Unilateral macroglossia as sole presenting manifestation of internal carotid artery dissection

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Abstract

A patient presented with acute-onset, painless, unilateral enlargement of the tongue. Steroid treatment for angioedema was ineffective, and a biopsy of an apparent mass of the tongue base showed normal tissue. Subsequent magnetic resonance imaging showed enlargement, enhancement, asymmetric T2 hyperintensity of the left half of the tongue, and dissection of the left cervical internal carotid artery (ICA) at the skull base. Unilateral enlargement of the tongue due to acute neurogenic denervation may be the sole clinical presentation of a spontaneous arterial dissection at the skull base. The hypoglossal neuropathy resulted from compression by the pseudoaneurysmal ICA dilatation or ischemia from interruption of the vasa nervorum.

Introduction

Painless macroglossia may be caused by amyloidosis (primary or secondary), hypothyroidism, lymphangioma, lipomatosis, reticulum-cell sarcoma, and non-Hodgkin lymphoma. Hypoglossal neuropathy paralyzes both extrinsic and intrinsic muscles of the tongue, and may result from many pathologic processes such as primary skull base neoplasms (e.g., paragangliomas), bony skull base metastases (e.g., from prostate cancer), traumatic skull base fractures, high cervical lymphadenopathy (e.g., tuberculosis), and internal carotid artery (ICA) dissection.

Cervical ICA dissection typically presents with ipsilateral laterocervical or facial pain, or Horner syndrome with or without cerebral hemisphere stroke. However, 3 to 12% of patients may present with isolated or multiple lower cranial neuropathies. As many as 95% of patients with ICA dissection complain of pain at onset, and pain is sometimes the only clinical manifestation; 0.5% of patients suffer no pain.

To the best of the author’s knowledge, this is the first report of a painless, unilateral enlargement of the tongue caused by acute hypoglossal nerve denervation as the sole presenting feature of a painless, spontaneous dissection of the distal cervical ICA at the skull base.

Case report

A 56-year-old man presented with an overnight-onset, painless enlargement of the left side of the tongue (figure 1). He complained of slurred speech and occasionally bit his tongue because of the swelling and perceived impaired control. He was initially unsuccessfully treated for angioedema with a 2-week course of oral corticosteroids. A subsequent indirect laryngoscopy-guided biopsy of an apparent submucosal infiltrative mass of the left side of the base of the tongue showed normal tissue. About 6 weeks after the onset of symptoms, a neurologic cranial nerve evaluation revealed subtle asymmetry of the soft palate arches with early atrophy and slight weakness of the left half of the tongue.

Dedicated high-resolution magnetic resonance imaging (MRI) was performed according to an oral cavity-neck protocol with extension to the skull base, utilizing axial and coronal, fat-saturated, T2-weighted; coronal T1-weighted; and post-gadolinium, axial and coronal, fat-saturated, T1-weighted sequences. MRI showed prominence of the left side of the tongue base in the oropharynx, best appreciated on T1-weighted images; asymmetric T2 hyperintensity; mild, diffuse enhancement of the left half of the tongue; and atrophy of the left levator veli palatini and tensor veli palatine muscles. The distal cervical portion of the left ICA directly below the skull base contained a luminal,
associated with noticeable weakness or atrophy of the tongue muscles. Furthermore, the underlying arterial dissection was spontaneous and painless and without evidence of cerebral ischemia. Nonrecognition of this phenomenon initially led to inappropriate treatment and a nondiagnostic, minor invasive procedure.

Unilateral hypoglossal neuropathy is clinically recognized by deviation of the protruded tongue toward the side of weakness. Additionally, when lying in the floor of the mouth, the tongue may seem enlarged and higher on the weakened side. In this patient, acute hypoglossal neuropathy caused unilateral tongue enlargement without paralysis, and it was mistaken as an infiltrative tongue lesion. Presumably, the acute and incomplete denervation did not cause tongue weakness; atrophy may take a few weeks to manifest. The pathophysiology of this partial hypoglossal nerve injury due to ICA dissection likely rested on compression by mild pseudoaneurysmal vessel dilatation. Alternatively, the expanding ICA-wall hematoma compressed the branches of the ascending pharyngeal artery, thereby interrupting the blood supply via vasa nervorum of the hypoglossal nerve.

Two pathogenetic mechanisms have been proposed to explain the painless nature of the ICA dissection. First, visceral nociceptor adaptation occurs if a dissection develops slowly. Un. Like traumatic dissections, spontaneous dissections can sometimes develop gradually and therefore cause no pain. A second likely explanation involves structural anomalies in the vessel wall that increase distensibility, thus reducing nociceptor stimulation.

Semilunar, peripheral T1- and T2-hyperintense signal abnormality compatible with a subacute mural thrombus, with an intimal flap indicative of a carotid dissection that measured 2.5 cm in length. Mild pseudoaneurysmal enlargement of the left ICA was noted; the vessel lumen was not compromised (figure 2).

Subsequent brain images showed no evidence of an ischemic complication in the left ICA distribution. These radiographic findings were all compatible with subacute neurogenic denervation of the left half of the tongue from a hypoglossal nerve injury due to ICA dissection.

Treatment was started with an antiplatelet regimen. The patient reported full recovery of tongue bulk and function during a telephone interview 1 year after presentation.

Discussion
To the best of the author’s knowledge, this is the first report of a painless, unilateral pseudomass of the tongue as the sole presenting clinical feature of a spontaneous arterial dissection at the skull base. The neurogenic nature of this unilateral tongue enlargement was initially not appreciated, as the hypoglossal nerve injury was not associated with noticeble weakness or atrophy of the tongue muscles. Furthermore, the underlying arterial dissection was spontaneous and painless and without evidence of cerebral ischemia. Nonrecognition of this phenomenon initially led to inappropriate treatment and a nondiagnostic, minor invasive procedure.

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![Figure 1](image1.png)
Figure 1. Photo shows the midline protrusion with increased bulk of the left side of the tongue, suggesting an infiltrative lesion.

![Figure 2](image2.png)
Figure 2. A: Left axial T2-weighted MRI of the oropharynx shows asymmetric T2 hyperintensity and enlargement and enhancement of the tongue, suggestive of subacute neurogenic denervation. B: Right axial T2-weighted MRI image of the skull base shows neurogenic atrophy of the left levator veli palatini muscle (arrow) and a semilunar intramural clot of the left distal internal carotid artery dissection (arrowhead).
In this patient, the MRI soft-tissue study of the oropharynx and neck showed increased signal intensity on T2-weighted images on the left half of the tongue, compatible with acute or subacute denervation.\textsuperscript{16,17} Denervation was associated with diffuse unilateral enhancement with gadolinium, which could create the false impression of an infiltrative tongue lesion. However, closer inspection of the imaging studies showed presumed neurogenic atrophy of the left-sided soft-palate muscles, implicating additional partial nerve injuries to the ipsilateral pharyngeal branch of the left vagus nerve and the otitic branch of the mandibular division of the left trigeminal nerve. Furthermore, cephalic extension of the imaging study protocol to include the skull base permitted the detection of an intramural ICA clot and intimal flap compatible with the causative, presumably spontaneous, distal cervical ICA dissection.

Knowledge of the relevant anatomy is vital to understanding the MRI patterns of muscle denervation caused by various cranial neuropathies. Furthermore, the signal changes seen in denervated muscle depend on the time course of the underlying pathology.\textsuperscript{18,19}

Acute and subacute denervation results in prolonged T2 relaxation time, producing increased signal in muscle on STIR (short tau inversion recovery) and T2 fat-saturated images. This signal change starts within 48 hours of denervation of muscle and increases with intensity over time. Signal change is thought to result from an enlargement of the capillary bed in the muscle or shift of fluid to the extracellular space. This increased fluid signal in muscle may also be seen in other pathologic processes such as injury, infection, or inflammation. Chronic denervation of the tongue is characterized by fatty atrophy of muscle with increased fat content between muscle fascicles.\textsuperscript{20} This results in increased muscle signal on T1-weighted images with decreased overall muscle bulk.

In conclusion, this report draws attention to the phenomenon of painless, unilateral enlargement of the tongue as the sole clinical presentation of an acute hypoglossal neuropathy due to a spontaneous cervical ICA dissection at the skull base. Appropriate imaging sequences with attention to anatomic detail and altered signal characteristics of structures may help avoid inappropriate treatment and more-or-less invasive diagnostic procedures.

References