Unilateral hypoglossal nerve palsy caused by an intraneural ganglion cyst

Case report

Yoichi Nonaka, M.D., Ph.D.,1 Peter M. Grossi, M.D.,1 Carol A. Filomena, M.D.,2 Allan H. Friedman, M.D.,1 Takanori Fukushima, M.D., D.M.Sc.1

Divisions of 1Neurosurgery and 2Pathology, Duke University Medical Center, Durham, North Carolina

The authors describe a rare case of unilateral hypoglossal nerve palsy caused by an intraneural ganglion cyst. Three similar cases have been reported with pathological classification still under consideration. One case was classified as an intraneural ganglion cyst and 2 cases were classified as atlantooccipital joint synovial cysts. (DOI: 10.3171/2010.1.JNS091526)

Key Words • intraneural ganglion cyst • synovial cyst • atlantooccipital joint • hypoglossal nerve palsy • transcondylar approach

The authors report on the rare case of an intraneural ganglion cyst and discuss the lesion’s histopathological features and surgical management.

There are only 3 previously reported cases of intraneural ganglion cysts that have resulted in isolated unilateral hypoglossal nerve palsy.1–3 Mujic et al.3 and Elhammady et al.2 concluded that their 2 cases were best classified as atlantooccipital joint synovial cysts. Baldauf et al.1 presented a case of hypoglossal nerve palsy caused by an intraneural ganglion cyst unrelated to the atlantooccipital joint. An intraneural ganglion cyst is most commonly seen as a peroneal nerve lesion.4–6 The present case is the fourth reported case of unilateral hypoglossal nerve palsy resulting from an intraneural ganglion cyst.

Case Report

History. This 54-year-old man originally presented with left-sided tongue atrophy and lower CN dysfunction including, choking, swallowing difficulty, hoarseness, and mild shoulder weakness. The symptoms slowly progressed over the past 7 years and recently became worse.

Examination. Except for tongue atrophy and dysfunction of the IX, X and XI CNs, the patient had no other neurological deficit. The MR images obtained 7 years prior and initially interpreted as negative retrospectively revealed a small 3–4-mm mass lesion located anterolateral to the medulla (Fig. 1). On a recent MR imaging (Fig. 2), we identified a 14 × 15–mm mass adjacent to the left side of the medulla that compressed and displaced the medulla and extended into the left hypoglossal canal. This mass was best demonstrated on the high-resolution and routine T2-weighted axial and coronal MR images. It has low signal on T1-weighted images, similar to CSF, with no convincing enhancement.

Operation. The patient was placed in the left-side-up lateral position with his head supported with a 3-pin head clamp. A C-shaped scalp incision and a subgaleal scalp elevation were made, suboccipital fascia was taken for watertight closure, and the muscle layer was split to expose the left mastoid and suboccipital fossa. Presigmoid dura mater and retrosigmoid dura mater were confirmed, and suboccipital bone was removed. After the craniotomy and completion of the far-lateral transcondylar approach, the dura was opened in a curvilinear fashion. When CNs IX and X were identified, they did not appear to be compressed by the tumor. Tumor compression involved the lateral medulla. The tumor was white, transparent, and

Abbreviations used in this paper: CN = cranial nerve; PICA = posterior inferior cerebellar artery.
Hypoglossal nerve palsy caused by an intraneural ganglion cyst

appeared as a gelatinous lesion originating from the full width of the left hypoglossal canal. The 11th spinal nerve and PICA were compressed posteriorly by the tumor (Fig. 3 left). The 11th spinal nerve was dissected and retracted medially to maintain the medullary branch of this nerve axis. The PICA was also separated carefully and retracted medially to expose the entire tumor and to clarify its origin. The IX, X and XI CNs were intact at the entrance of the jugular foramen. The superior edge of the tumor was 2–3 mm away from the jugular foramen. After gradual elevation of the tumor, severe adhesion was observed at the anterior wall of the hypoglossal canal (Fig. 3 right), and the tumor was observed to originate from the hypoglossal canal. The origin of the tumor was coagulated and the tumor was excised. Osseous distraction was not identified inside of the hypoglossal canal. The cisternal segments of CN XII were fused into the tumor. The last part of the tumor was separated from the hypoglossal nerve pontine stump.

Postoperative Course. The patient was able to swallow without difficulty and his hoarseness improved slightly over the next several days. His swallowing disturbance and hoarseness were attributed to brainstem compression rather than CN IX and X compression. Tongue atrophy and deviation did not resolve, but the patient is otherwise neurologically asymptomatic. He was discharged on postoperative Day 4. Six months after surgery, follow-up MR imaging demonstrated no recurrence of the tumor (Fig. 4 right).

Discussion

The most common location of an intraneural ganglion cyst is in the peripheral nerves, which course near joints and tendon sheaths. Ganglion cysts become symptomatic when they compress or invade adjacent nerves. The presence of a CN ganglion cyst is uncommon. Three cases have been previously reported (Table 1).

Mujic et al. have reported a case of an atlantooccipital joint synovial cyst that involved the left hypoglossal

Fig. 1. Initial axial T1-weighted MR image acquired 7 years prior to our treatment, demonstrating a hypointense small lesion located anterolateral to the left medulla.

Fig. 2. Preoperative axial T2-weighted MR image revealing a hyperintense mass lesion compressing the left medulla.

Fig. 3. Intraoperative photographs. Left: View through the microscope demonstrating the transparent multilobular cystic lesion arising from the left hypoglossal canal and compressing the PICA and spinal accessory nerve. Right: The white part of the tumor is adherent to the inner wall of the left hypoglossal canal.

Fig. 4. Left: Intraoperative photograph obtained after total resection of the tumor. Right: Six-month postoperative axial T2-weighted MR image demonstrating complete resection of the lesion and no recurrence.
canal and extended inferiorly adjacent to the C-1 lateral mass. Baldauf et al.\(^1\) reported a case of isolated hypoglossal nerve palsy caused by an intraneural ganglion cyst at the entrance of the hypoglossal canal. Elhammady et al.\(^2\) have reported on an atlantooccipital joint synovial cyst. There are many similarities including symptoms, neurological deficits, patient age, MR imaging findings, and tumor appearance in all of these cases. The histopathological classifications were slightly different. Mujic et al. and Elhammady et al. reported their cases as “atlanto-occipital joint synovial cysts.” Baldauf et al. classified their case as an “intraneural ganglion cyst.”

In their report, Mujic et al.\(^3\) described the histopathological features as a cyst wall consisting entirely of loose fibrous connective tissue with focal myxoid change. No epithelial lining and no associated neural tissue were identified. These findings were consistent with a synovial cyst formed from the penetration of the dura by synovial contents from the left atlantooccipital joint and resulting in the formation of mucoid luminal material, a myxoid edematous capsular wall, and a topographical association with the occipitocervical joint. Elhammady et al.\(^2\) have reported a case of a juxtafacet cyst of the left atlantooccipital joint that can be classified as a synovial cyst lined with synovial cells and containing clear or xanthochromic fluid, or as a ganglion cyst without a synovial lining and with gelatinous content. Occasionally a ganglion cyst may invade and compress a nerve and produce an intra-

### TABLE 1: Summary of published case reports on isolated hypoglossal nerve palsy caused by atlantooccipital joint synovial cysts*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Mujic et al., 2003</th>
<th>Baldauf et al., 2005</th>
<th>Elhammady et al., 2009</th>
<th>Present Case</th>
</tr>
</thead>
<tbody>
<tr>
<td>age (yrs)</td>
<td>52</td>
<td>51</td>
<td>67</td>
<td>54</td>
</tr>
<tr>
<td>sex</td>
<td>male</td>
<td>female</td>
<td>female</td>
<td>male</td>
</tr>
<tr>
<td>side</td>
<td>lt</td>
<td>lt</td>
<td>lt</td>
<td>lt</td>
</tr>
<tr>
<td>Sx</td>
<td>3-wk hx of dysarthria &amp; difficulty swallowing</td>
<td>6-mo hx of slurred speech</td>
<td>3-mo hx of lt jaw &amp; facial pain, neck discomfort, difficulty w/ speech &amp; swallowing, 7-yr history of tongue atrophy, swallowing disturbance, hoarseness, &amp; worsening shoulder weakness</td>
<td></td>
</tr>
<tr>
<td>signs</td>
<td>lt tongue hemiatriphy</td>
<td>lt tongue hemiatriphy</td>
<td>lt tongue hemiatriphy &amp; gait ataxia</td>
<td>tongue atrophy</td>
</tr>
<tr>
<td>MRI</td>
<td>T1: no enhancement; T2: high</td>
<td>T1: iso, no enhancement; T2: high (CT: hypoglossal canal enlargement)</td>
<td>T1: iso, no enhancement; T2: high</td>
<td>T1: no enhancement; T2: high</td>
</tr>
<tr>
<td>approach</td>
<td>far-lateral</td>
<td>far-lateral</td>
<td>juxtacondylar</td>
<td>transcondylar suboccipital</td>
</tr>
<tr>
<td>findings</td>
<td>clear, jellylike content</td>
<td>gelatinous</td>
<td>jellylike, amorphous mucoid</td>
<td>gelatinous, multicystic</td>
</tr>
<tr>
<td>location</td>
<td>extra- &amp; intradural atlantooccipital joint</td>
<td>extra- &amp; intradural (intraneural)</td>
<td>extradural</td>
<td>intradural</td>
</tr>
<tr>
<td>pathology</td>
<td>synovial cyst (synovial substance from the AO joint infiltrated the nerve)</td>
<td>intraneural ganglion cyst (nerve fibers infiltrated &amp; surrounded by myxoid connective tissue)</td>
<td>similar to intraneural ganglion cyst (fragments of nerve w/ intervening strips of fibrous tissue; surrounding fibrous tissue &amp; adherent nerve fibers wrapping around tumor)</td>
<td></td>
</tr>
</tbody>
</table>

* Adapted and expanded with permission from Elhammady MS et al: J Neurosurg Spine 10:234–239, 2009. Abbreviations: AO = atlantooccipital; iso = isointense; T1 = T1-weighted; T2 = T2-weighted.

**Fig. 5.** Photomicrographs. **Left:** Low-power image showing the nerve with intervening strips of fibrous tissue. The surrounding fibrous tissue and adherent nerve fibers wrap around the lesion. The fibrous wall of the ganglion cyst is adjacent to peripheral nerve (lower-right area in image). **Right:** High-power magnification of the fibrous wall of the ganglion cyst and adjacent peripheral nerve. H & E, original magnification × 20 (left) and 100 (right).
neural cyst. A photomicrograph revealed a cystic space lined by synovium in an H & E–stained slide.

Baldauf et al. have classified their case as an intraneural ganglion cyst and concluded that the anatomical relationship of the hypoglossal nerve and the hypoglossal canal to the atlantooccipital joint was responsible for the formation of the synovial cyst. The lower branch of the hypoglossal nerve was infiltrated and expanded by the cyst. A photomicrograph demonstrated myxoid connective tissue characteristics of intraneural ganglion cyst, and the immunohistochemical stain for S100 positively stained the involved hypoglossal nerve fibers. The authors concluded that the origin of the cyst was intraneural.

In the present case, the cystic lesion was localized to the hypoglossal canal under the jugular bulb with medial and inferior extension with brainstem compression. The tumor was firmly attached to the dura at the entrance of the hypoglossal canal without dural adhesion at the condyle. The remaining branches of the hypoglossal nerve were fused to the tumor. Bone erosion and hypoglossal canal enlargement were not present. The findings confirmed that the tumor originated from the hypoglossal nerve rather than from the atlantooccipital joint. Microscopically, there were fragments of nerve with intervening strips of fibrous tissue (Fig. 5). The trichrome stain highlighted surrounding fibrous tissue and adherent nerve fibers wrapped around the tumor. Based on these findings, this lesion was classified as an intraneural ganglion cyst of the hypoglossal nerve. Although many theories have been proposed to explain the pathogenesis of juxtafacet cysts and intraneural ganglia, the origin and pathogenesis of these cystic tumors remain unclear.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Y Nonaka, T Fukushima. Acquisition of data: Y Nonaka, PM Grossi, CA Filomena, T Fukushima. Analysis and interpretation of data: Y Nonaka, T Fukushima. Drafting the article: Y Nonaka. Critically revising the article: CA Filomena, T Fukushima. Reviewed final version of the manuscript and approved it for submission: Y Nonaka, PM Grossi, CA Filomena, AH Friedman, T Fukushima. Administrative/technical/material support: PM Grossi, CA Filomena, T Fukushima. Study supervision: AH Friedman, T Fukushima.

References