Hypoglossal Nerve Schwannoma Mimicking a Vagal Nerve Schwannoma

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Introduction

- Schwannomas are relatively common benign tumors of peripheral nerve sheath; rarely arising from purely motor nerves such as cranial nerve XII(1).
- As of 2018, only 94 case reported
- Even though histopathology has not been proven to be effective, it led us to the correct diagnosis. This was ultimately confirmed with post-excisional pathology (2).
- MRI can demonstrate a characteristic enhancement pattern (7) and in this case, origin of the tumor was presumed to be cranial nerve X due to anatomical location on imaging.

We present a case where symptoms, imaging and histopathology raised suspicion for vagal nerve schwannoma, but intra-operatively was found to be emanating from the hypoglossal nerve.

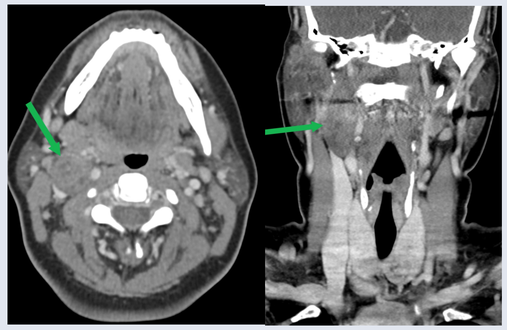


Figure 1 - Axial (left) and coronal (right) post-contrast CT images of the neck demonstrate a heterogeneous, well-circumscribed mass (**arrows**) within the right carotid space.

Case Presentation

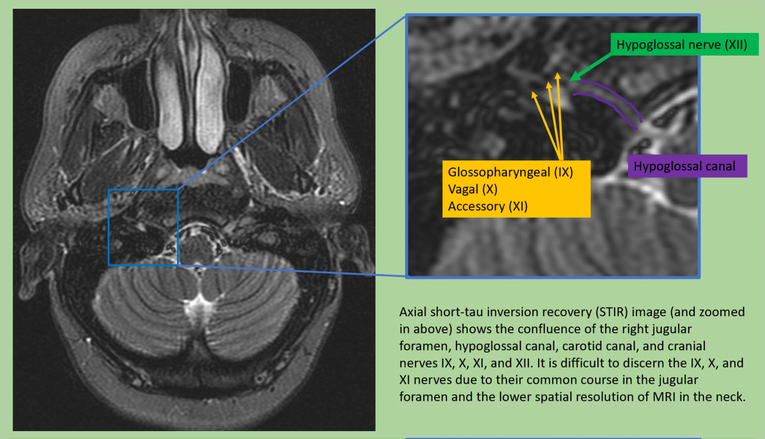
- 36-year-old woman with incidentally found soft tissue mass on MRI associated with right neck fullness and anxiety attacks
- Firm, immobile, level IIA neck mass on exam, otherwise normal
- MRI showed a 1.9 x 2.2 x 3.2 cm mass within the right post-styloid space
- 2 years later with no treatment (patient choice), symptoms included odynophagia, dysphagia, and dysphonia
- Repeat imaging showed that the mass had grown to 2.8 x 2.7 x 6.6 cm

Intra-operatively

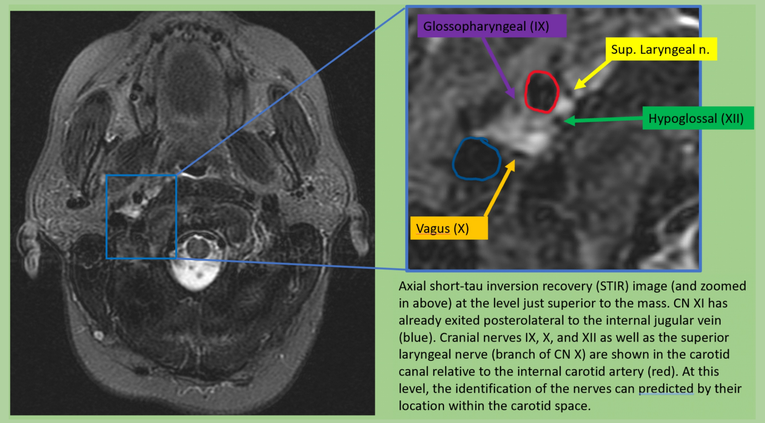
- Dissection was carried out to expose carotid sheath and identification of CN XI-XII.
- As dissection continued posterior to the hypoglossal nerve, it became evident that the mass was emanating from this nerve.
- The mass was successfully removed en-bloc from the nerve sheath, however due to involvement of the main body of the nerve, the hypoglossal nerve was sacrificed.
- There was insufficient length for primary approximation, therefore a cervical branch of the facial nerve was used for neurorrhaphy.

Post-operative course and follow up:

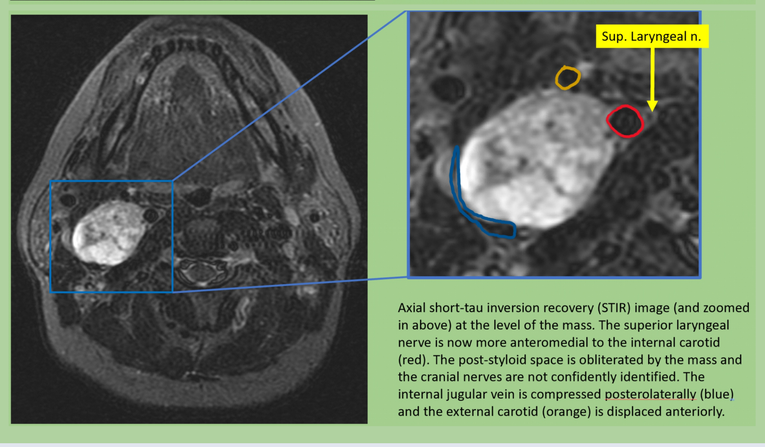
- • Mild right sided tongue weakness and difficulty articulating immediately post-op.



Axial short-tau inversion recovery (STIR) image (and zoomed in above) shows the confluence of the right jugular foramen, hypoglossal canal, carotid canal, and cranial nerves IX, X, XI, and XII. It is difficult to discern the IX, X, and XI nerves due to their common course in the jugular foramen and the lower spatial resolution of MRI in the neck.



Axial short-tau inversion recovery (STIR) image (and zoomed in above) at the level just superior to the mass. CN XI has already exited posterolateral to the internal jugular vein (blue). Cranial nerves IX, X, and XII as well as the superior laryngeal nerve (branch of CN X) are shown in the carotid canal relative to the internal carotid artery (red). At this level, the identification of the nerves can predicted by their location within the carotid space.



Axial short-tau inversion recovery (STIR) image (and zoomed in above) at the level of the mass. The superior laryngeal nerve is now more anteromedial to the internal carotid (red). The post-styloid space is obliterated by the mass and the cranial nerves are not confidently identified. The internal jugular vein is compressed posterolaterally (blue) and the external carotid (orange) is displaced anteriorly.

References

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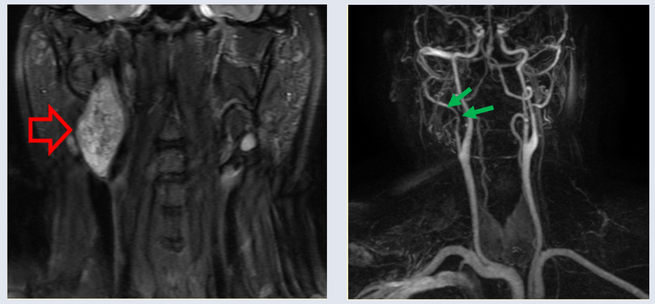

Coronal T2 fat saturated (left) and coronal maximum intensity projection (MIP) of the Time-Resolved Imaging of Contrast KineticS (TRICKS) sequence in the later arterial phase (right). The mass is heterogeneously T2 hyperintense and appears as a spindle-shaped, well-circumscribed mass in the post-styloid space (thick arrow).
Note the medial displacement of the right ICA and ECA on the TRICKS image (arrows). There is absence of contrast enhancement in the area of the mass characteristic of the venous or delayed pattern of a Schwannoma as opposed to the intense arterial pattern expected of a paraganglioma.

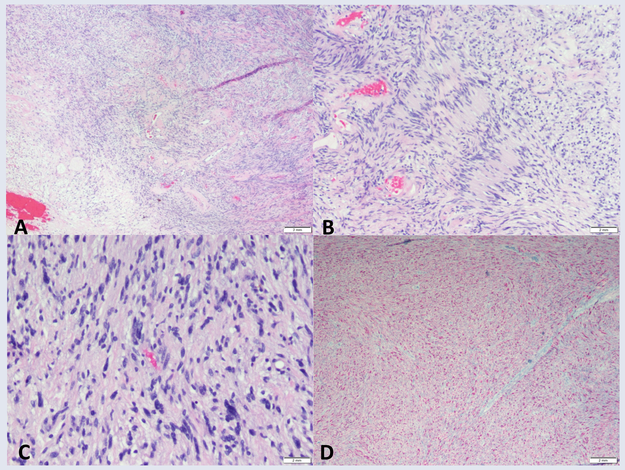

Figure 6 – Pathologic assessment. Biphasic hypercellular Antoni A region and hypocellular Antoni B region (Image A). Vercoay bodies with nuclear palisading (image B). Degenerating Schwan cell (Image C). S100 (nuclear and cytoplasmic), confirming neural origin (image D)

Literature review

- 93% of cases undergo surgical excision and 67% of these have permanent mild and well tolerated neurological deficits.
- 94 reported cases of which 6 patient underwent resection with need for nerve neurorrhaphy or grafting. All with post-operative dysarthria and tongue weakness, but with resolution of symptoms within 6 months.

Conclusion

Hypoglossal nerve schwannomas are rare head and neck tumors with difficult diagnosis due to its location, overlapping features, and function. Histopathology and imaging characteristics of our patient's lesion was suggestive of vagal origin due to this being more likely than hypoglossal nerve schwannomas. However, pinpointing the correct nerve was difficult prior to surgery. Despite successful en-block resection of the mass, neurorrhaphy had to be performed intra-operatively due nerve compromise with a good clinical outcome.