

A Rare Cause of Unilateral Hypoglossal Nerve Palsy: Intraneural Ganglion Cyst of the Hypoglossal Nerve

Jeremie Oliver, BS BA¹, Vijay Agarwal, MD², Jeffrey Janus, MD³, Jamie Van Gompel, MD^{2,3}

¹Mayo Clinic School of Medicine; ²Department of Neurosurgery; ³Department of Otorhinolaryngology-Head and Neck Surgery
Mayo Clinic, Rochester, MN

Case Presentation

A 56-year-old male presented to our department with a reported history of seven years of left tongue weakness and progressive atrophy, as well as left palatal numbness, with no pain reported.

Biting his tongue more on the left-hand side.

No change in the quality or character of his voice or articulation.

Previous MRI studies performed at an outside institution revealed increased T2 signal enhancement of the hypoglossal nerve in the area of the hypoglossal canal. No intervention was performed at that time.

Follow-up imaging several years later clearly demonstrated a progressively enlarging lesion in the left hypoglossal foramen being T2 hyperintense with no gadolinium contrast enhancement.

Ultimately MRI studies one-year later showed substantial expansion of the lesion to approximately 2.5-cm.

Further CT imaging at that time demonstrated widening of bone in the region of the hypoglossal canal, as well as a very small amount of enhancement on the anterior portion of the lesion.

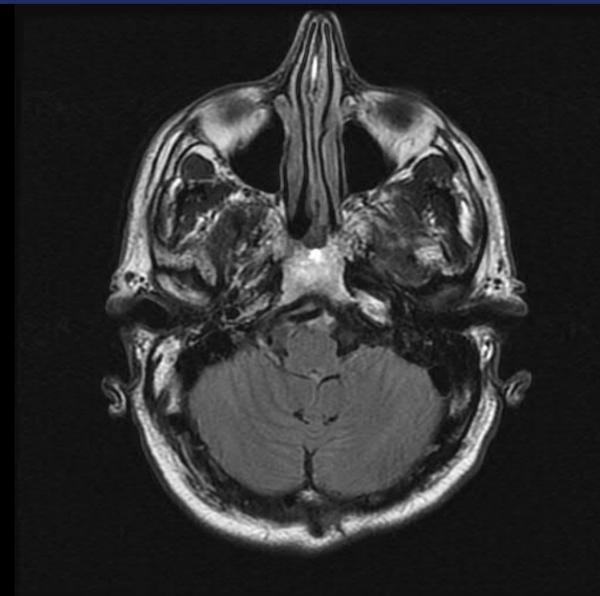
Brainstem compression was notable, as well as hypoglossal and cerebellar tonsillar compression, at the time of meeting the patient, surgery was offered.

Pre-Op Axial T2 Flair



Axial, T2-weighted MRI: Hyperintense, cystic lesion related hypoglossal canal

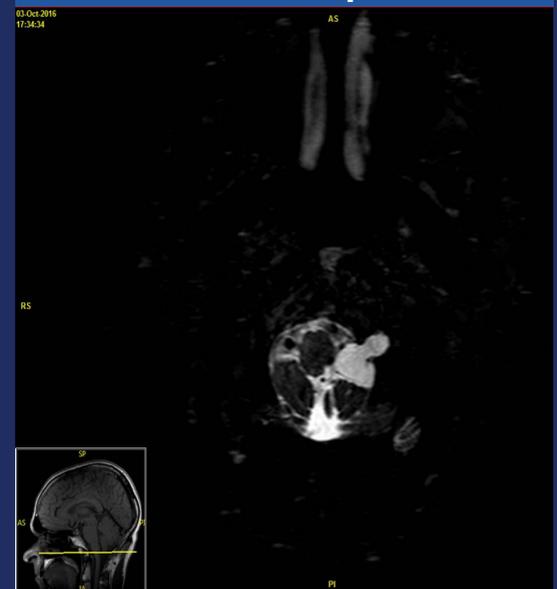
Post-Op Axial T2 Flair



All Reported Intraneural Ganglion Cysts Involving Hypoglossal Nerve

Characteristic	Bilgin-Freiert et al 2015	Gambhir et al 2011	Nonaka et al 2010	Baldauf et al 2005	Present Case
Age (years)	Not reported	70	54	51	56
Sex	M	M	M	M	M
Laterality	Left	Left	Left	Left	Left
Symptoms	Slurred speech	CNXII palsy, mildly slurred speech	Dysphagia, hoarseness, shoulder weakness	Slurred speech	Palatal weakness, numbness
Signs	Left tongue atrophy	Tongue deviation (left), atrophy	Left tongue atrophy (7-yr hx)	Left tongue atrophy	Left tongue atrophy (7-yr hx)
Imaging (MRI)	T1: no; T2: high	T1: iso; T2: high	T1: no; T2: high	T1: iso, no; T2: high (CT: hypoglossal canal enlargement)	T1: no; T2: high (CT: widening of bone in the region of the hypoglossal canal; brainstem, hypoglossa, and tonsillar compression)
Surgical Approach	Transcondylar	Stereotactic suboccipital craniotomy	Transcondylar suboccipital	Far-lateral	Supra-condylar far-lateral

Pre-Op Axial T2 3D Brainstem Compression



Surgical Approach

Surgical resection of the hypoglossal nerve lesion included a left sub-occipital craniotomy and C1 laminectomy to facilitate the supracondylar far lateral left-sided approach. Intraoperative neural monitoring performed on CNX, XI, and XII on the left side, including somatosensory evoked potential (SSEP) monitoring, yielded no changes during resection.

A midline incision was made along the avascular plane down to the C1 and C2 region to expose the sub-occipital region. The sub-occipital region was eccentrically exposed to the left side.

Cystic tumor of CNXII was found filling the foramen of Lushka. The vertebral artery was dissected away from the tumor as well as the spinal portion of the spinal accessory nerve. The tumor itself was found to be mostly cystic, decompressing quite easily.

Working around the margins of the extra-foraminal portion of the tumor, all nerve rootlets were dissected away, and ultimately removed the tumor completely. Foramen of Lushka was expanded, and given the position, we were able to look directly out it. Approximately three nerve rootlets of the hypoglossal nerve were preserved in the inferior aspect of the canal.

Continued aggressive removal of tumor was performed until there was no evidence of further tumor. The jugular foramen contents were intact without any evidence of disruption. The spinal portion of the spinal accessory nerve was intact.

Intraoperative and immediate postoperative course uneventful. Final pathology returned a ganglion cyst of the hypoglossal nerve. Follow-up visit at 4-months postoperatively was completely benign, with revealed absence of ganglion cyst on MRI.

Conclusions

We present a case of intraneural involvement of the cystic lesion within the hypoglossal nerve, which constitutes only the fifth reported case to date.

We suspect the origin of the cyst to have been the occipital condylar joint.

This case represents the only in which palatal numbness was described as a symptom, and only the second case to describe hypoglossal canal widening on CT.

First to describe microdissection of accessory nerve rootlets encompassing the vertebral artery during removal of the intraneural ganglion cyst.

Given the intra-dural location of this patient's lesion, our approach involved a lateral sub-occipital exposure, combining a left sub-occipital craniotomy and C1 laminectomy to facilitate the supracondylar far lateral left-sided approach. This approach provided excellent exposure of the cerebellomedullary midline raphe, thereby allowing complete tumor removal without risk of instability.