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Hemifacial Spasm Due to Contralateral Supratentorial Tumor: A Case Report

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ABSTRACT

Introduction: A rare neurological disorder known as hemifacial spasm (HFS) is characterized by involuntary, unilateral facial muscle contractions, typically caused by facial nerve irritation. In this report, we looked at a rare and fascinating case of hemifacial spasm (HFS) caused by a large contralateral supratentorial meningioma. The HFS went away amazingly after the tumor was removed and the patient was treated with corticosteroids. The complex connection between neurological disorders and intracranial tumors is highlighted in this case, along with potential treatment approaches for the patient.

Case: A 48-year-old woman complained of uncontrolled movements and spasms in her right eyelid and right lip corner for the past two years. The symptoms were accompanied by headaches, cognitive impairment, and a history of syncope. A neurological examination revealed right-sided central facial palsy. An MRI revealed a large atypical meningioma in the left frontal lobe. This resulted in significant brain displacement and perifocal edema, but no neurovascular contact with the 7th cranial nerve. Her hemifacial spasms significantly decreased following surgical resection, even with medication reduction.

Conclusion: As an uncommon manifestation of contralateral supratentorial tumors, HFS requires careful evaluation of the patient's history, physical examination, and radiological findings, as well as consideration of a wide range of potential causes. Accurate diagnosis and effective management can help reduce the risk of poor prognosis.

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INTRODUCTION

Hemifacial spasm is an uncommon involuntary movement in neurological disorders. It is characterized by sudden, involuntary twitching of the facial muscles on one side of the face, which are innervated by the same side's facial nerve.[1,2] Symptoms include facial muscular contractions that are paroxysmal, clonic, or tonic and occur irregularly and involuntary.[3,4] These spasms are often rapid, brief, and repetitive (clonic), but they can also be longer-lasting and continuous (tonic), with intervals of inactivity in between. These contractions often begin with the periorbital muscles and can progress to affect the perioral muscles, platysma, and other muscles involved in facial expression.[3,5] A compressive lesion of the facial nerve is likely to cause it. A tortuous branch of the basilar artery, which curves around the ventral surface of the pons and under the proximal seventh nerve, usually cause this.[6]

This disorder affects 14.5 per 100,000 women and 7.4 per 100,000 men. The distribution by gender was 2:1. Also, the disease usually initially appears in adulthood, with an average onset age of between 45 and 50 years. Only about 1% to 6% of patients are under the age of 30.[3,7,8] Some studies have shown a slightly higher prevalence of this disease among Asian populations, although the reason for this difference is unclear.[7,8]

An abnormal artery usually causes primary hemifacial spasm by putting pressure on the facial nerve in the posterior cranial fossa. This result in nearly all patients experiencing hemifacial spasm at the root exit zone. Aneurysms, parotid gland tumors, brainstem lesions, trauma, Bell's palsy, and other anatomical abnormalities in the posterior cranial fossa.[9,10] In fewer than 5% of cases, tumors and other space-occupying lesions may be the cause of hemifacial spasm. The tumors that cause this are usually cerebellopontine angle tumors or tentorial meningiomas on the same side. It was extremely rare for the tumor to be located in the supratentorial region.[11]

A clinical history and a neurological physical examination establish the diagnosis clinically. Some tests, such as the EMG and radiology examination, may be considered in addition to the examination.[12] For HFS, radiological test options include a head MRI and an MRA, which may detect around 80% of vascular abnormalities. MR diffusion tensor tractography makes 3D reconstructions of the facial nerve, which can help find any displacement of cranial nerve fibers.[13,14]

Lesions located distant from the facial nerve's root exit zone are an uncommon cause of HFS. We present a unique case of a large contralateral supratentorial meningioma that did not affect the root entry zone (REZ) area. The patient experienced HFS, which improved with tumor surgery and corticosteroid treatment.

CASE

The outpatient clinic received a 48-year-old woman who had experienced uncontrolled movement and spasms of the right eyelid and right lip corner for the previous two years. The condition has gotten worse over the past year. Her family reported that the movement also occurred during her nighttime sleep, occasionally disrupting her sleep. This condition leaves her feeling weak during the day and prevents her from getting enough sleep. Prior to her hospital admission, she had been suffering from a headache for a year. It is getting worse day by day. The headache was intense, causing throbbing pain throughout her head, particularly on the left side. She reported that the headache's pain scale (NRS) was 8-9 for the initial time. Her NRS was 8-9 while she wasn't taking any medicine, but it somewhat improved to 5-6 after taking an analgesic and getting some rest. Over the past month, NRS's headaches did not improve with medication, instead becoming more frequent.

Her family reported that she frequently forgets small details in the past year and sometimes takes a long time to respond to simple questions like where she put her belongings or what she did the day before. She also has a history of syncope, which began with a sudden headache while she was at home in April 2022. Once the patient regained consciousness, she was unable to recall the details of the accident. She had previously received contraceptive injections, which she discontinued 15 years ago. During the neurological examination, we identified the right facial palsy (Upper Motor Neuron type) and found that her motor and sensory strength were within normal limits.

A brain CT scan with contrast revealed a high-grade glioma at the front-temporal lobe of the brain, causing a midline shift to the right around 13.4 mm (Figure 1). A brain MRI with contrast revealed an atypical meningioma at the left frontal lobe of the brain, with a size of ± 6.0 x 5.4 x 4.9 cm, causing a midline shift to the right as far as ± 2.0 cm and accompanied by non-communicating hydrocephalus.

Her EEG result was normal. There was no EEG seizure during the spasmic movement on the right side of her face. The spasmic movement decreased after 1 month's therapy with clonazepam (1 mg twice daily) and a low dosage of dexamethasone, and the headache was also improved. In August 2022, the patient underwent surgery for her tumor, and the anatomical pathology results were meningioma angiomatous type WHO grade I. After surgery, her hemifacial spasms improved; even when dexamethasone was stopped and

clonazepam was reduced to 1 mg once daily, the frequency and severity of spasms remained reduced.

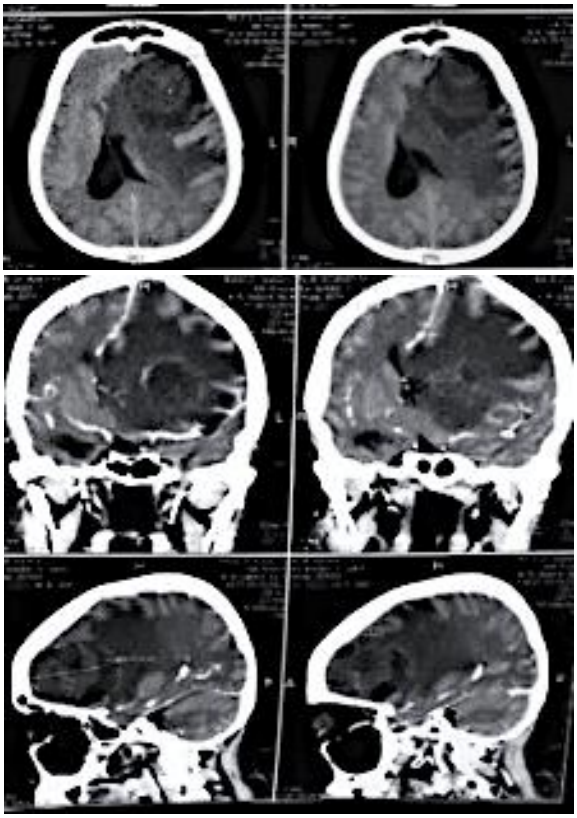

Figure 1. Preoperative head CT scan with contrast. Suspect of high-grade glioma at left cerebral hemisphere that causes midline shift to the right around 13.4 mm.

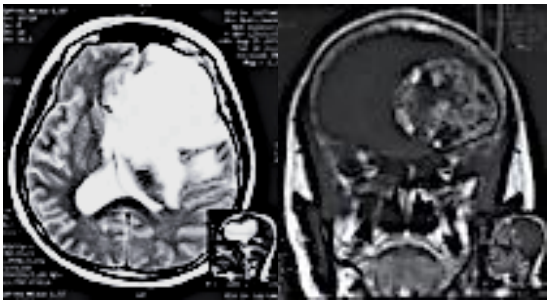

Figure. 2. Preoperative head MRI with contrast. Atypical meningioma at left frontal lobe of brain that causes midline shift to the right.

DISCUSSION

In this case, we talked about a large meningioma originating in the contralateral cortical lobe, specifically in the frontotemporal region, without direct contact with the brainstem or facial nerve. The MRI showed that there was no direct neurovascular contact at REZ, but the hemifacial spasm still occurred. However, the significant brain shift induced by the tumor and perifocal edema raised the possibility that this was the underlying cause of our patient's hemifacial spasm. Hemifacial spasm (HFS) is largely diagnosed based on clinical symptoms. HFS is characterized by recurrent, sudden, involuntary twitching of the facial muscles, which typically occurs on one side of the face but less frequently on both.[15]

The study found that local demyelination of the facial nerve caused by compression by an expanded or aberrant blood vessel is usually the main cause of hemifacial spasm. This condition is rarely caused by diseases other than vascular compression around the facial nerve's REZ.[16] This one-sided hyperactivity of the facial nerve starts with spasms in the orbicularis oculi muscle. These spasms spread to the orbicularis oculi muscle and eventually affect all the muscles that make facial expressions, including the platysma.[3] At first, the seizures are typically mild and do not cause significant distress to the patient, but as the disease worsens, they become severe and cause clonic movements, which are sometimes followed by tonic movements.[17]

Several hypotheses have been proposed to explain the etiology, but the ephaptic transmission concept is generally accepted. Without myelin, the axonal transmission might hop to an adjacent neuron when it goes through an unprotected area; deviating from its intended path causes incorrect stimulation of nerves that are not engaged in the intended process. Another theory that may explain the etiology of this disorder is that when the facial nerve is injured, this leads to degenerative changes in the medulla and functional reorganization of the connective tissue. This, in turn, causes hyperexcitation of the nucleus and leads to the formation of new nerve cell bodies. The last theory is called the sympathetic hypothesis, and it says that damage to the endings of sympathetic nerve fibers in the adventitia layer of an artery causes neurotransmitter imbalances, which in turn cause abnormal action potentials. [8,17]

In certain cases, HFS can be caused by tumors, trauma, or infection, which is known as secondary HFS. Research by Zhang et al. reported that the prevalence of secondary HFS is just 0.3%.[18] However, it is rare for tumors located on the opposite side and distant from the cerebellopontine angle to cause hemifacial spasm, and the underlying cause remains unclear. Only five cases have previously been reported. Some lesions that have been reported are vestibular schwannoma, a contralateral CPA tumor, a huge tentorial meningioma, tumors in the fourth ventricle, and an occipital falcine meningioma.[19] However, the role of supratentorial tumors in causing hemifacial spasm remains unclear.

The patient had surgery to remove the tumor, and glucocorticoids helped to relieve her hemifacial spasms thereafter. In cases of meningioma, studies advise resecting as much tumor tissue as possible,

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including any affected bone and dura mater. The patient was diagnosed with a WHO Grade I meningioma; therefore, a total resection was performed without postoperative radiotherapy.[20] The patient's decision to underwent surgery was influenced by her worsening hemifacial spasms, increasing headache intensity, and the large size of the tumor with extensive surrounding edema. In this case, surgery was needed because the tumor and perifocal edema had caused the significant brain displacement, as well as the existence of hemifacial spasms that persisted despite no direct neurovascular contact at the REZ, as shown on MRI.[7]

A month after surgery, we reduced the clonazepam dosage and discontinued the steroids. Three weeks after this adjustment, the hemifacial spasms continued to decrease but were not fully gone. We concluded that the hemifacial spasms were not totally caused by the edema or the tumor, but they worsened the symptoms. To find the exact cause of the hemifacial spasms, more tests are needed, like MR DTI tractography, to see if there are any problems with the nerve pathways in the face.

This case highlights the unique presentation of a large supratentorial tumor resulting in HFS on the contralateral side—a rare and complex phenomenon. The scarcity of documented cases poses significant challenges in elucidating the underlying mechanisms. Such cases necessitate a detailed investigation of potential etiologies, incorporating meticulous interpretation of brain imaging to identify subtle or indirect factors contributing to HFS.

This report's primary limitation lies in its focus on a single case, which limits its generalizability to other patients with similar conditions. Further studies, including case series and long-term follow-ups, are needed to establish broader insights. Nonetheless, the strength of this report lies in its rarity, which serves to raise awareness among healthcare professionals about the potential association between hemifacial spasm (HFS) and supratentorial tumors. Such awareness may inspire further research and contribute to the development of more effective diagnostic and management strategies for similar cases in the future.

CONCLUSION

HFS is an uncommon manifestation of contralateral supratentorial tumors, especially when there is no direct neurovascular contact of the 7th cranial nerve. It is crucial to consider a wide range of potential causes of HFS and to conduct a thorough assessment of the patient's history, physical examination, and radiological findings. This comprehensive approach is essential for accurate diagnosis and effective management, which can help mitigate the risk of a poor prognosis.

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Conflict of Interest
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