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A Rare Cause of Hemifacial Spasm: Papillary Oncocytic Cystadenoma

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Background: Hemifacial spasm is a sudden, involuntary and synchronous spasm of the facial muscles. The most frequent cause of this condition is compression of the facial nerves due to vascular pathologies. The most commonly used method of treatment is Botulinum toxin injection. However, the gold standard treatment is surgical treatment.

Case Report: A 64-year-old male patient with hemifacial spasms, which had occurred due to a rare parotid mass that had been surgically treated, is presented in this case.

Conclusion: This case report demonstrates that long-standing parotid gland masses may compress the facial nerves and cause demyelination in the nerve and thus may cause spasms in the facial muscles.

Keywords: Hemifacial spasm, parotid tumor, facial nerve

Hemifacial spasm (HFS) is a movement disorder of the muscles that are innervated by the ipsilateral facial nerve and develops as an irregular, tonic-clonic spasm of those muscles. It may appear around 50 years of age in an individual with no prior complaints. It is more frequently seen in women. The unilateral nature and continuation during sleep is characteristic of the spasm. Complaints are more severe with stress. The most common cause is the compression of the facial nerves due to abnormalities in the vascular structures in the brain stem. HFS may also develop secondary to trauma and peripheral facial paralysis (1). Rarely, it may be secondary to a mass of the parotid gland (1,2).

The current study presents the case of a 64-year-old male patient who continued to have complaints of hemifacial spasm, in spite of long-term treatment with Botox. The patient had undergone a total parotidectomy due to a progressively enlarging mass that was previously present at a location anterior to his left ear. His complaint of spasm greatly resolved after surgical treatment.

CASE PRESENTATION

A 64-year-old male patient presented with swelling in front of his left ear and a twitch on the left side of his face. The patient, whose face twitching gradually increased over the previous two years, received carbamazepine (Tegretol, Novartis Pharma AG; Basel, Switzerland) therapy for six months, but the symptoms persisted. Twitches during sleep were particularly present around his eyes and mouth and were occurring almost 20-30 times a day. The frequency increased during stressful conditions.

The patient was administered Botox injections for seven sessions using the guidance of an electromyogram (EMG). As a result of the EMG (left orbicularis oris, orbitalis oculi and frontalis), spontaneous, high-frequency motor unit discharges and isometric contractions were reported. Furthermore, except for the stimulated branch, delayed stimulations were present on the facial nerve branches. Following the Botox injections (of unknown doses), the patient felt relieved for approximately three months. However, the symptoms returned. During one of the Botox injections around the eyes, the left eyelid drooped. Nonetheless, the patient did not suffer from any other significant complication.

His anamnesis revealed that the swelling in front of his ear had been present for a long time; however, its dimensions increased during the preceding year. Upon physical examination, a lobulated hard mass measuring approximately 3 cm in diameter at the parotid area, posterior to the angulus mandibula on the left side of the neck was detected. Saliva output from the

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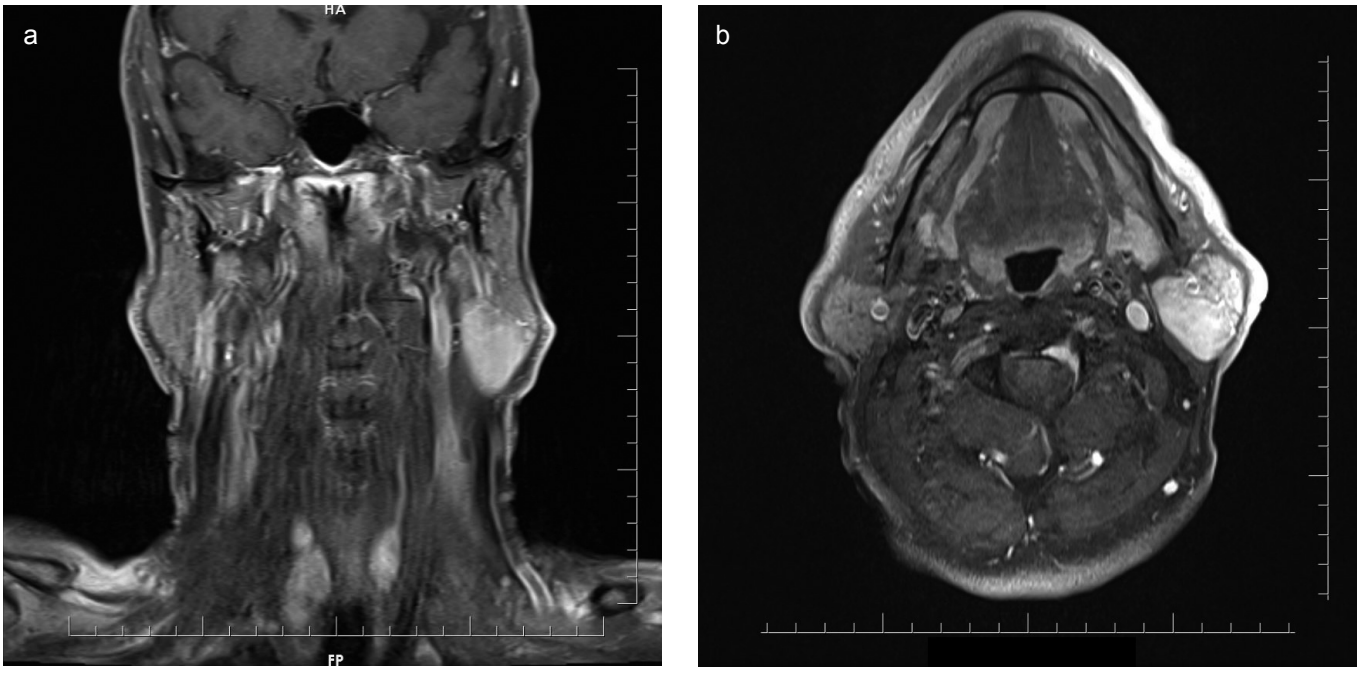

FIG. 1. a, b. MRI of the head and neck, axial (a) and coronal (b) views of left parotid gland. The lesion measuring 32x20 mm with no marked uptake of contrast material in the inferior and posterior neighborhood of the left parotid gland, including a cystic component, which was thought to be a lymphadenopathy.

left parotid gland had decreased. The neurological examination was normal. An elliptical image compatible with lymphadenopathy at the lower pole of the left parotid gland measuring 29x24 mm with a central echogenic hilus and a thick cortical rim was observed in a neck ultrasound performed at another center. Magnetic resonance imaging of the neck revealed a lesion measuring 32x20 mm with no marked uptake of contrast material in the inferior and posterior neighborhood of the left parotid gland, including a cystic component, which was thought to be a lymphadenopathy (Figure a,b).

The patient underwent a left total parotidectomy under general anesthesia. The mass located at the tail of the parotid gland extended to the deep part of the parotid gland. The lesion, which was easily dissected from the surrounding tissues, was excised together with all the parotid gland tissue while preserving the facial nerve. The histopathological examination revealed a "papillary oncocytic cystadenoma."

During the postoperative period, the twitching complaints of the patient largely resolved. No additional treatment was necessary during the six-month follow-up. Written informed consent was obtained from the participant of this study.

DISCUSSION

Hemifacial spasm is a unilateral, involuntary and painless contraction of the facial muscles that are innerved by the seventh cranial nerve (1). It was first described by Schultze in 1875. It was first mentioned in English in a book by Gowers titled, "Manual of Diseases of the Nervous System" in 1888. Gardner and Sawa reported in 1960 that this condition may be reversible when the etiologic factor causing the disease is removed (3). In the beginning, spasms are usually mild and clonic, while they may become severe and sometimes tonic with progression of the disease (4). Symptoms tend to be aggravated with changes in the emotional state and stress, as was seen in our case. It is generally seen in middle aged women and in the fifth decade. The female to male ratio is approximately 3/2 (4).

The most common cause of HFS is association of the facial nerves at the entry region of the nerve root of the brain stem with pathological vascular structures. Other rare causes include tumors (most frequently cerebellopontine angle tumors), bone diseases (Paget's disease), nerve injuries, trauma, hypothyroidism, idiopathic intracranial hypertension, and multiple sclerosis (4). No etiologic factors can be detected in the vast majority of patients. However, a detailed anamnesis should be obtained and a physical examination including a neurological examination and magnetic resonance imaging should be performed to make the diagnosis and define the etiological factors (5).

One of the rare causes of HFS is lesions of the parotid gland. Several cases of hemifacial spasm have been reported in the literature, secondary to lesions of the parotid glands. Papillary oncocytic cystadenoma was present in our case. Cystadeno-

mas of the salivary gland are rarely seen encapsulated tumors with regular borders and possible intracystic papillary extensions as well as oncocystic features. They are benign and multicystic tumors originating from the epithelium of the salivary gland canals that could not be differentiated. The past medical history of the patient generally reveals painless swelling in the neck, which is present for a long time, as was the case in this case reported here (5,6).

Although there are several theories related to the pathophysiology of HFS, ephaptic transmission is most commonly accepted. Transmission in the axon that has lost its myelin may skip to a neighboring nerve when passing through a region that is not enveloped, when deviating from its original pathway. This condition is called ephaptic transmission. Therefore, mistaken transmission of a stimulus occurs in a nerve unrelated to that process. This pathological condition is thought to cause the pathophysiology of HFS. A lesion on the nerve with unknown etiology is generally thought to cause ephaptic transmission between the facial motor nerves in that region, which creates spasms by carrying intense and spontaneous impulses episodically to the peripheral muscles. Another theory is the development of abnormal and multiple branching after axonal regeneration. Increased excitability of the facial motor neurons is thought to occur secondary to this condition (5-7).

Facial twitches, which are one of the diseases included in the differential diagnosis of HFS, are involuntary movements in other muscles of the body in addition to the facial muscles. The difference of this condition from hemifacial spasm is the partial inhibition of the movements. Blepharospasm, on the other hand, is differentiated from hemifacial spasm by bilateral symmetrical and synchronous involvement of the eye muscles. In oromandibular dystonia, repetitive and continuous twitches are present in the muscles of the lower face, jaw, tongue, pharynx and oral muscles, with no involvement of the ophthalmic muscles. Facial myokymia is a continuous and wave-like involuntary spasm of the facial muscles. Stereotypical movements of the face, neck, and arms occur in tardive dyskinesia. Rotation of the body and wrinkling in the face are characteristic. Focal seizures affecting half of the face in focal epileptic seizures are confused with facial twitches and electroencephalogram is important in the differential diagnosis. A characteristic feature of psychogenic facial twitches, on the other hand, is the absence of twitches during sleep (8).

Hemifacial spasm can be treated medically or surgically. The most important point is to define the underlying pathology. The purpose of the drugs used for medical treatment such as carbamazepine, anticholinergics, baclofen, haloperidol, and gabapentin is sedation. These drugs are considered only in mild cases and in inoperable cases, since their treatment effects are limited. Other than this indication, they should not be considered as an option in the treatment of facial twitches.

Currently, Botulinum toxin (Botox) injection is frequently used as an alternative medical treatment. It was first used for facial twitches in 1985. Its success rate reaches 75-90% according to different reports; its duration of effect is limited to several months and its application should absolutely be repeated. Its complications include dry eyes, paralysis in the eyelids and facial muscles, diplopia and excessive tear secretion. The most common complication is eyelid ptosis. The most important disadvantages of this treatment are high cost and the requirement of repeated injections (8).

Surgical treatment is the gold standard for HFS treatment. Microvascular decompression is a frequently employed surgical procedure. It is a serious surgical intervention that requires experience and the use of a microscope and microsurgical instruments. Nevertheless, it provides long term and permanent improvement in 90% of the patients in contrast to botulinum toxin injection and drug treatments. It can be applied when repeated Botox injections fail or as a first line treatment option. In addition, surgical treatment should be considered as a first-line treatment option when a mass lesion is detected on the trajectory of the facial nerve (7-9). The anamnesis of the case presented here revealed a persistent hemifacial spasm in spite of long-term treatment with Botox. The progressive increase in the size of the swelling in front of the left ear and the facial twitches in the last one year suggested that the mass in the parotid gland might have compressed the facial nerve. During the first-week post-operative control of our patient, it was observed that the spasms ceased completely. However, during the subsequent period, there were spasms with one or two contractions every two or three days, which did not affect the daily life of the patient. There were also periods of several weeks in which no spasms were observed. These findings have led us to consider that the symptoms suffered by the patient were due to ephaptic transmission. Nonetheless, they support the hypothesis that the main etiological factor is a mass effect or local irritation. The substantial reduction in complaints of facial twitching following total parotidectomy proves the efficacy of etiology-oriented treatment planning and surgical treatment.

Hemifacial spasm is an irritating and disturbing condition, yet it is a treatable disease that affects the daily life of a patient. Botulinum toxin injection into the affected muscles is the most commonly used method in conservative treatment. However, surgical treatment is the gold standard. Etiological conditions should be sought out, and treatment should be planned accordingly.

This case report demonstrates that long-standing parotid gland masses may compress the facial nerves and cause demyelination in the nerve, which in turn may cause spasms in

the facial muscles due to ephaptic transmission. Therefore, although they are seen very rarely, ear, nose, and throat specialists should always keep in mind that parotid lesions may cause hemifacial spasm.

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Informed Consent: Written informed consent was obtained from patient who participated in this study. .

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