facial palsy case. Furthermore, the clinical approach and therapeutic options for patients with Bell's palsy are also discussed.

Keywords: Bell's palsy, Functional Disorders, Basrah, Family history, Genetics

I. INTRODUCTION

1.1 BASIC ANATOMY OF FACIAL NERVE

The facial nerve is the seventh cranial nerve and, although predominantly motor, also serves an important parasympathetic and sensory function. It exits the brainstem ventrally via the internal acoustic meatus in the petrous part of the temporal bone near the pontomedullary junction (Elan et al. 2022).

1.2 FACIAL NERVE LESIONS

Lesions near the origin of the nerve, or in the vicinity of the geniculate ganglion, are accompanied by a loss of motor, gustatory, and autonomic functions, while lesions between the geniculate ganglion and the origin of the chorda tympani typically spare lacrimation, Furthermore, lesions near the stylomastoid

foramen spare taste and lacrimation, causing only ipsilateral facial paralysis of the upper and lower face. Moreover, lesions of the facial nerve nucleus in the brainstem also cause ipsilateral paralysis of all facial muscles, both upper and lower. These types of lesions are of the lower motor neuron type (Elan et al. 2022).

The pattern of peripheral or nuclear injury (the peripheral seventh nerve lesion) must be distinguished from that associated with central motor pathway lesions above the level of the nucleus. Central facial weakness, or supranuclear palsy, causes weakness and paralysis in the lower half of the face while sparing forehead wrinkles because of the redundancy of central pathways subserving upper-face muscles (Elan et al. 2022).

1.3 CAUSES OF FACIAL PALSY

Long lists of causes are implied in the causality of facial plays. The intracranial course of the nerve can be affected by diseases such as cerebral tumours, multiple sclerosis, and strokes, as well as diabetes mellitus and connective tissue disease. Furthermore, its extracerebral course may be affected in the temporal bone region by Ramsay Hunt syndrome, while in the middle ear region, cholesteatoma and mastoiditis can also lead to facial palsy. Additionally, in the parotid region, its course can be affected by malignancy or trauma to the facial nerve by surgery. Other diseases can also lead to facial palsy, such as Lyme disease and sarcoidosis, in addition to myasthenia gravis due to muscle weakness, and in some cases, no cause can be identified, which is formerly known as idiopathic facial palsy or bell palsy. Familial bell palsy is also reported worldwide (Kubik et al. 2012). Finally, patients suffering from conversion disorder or malingering may also present with facial palsy (Zandian et al. 2014).

II. CASES PRESENTATION

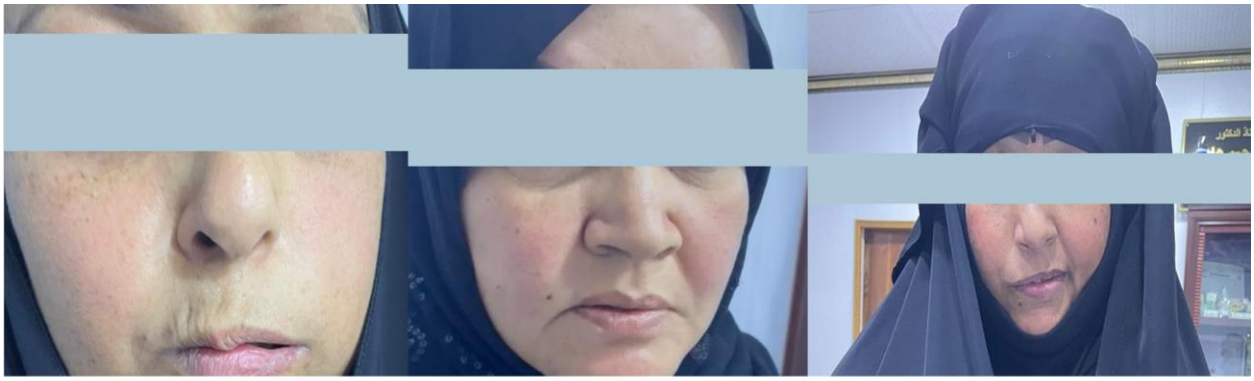

Figure (1) Figure (2) Figure (3)

2.1 FIRST CASE SCENARIO : FAMILIAL BELL'S PALSY

A 51-year-old female with recurrent left-sided facial palsy of the lower motor neuron lesion pattern. She has no personal or family history of diabetes; her blood glucose level was 87 mg/dl, and her HbA1C was 5.3; and neuroimaging revealed no evidence of a brain lesion. Her otoscopic examination was also normal, and the initial screening for connective tissue disease was negative, but her son complained of the same symptoms, and she also provided a family history of facial palsy in her father and uncles in their fifties. She refused to perform electrodiagnostic studies, but she was treated with oral prednisone with a partial response (Figure 1).

2.2 SECOND CASE SCENARIO : FAMILIAL BELL'S PALSY

A 47-year-old female with two episodes of lower motor neuron unilateral facial palsy and an approach for secondary causes including diabetes mellitus, brain pathology, otological disorder, and connective tissue screen was negative, but her mother and sister also gave a recurrent history of facial palsy, and she achieved a full and complete recovery with oral prednisolone (Figure 2).

2.3 THIRD CASE SCENARIO : FUNCTIONAL FACIAL PALSY

A 42-year-old female with recurrent attacks of facial palsy, other neurological exams, and relevant investigations showed no significant findings. Therefore, the patient was diagnosed with idiopathic Bell's palsy and treated with steroids and acyclovir, but there was no significant improvement. The patient had a history of family problems and an unstable psychological background. The patient is treated with a short course of antidepressants, and the facial asymmetry is reversed immediately upon waking from bed after ten days of treatment (Figure 3).

III. DISCUSSION AND CLINICAL APPROACH

Bell's palsy is named after Charles Bell, a Scottish anatomist. Acute, unilateral paresis or paralysis of muscles innervated by the facial nerve arises spontaneously over hours to days in this common clinical condition of unknown cause. It is the most common cause of facial nerve injury and acute mononeuropathy. It affects people of all ages but is significantly more common in their third to fifth decades. It can affect either the right or left side. Recurrence, whether on the same or opposite side, is uncommon and raises the possibility of a more widespread illness (Elan et al. 2022).

Although some patients describe exposing the affected side to a continuous wind or fan for several hours just prior to the start, risk factors are unknown. Diabetes mellitus, pregnancy, and hypertension have all been linked to it. Some speculate that reactivation of a latent herpes simplex virus, or varicella-zoster, and human herpesvirus 6 are to blame. In cases of bilateral facial palsy, Lyme disease is frequently implicated. Familial Bell's palsy is reported but is also rare (Zandian et al. 2014).

Despite there is no firm hereditary foundation for idiopathic facial palsy, familial clustering has long been observed in the literature. Since the first case series of familial Bell's palsy was published in 1887, A cohort of 230 people with idiopathic peripheral seventh nerve palsy, 6 percent of whom had a family history. Surprisingly, one family had a total of 29 occurrences of Bell's palsy and a high degree of consanguinity. According to estimates, about 4–14 percent of familial Bell's palsy cases have a positive family background (Kubik et al. 2012).

There was no difference in the severity of the palsy, residual impairments, recovery duration, resp onse to medical management, or recurrence among the family cases compared to nonfamilial instances. A utosomal dominant with variable penetrance is the most commonly stated mode of inheritance (Kubik et al. 2012).

Several research have attempted to show that idiopathic facial palsy has a hereditary basis. The human leukocyte antigen (HLA) system, which has strong objective links to a variety of autoimmune disorders, has received a lot of attention thus far. A Mexican study of 92 patients with idiopathic facial palsy discovered a significant decrease in the HLA class 2 DR antigen and acutely decreased levels of CD3/CD4 T cells, raising the prospect of HLA-DR linked "resistance gene." In 46 percent of people with low DR antigen and idiopathic facial palsy, a family antecedent was discovered (Kubik et al. 2012).

3.1 CLINICAL FEATURES AND INVESTIGATIONS OF BELL'S PALSY

Patients usually present with complete facial paralysis or unilateral facial weakness. Facial paralysis may be linked with decreased lacrimation and/or poor taste in the anterior two-thirds of the tongue on the affected side, and a weaker or absent blink reflex is always present, depending on the site of the lesion. Except in the Ramsay Hunt syndrome, which is caused by herpes zoster and is followed by a vesicular eruption in the sensory distribution of the seventh cranial nerve in the ipsilateral ear, pain is uncommon (Zandian et al. 2014).

Because the stapedius muscle is innervated, there may be an enhanced sensitivity to noise on the afflicted side. There should be no problems with eye movements, visual changes, or other bulbar symptoms like dysphagia or facial numbness. Any of those signs or symptoms, if present, should prompt a search for a different diagnosis. Laboratory investigation, diagnostic imaging, and electrodiagnostic testing have no place in this scenario unless there is complete facial paralysis (Zandian et al. 2014).

3.2 ROLE FOR NERVE CONDUCTION STUDIES (NCS) AND ELECTROMYOGRAPHY (EMG)

Nerve conduction studies help assess the nature and degree of injury; examinations of the extracranial section of the facial nerve are sometimes undertaken, especially in cases of full paralysis, combined with needle recordings from its myotomes, such as facial muscle EMG. Blink reflex testing can reveal injury to the intracranial area of the brain (Hassan et al. 2021) and (Elan et al. 2022).

3.3 OUTCOME AND TREATMENT

Although severe cases of bell's palsy might result in lasting disabilities, the vast majority of people with bell's palsy recover fully functionally with few to no residual abnormalities. Acute treatment options include steroids, antiviral medicine (acyclovir), and surgical decompression (Elan et al. 2022).

In a large, randomised controlled study, 94 percent of those who took prednisolone 25 mg twice a day for ten days recovered well at nine months, compared to 82 percent of those who did not take steroids. In the same research, the antiviral medication acyclovir showed no benefit at doses of 400 mg five times per day for ten days. In another large research study, however, valacyclovir demonstrated benefit at one g/d for five days. Surgical intervention has also been studied, and a recent assessment of the Cochrane database found insufficient evidence to determine whether it is beneficial or detrimental. Physical treatment may be effective early on, especially if the paresis or paralysis is severe (Hato et al. 2007) and (Sullivan et al. 2007).

Within 72 hours of symptom onset, the American Academy of Otolaryngology—Head & Neck Surgery Foundation practise guidelines prescribe antiviral therapy in addition to oral steroids, but make no recommendations for physical therapy, acupuncture, or surgical referral. The American Academy of Neurology issued an update on steroid and antiviral medication recommendations, stating that steroids should be administered to patients with new-onset palsy and that antiviral therapy may be offered, though it is likely to be of minimal utility at most (Baugh et al. 2013) and (AAN Guidelines - 2012).

Consider administering prednisolone to people who are present within 72 hours of the onset of symptoms, according to NICE guidelines in the United Kingdom. There is no consensus on the best dosing schedule, but if a lowering dose is preferred, possibilities include giving 50 mg daily for 10 days or 60 mg daily for five days, followed by a daily drop in dose of 10 mg for a total treatment length of 10 days. Antiviral medications alone are not advised. Combining antiviral therapy with a corticosteroid may provide some benefit, but this is rarely considered (NICE Guidelines - 2019)

The prognosis of facial nerve injury following Bell's palsy has also gotten a lot of attention. In general, the preservation of motor amplitudes on nerve conduction studies after 7 to 10 days indicates that axonal integrity has been preserved and that recovery is likely. Rapid loss of motor amplitudes, on the other hand, indicates significant axonal involvement, Wallerian degeneration, and a lower probability of functional rehabilitation. Needle electromyography can also help detect denervation changes, which supports the theory of axonal damage. The utility of electroneurography as a prognostic tool has recently been investigated (Kubik et al. 2012) and (Elan et al. 2022).

IV.CONCLUSION

Keep in mind and remember the top tips for dealing with facial palsy, which include asking about any history of diabetes (DM)? Any symptoms suggestive of DM? Any family history of DM? Did the patients previously test positive for diabetes? And order a test for DM (FBS and/or HbA1C) to exclude it first. Then, ask about any family history of the same condition. Father and mother? Brothers and sisters? siblings? Also dig into the details of their psychological status. depression? Anxiety disorder? What about family issues? After that, examine the ear, especially in immunocompromised individuals. Furthermore, think about structural or central causes and get a brain image if the history is suspicious. Finally, follow your patient with neurophysiological studies to predict their prognosis.

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CONFLICTS OF INTERESTS

Not reported.

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