Published in final edited form in: Neurosurg focus. 2005 Jul;21(7):524-27.

Far Lateral Transcondylar Approach for Resection of Neurenteric Cysts of the Cervicomedullary Junction

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Abstract

Neurenteric cysts are rare benign cysts of the central nervous system that are lined by endodermal-derived epithelium. Although they mostly occur in the spine, they can occur intracranially, most often in the posterior fossa. Neurenteric cysts that are located in the anterior cervicomedullary junction are even rarer lesions and often require a skull base approach for adequate surgical resection. The authors describe two cases of neurenteric cysts arising from the cervicomedullary junction resected by a far lateral transcondylar approach. They discuss the surgical approach and operative nuances in removing these lesions and review the clinical presentation of neurenteric cysts in this region, the neuroimaging characteristics, histopathologic findings, and surgical management. Operative videos are presented.

Key Words: neurenteric cyst, cervicomedullary junction, far lateral transcondylar approach, enterogenous cyst
Neurenteric cysts, also known as enterogenous, endodermal, neuroenteric, respiratory, or bronchogenic cysts, are rare benign cysts of the central nervous system that are lined by endodermal-derived epithelium. They are most frequently found in the intradural extramedullary space in the lower cervical and upper thoracic spine. Intracranial neurenteric cysts are even more rare, with the majority located in the posterior fossa. Spinal neurenteric cysts are often connected by a fibrous tract, fistula, or cleft to structures derived from the primitive gut in the thoracic or abdominal cavities and are commonly associated with vertebral anomalies, such as vertebral body dysgenesis, split cord malformations, hemivertebra, segmentation abnormalities, and spina bifida. This is not the case with intracranial neurenteric cysts. Although the exact pathogenesis remains unknown, intracranial neurenteric cysts are thought to arise from a failure of dissolution of the transient neurenteric canal between the foregut or the respiratory buds and the notochord during notochordal development.

Posterior fossa neurenteric cysts constitute more than 90% of the intracranial form of these cysts, and they occur mostly along the midline. In a review by Bejjani, et al., the most common locations of posterior fossa neurenteric cysts were anterior to the brainstem (51%) or within the fourth ventricle (21%). Eighteen percent of the cysts extended into the cervical canal and 17% were primarily in the cerebellopontine angle. Neurenteric cysts that are located in the anterior cervicomedullary junction are even rarer lesions and often require a skull base approach for adequate surgical resection. We describe two cases of neurenteric cysts arising from the cervicomedullary junction resected by a far lateral transcondylar approach. We discuss the surgical approach and
operative nuances in removing these lesions, and we review the clinical presentation of neurenteric cysts in this region, the neuroimaging characteristics, histopathologic findings, and surgical management.

Case Reports

Case 1

History and Examination. This 46-year-old woman presented with pain in the right shoulder, right ear, and interscapular area. Neurologic examination was normal, with the exception of a markedly diminished gag reflex on the right side. Laryngoscopic examination by the otolaryngology service demonstrated normal vocal cord movement.

Neuroimaging. Magnetic resonance imaging (MRI) demonstrated a lesion in the cervicomedullary cistern involving the ninth and tenth nerves on the right side (Fig. 1). The lesion was isointense on T1-weighted images and hyperintense on T2-weighted and FLAIR images. No enhancement was observed after gadolinium administration.

Operation. The patient underwent a far lateral transcondylar approach for gross total resection of the lesion (approach described in detail below). Intraoperatively, a white cystic lesion was identified beneath the flocculus, deep to the ninth and tenth cranial nerve complex (Fig. 1, Video Clip 1). The structure appeared gelatinous in consistency and was covered by a thin translucent membrane. The lesion was dissected from the lower cranial nerves and removed entirely.
**Histopathological Examination.** Grossly, the specimen appeared as a white, translucent mucoid substance. Histological examination revealed a cyst wall lined by ciliated columnar epithelium (Fig. 2) that stained positive for AE 1.3 and Cam 5.2 keratin markers, suggesting an endodermal origin. The final diagnosis was neurenteric cyst.

**Postoperative Course.** The patient did well postoperatively and had an uneventful course. She had no difficulty with swallowing, and sensation in her right posterior pharynx improved after surgery. Postoperative MRI demonstrated a complete resection of the lesion (Fig. 1).

**Case 2**

**History and Examination.** This 26-year-old woman presented with headaches. Neurologic examination was normal.

**Neuroimaging.** MRI demonstrated a smoothly margined mass occupying the right cervicomedullary junction extending anterior to the brainstem with mild mass effect (Fig. 3). It was hyperintense on T1-weighted images, slightly hyperintense on T2-weighted images, and minimally enhanced after gadolinium administration.

**Operation.** This lesion had been followed, and showed some interval growth. Because of the size of the lesion and the fact that it was already showing some mass effect on the brainstem, the patient underwent a far lateral transcondylar approach for gross total resection of the lesion (approach described in detail below). Intraoperatively, a
yellowish-white, custardly-appearing cyst was identified surrounding and enveloping the ninth and tenth nerves (Fig. 3, Video Clip 2). The inferior aspect of the cyst was mobilized from the twelfth nerve and the cyst was opened to remove some of the colloid material to decompress the cyst. The remaining cyst wall was dissected off the anterior aspect of the brainstem and the vertebral artery up to the vertebrobasilar junction. All of the cranial nerves were left intact.

*Histopathological Examination.* Histological examination revealed a cyst wall lined with ciliated simple cuboidal-to-columnar epithelium (Fig. 4). The cyst contents consisted of anucleated, keratinous debris. The final diagnosis was a neurenteric cyst.

*Postoperative Course.* Postoperative MRI demonstrated a complete resection of the lesion (Fig. 3). The patient did well postoperatively with no neurological deficits. The remainder of the hospital course was uneventful.

**Far Lateral Transcondylar Approach: Operative Technique**

*Preoperative Considerations*

The far lateral transcondylar approach is performed on the side of the lateral extension of the lesion. If the lesion is strictly midline, the laterality of the approach can be chosen based on the anatomy of the vertebral artery, the sigmoid sinus, and the jugular bulbs. Al-mefty, et al., have suggested approaching from the side of the nondominant vertebral artery or nondominant jugular bulb in these cases. Intraoperative monitoring,
including somatosensory evoked potentials, auditory evoked responses, facial nerve monitoring, and tenth, eleventh, and twelfth cranial nerve monitoring, should be implemented. The tenth cranial nerve is monitored with an electromyographic endotracheal tube, and the eleventh and twelfth nerves are monitored via electrodes placed directly into the sternocleidomastoid muscle and the tongue, respectively.

*Positioning and Skin Incision*

The patient is placed in the lateral position and the head is held in three-point pin fixation with the neck slightly flexed, the vertex angled slightly down, and the face rotated slightly ventrally, so that the ipsilateral external auditory meatus and the mastoid bone are at the highest point (Fig. 5). Positioning the head in this manner allows improved exposure of the occipitocervical region and improves the inferior-to-superior viewing angle for the surgeon. An axillary roll is placed, and the patient’s contralateral arm rests on a Krauss armrest. The elevated arm is distracted inferiorly toward the foot of the table to provide more room for the surgeon above the shoulder. All pressure points are carefully padded with foam or gel pads. The patient is secured to the operating table with adhesive tape to allow safe rotation of the table during the operation to improve the surgeon’s line of sight. Intravenous glucocorticoids, antibiotics, and mannitol are administered at the time of the skin incision.

A retroauricular curvilinear skin incision starts approximately 2 to 3 cm behind the ear and continues inferiorly into the neck over the posterior border of the sternocleidomastoid muscle to about C3 or C4 (Fig. 5). The skin flap is elevated in two layers. The incised skin and galea are first elevated to expose the underlying pericranium
above the superficial neck fascia, which may be harvested as a fascial graft for later watertight dural closure. The pericranium and the superficial fascia are then elevated to expose the underlying musculature.

Muscle Dissection and the Suboccipital Triangle

Anatomically, three layers of muscle are identified during the dissection. The superficial layer (trapezius and sternocleidomastoid) and the middle layer (splenius capitis, longissimus capitis, semispinalis capitis) of muscles are incised and reflected as a single layer to expose the suboccipital triangle (Fig. 6), which is bound by the deep layer of muscles (medially by the rectus capitis posterior major; inferiorly by the inferior oblique; and superolaterally by the superior oblique muscle). The rectus capitis major muscle inserts superiorly on the inferior nuchal line and inferiorly on the spinous process of C2; the inferior oblique muscle inserts superiorly on the transverse process of C1 and inferiorly on the spinous process of C2; the superior oblique muscle inserts superiorly at the temporo-occipital suture and inferiorly on the transverse process of C1. The suboccipital triangle, which involves the dorsal ramus of the C1 nerve root and the V3 horizontal segment of the vertebral artery, can be opened by detaching the insertions of the superior and inferior oblique muscles from the transverse process of C1 and reflecting them posteriorly. The rectus capitis major is detached from the inferior nuchal line and reflected posteriorly. The C1 lamina and vertebral artery will become more apparent. The vertebral artery is covered by a venous plexus, sometimes referred as the suboccipital cavernous sinus. Further exposure of the laminae of C2 or C3 may be performed if more inferior exposure is needed.
Exposure and control of the extradural vertebral artery is important and can be achieved by identifying its extradural course from the foramen transversarium of C2 to the occiput. The ventral ramus of the C2 nerve root, found between the laminae of C1 and C2, can be traced laterally until it crosses dorsally to the vertical segment of the vertebral artery, coursing between the foramen transversarium of C2 and C1 (Fig. 6). As the vertebral artery exits the foramen transversarium of C1, it is encased in a venous plexus and courses posteriorly behind the lateral mass of C1 in the vertebral groove and turns medially to pierce the atlanto-occipital membrane and dura. Several small muscular branches and the posterior meningeal artery arise from the horizontal segment of the vertebral artery, which can be safely coagulated. In some cases, the posterior spinal artery and PICA can arise extradurally and can potentially be injured. Subperiosteal dissection of the vertebral artery from the vertebral groove reduces bleeding from the venous plexus by leaving the periosteal sheath around the artery intact. The atlanto-occipital membrane is sharply divided to expose the underlying dura.

Suboccipital Craniectomy and C1 Hemilaminectomy

A lateral suboccipital craniectomy or craniotomy is initially performed with a high-speed drill and rongeurs. The craniectomy usually extends towards the midline medially and to the inferior nuchal line superiorly (depending on the extent of exposure needed), to the posterior rim of the foramen magnum inferiorly, and up to the occipital condyle laterally (Fig. 7). To provide more superior access to the cerebellopontine angle,
the craniectomy can be extended up to the transverse sigmoid junction. The sigmoid sinus
and jugular bulb are exposed with rongeurs and a high-speed drill. The posterior condylar
emissary vein will be encountered as it travels from the jugular bulb and exits the
condylar fossa via the condylar canal to join the extradural venous plexus. Hemostasis
can be achieved by packing the vessel with Surgicel. An ipsilateral hemilaminectomy of
C1 improves the dural exposure inferiorly. More inferior exposure for lower-lying lesions
can be created by removing the hemilamina of C2 and C3, if desired.

Transcondylar Resection

Extradural reduction of the occipital condyle is one of the key maneuvers in
maximizing exposure to the ventral aspect of the craniovertebral junction while avoiding
brain stem retraction (Fig. 7). Anatomic morphometric studies have demonstrated that
partial condylar resection increases the angle of exposure, the working space at the level
of the foramen magnum, and the visualization of the ventral and ventrolateral aspect of
the craniovertebral junction and the contralateral aspect of the inferior clivus.11,31,33 The
recommendations for the degree of occipital condyle removal vary greatly in the
literature, ranging from no resection up to complete condyle resection.5,23,27-31 In our
experience, removal of the posterior and medial one-third of the condyle is generally
adequate if more ventral exposure is needed. If 50% or more of the condyle has been
resected or destroyed by the lesion, instability of the craniovertebral junction increases
and an occipitocervical stabilization should be strongly considered.32 Although extradural
reduction of the jugular tubercle aids in maximizing intradural exposure across the
anterior surface of the brain stem and mid-clivus, this maneuver is probably not necessary
for neurenteric cysts because they are soft and suckable lesions.

*Intradural Exposure*

A curvilinear incision of the dura is made several millimeters posterior to the sigmoid sinus, extending inferiorly towards the C2 lamina, staying posterior to the vertebral artery where it pierces the dura (Fig. 7). The dural opening may be extended anteriorly in a “T” fashion, and we prefer to do this just superior to the vertebral artery to enable greater exposure. A dural cuff is preserved around the vertebral artery for later watertight closure. The incision can be extended up to the junction of the transverse-sigmoid sinus if more exposure of the cerebellopontine angle is needed. The anterior leaflet of dura is reflected laterally and held with tacking sutures for maximal exposure. Adequate reduction of the occipital condyle should provide a straight surgical trajectory to the craniovertebral junction parallel to the intracranial course of the vertebral artery. Structures of the inferior aspect of the cerebellopontine angle and the cerebellomedullary angle are visualized. Sharp arachnoid dissection is performed and the following structures can be visualized: the fifth through twelfth cranial nerves, the basilar artery, the vertebral artery, the vertebrobasilar junction, the posterior-inferior cerebellar artery (PICA), and the anterior-inferior cerebellar artery (AICA) (Fig. 8). Complete resection of the cyst wall and contents is then performed, with strict adherence to microsurgical technique and preservation of all cranial and spinal nerves and vascular structures.

*Closure*

A primary watertight closure of the dura should be performed. If necessary, an
autologous pericranium or fascial graft can be harvested from the neck wound. This can be supplemented with autologous fat and fibrin glue. The exposed mastoid air cells are closed with bone wax. The muscle layers are carefully reapproximated to avoid postoperative CSF leakage. Temporary CSF diversion with a lumbar drain can promote sealing of the wound and reduce the risk of the patient developing a pseudomeningocele.

Discussion

Clinical Presentation

These lesions are frequently incidental radiographic findings, but occasionally they may produce symptoms of progressive neurological impairment and chronic increased intracranial pressure. Neurenteric cysts of the cervicomedullary junction can cause symptoms by an inflammatory reaction or by mass effect. Active secretion by goblet cells can result in cyst expansion and compression of neighboring neurovascular structures. The most frequent complaint is headaches followed by gait disturbance and motor and sensory disturbances. Intermittent leakage of cyst contents can result in recurrent aseptic meningitis. Involvement of the cranial nerves can result in facial numbness, hemifacial spasm, hearing loss, and swallowing difficulties.

Neuroimaging Characteristics

The imaging characteristics of neurenteric cysts can be quite variable. This may be attributed to the presence of proteinaceous or other hydrophilic contents. Computed tomography (CT) usually reveals a hypodense lesion in most cases, although occasionally
the lesions can be hyperdense. Sometimes the lesion may be undetectable on CT. MRI is the study of choice for evaluating neurenteric cysts. These extramedullary cystic lesions may appear hypointense, isointense, or slightly hyperintense on T1-weighted images and mostly hyperintense on T2-weighted and FLAIR images without edema. The cyst wall usually does not enhance after gadolinium injection in most cases.²⁶ In the patient presented in Case 2, however, the cyst enhanced after gadolinium. Unlike epidermoid cysts, neurenteric cysts do not restrict on diffusion-weighted images.

**Histopathological Findings**

Intraoperatively, neurenteric cysts appear as yellowish, milky-white lesions filled with gelatinous or mucoid fluid. Histologically, they are benign lesions characterized by a cyst lined with simple or pseudostratified cuboidal-to-columnar epithelium with a basement membrane resembling those of the respiratory and intestinal tracts.¹⁸ The epithelium is sometimes ciliated and often contains mucin-secreting goblet cells that exhibit periodic acid-Schiff (PAS)-positive staining. The intracystic fluid is usually milky, xanthochromic, or transparent.¹⁴ Immunohistochemistry is often useful in the diagnosis. Neurenteric cysts usually exhibit positive staining for keratin, carcinoembryonic antigen (CEA), and epithelial membrane antigen (EMA). They are negative for glial fibrillary acidic protein (GFAP), neuron-specific enolase, S-100, and vimentin. This pattern of immunoreactivity confirms an endodermal origin of neurenteric cysts.⁶,¹⁸,²⁶

**Surgical Management**
Because neurenteric cysts are benign lesions, the goal of surgery should be complete excision of both the cyst contents and the cyst wall if the dissection can be performed without injury to the neighboring neurovascular structures.²,⁸,¹⁴ If the cyst wall appears strongly adherent to the brainstem or vasculature, subtotal resection is a reasonable alternative to avoid injury to these structures. In these cases, patients should be carefully monitored with serial MRI to detect cyst recurrence. Neurenteric cysts of the cervicomedullary junction often require a skull base approach to gain access to the lesion. We prefer the far lateral transcondylar approach because it allows access to ventrally and ventrolaterally located lesions of the cervicomedullary junction. Access to the entire cerebellopontine angle up to the tentorium can be achieved, if needed.
References


Fig. 1. Case 1. Preoperative MRI (A: T1-weighted axial image; B: FLAIR axial image) demonstrating a neurenteric cyst in the cervicomedullary cistern involving the ninth and tenth nerves on the right side. Note that the cyst is isointense on the T1-weighted image and hyperintense on the FLAIR image. There was no enhancement after gadolinium administration. C: Intraoperative photograph demonstrating a white cystic lesion next to the ninth and tenth cranial nerve complex. D: Postoperative MRI (FLAIR axial image) shows gross total resection of the neurenteric cyst.

Video Clip 1. Intraoperative video of patient in Case 1. A right-sided far lateral transcondylar approach was performed for resection of a neurenteric cyst in the cervicomedullary junction.
Fig. 2. Photomicrograph of the specimen in Case 1 showing the cyst wall lined by ciliated columnar epithelium. H&E, original magnification 400x.

Fig. 3. Case 2. Preoperative MRI (A: T1-weighted axial image; B: T1-weighted sagittal image; C: post-gadolinium T1-weighted coronal image) demonstrating a neurenteric cyst occupying the right cervicomedullary junction extending anterior to the brainstem with mass effect. In this case, the cyst is hyperintense on T1-weighted images and enhances after gadolinium administration. D: Intraoperative photograph demonstrating a white
cystic lesion surrounding and enveloping the ninth and tenth nerves. Postoperative MRI (E: post-gadolinium T1-weighted axial image; F: T1-weighted sagittal image) shows gross total resection of the neurenteric cyst.

Video Clip 2. Intraoperative video of patient in Case 2. A right-sided far lateral transcondylar approach was performed for resection of a neurenteric cyst in the cervicomedullary junction. Note the vertebrobasilar junction and the lower cranial nerves that are preserved at the end of the operation.

Fig. 4. Photomicrograph of the specimen in Case 2 showing the cyst wall lined by ciliated simple cuboidal-to-columnar epithelium. H&E, original magnification 400x.
Fig. 5. A. Illustration of lateral positioning. B. Retroauricular curvilinear skin incision (dotted line).

Fig. 6. A. Inferior view of the base of the skull demonstrating the anatomical relationship of the jugular tubercle, hypoglossal canal, occipital condyle, and foramen magnum. B. Surface anatomy and key landmarks. C. Course of the vertebral artery. Note that the
ventral ramus of the C2 nerve root, found between the laminae of C1 and C2, can be traced laterally until it crosses dorsally to the vertical segment of the vertebral artery, coursing between the foramen transversarium of C2 and C1. D. The suboccipital triangle, which is bound medially by the rectus capitis posterior major (RCM) muscle, inferiorly by the inferior oblique (IO) muscle, and superolaterally by the superior oblique (SO) muscle, serves as an anatomical landmark for identifying the dorsal ramus of the C1 nerve root and the V3 horizontal segment of the vertebral artery (VA).

Figure 7. A. Retrosigmoid lateral suboccipital craniectomy (dotted line). B. The dural incision (dotted line) is made in a curvilinear fashion several millimeters posterior to the sigmoid sinus and extends inferiorly towards the C2 lamina, staying posterior to the vertebral artery where it pierces the dura.
Fig. 8. Diagram illustrating the intradural exposure of the far lateral transcondylar approach.