Management of rare atlantoaxial synovial cyst case with extension to the cerebellopontine angle: illustrative case

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BACKGROUND Synovial cysts are a common finding in degenerative spine disease, most frequently involving the facet joints of the lumbar spine. Synovial cysts are less common in the cervical spine and rarely involve the atlantoaxial junction.

OBSERVATIONS In this case report, the authors detail a unique presentation of a left atlantoaxial synovial cyst with large intracranial extension into the cerebellopontine angle causing progressive cranial nerve palsies resulting in tinnitus, vertigo, diminished hearing, gait imbalance, left trigeminal hypesthesia, left facial weakness, and dysarthria. The patient underwent a retromastoid craniectomy for resection of the synovial cyst, resulting in improvement and resolution of symptoms. Follow-up occurred at 6 weeks, 3 months, and 5 months postoperatively without recurrence on imaging.

LESSONS The authors describe acute and long-term management of a unique presentation of an atlantoaxial synovial cyst including retromastoid craniectomy, intervals for follow-up for recurrence, and possible treatment options in cases of recurrence. A systematic literature review was also performed to explore all reported cases of craniocervical junction synovial cysts and subsequent surgical management.

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KEYWORDS atlantoaxial cyst; synovial cyst; juxtafacet cyst; cerebellopontine angle mass

Synovial cysts have been reported since the late 19th century, when they were reported during an autopsy by von Gruker, with the clinical diagnosis first reported in 1968.1 Their exact etiology remains unknown but is believed to be from micromotion-associated degeneration of the capsular ligament, leading to progressive fluid accumulation within the collagenous capsule. Synovial cysts occur primarily in the lumbar spine, with a lower frequency reported in the cervical and thoracic spine. Frequently, they can be asymptomatic, or they present with signs and symptoms of neural element compression. After a thorough review of the literature, we believe that this is the only case with a synovial cyst extending to the cerebellopontine angle. The preoperative workup, surgical treatment, and outcome are discussed.

Illustrative Case

A 68-year-old female with a prior medical history of hypertension, hyperlipidemia, diabetes, epilepsy, mild cerebral palsy, and a significant smoking history presented to her primary care physician for an annual visit with complaints of balance difficulty, vertigo, ear fullness, and decreased hearing on the left side, with symptoms developing over a few weeks. She was evaluated by ear, nose, and throat personnel and was noted to have had several weeks of tinnitus prior to developing diminished hearing on her left side along with long-standing right-sided diminished hearing due to a history of head trauma. Additionally, she developed left-sided facial droop, left-sided sharp pain along the skull, drooling, and speech changes. She underwent audiography, which demonstrated sensorineural hearing loss on the left and moderately severe hearing loss on the right. She was prescribed prednisone, and magnetic resonance imaging (MRI) of the brain and internal auditory canal were ordered. In the interim, her facial droop, drooling, and speech were noted to worsen, so she presented to the emergency department, where a noncontrast head computed tomography (CT) revealed a 1.6 × 3.2 cm hypodense cystic lesion at the left cerebellopontine angle (CPA), with mass effect on the brainstem.
The lesion was new compared to a previously unremarkable head CT 2 years prior. She was therefore urgently referred to the neurosurgery clinic, where neurological examination revealed a House-Brackmann grade 3 facial palsy (inability to raise left eyebrow, weakness of left eyelid closure, and inability to puff out left cheek), diminished sensation in the left V2 and V3 dermatomes, and left-sided hearing loss with intact cranial nerve function on the right. Visual acuity and extraocular movements were intact. Motor and sensory examination in the extremities revealed symmetric strength and no signs of hyperreflexia.

She underwent MRI of the brain, which revealed a 3.2 × 2.1 × 4.4 cm bilobed cystic lesion centered at the left CPA (Fig. 1). The cystic component was isointense to cerebrospinal fluid (CSF) on both T1 and T2 sequences, with no enhancement of the cyst wall after gadolinium administration. Three-dimensional constructive interference in steady state (CISS) imaging revealed the mass continued inferiorly with exertion of local mass effect on the ventral medulla, left pons, left cerebellar peduncle, left cranial nerves (CNs) VII and VIII, cisternal segment of the left CNs V and VI, with minimal extension through the left foramen lacerum into the proximal internal auditory canal without expansion of the canal (Fig. 2). The cyst extended inferiorly to the left side of the median atlantoaxial joint. Given the patient’s age and longstanding smoking history, a chest radiograph was obtained to rule out malignancy.

Based on imaging findings, rapid progression of symptoms, and the presence of brainstem compression, a left retromastoid craniectomy and resection was planned for preliminary diagnosis of an arachnoid cyst. The patient was given mannitol, dexamethasone, and antibiotics preoperatively. Craniectomy was performed, and the dura was opened. The cisterna magna was opened for the release of CSF and cerebellar relaxation. Meticulous arachnoid dissection was then performed to visualize the CNs and the CPA cyst. Once the cyst was identified, needle aspiration was performed, which extracted xanthochromic, mucoid-gelatinous, yellow-tinged cystic contents that were subsequently sent to pathology (Fig. 3). Additional cyst fenestration and partial resection of the cyst wall were performed, and additional cystic contents were removed safely. A portion of the cyst wall could not be safely removed because it was beyond the visualized area with the retromastoid craniotomy. However, all the gelatinous synovial fluid was removed to minimize the possibility of postoperative aseptic meningitis.

Pathology showed a predominance of macrophages, glial cells, debris, and mucoid material with the absence of malignant cells, consistent with a benign cyst. Definitive classification could not be obtained because of no identifiable cyst wall cells in the pathology sample; however, preoperative radiological features at the atlantoaxial joint and intraoperative findings of cystic contents were most consistent with synovial cyst. Therefore, postoperative MRI of the cervical spine was performed to characterize the degenerative changes at the atlantoaxial joint, revealing several levels of osteophytic growths and bilateral extradural juxtafacet cysts at C1 extending to adjacent joint spaces (Fig. 4). The patient was discharged home on postoperative day 1 in stable condition.
By the 6-week follow up, the patient had recovered most of her left facial nerve function and experienced complete relief of the left trigeminal pain. However, there was persistent diminishment of left-sided hearing and sensation in the V2-V3 dermatomes. No evidence of cyst recurrence was seen on 3-month postoperative MRI. At the 5-month follow-up, the patient’s facial and trigeminal nerve function had recovered, although she continued to have diminished hearing on the left.

Methods for Literature Review

An extensive literature search conducted on PubMed from April 10–14, 2023, regarding synovial cysts involving the cervical spine, revealed a limited number of cases specifically involving the atlantoaxial or atlantooccipital junction. Search terms included: “synovial cyst of the atlanto-axial joint,” “atlantooccipital junction cyst,” “atlantoaxial juxtafacet cyst,” “C1–C2 synovial cyst,” “C1–C2 synovial cyst with cranial nerve palsy,” “atlantooccipital junction cyst with craniotomy,” “atlantoaxial cyst craniotomy,” “C1–C2 synovial cyst with craniotomy,” “management of atlantoaxial synovial cyst,” and “management of atlantooccipital synovial cyst.” Of 43 articles found on the topic from 1988 to 2023, only 30 cases were relevant to the atlantoaxial/atlantooccipital junction and only 13 of these cases were treated with craniotomy/craniectomy and/or involved cranial nerve palsy (Table 1).5–14 Articles were excluded if pathology was found to be anything other than synovial cyst, if the cyst was at a subaxial cervical level, or if either full text was unavailable or the abstract did not provide sufficient information for the literature review.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

The exact cause of synovial cysts is unknown, but they are believed to develop as a result of increased pressure within the zygapophyseal joints of the spine, causing fluid to leak out and form a cyst.5 Recent studies have shown a link with degenerative spondylosis, spinal instability, or trauma as inciting factors.16–19 The aging population has also revealed an increase in the prevalence of synovial cysts with a mean age of 61 to 66 years, thereby highlighting the key role that degeneration plays in the pathophysiology of the disease.20–22

The majority of synovial cysts are asymptomatic and typically go undiagnosed. In a study by Janssen et al.20 exploring the epidemiology of lumbar synovial cysts (n = 19,010), 42% of patients with lumbar synovial cysts were asymptomatic. However, of the spinal levels from which cysts can arise, the lumbar spine is far more common because of its mobility and increased risk for degenerative changes. Cervical cysts are infrequent. If present, they most commonly affect the C7–T1 levels.21 Even rarer are cysts at the craniovertebral junction including the atlantooccipital and atlantoaxial joints. The atlantoaxial joint consists of two types of synovial lined joints, the lateral atlantoaxial joints and the medial joint, with cysts arising more commonly from the former.22

Common presenting symptoms include neck pain, discomfort, stiffness, and headaches. Variations in symptoms are typically contingent on the migratory direction of the cyst such that lateral migration results in radiculopathy, medial migration results in myelopathy, and, as we saw in our case, superior migration can involve CN palsy. Of the three migrations, the lateral migration is most commonly presented by patients. Superior migrations involving the CNs are highly uncommon. In our literature search, only four cases involved superior cyst migration and resulted in hypoglossal nerve palsy.6–8,10 To our knowledge, the present case is the first atlantoaxial synovial cyst with superior migration extending to the CPA causing brainstem compression and multiple CNs dysfunction including CNs V, VII, and VIII.

Management of these lesions is individualized and has included conservative observation, fixation/fusion, aspiration, and resection/fenestration. However, resection/fenestration is the treatment of choice for synovial cysts causing neurological symptoms related to brainstem, CN, or spinal cord compression, as it can achieve rapid neurological symptom resolution. Factors to consider in surgical planning include not only selecting a surgical approach with adequate access to the cyst, but also considering the possible need for arthrodesis, because these lesions frequently arise in the setting of subclinical or overt instability of the joint. This was illustrated by a case reported by Tangyiriyapaiboon et al.23 who successfully managed a retro-odontoid cyst with only posterior atlantoaxial fusion with spontaneous regression, significant neurological improvement, and no recurrence. The surgical plan should also take into consideration the possibility of iatrogenic instability related to bone or ligament removal for the surgical approach.12

For cysts involving superior migration and CN palsy, craniotomy is typically planned. However, the exposure differs based on the anatomical location of the cyst. For example, cysts located lateral to the medulla, a juxta-condylar approach may be optimal, whereas an anterior cyst may necessitate a transcondylar approach.6 Complications of each route must also be taken into consideration. The transoral route, although popular in the late 1990s for the treatment of cervical synovial cysts (retro-odontoid), introduces a risk of infection from oral flora. Now other approaches have come into favor such as the far lateral and lateral suboccipital approaches done by Adada et al.12 and Miyazawa et al.13 respectively.

Intraoperatively, the extent of safe resection is taken into consideration. Cysts can be accompanied by significant inflammation and adhesions to surrounding tissue, CNs, or dura. Forceful resection could lead to significant or permanent neurological complications giving the proximity to critical structures.13,14 Fortunately, in cases in which complete resection was not possible, significant improvement has been reported with partial resection/decompression and a low frequency of recurrence.5,9,13
### Table 1. Summary of Management and Outcome in Cases of Atlantoaxial/Atlantooccipital Junction Synovial Cyst Requiring Craniotomy and/or Presenting with Cranial Nerve Palsy

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases/Age (yrs)/Sex</th>
<th>Clinical Presentation</th>
<th>Imaging Findings (T1/T2/contrast)</th>
<th>Cyst Location</th>
<th>Surgical Approach</th>
<th>Reasoning for Surgical Plan</th>
<th>Outcome (EOR/ROS/R)</th>
</tr>
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<tbody>
<tr>
<td>Onofrio &amp; Mih, 1988</td>
<td>1/73/M</td>
<td>Bilat hand weakness</td>
<td>−/−</td>
<td>Retro-odontoid, transverse ligament</td>
<td>Suboccipital craniectomy w/ C1–2 laminectomy</td>
<td>Cyst present on transverse ligament requiring wide C1–2 laminectomy, craniectomy, &amp; transdural approach; although mild C2–3 anterior instability seen postoperatively, stabilization not required</td>
<td>~SR/NR</td>
</tr>
<tr>
<td>Miller et al., 1989</td>
<td>1/67/F</td>
<td>Neck pain, decreased ambulation, rt arm pain, quadriaparesis, hyperreflexia</td>
<td>I/−/−</td>
<td>Anterolat to cervical spinal cord</td>
<td>Laminectomy of C1–2 w/ foramen magnum craniotomy</td>
<td>Anatomically optimal approach for cyst localization &amp; resection</td>
<td>CR/SI/NR</td>
</tr>
<tr>
<td>Fransen et al., 1997</td>
<td>1/75/F</td>
<td>Spastic quadriaparesis, hyperreflexia, stereognosis, graphesthesia, paresthesia</td>
<td>L/H/RE</td>
<td>Retro-odontoid</td>
<td>Posterolat approach w/ lt suboccipital craniectomy</td>
<td>Allowed for control over the VA, spinal root of accessory nerve, &amp; spinal cord/medulla</td>
<td>CR/SR/−</td>
</tr>
<tr>
<td>Eustacchio et al., 2003</td>
<td>1/75/M</td>
<td>Occipital pain, gait abnormalities, quadriaparesis, hyperreflexia</td>
<td>−/−</td>
<td>Adjacent to odontoid process</td>
<td>Suboccipital craniectomy &amp; C1 hemilaminectomy</td>
<td>Less invasive compared to anterior approach; although partial resection anticipated, literature has shown neurological symptom resolution &amp; minimal recurrence in cases of partial resection</td>
<td>PR/SI/NR</td>
</tr>
<tr>
<td>Mujic et al., 2003</td>
<td>1/52/M</td>
<td>Dysarthria, dysphagia, lt CN XII palsy</td>
<td>−/H/NE</td>
<td>Lt atlanto-occipital joint</td>
<td>Far lat suboccipital craniectomy</td>
<td>Anatomically optimal approach for cyst localization &amp; resection</td>
<td>CR/PI/NR</td>
</tr>
<tr>
<td>Baldauf et al., 2005</td>
<td>1/51/F</td>
<td>Dysarthria, lt CN XII palsy</td>
<td>I-L/H/NE</td>
<td>Atlanto-occipital joint</td>
<td>Far lat suboccipital craniotomy w/ endoscopy</td>
<td>Anatomically optimal approach for cyst localization &amp; resection</td>
<td>CR/NI/NR</td>
</tr>
<tr>
<td>Elhammady et al. 2009</td>
<td>1/67/F</td>
<td>Lt lower facial pain, neck pain, dysarthria, dysphagia, gait abnormalities, lt CN XII palsy</td>
<td>I/H/RE</td>
<td>Abutting lat medulla just medial to lt hypoglossal canal w/o extension into canal</td>
<td>Juxta-condylar approach</td>
<td>Approach chosen due to lat location of lesion w/ respect to medulla, whereas far lat approach (transcondylar) preferred for anterior lesions; fusion only necessary if concerned for neck instability</td>
<td>CR/W/NR</td>
</tr>
<tr>
<td>Harries et al., 2010</td>
<td>1/75/F</td>
<td>Paresthesias, upper extremity paresis, gait abnormalities, hyperreflexia</td>
<td>−/−/−</td>
<td>Mass at C1–2 causing compression of cervico-medullary junction</td>
<td>Far lat approach</td>
<td>Difficult to determine nature of mass due to patient contraindication for MRI; resulted in preliminary diagnosis of meningioma, leading authors to choose far lat approach for optimal access</td>
<td>PR/SI/NR</td>
</tr>
</tbody>
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TABLE 1. Summary of management and outcome in cases of atlantoaxial/atlantooccipital junction synovial cyst requiring craniotomy and/or presenting with cranial nerve palsy

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<th>Authors &amp; Year</th>
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<tbody>
<tr>
<td>Mendes-Arajuo et al., 2010&lt;sup&gt;10&lt;/sup&gt;</td>
<td>1/51/F</td>
<td>Dysarthria, tongue fasciculation, Lt CN XII palsy</td>
<td>L/H/NE</td>
<td>Lt atlantoaxial joint extending superiorly &amp; insinuated through ipsilateral hypoglossal canal</td>
<td>Patient refused surgical intervention, no progression of symptoms at 1-yr FU</td>
<td>—</td>
<td>–/NI/–</td>
</tr>
<tr>
<td>Hartmann et al., 2016&lt;sup&gt;11&lt;/sup&gt;</td>
<td>1/74/M</td>
<td>Headache, neck pain, gait abnormalities, LUE paresis, hyperreflexia, BLE hypoesthesia</td>
<td>–/H/–</td>
<td>Medial atlantoaxial joint spanning clivus to C2</td>
<td>Lt suboccipital approach w/ C1 laminectomy &amp; C2 cranial laminotomy</td>
<td>Dorsal decompression w/o instrumentation guided by preoperative imaging showing no atlantoaxial instability</td>
<td>CR/P/I/–</td>
</tr>
<tr>
<td>Adada et al., 2019&lt;sup&gt;12&lt;/sup&gt;</td>
<td>1/88/M</td>
<td>Gait abnormalities, neck pain, mild quadripareis, hyperreflexia</td>
<td>–/H/–</td>
<td>Retro-odontoid</td>
<td>Modified far lat, transatlas extradural approach w/ decompression and fusion</td>
<td>Approach offers multiple advantages: extradural, provides extensive exposure, avoids complications related to oral surgical field, &amp; facilitates fusion to address instability</td>
<td>CR/S/I/NR</td>
</tr>
<tr>
<td>Miyazawa et al., 2020&lt;sup&gt;13&lt;/sup&gt;</td>
<td>2/71,78/M</td>
<td>Hemiparesis, rt thermal pain, dysarthria, dysphagia</td>
<td>L/L/RE (1/2)</td>
<td>Retro-odontoid/retro-odontoid</td>
<td>Lat suboccipital approach (w/ C1 laminectomy of 1 case)</td>
<td>Mass reduction w/o forcible detachment from brain provided safer &amp; sufficient alternative to oral approach (risk of infection &amp; instability from bone resection)</td>
<td>PR/S/I/NR; PR/ S/I/NR</td>
</tr>
<tr>
<td>Fana et al., 2022&lt;sup&gt;14&lt;/sup&gt;</td>
<td>1/57/M</td>
<td>Neck pain, hyporeflexia</td>
<td>–/H/–</td>
<td>Retro-odontoid</td>
<td>Far posterolateral approach w/ endoscopy &amp; Lt hemilaminectomy of C1–2</td>
<td>Approach avoided VA transposition, bony instability, &amp; fusion; endoscopy aided in visualization of dural attachment to prevent sacrificing bone</td>
<td>PR/U/–</td>
</tr>
</tbody>
</table>

A = aspiration; BLE = bilateral lower extremities; CN = cranial nerve; CR = complete resection; EOR = extent of resection; FU = follow-up; H = high signal intensity; I = isointense; L = low signal intensity; LUE = left upper extremity; NE = non-rim enhancing; NI = no improvement; NR = no recurrence; PI = partial improvement; PR = partial resection; R = recurrence; RE = rim enhancing; ROS = resolution of symptoms; SI = significant improvement; SR = spontaneous regression; VA = vertebral artery; W = worsening.

For our case, retromastoid craniectomy provided optimal exposure of the cyst. Because the cervical vertebral complex was not disrupted and instability was not appreciated preoperatively, fusion was not performed. Finally, with the cyst’s extensive superior migration into the CPA, partial resection with wide fenestration and complete aspiration of cyst contents were performed. In this case, we continue to monitor the patient with serial MRI. If the cyst were to recur, arthrodesis would be considered.

Observations
This case demonstrates imaging characteristics typical of a synovial cyst, with a uniformly hypodense center on CT, uniformly hypointense center on T1 sequences, and uniformly hyperintense center on T2 sequences (Fig. 1). Intraoperative extraction of cystic contents revealed xanthochromic, mucoid-gelatinous substance with pathological confirmation of benign cystic contents, classic for synovial cysts and further supporting our final diagnosis (Fig. 3).

Lessons
Synovial cysts involving the atlantoaxial junction are infrequent lesions. However, they can undergo rapid enlargement and cause severe deficits related to compression of brainstem structures or CNs. Diagnosis is confirmed with CT and/or MRI, which should characteristically demonstrate a uniformly hypodense center on CT, uniformly hypointense center on T1 sequences, and uniformly hyperintense center on T2 sequences. Retromastoid craniectomy allows for decompression of the brainstem, cerebellum, and CNs with significant clinical improvement. Resection of the entire cyst is...
preferred; however, decompression of cyst material with maximal safe resection of the capsule wall will likely suffice in most cases. Regular imaging follow-up should be considered to monitor for cyst recurrence. A recurrence of the cyst should warrant consideration of arthrodesis of the affected joint.

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References

Disclosures
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
Conception and design: Graham, Seshadri, Shah, Hachmann. Acquisition of data: Graham, Seshadri, Shah, Hachmann. Analysis and interpretation of data: Graham, D’Souza, Shah, Hachmann. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Graham. Statistical analysis: Seshadri. Administrative/technical/material support: Graham, Seshadri. Study supervision: Graham.

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