Isolated Hypoglossal Nerve Paralysis

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CASE REPORT
The present case involved a 54-year-old woman who was diagnosed with locally advanced breast cancer in 2011. The patient subsequently received neoadjuvant chemotherapy and underwent a modified radical mastectomy. After surgery, she was regularly followed up, and no distant metastasis or local recurrence was detected by serial imaging studies or laboratory examinations.

Approximately 5 months before the present event, she started to experience slightly slurred speech and difficulty in moving food to the back of her mouth. She then presented to our department with progression of the above-mentioned symptoms and easy choking on food and water. Physical examination revealed isolated muscle atrophy as well as wasting and fasciculation on the right side of the tongue compared with the left side, which resulted in the tongue’s deviation to the right side (Figure 1). Other neurological examinations, including primitive neurological reflexes, deep tendon reflex, and peripheral muscle strength of the extremities, were unremarkable.

Computed tomography demonstrated an osteolytic lesion with a contrast-enhancing soft tissue component in the right clivus bone (Figure 2), which directly invaded the right hypoglossal canal (Figure 2B, arrow). The patient’s symptoms and deviated tongue improved with local irradiation, monthly subcutaneous administration of denosumab, and systemic chemotherapy (Appendix, available online).

DISCUSSION
Common primary tumors with metastases to the base of the skull include those of the breast, lungs, and prostate, whereas relatively rare tumors involving such a complication are renal cell carcinoma, thyroid cancer, and melanoma. In most cases, tumor cells metastasize to the base of the skull or the calvarium via hematogenous arterial circulation. Alternatively, direct extracranial extension through the skull base in head and neck malignancy could be another differential diagnosis. Although skull metastases from malignancy are not uncommon, they are often neglected owing to their asymptomatic nature and limited localized pain. Most are not detected until signs and symptoms from the stretching of the dura, compression of the cranial nerves, irritation (with resulting edema) of the adjacent brain tissue, and, less commonly, occlusion of the dural venous sinuses are observed.
Furthermore, results of previous studies suggest that the clinical manifestations of cranial neuropathy often vary from clinical syndromes involving multiple cranial nerves to isolated cranial nerve lesions.

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Figure 1 Obvious muscle atrophy of the right side of the tongue with deviation to the right side.
In general, it is not uncommon for hypoglossal nerve palsy to concurrently occur with other cranial nerve symptoms such as facial nerve paralysis and ophthalmic manifestations. However, isolated hypoglossal nerve palsy is relatively rare and considered a diagnostic challenge. In most etiologies of isolated hypoglossal nerve paralysis, malignancy is the leading cause, followed by gunshot injury and neurological disorders. Unlike cerebral vascular accidents, the metastatic lesions that cause hypoglossal paralysis are usually unilateral, resulting in isolated paralysis and tongue deviation.

The hypoglossal nerve is responsible for the tongue’s movement and, as a result, pronunciation. Injuries to this nerve cause the tongue to deviate toward the weaker side. Lesions of the hypoglossal nerve can be classified into 3 categories—supranuclear, nuclear, and infranuclear—based on the location. The clinical manifestations often involve mild weakening/paralysis of the tongue or could even be asymptomatic in patients with supranuclear lesions, which can be explained by the unaffected side compensating for the lesion’s effects. However, nuclear or infranuclear lesions often result in ipsilateral tongue muscle atrophy and fasciculation, subsequently causing the tongue to deviate toward the side of the lesion. Additionally, nuclear lesions usually involve other neurologic deficits and present with various features of brainstem involvement. On the other hand, the manifestations of infranuclear diseases often include fewer upper motor neuron signs and other associated symptoms (similar to medulla symptoms) than that of nuclear lesions.

A systemic approach should be considered when evaluating infranuclear lesions to sequentially include the medullary, cisternal, skull base, and extracranial segments. However, the most common etiology involving all the above-mentioned sites is still neoplasm, which could involve metastatic or primary lesions with direct invasion. Other rare causes include neurovascular issues, multiple sclerosis, demyelinating diseases, or infection. In the skull base segment, metastatic lesions from a primary breast, lung, or prostate tumor should be prioritized over a head and neck tumor with direct invasion. However, owing to the chronic disease process, the initial symptoms are often ignored. Most patients notice the symptoms only when obvious tongue deviation, choking, and dysarthria occur. Although magnetic resonance imaging remains the modality of choice to diagnose metastatic lesions, computed tomography of the skull base can be utilized as an initial procedure. Further management should otherwise focus on the underlying causes of hypoglossal nerve palsy. In conclusion, careful physical examination with appropriate imaging studies is essential in investigating isolated hypoglossal nerve palsy, allowing physicians to properly detect occult metastatic lesions in the skull base.

References

Appendix
A video showing the condition of the tongue posttreatment is available as supplementary material, in the online version of this article, at http://dx.doi.org/10.1016/j.ajnmed.2014.03.040.