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# Surgical Management of Intracranial Neuroenteric Cysts: The UCSF Experience

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## Abstract

**Objective** Modern surgical experience with intracranial neuroenteric cysts is limited in the literature. We review our 15-year institutional experience with these rare lesions.

**Design** Single-institution retrospective study.

**Setting** Large North American tertiary care center.

**Participants** Histologically confirmed cases of intracranial neuroenteric cyst from January 2000 to September 2014.

**Main Outcome Measures** Pre- and postoperative modified Rankin Scale (mRS) scores, extent of resection, and postoperative complications are reported. Clinical presentation, imaging features, pathology, and operative approach are discussed.

**Results** Five spinal and six intracranial neuroenteric cysts were surgically treated over a 15-year period. Median age at presentation for the intracranial cysts was 38.5 years. Mean cyst diameter was 3.8 cm. Five cysts were located in the pre-pontomedullary cistern, and one was located in the third ventricle. Gross total resection was achieved in four of the five posterior fossa cysts through a far lateral transcondylar approach. Postoperative complications included aseptic meningitis (one), transient abducens palsy (one), and pseudomeningocele requiring reoperation (three). Postoperative mRS scores improved to $\leq$1 by 6.5 months median follow-up.

**Conclusions** Intracranial neuroenteric cysts are rare lesions with a variable imaging appearance. Complete surgical resection through a far lateral transcondylar approach is possible and usually results in symptom improvement or resolution.

**Keywords**
- neuroenteric cyst
- neurenteric cyst
- enterogenous cyst
- bronchogenic cyst

## Introduction

Neuroenteric cysts are rare and benign congenital lesions of endodermal origin that can arise throughout the neuraxis. Although they have been reported to occur most commonly in the thoracic spine, they can be found rarely intracranially. Examples in the posterior fossa are ventral to the brainstem or in the cerebellopontine angle, and they present with headache, hydrocephalus, or cranial nerve deficits.[1] Less frequently they have been reported supratentorially. Overall they have been estimated to comprise $\sim$0.03% of intracranial lesions.[2,3] Historically, these lesions have had various designations including teratomatous cyst, intestinome, gastrocytoma, enterogenous cyst, respiratory cysts, and bronchogenic cyst.[1] Although the precise origin of these lesions is not completely

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understood, histologically they are composed of cuboidal or columnar epithelium that resembles that of the gastrointestinal or respiratory tract. Thus intracranial neuroenteric cysts in the posterior fossa are thought to arise from rostrally located vestigial remnants of the neuroenteric canal. In extremely rare cases they can arise supratentorially, where remnants of the Seessel pouch have been postulated as the origin.[1]

Radiographically, the lesions have a variable appearance on magnetic resonance (MR) imaging, making definitive preoperative diagnosis difficult.[1,3–5] They can have highly variable signal intensity on T1- or T2-weighted images, rarely can enhance on contrast-enhanced T1-weighted images, and do not show reduced diffusion on diffusion-weighted imaging (DWI). The MR imaging features of neuroenteric cysts can be similar to the more common arachnoid cysts. In $>$ 150 cases reported in the literature since the 1950s, the differential diagnosis has included many other cystic lesions such as epidermoid and dermoid cyst, ependymoma, cystic schwannoma, and arachnoid cyst.[1,3,5,6]

The current treatment recommendation for neuroenteric cysts is complete surgical resection when possible.[3,5] However, due to their rarity, there is a lack of modern surgical experience with these lesions in the literature.[3–5] The purpose of this study was to report our single-institution experience with these rare intracranial lesions over a 15-year period including their clinical presentation, radiographic appearance, and surgical treatment.

## Methods

This retrospective study was approved by the University of California, San Francisco, Committee on Human Research (14–14211). An institutional database maintained by the Department of Pathology was queried to identify all patients with a histologically confirmed diagnosis of neuroenteric cyst between January 2000 and December 2014. Histologic criteria for diagnosis of neuroenteric cyst included cuboidal or columnar epithelium with cilia and/or mucin-producing cells indicative of gastrointestinal epithelium. Spinal neuroenteric cysts centered below the foramen magnum were excluded.

Demographic, clinical, and imaging information was obtained including age, gender, presenting symptoms, duration of symptoms, and pre- and postoperative detailed neurologic examination. Pre- and postoperative modified Rankin Scale (mRS) scores were determined based on clinical documentation. Operative reports, pre- and postoperative imaging, and clinical follow-up information were also reviewed to identify the radiographic features, surgical approach, the extent of cyst wall resection, postoperative complications, and cyst recurrence.

## Results

During the 15-year study time period, 20,896 specimens were obtained from neurosurgical cases at the Neuropathology Division (excluding legal cases and consultations). A query of this database identified 11 cases of histologically confirmed neuroenteric cysts between 2000 and 2014 (0.053%). Six of these lesions were intracranial above the foramen magnum. The remaining five patients with spinal neuroenteric cysts were excluded from analysis. The study patients had a median age of 38.5 years at presentation (range: 10–71 years) and included one male (►**Table 1**). The most common presenting symptom was headache, followed by nausea and vomiting, cranial nerve deficits, and weakness. One supratentorial lesion was identified in the third ventricle of a 10-year-old female patient who presented with headaches and precocious puberty. The duration of symptoms ranged from 4 months to 4 years (mean: 19.6 months).

The representative imaging appearance of neuroenteric cysts can be seen in ►**Figs. 1**, **2**, and **3**. On computed tomography (CT), cyst contents ranged from hypo- to hyperdense. On MR imaging, the neuroenteric cysts were either T1

**Table 1** Demographic, clinical, and radiographic information for six cases of intracranial neuroenteric cysts

| Case | Age, y/ Gender | Presenting symptoms | Location | T1 | T2 | DWI | Contrast | mRS score, pre/postop |
|---|---|---|---|---|---|---|---|---|
| 1 | 10/F | Headache, precocious puberty | Third ventricle | − | + | NR | Thin rim | 1/0 |
| 2 | 25/M | Headache, facial numbness, hearing loss | Anterior pontomedullary junction | − | + | NR | Thin rim centrally | 1/0 |
| 3 | 55/F | Headache, nausea/vomiting, ataxia | Right paramedian pontomedullary junction | − | + | NR | NE | 4/0 |
| 4 | 31/F | Headache, nausea/ vomiting | Right paramedian pontomedullary junction | + | − | NR | NE | 1/0 |
| 5 | 46/F | Headache, facial paresthesias | Anterior pontomedullary junction | + | + | NR | NE | 1/0 |
| 6 | 71/F | Leg weakness, unsteady gait | Right paramedian pontomedullary junction | + | + | NR | NE | 2/1 |

Abbreviations: −, hypointense; +, hyperintense; DWI, diffusion-weighted imaging; F, female; M, male; mRS, modified Rankin Scale; NE, nonenhancing; NR, no restricted diffusion.

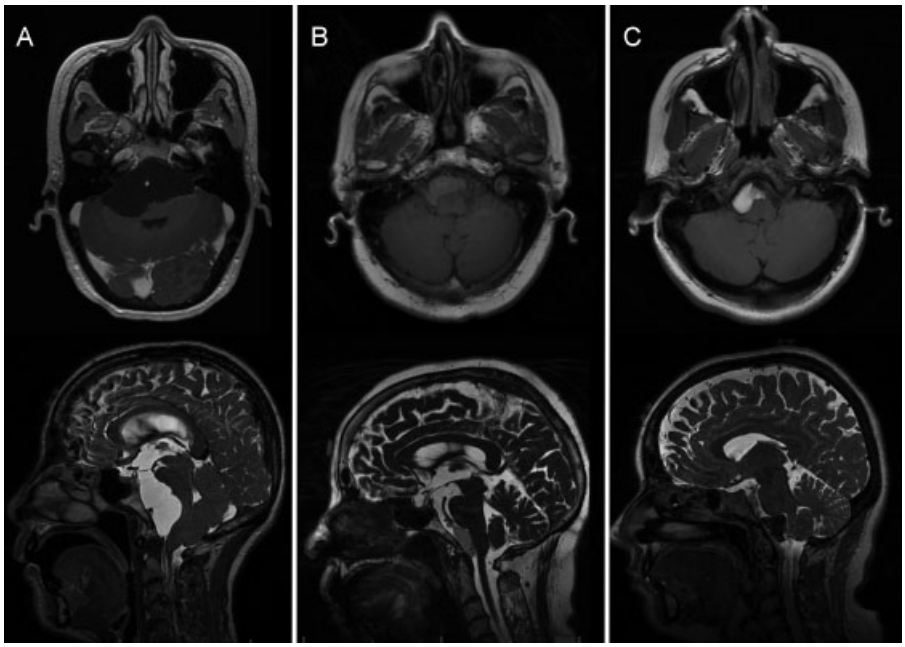


**Fig. 1** Magnetic resonance imaging from three cases of neuroenteric cyst illustrating the variable appearance, typical location, and anterior displacement of the vertebrobasilar complex (VBC). (A) Case 3. T1 postgadolinium axial (top) and T2 sagittal (bottom) showing a nonenhancing T1 dark, T2 bright cyst in the prepontine cistern with anterior displacement of the VBC. (B) Case 5. T1 axial (top) and T2 sagittal (bottom) showing a T1 slightly hyperintense, T2 hypointense to cerebrospinal fluid premedullary lesion. (C) Case 4. T1 axial (top) and T2 sagittal (bottom) showing a T1 bright, T2 dark cyst with anterior displacement of the VBC.

hypointense (three cases) or hyperintense (three cases). On T2-weighted or fluid-attenuated inversion recovery sequences the lesions were primarily hyperintense, with only one of six cases demonstrating a low signal. Two cysts had a thin rim of enhancement. None of the lesions demonstrated reduced diffusion on DWI. The most common location was the prepontine/premedullary cisterns of the posterior fossa (five cases). The rostral-caudal extent ranged from the dorsum sella to the ring of C1 inferiorly. The mean maximal cyst size was 3.8 cm (range: 2.2–6.2 cm). All posterior fossa lesions crossed the midline, and three lesions were eccentrically located on the right side. In four cases there was a mass effect on the brainstem, with associated hydrocephalus in two cases. The vertebrobasilar arterial complex was displaced anteriorly away from the brainstem in all five posterior fossa cases. One lesion was located supratentorially within the third ventricle, causing hydrocephalus and endocrine dysfunction.

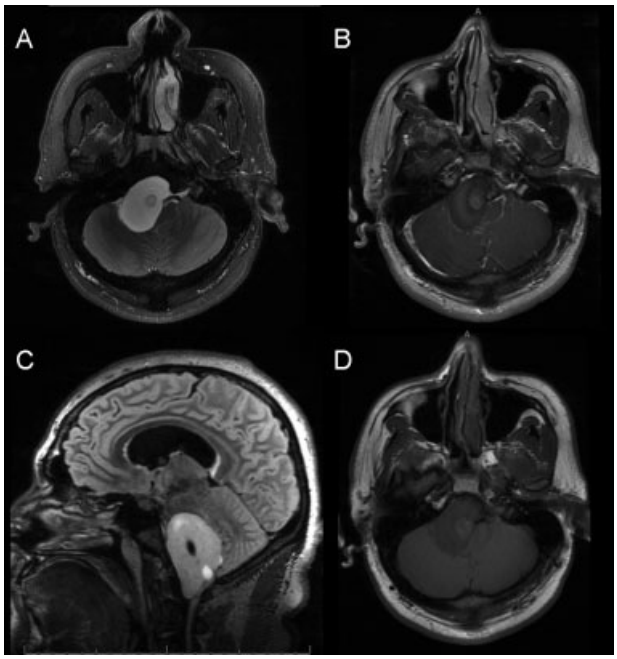


**Fig. 2** Preoperative magnetic resonance (MR) imaging from case 2, a 25-year-old man initially thought to have racemosus neurocysticercosis. However, histology showed a neuroenteric cyst. (A) Axial fluid-attenuated inversion recovery (FLAIR). (B) Axial T1 postgadolinium. (C) Sagittal FLAIR. (D) Axial T1 without contrast.

The preoperative differential diagnosis considered for the posterior fossa lesions included arachnoid cyst, epidermoid cyst, and neuroenteric cyst (in three cases). In one case, the preoperative diagnosis was racemosus neurocysticercosis. This patient was a 25-year-old man (►**Table 1**, case 2), originally from Mexico, who presented with 3 to 4 months of dull headaches, hoarse voice, and facial numbness. His MRI showed a loculated cystic lesion with proteinaceous content ventral to the right pons and medulla extending inferiorly through the foramen magnum, with a ring-enhancing central structure (►**Fig. 2**). The preoperative diagnosis was strongly favored to be racemosus neurocysticercosis. He was evaluated by the infectious diseases service and treated empirically with albendazole prior to surgical removal and the final pathology diagnosis of neuroenteric cyst.

In the case of the third ventricular lesion, the preoperative diagnosis was favored to be craniopharyngioma, colloid cyst,

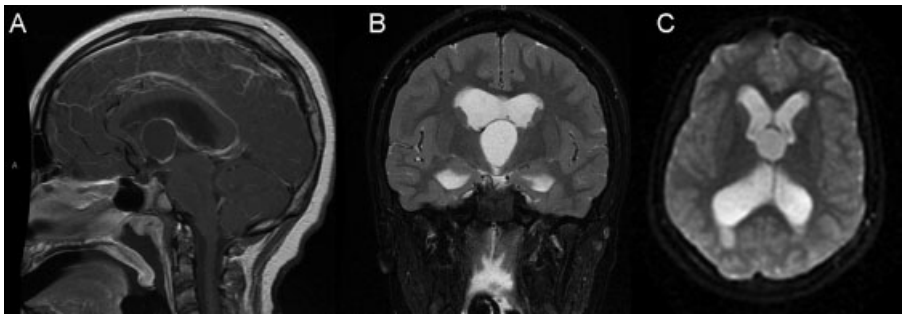


**Fig. 3** Case 1, a 10-year-old girl who presented with precocious puberty and headaches. (A) Sagittal T1 postgadolinium, (B) coronal T2, and (C) axial diffusion-weighted images demonstrating a third ventricular cystic lesion. Final pathology was neuroenteric cyst.

or cystic glioma given its location and imaging appearance (►**Fig. 3**). Histologically the cyst was composed of cuboidal/ciliated columnar epithelium with mucinous differentiation and goblet cells, and focal squamous metaplasia. Of note, this patient was also later diagnosed with a midline nasopalatine canal cyst.

The five posterior fossa cysts were approached through a far lateral transcondylar craniotomy, as suggested recently by other groups.[3,5] In one case, a portion of the cyst wall was left behind. Gross total resection by postoperative MRI was achieved in four of five posterior fossa cases. Postoperative complications included aseptic meningitis (one case), temporary abducens palsy (one case), and pseudomeningocele requiring reoperation (three cases). There were no delayed postsurgical complications after a mean follow-up time of 6.5 months (range: 0.5–16 months). All patients had symptom improvement or resolution. Pre- and postoperative mRS scores are shown in ►**Table 1**. At a mean follow-up time of 10 months, there was no radiologic evidence of cyst recurrence in three cases for which follow-up MR imaging was available.

## Discussion

Intracranial neuroenteric cysts are rare and benign congenital lesions most commonly arising in the posterior fossa. Due to their rarity, our knowledge about these lesions is primarily derived from case reports and small series. The embryology, as well as the clinical, radiographic, and histological characteristics of these lesions were recently reviewed by Gauden et al and others.[1,3,4] The goal of this study was to provide additional insight on the clinical characteristics of these rare lesions and report the 15-year single institutional experience with surgical treatment of these rare lesions.

The incidence of neuroenteric cyst among new intracranial lesions is quite low. Although there are no actuarial incidence studies related to neuroenteric cysts, we have observed a rate of 0.053% among in-house neuropathology cases at a high-volume referral center over a 15-year period.[3] Five of the six cases had a typical location in the posterior fossa, anterior to the brainstem.

In general, neuroenteric cysts are thought to arise from ectopic endodermal elements within the central nervous system. Their occurrence anterior to the brainstem and caudal to the dorsum sella has been explained by vestigial remnants of the neuroenteric canal.[1] Uschold and colleagues identified anterior displacement of the vertebrobasilar vessels away from the pial surface of the brainstem as a unique radiographic feature of neuroenteric cysts.[5] This feature was seen in all five posterior fossa cases. The far lateral transcondylar approach to these lesions allows for complete excision in most cases.

There was one case of a third ventricular cyst in a 10-year-old patient. Neuroenteric cysts are exceptionally rare in this location and age group. To our knowledge, there have only been two other cases of third ventricular neuroenteric cysts reported in the literature.[7,8] Seessel pouch, an endodermal diverticulum of the respiratory tract, has been proposed as the possible origin of these suprasellar neuroenteric cysts.[9,10] Histologically, Preece et al described two major histologic patterns, differentiated primarily by the presence of mucin-producing cells.[4] The cyst walls are composed of simple or pseudostratified columnar or cuboidal epithelium on a basement membrane (►**Fig. 4**). The epithelium may also be ciliated. Extremely rare cases of malignant degeneration have also been reported.[11]

This retrospective study has several important limitations. First, the small number of cases limits the strength of the conclusions. Due to the rarity of intracranial neuroenteric

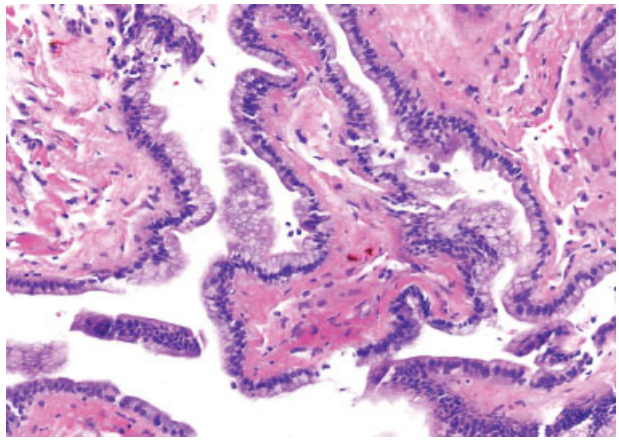

**Fig. 4** Hematoxylin and eosin stain of the cyst in case 3. Simple columnar epithelium with goblet cells is suggestive of intestinal epithelium and diagnostic for neuroenteric cyst (original magnification ×200).

cysts, a prospective study is unlikely. Our experience with six cases constitutes one of the largest single-institution series. A second limitation is the short follow-up time. Neuroenteric cysts are known to be slow-growing benign lesions that recur on the order of 7.8 years (range: 2 months to 32 years) in the literature.[12] Therefore longer follow-up times, on the order of many years, are needed to have a meaningful understanding of the intracranial recurrence rate.

## Conclusions

Intracranial neuroenteric cysts are rare and can have a variable imaging appearance on both CT and MR imaging. They should be considered in the differential for cystic posterior fossa lesions, and ventral displacement of the vertebrobasilar complex appears to be a characteristic imaging feature.[5] In many cases, complete surgical resection is possible—often using a far lateral transcondylar approach given their frequent location ventral to the pons and medulla.[3] In this series, surgical resection resulted in resolution of symptoms with no delayed complications.

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