Chapter

Introductory Chapter: Facial Nerve - An Overview

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1. Introduction

The facial nerve (seventh cranial nerve—CNVII) is the nerve of facial expression. It innervates all superficial muscles of the face and scalp, the contraction of which is responsible for all our numerous facial expressions like anger, pain, fear, smile, etc. Facial disfigurement resulting from facial nerve disorders can affect the physical, psychological, and emotional integrity of an individual. This might result in social, occupational, and educational handicap. The facial nerve is one of the most common cranial nerves implicated by disorders. It is a mixed nerve, which carries motor, sensory, and parasympathetic fibers. The motor fiber-innervated muscles developed from second branchial arch, the sensory fibers transmit the special sense of taste, and the parasympathetic fibers supply the submandibular, sublingual, and lacrimal glands [1]. Embryologically speaking, it is formed very early within the acousticofacial complex from the second branchial arch [2]. Facial nerve consists of the juxtaposition of somatic and branchial elements of the cranial nerve nuclei, in particular, accounting for trigeminal and facial nerve anastomosis [3]. A wide variety of disorders can involve the facial nerve including congenital, traumatic, infectious, inflammatory, and neoplastic disorders.

2. Anatomy

The anatomy of the facial nerve is the most complex among other cranial nerves. It composed of approximately 10,000 neurons. Seven thousands of these fibers are myelinated and innervate the muscles of facial expression and the stapedial muscle. The other 3000 nerve fibers form the nervus intermedius with a secretary and somatosensory component. These include the afferent taste fiber from the chorda tympani nerve, the afferent taste fiber from the soft palate, the parasympathetic secretory innervation to sublingual, submandibular and lacrimal glands, and the cutaneous sensory component from afferent fibers originating from the skin of the auricle and postauricular areas [4]. The brain stem contains the *intraaxial segment*, which consists of the motor nucleus, superior salivary nucleus (parasympathetic), and nucleus of tractus solitarius (sensory). On the other hand, the *cisternal segment* consists of the motor root and nervus intermedius (nerve of Wrisberg), which emerge from the brain stem and pass into the internal auditory canal. These two segments merge at the internal auditory canal to form the *canalicular segment*. It runs in the internal auditory canal between the cochlea and vestibule of the inner ear to the geniculate ganglion forming the *labyrinthine segment*. From the geniculate ganglion, three nerves arise, the greater superficial petrosal nerve and lesser petrosal nerve carrying the parasympathetic fibers to the lacrimal and parotid gland

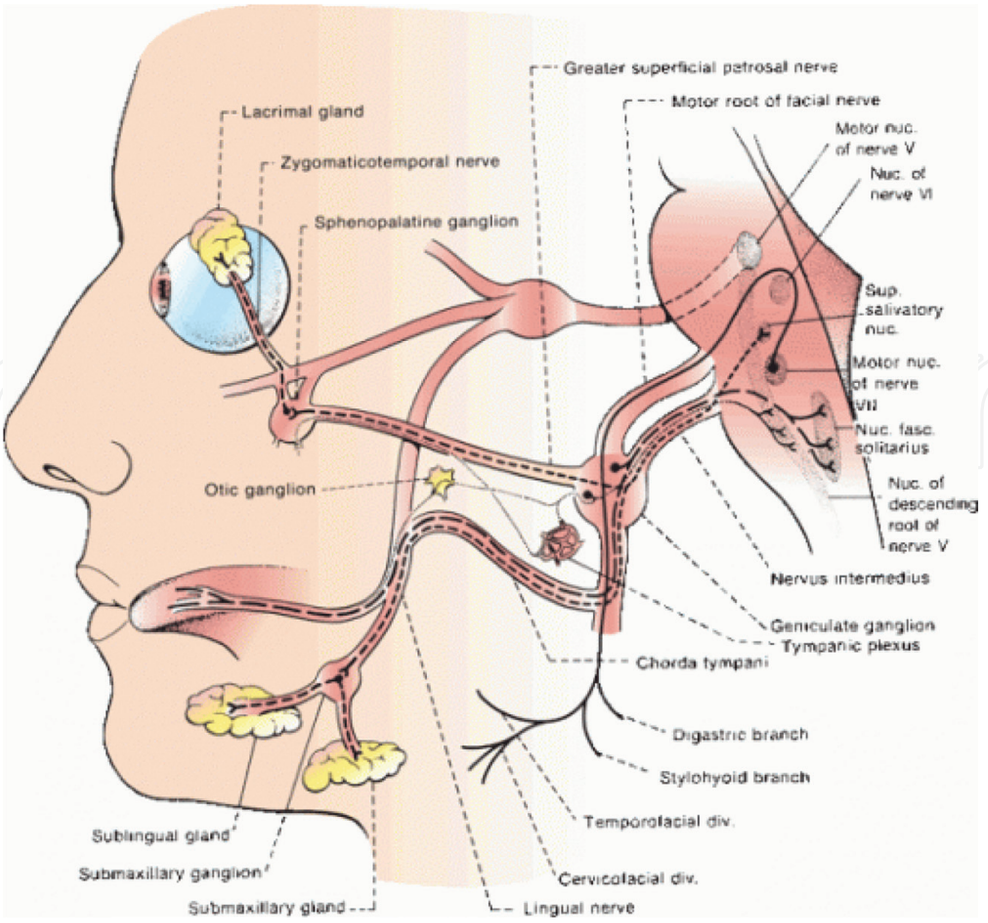

Figure 1.
The anatomy of the facial nerve. Adopted from the free domain: https://myneurosurg.com/cranial-anatomy/cranial-nerve-7-facial-nerve-and-its-pathology/.

and the external petrosal nerve supplying the sympathetic fibers to the middle meningeal artery. The nerve coursing from the geniculate ganglion to the pyramidal eminence forming the *tympanic segment*. From the pyramidal eminence, it passes down to the stylomastoid foramen forming the *mastoid segment* with branches to the stapedius muscle and chorda tympani. The *extracranial segment* started with emerging from the stylomastoid foramen, and it runs through the parotid gland giving rise to the superior temporozygomatic, inferior cervicofacial, temporal, zygomatic, buccal, mandibular, and cervical branches [5] (**Figure 1**).

Knowledge about the normal anatomy, vascular relationship, and the wide spectrum of the abnormalities is vital for the radiologist and neurologist in correctly diagnosing and managing facial nerve disorders.

3. Clinical disorders of facial nerve

3.1 Facial palsy

Bell's palsy is the idiopathic variety of facial nerve palsy where the patient has signs and symptoms of paralysis of facial muscles without known etiology. The etiologies that should be excluded in diagnosing Bell's palsy include intracranial and extracranial malignancies, infections, trauma, cerebrovascular accident, etc. Previously Bell's palsy was regarded as a diagnosis of exclusion, but in 1984, May et al. emphasized that Bell's palsy is a positive diagnosis based on specific clinical features [5]. The clinical features that may help to distinguish it from

other causes of facial palsy include sudden onset of usually unilateral facial palsy with the absence of signs and symptoms of CNS, ear, and cerebellopontine angle disease. The annual incidence of Bell's palsy globally is estimated to be 11.5–53.3 per 100,000 in different population, and generally, the percentage is increasing as individuals get older [6–8], although some studies suggest that the highest incidence occurs among young and middle-aged individuals [9]. Bell's palsy thought to account for approximately 60–75% of cases of acute unilateral facial paralysis with the right side being affected in 63% of the time. It can also be recurrent with a reported recurrence range of 4–14% [10]. It affects both sexes equally but might affect women more than men in a certain age group. The lowest incidence is reported in persons younger than 10 years, and the highest incidence is in a person aged 70 years or older. Rarely it might occur bilaterally (0.3%), and positive family history is found in 4–14% of patients [11].

The pathophysiology of Bell's palsy remains unclear. Compression of the facial nerve while it course through facial canal, especially the narrow labyrinthine segment, is the most acceptable theory. The compression has been demonstrated in MRI scan with facial nerve enhancement [12]. Any inflammatory, demyelinating, and ischemic or compressive process in this area may impair the neural conduction. Viral infection is the most commonly accepted theory behind Bell's palsy, and the Herpes simplex virus is the most commonly implicated virus. Other viruses include Mumps virus, cytomegalovirus, and HIV. Other suggested theories include autoimmunity, mycoplasma infection, inflammation, microvascular disease such as diabetes mellitus, and many other mechanisms. It is apparent that none of these theories stands on a solid base.

Bell's palsy typically occurs suddenly, and the symptoms peak in less than 48 hours. The paralysis must include the upper and lower aspects of the face, otherwise, if it involves the lower portion of the face, a central cause (supranuclear), such as stroke, should be suspected. If the onset of the facial paralysis is insidious, associated with weakness of the contralateral side, or there is a preceding history of trauma or infection, other causes of facial paralysis must be strongly considered.

In addition to the sudden onset of unilateral upper and lower facial muscle paralysis, the patient may have hyperacusis, posterior auricular pain, otalgia, incomplete eye closure, excessive salivation, and taste disturbances. Many patients report numbness on the affected side. Whether this numbness is due to the involvement of trigeminal nerve or lack of movement of facial muscle is not clear. Ocular pain, epiphora or decreased tearing, and blurred vision are other possible associated symptoms.

On examination, in addition to demonstrate weakness and/or paralysis of the entire side of the face, we find a flattening of the nasolabial fold on the affected side, inability to elevate eyebrow, and distorted face when the patient asked to smile (**Figure 2**). The patient should be examined in details for full neurological examination, any skin lesion, ear or eye problem, and parotid diseases.

The goal of treatment of Bell's palsy is to improve the facial nerve function and to reduce neuronal damage. Steroids (prednisone) and antiviral therapy (acyclovir) are the recommended therapy by the American Academy of Neurology (AAN) and American Academy of Otolaryngology [13]. Additionally, a variety of non-pharmacological therapy is suggested for the treatment of Bell's palsy including physical therapy (facial exercises, neuromuscular training) and acupuncture. A variety of pharmacological and non-pharmacological treatments to protect the eyes of patient with Bell's palsy have been used. These include the use of topical ocular lubrication (with artificial tear during the day and lubricating ophthalmic ointment at night), occluding the eyelids by tape or patch, inducing ptosis by botulinum toxins, and many other procedures aiming to protect the patient's eye from drying, corneal abrasion, and corneal ulcers [14, 15].

Figure 2.
Seven-year-old child with left facial palsy asked to smile (published with parent permission).

Surgical treatment of Bell's palsy with facial nerve decompression is also used, but it still carries a controversy, and it may be considered in patients with poor prognosis as predicted by facial nerve testing (EMG shows greater than 90% axonal degeneration within 3 weeks of the onset of paralysis) or complete persistent paralysis not responding to treatment [16, 17].

Other possible causes of facial palsy include congenital facial palsy (Möbius syndrome, hemifacial macrosomia), polyneuritis (Guillain-Barre syndrome, Ramsay-Hunt syndrome due to herpes zoster, autoimmune) malignancy (parotid tumors, brain tumor), trauma (temporal bone fracture, birth injury), sarcoidosis, infections (otitis media, cholesteatoma), and many other rare causes [18].

3.2 Tumors

Facial nerve tumors are rare and represent uncommon cause for facial palsy accounting for only 5–10% of all cases of facial nerve palsy [19]. Hearing defects and facial paresis are the most common presenting features. The most commonly identified tumor are schwannomas. Facial nerve schwannomas are benign tumors, which arise from Schwann cells. They mostly occur sporadically; some occur as a part of genetic syndromes as neurofibromatosis types 1 and 2. The facial nerve is the third common site for intracranial schwannomas. The patient usually presents with sudden onset or progressive facial weakness with or without conductive or sensorineural hearing loss, tinnitus, vertigo, facial pain, hemifacial spasm, facial palsy, and otalgia. Workup may involve detailed hearing test, MRI scan of the brain, and CT scan of the temporal bone. Other tumors involving the facial nerve include hemangiomas (accounting for 18% of facial nerve tumors and 0.7% of intratemporal bone tumors) [20], neurofibromas, and meningiomas. Most of these tumors present with facial palsy with or without hearing defects.

3.3 Hemifacial spasm (HFS)

It is a segmental myoclonus of muscles innervated by the facial nerve. Gowers first described it in 1884. It affects 11 per 100,000 of the population mostly in the fifth and sixth decade of life with a slight female predominance. The majority of cases are unilateral, although bilateral involvement might occur rarely in severe cases. It generally starts as brief clonic movements of the orbicularis oculi that spread over the years to other facial muscles (corrugator, frontalis, orbicularis oris, platysma, and zygomaticus), and it may become more sustained with time. Although it is a benign condition, it often leads to social embarrassment resulting in psychosocial stress and social withdrawal (**Figure 3**). HFS is characterized by progressive involuntary clonic or tonic movements of the muscle supplied by the facial nerve. It typically started in the orbicularis oculi muscle and progressively extends to involve the other facial muscle, involving the platysma muscle in severe cases. Most of the cases persist during sleep, and some patients report clicking sound in the ear, which presumed to be due to contraction of the stapedius muscle. In severe cases, impairment of vision might occur because of severe spasm of the orbicularis oculi muscle. The symptoms are usually exaggerated by psychological tension and speech. The main differential diagnosis of HFS includes blepharospasm (occur bilaterally symmetrically), oromandibular dystonia (sustained contraction of the lower part of the face, mouth, mandible and maxilla, tongue and pharynx), facial nerve tic (complex coordinated multifocal movement that switches between the right and left sides of the face), hemimasticatory spasm (painful contractions of the muscles of mastication), focal seizures, and synkinesias after facial nerve paralysis (activation of several muscles innervated by the facial nerve).

The underlying cause of hemifacial spasm in most cases is an ectatic or atypically aberrant blood vessel, which compresses the facial nerve at the place where it exits the brain stem. This area is the most susceptible part of the facial nerve to external stimuli since it ensheathed only by an arachnoidal membrane without the epineurium. Furthermore, this area represents the transition zone between central (oligodendroglial cells) and peripheral (Schwann cells) myelinations, and there is no connective tissue septa that traverse the individual fascicles. In the majority of cases, inferior posterior cerebellar artery or the inferior anterior cerebellar artery is the cause for vascular compression. Rarely, vertebral artery or a combination of these arteries is responsible for this compression. Very rarely, a vein might be the cause. HFS diagnosis is mainly clinically supported by investigations such as electromyography (EMG) and MRI. These investigations are useful also to exclude pathological changes in the cerebellopontine angle such as tumors or brain stem lesions. A high-resolution, T2-weighted sequence is particularly useful in demonstrating vascular compression.

Figure 3.
The editor of this book is afflicted by right-sided HFS started at the age of 45; unfortunately, vascular decompression failed to resolve the problem. (A) Spasm of the right orbicularis oculi and right facial muscle and (B) spontaneous resolution of the spasm.

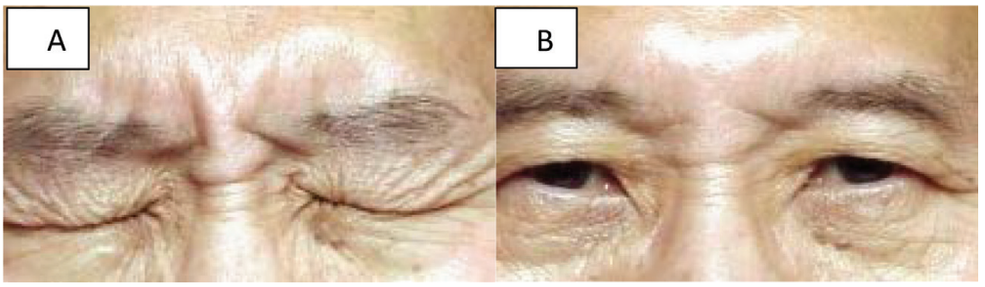

Figure 4.
(A) Patient with blepharospasm and (B) same patient after treatment.

The therapeutic options for HFS are variable ranging from simple applications of heat to pharmacological treatment (carbamazepine, clonazepam, baclofen, and gabapentin) and botulinum injections to microvascular decompression surgery. The response to medications, in general, is poor and not sustained and thus is devoted to milder cases. Botulinum toxin (BOTOX, BTX) became the standard treatment since its use in the early 1980s. It alleviates symptoms in 85–95% of patients. The major drawback is its temporary effects and the need to repeat its administration every 3–4 months [21–23]. The major advantages of BTX are that it is a noninvasive technique and can be done in an outpatient setting. The only curative therapeutic option is microvascular decompression of the facial nerve by placing a Teflon sponge between the vessel and the brain stem. Using an endoscopic technique makes the operation safe, but still, there is a risk of less than 1% of intraoperative life-threatening complications such as life-threatening hemorrhage or cerebellar or brain stem infarctions. Additionally, postoperative complications including temporary or permanent hearing impairment and permanent facial paralysis may occur in up to 8 and 0.9%, respectively [24].

3.4 Blepharospasm

It is the idiopathic progressive bilateral involuntary spasm of the orbicularis oculi and upper facial muscle (**Figure 4**). Involvement of the lower face by spasm is not uncommon, and the patient might be functionally blind as the eyes might be closed most of the day. The cause of the blepharospasm is thought to be central, yet the exact mechanism is not known. Treatment options include selective destruction of facial nerve branches that innervate the orbicularis oculi muscle, inducing paralysis of orbicularis oculi muscle by BOTOX, and pharmacological therapy by anticholinergics drug (Artane), benzodiazepine (clonazepam), GABAB receptor agonist (baclofen), anticonvulsant (levetiracetam), and many other drugs [25].

4. Investigations

In addition to a detailed history and clinical examination, investigations are an essential part of the workup of patients with facial nerve disorders. Still, there are controversies about the appropriate investigations that are needed for patients present with facial nerve disorders. The most commonly used investigations include:

4.1 Audiometry and brain stem auditory evoked response

These tests used to detect any associated hearing defect and diminished stapedial reflex (paresis of the stapedial branch of the facial nerve). Brain stem auditory evoked response (BAER) in particular is effective in detecting retrocochlear lesion. These tests are not used routinely unless there are multiple cranial nerve palsies [26, 27].

4.2 Full blood count

Facial paralysis is a recognized feature of leukemia relapses in both children and adult.

4.3 Imaging studies

Imaging studies play an important role in the evaluation of facial nerve disorder. MRI is especially helpful in recognizing brain stem pathology, and high-resolution CT scan is helpful in identifying bony abnormalities of the intratemporal facial nerve like congenital anomalies, trauma, and cholesteatoma. MRI is especially helpful in the detection of soft tissue abnormalities around facial nerve as in neoplasms, inflammatory processes, and hemifacial spasm. Recently, facial nerve ultrasound and diffuse tensor tractography (three-dimensional reconstruction of facial nerve using MRI) are used to identify cranial nerve fiber displacement by vestibular schwannomas [28, 29]. Contrast-enhanced MRI is helpful when the facial palsy cannot be definitively localized [30]. The patient's symptoms and the proposed differential diagnosis will determine the choice between these investigations.

4.4 Neurophysiological studies

Neurophysiological studies are of diagnostic and prognostic value, especially in chronic facial nerve problem. Most of these tests require the cooperation of patients. Examples of these tests include measurement of fibrillation potentials and recording of the blink reflex. The presence of fibrillation potentials, which is part of the electromyography, in the muscles implies the loss of muscle innervation, which indicates a significant axonal degeneration. Blink reflex tests, on the other hand, utilize the polysynaptic nature of this reflex. Trigeminal nerve carries a unilateral stimuli (electric stimulation of the supraorbital nerve or a puff of air on the cornea), producing an early ipsilateral facial motor response followed by a bilateral late response. Absence or delay in the late response can be used to assess facial innervation [31].

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