A tardy tongue palsy

Delayed post-traumatic isolated unilateral hypoglossal neuropathy

Cullen M. O'Gorman, MBBS; E. Paul Lindell, MD; Jamie J. Van Gompel, MD; James P. Klaas, MD

A 76-year-old man presented with insidious onset of slurred speech and difficulty swallowing over 1 year. Initial symptoms were episodic imprecise articulation, tongue swelling, tongue deviation, and mild right-sided headache. Progressive deterioration of speech and swallowing function had occurred; at the time of presentation, the patient was struggling to swallow food and saliva. He had undergone C1-C5 posterior fusion following a C2 fracture 50 years prior. There was no relevant medical history and the patient was systemically well. Examination found hemiatrophy of the right side of the tongue with visible fasciculations. The tongue protruded to the right and there was associated dysarthria. There was no evidence of other cranial nerve involvement, cervical myelopathy, or a diffuse motor neuron disorder.

Imaging of the head found evidence of right atlanto-occipital arthritis with an osteophyte impinging upon the right exiting hypoglossal nerve at the hypoglossal canal. The right genioglossus was denervated and atrophic (figure). Vascular imaging excluded dissection of cervical or intracranial vessels. Needle EMG of the tongue found fibrillations and small, polyphasic varying motor units with markedly reduced recruitment in the right genioglossus, suggesting active denervation and reinnervation. EMG of left genioglossus and upper and lower limbs was unremarkable.

Extension of the cervical fusion was considered, but the patient elected to pursue conservative treatment and underwent a course of speech therapy.

DISCUSSION

The twelfth cranial nerve is a pure motor nerve primarily responsible for innervation of the intrinsic and extrinsic muscles of the tongue. Nerve injury causes speech and swallowing dysfunction, ipsilateral tongue paresis, and deviation to the side of the lesion. Hypoglossal nerve lesions are rare and result from diverse etiologies, most commonly benign or malignant tumors, trauma, stroke, vascular dissection, brainstem demyelination, infection, and iatrogenic injury.¹

In this case, the hypoglossal neuropathy most likely resulted from mechanical compromise of the nerve by an osteophyte arising from the right atlanto-occipital joint. There is heterotopic ossification outside of the hypoglossal canal; therefore direct nerve impingement is likely responsible for ongoing denervation. Arthritis of this joint typically presents with unilateral unremitting occipital-cervical pain exacerbated by neck movement and neurologic complications are rare. However, dynamic motion between the occipital cervical junction may also contribute to nerve injury. The history of trauma is relevant, since post-traumatic arthritis of the joint, or arthritis secondary to cervical spine fixation (i.e., adjacent segment disease), may be contributory. The long delay between the original neck injury and the onset of tongue weakness

Practical Implications

Hypoglossal neuropathy is a rare complication of neck trauma and degenerative spine disease. Atlanto-occipital arthritis should be considered as a possible cause for isolated hypoglossal nerve palsy following a careful evaluation for more common etiologies such as tumors, vascular dissection, or demyelinating disease.

Departments of Neurology (CMO, JPK), Neuroradiology (EPL), and Neurosurgery (JJVG), Mayo Clinic, Rochester, MN.

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Correspondence to: klaas.james@mayo.edu

Figure CT scan (selected images)

(A) Hemiatrophy of the right genioglossus muscle (arrow), (B) the hypoglossal canal (arrow) and associated degenerative joint disease and hypertrophic change causing narrowing of the canal exit (arrowhead), and (C) the degenerative right hypoglossal canal (arrowhead) compared to the left hypoglossal canal (arrow).

probably reflects slow growth of the osteophyte, although the cervical fixation may also have delayed the development of arthritis.

Traumatic hypoglossal neuropathy is usually associated with decelerative head injury and occipital condyle fracture.² Presentation of tongue weakness can be missed in the setting of other injuries.³ One study⁴ reported delayed hypoglossal neuropathy at 3 months following occipital condyle fracture, attributed to formation of a callus at the fracture site.

Isolated hypoglossal neuropathy has also been reported in association with synovial cysts arising from the atlanto-occipital joint⁵ or atlanto-axial joint,⁶ in the absence of neck trauma. Surgical fixation of the cervical spine, through an anterior or posterior approach, can also be associated with hypoglossal nerve injury.⁷

Arthritis of the atlanto-occipital joint may be missed by MRI; typical protocols omit coronal views, and axial images are taken caudal to C2. CT imaging includes reconstruction in 3 planes and is better suited to detect bony pathology at this level. Dedicated MRI of the craniocervical junction can also be performed to further evaluate for soft tissue or disc pathology.

Treatment of atlanto-occipital osteoarthritis includes rest, soft cervical collars, nonsteroidal anti-inflammatories, muscle relaxants, or tricyclic antidepressants. Occipitocervical arthrodesis can be considered in refractory cases, but results in considerable limitation of cervical mobility. In this case, nascent motor units were present in the right genioglossus muscle, suggesting that some reinnervation has occurred. Functional recovery will depend upon the extent of successful reinnervation.

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