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Case Report

# Intracranial neurenteric cyst: A case report and differential diagnosis of intracranial cystic lesions ☆

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ABSTRACT

Neurenteric cysts are uncommon, benign lesions that are rarely located intracranially. These cysts are likely due to aberrant embryologic development of the notochord. Clinically, neurenteric cysts may present with symptoms of mass effect, or they can be asymptomatic and incidentally discovered. Imaging features of neurenteric cysts have significant overlap with other intracranial cystic lesions, which can make diagnosis difficult. We present a case of a 35-year-old female with a histopathologically confirmed neurenteric cyst in the premedullary and left cerebellomedullary cistern, with associated symptoms of headache, dizziness, tinnitus, and dysphagia. The patient underwent surgical resection, with improvement in symptoms. We present a review of literature, and a discussion of typical features of multiple intracranial cystic lesions. We hope to promote accurate preoperative diagnosis, to allow for appropriate surgical technique to reduce the risk of recurrence.



## Introduction

Neurenteric cysts (NC), also known as endodermal or enterogenous cysts, are rare benign lesions that can occur anywhere along the central neuraxis. They are typically located ventral to the cervical or thoracic spinal cord, though there is a small minority of reported intracranial lesions [1,2]. The imaging of these intracranial lesions has significant overlap with other intracranial cystic lesions. Due to their rarity and nonspecific imaging characteristics, intracranial neurenteric cysts are often misdiagnosed prior to surgical resection. We present a case of a pathologically proven intracranial neurenteric cyst, with a review of the literature and overview of the radiologic differential diagnosis for intracranial cysts.

☆ **Patient Consent Statement:** Informed consent was obtained from the patient for the purpose of publishing a case report with use of their preoperative and postoperative imaging, as well as clinical history and course. The patient has consented to publication of their imaging, pathology, medical history, physical exam, and clinical course, as well as disclosure to Elsevier and use by Elsevier or its licensees in any work or product. Written consent is on file and will not be uploaded to maintain anonymity. All personally identifying material has been removed from submitted content.

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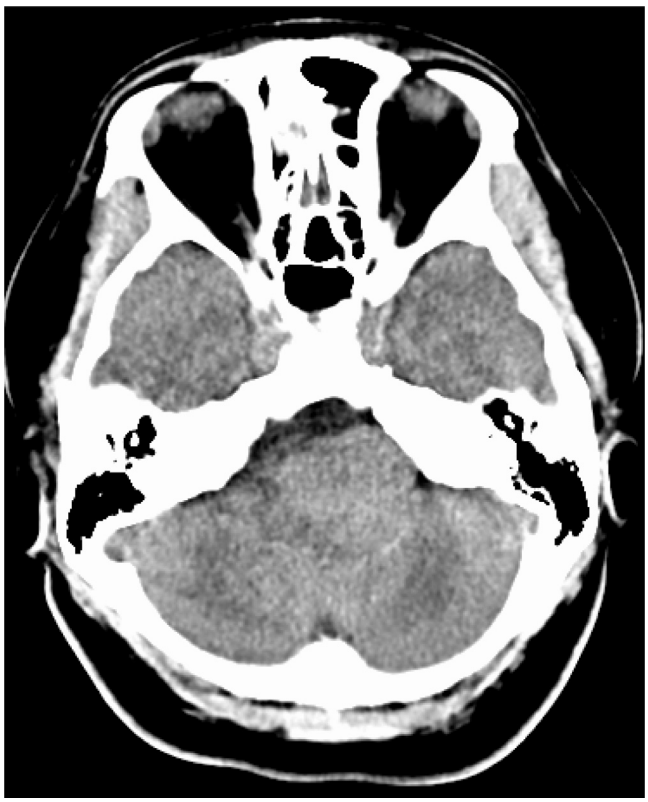

**Fig. 1 – Nonenhanced axial CT image shows the mass is isodense to gray matter. No calcifications were seen.**

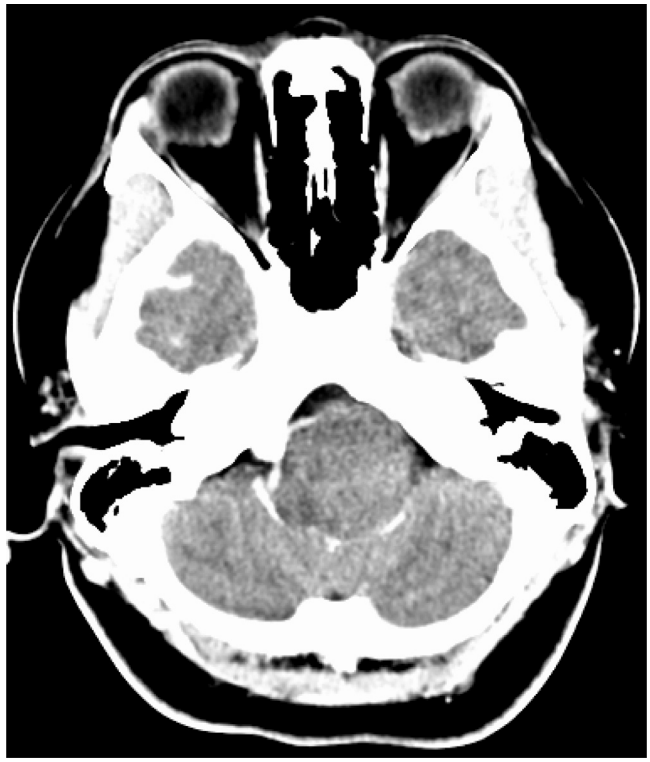

**Fig. 2 – Axial CT angiogram shows the typical displacement of the adjacent vasculature. No enhancement of the lesion was seen.**

## Presentation

A 35-year-old female presented with neck pain following a motor vehicle accident, and was found to have a cystic mass in the left cerebellomedullary cistern extending into the premedullary cistern on imaging of the cervical spine. Upon further questioning, she reported a 15-year history of migraine headaches. Since the delivery of her twins 1 year prior, her headaches had increased in severity and had also developed a positional quality, as they were exacerbated by lying down. Additionally, she reported a 3-month history of memory impairment and dizziness, along with a 1-year history of gradual-onset tinnitus and dysphagia. Physical exam was unremarkable.

### *Imaging*

Computed tomography (CT) head without contrast showed the mass was isodense to gray matter (Fig. 1). CT angiogram demonstrates displacement of the prepontine vasculature without encasement (Fig. 2). Magnetic resonance imaging (MRI) with contrast showed a T1 and T2 hyperintense mass in the left cerebellomedullary cistern extending into the premedullary cistern (Figs. 3 and 4). The mass measured 3.4 × 3.3 × 3.3 centimeters in transverse, anteroposterior, and craniocaudal dimensions, respectively. There was associated mass effect on the brainstem and cerebellum without associated edema (Fig. 3D). The mass did not restrict on diffusion-weighted imaging (DWI, Fig. 3E). There was a focus of signal loss on susceptibility-weighted imaging likely due to hemosiderin, as there were no calcifications on CT (Fig. 3F). There were no areas of enhancement (Fig. 3B).

### *Operative findings and pathology*

Due to symptomatology, the patient underwent left suboccipital craniotomy and stereotactic-guided volumetric resection of the tumor. The mass was located within the subarachnoid space and was adherent to the left spinal accessory nerve and left vertebral artery, from which it was dissected (Fig. 5).

Intraoperative cyst disruption revealed yellow, liquid cyst contents that did not yield any growth on bacterial or fungal cultures. The disrupted cystic lesion was sent for frozen section, which revealed a histiocyte-rich lesion with xanthogranulomatous change. The findings were consistent with a benign lesion, but definitive diagnosis was not possible during the time of frozen section.

Microscopic examination of permanent sections demonstrated a benign cystic lesion lined by pseudostratified, ciliated epithelium with focal squamous metaplasia. The adjacent tissue contained abundant histiocytes in the setting of xanthogranulomatous inflammation (Fig. 6).

Immunohistochemical studies were performed. Cytokeratin staining was positive in the epithelial cell lining, and

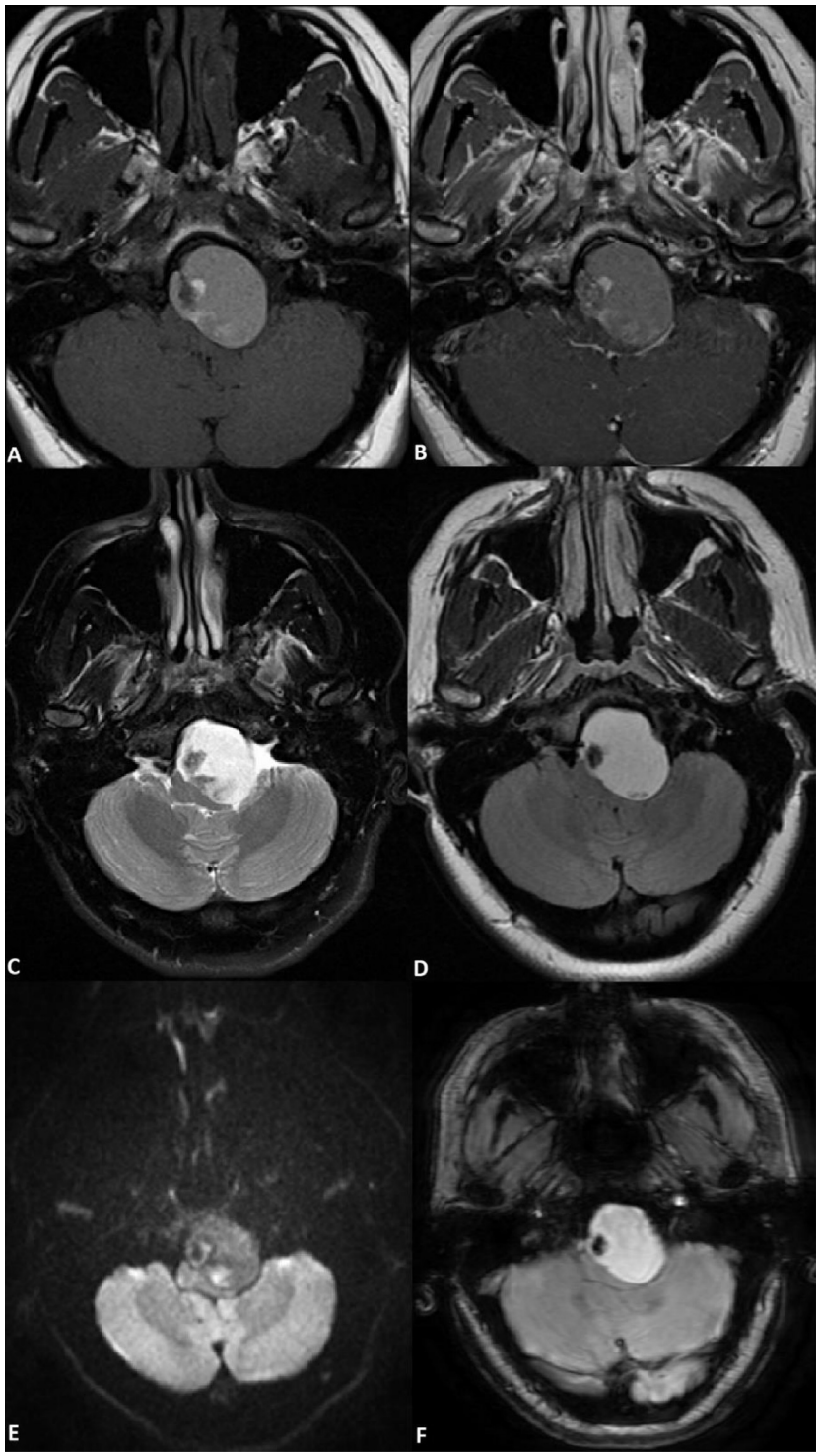


**Fig. 3 – (A) Axial T1 precontrast shows the well circumscribed mass in the left cerebellomedullary and premedullary cisterns with associated mass effect on the brainstem and cerebellum. The mass is hyperintense to CSF. The area of low T1 signal above corresponds to an area of low T2 signal, and loss of signal on SWI, likely representing a focus of hemosiderin. Foci of T1 hyperintensity above showed T2 hypointensity and likely represents areas of proteinaceous material. (B) Axial T1 postcontrast shows the mass does not have any areas of enhancement. (C) Axial T2 fat suppressed image shows the hyperintense mass. (D) Axial T2 fluid-attenuated inversion recovery (FLAIR) image shows the mass does not suppress and is hyperintense to CSF. There is no surrounding edema of the brain parenchyma. (E) DWI shows that the mass does not restrict diffusion. (F) Axial SWI shows a foci of signal loss. This corresponds to a region of T1 and T2 hypointensity and likely represents hemosiderin. No calcifications were seen on CT.**

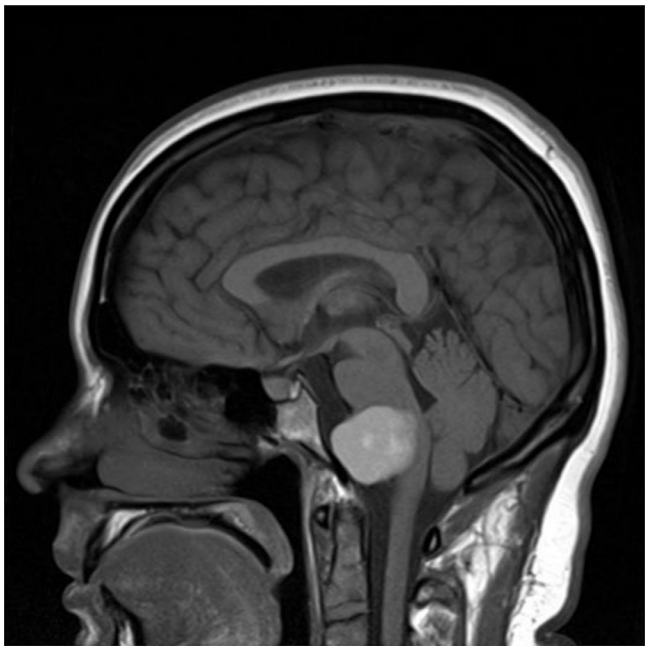

**Fig. 4 – Sagittal T1 image shows a well-circumscribed mass in the premedullary and left cerebellomedullary cisterns that is hyperintense to CSF. There is associated mass effect on the brainstem and cerebellum.**

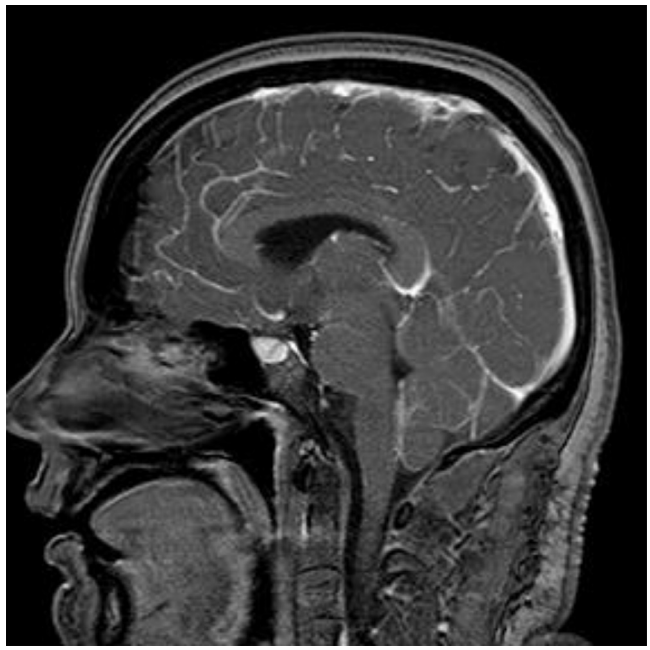

**Fig. 5 – Postoperative sagittal T1 with contrast image shows resection of the mass.**

CD68 highlighted the abundant histiocytes. Neuroglial markers, including glial fibrillary acidic protein and OLIG2, were negative. Cytopathologic analysis of the cyst fluid revealed macrophages and admixed ciliated epithelium. These morphologic, immunohistochemical, and cytologic features are consistent with a diagnosis of a benign endodermal cyst.

*Postoperative course*

Postoperatively, the patient experienced left cranial nerve VI palsy that gradually improved, as well as mild right hemibody weakness and sensory deficits that also improved over the course of the next few months. The patient also reported postoperative improvement of her subjective memory impairment.

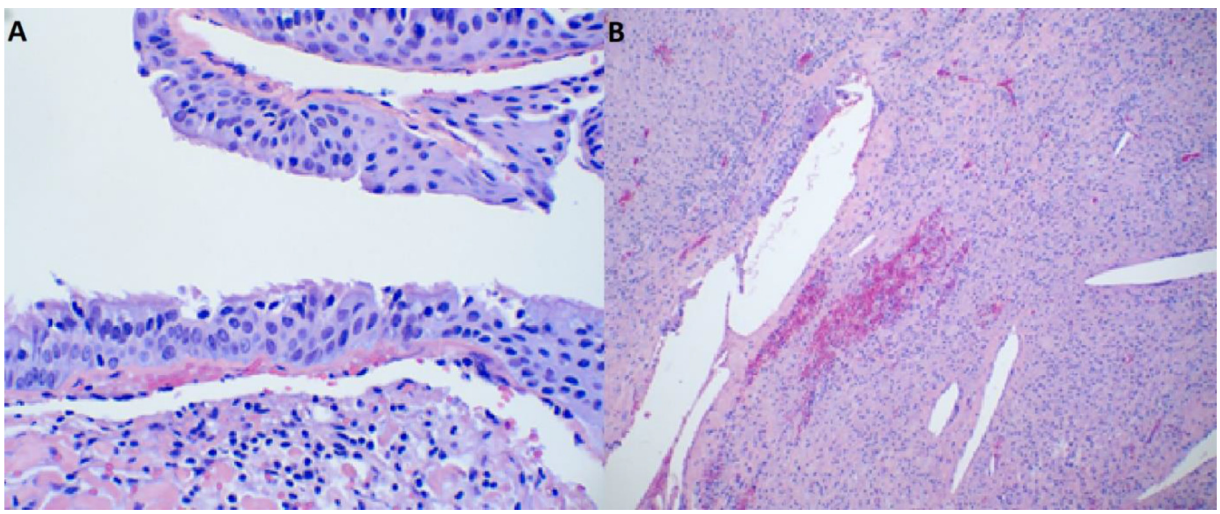


**Fig. 6 – Microscopic examination of the cyst lining reveals a pseudostratified ciliated epithelium, similar to typical respiratory-type mucosa (A). Adjacent xanthogranulomatous inflammation with cholesterol cleft formation, foamy lipid-laden macrophages, and multinucleated giant cell reaction (B).**

## Discussion

A neurenteric cyst is an endodermal lesion of the central nervous system, and has been previously known as an enterogenous cyst, enteric cyst, endodermal cyst, gastroenterogenous cyst, gastrocytoma, intestinoma, and archenteric cyst [3]. They are rare entities, and can occur anywhere within the central nervous system, most commonly within the spinal canal. Neurenteric cysts represent approximately 1.3% of all spinal tumors and 0.15%-0.35% of all intracranial tumors [2]. Spinal neurenteric cysts are typically ventral to the cord and are associated with vertebral abnormalities in 50% of cases [3]. Intracranial neurenteric cysts have been estimated to represent 17.9% of cases. Of those, 72.2%-90% arise in the posterior fossa, usually anterior to the brainstem or in the cerebellopontine angle [2].

Much less commonly they have been reported within the brainstem, in the fourth ventricle, and supratentorially [2–4]. These lesions have been found in patients of all ages.

### *Pathogenesis*

The exact pathogenesis of neurenteric cysts remains to be elucidated. However, the most common hypothesis is that these lesions arise after a failure of obliteration of the neurenteric canal (a transient connection that links the amniotic cavity to the yolk sac during the third and fourth weeks of embryogenesis, traversing both endoderm and ectoderm in the process). The result is a failure of complete separation of the notochord from the foregut, with displaced endodermal cells. Thought to arise by a similar mechanism, neurenteric cysts are associated with increased incidence of vertebral abnormalities, intra-abdominal cysts, and intrathoracic cysts [5]. Histologically, neurenteric cysts are defined as benign cystic lesions lined by respiratory-type or gastrointestinal-type epithelium. The cyst lining can range from simple to pseudostratified, cuboidal to columnar epithelium, with or without cilia [6,7]. While extremely rare, there are few reports of malignant transformation [7].

### *Presentation*

Clinical symptoms are due to mass effect, and presentation is therefore variable and related to lesion size and location. Headache is a common symptom of intracranial neurenteric cysts, present in 47.1% of cases in one case series [3], as well as in our patient. Neurenteric cysts in the posterior fossa often present with vertigo or imbalance, and less commonly hearing loss or tinnitus [3,5]. Symptoms from associated cranial nerve compression may also be present [1,3,5,8]. In the rare cases of supratentorial neurenteric cysts, symptoms are typically due to increased intracranial pressure, including headache, nausea, and vomiting [5]. Depending on location of the lesion, patients may also present with seizures or motor or sensory deficits [3,5,13]. Uncommonly, cyst contents may leak into the subarachnoid space and cause aseptic meningitis [3,9]. Symptoms are typically insidious, related to the slow growth of the cyst. Patients may also be asymptomatic, and these lesions may be identified incidentally.

### *Imaging characteristics*

Radiologic appearance is somewhat variable, likely related to protein content of the cyst. On CT, neurenteric cysts are typically hypodense without enhancement, although they can appear isodense, and rarely hyperdense [2,5,10]. The cysts do not show internal enhancement. Intracranial neurenteric cysts are much less likely to be associated with vertebral anomalies compared to spinal neurenteric cysts [3].

MRI is more helpful in diagnosis, typically showing isointense to hyperintense signal compared to cerebrospinal fluid (CSF) on T1-weighted images due to the protein content. T2-weighted images will show hyperintensity. On FLAIR sequences, the cyst will show hyperintense signal without surrounding edema. DWI may show mild restriction [3]. Mild wall enhancement may be present but is rare [2,3].

### *Differential diagnosis*

Due to the nonspecific imaging characteristics of neurenteric cysts, the differential diagnosis is broad. Other similar appearing intracranial lesions include arachnoid cysts, choroid plexus cysts, colloid cysts, dermoid or epidermoid cysts, ependymal cysts, notochordal remnants including ecchordosis physaliphora, parasitic cysts, primary or metastatic cystic tumors, and Rathke cleft cysts. Location of the lesion may be helpful in diagnosis, as neurenteric cysts are more common in the posterior fossa. In contrast, an arachnoid cyst is more commonly found in the middle cranial fossa. An arachnoid cyst, like a neurenteric cyst, usually appears as a rounded cyst that displaces adjacent structures. An arachnoid cyst typically follows CSF appearance on CT and MR, and characteristically has suppressed signal on FLAIR sequences without restriction on DWI. A choroid plexus cyst will be located within the ventricles, most commonly within the choroid plexus of the lateral ventricles, and less commonly in the third or fourth ventricles. In contrast to neurenteric cysts, prominent restricted diffusion is a common finding with choroid plexus cysts, as are peripheral calcifications [14]. An ependymal cyst has imaging features nearly identical to an arachnoid cyst, appearing as a thin-walled cystic structure with signal paralleling CSF. Location is key in distinguishing ependymal cysts, which will typically be located within the ventricles, most commonly the lateral ventricles. Epidermoid and dermoid cysts are typically more lobulated, in contrast to the rounded appearance of neurenteric cysts. Epidermoid and dermoid cysts are more likely to show calcification when compared to neurenteric cysts [5], although most epidermoid and dermoid cysts will not have calcifications. A key distinguishing factor is that irregular contours of a dermoid or epidermoid cyst are more likely to encase neurovascular structures, while the slowly expanding neurenteric cyst is more likely to displace adjacent structures. Both dermoid and epidermoid show more marked restriction on DWI when compared to NC. Rathke cleft cysts and colloid cysts have similar imaging characteristics as they are also thought to have an endodermal origin. Differentiation from NC is based on location of the lesion, with Rathke cleft cysts located in the sellar or suprasellar region, and colloid cysts located adjacent to the foramen of Monro. Primary cystic neoplasms and cystic metastases are differentiated by their

contrast enhancement, a feature not typically present with neurenteric cysts. Ecchordosis physaliphora will follow CSF signal and is typically located in the prepontine cistern. These lesions can be distinguished by a typical association with the clivus, commonly in the form of an osseous stalk or pedicle. Less commonly there may be a well-defined, scalloped bony defect in the adjacent clivus [11]. Parasitic infections such as neurocysticercosis show similar signal to CSF, but are typically multiple and may show leptomeningeal enhancement. Cystic lesions of neurocysticercosis will more commonly be located within brain parenchyma. Due to the variability in appearance of these cystic entities, definitive diagnosis can only be made by histologic analysis.

### *Management*

The current treatment recommendation for neurenteric cysts is surgical excision in symptomatic patients. Complete resection is ideal to reduce the risk of recurrence, although this may be precluded by adherence of the cyst to critical structures [15]. It has been shown that partial resection is more likely in cases of incorrect preoperative diagnosis, which may lead to suboptimal surgical approach [12]. Recurrence rates have been reported to range from 11.9% to 37%, with timeline of recurrence ranging from 4 months to 14 years postoperatively [12]. Because of the variability of recurrence, one source has recommended clinical and radiological follow-up for at least 10 years postoperatively [5].

## Conclusion

Neurenteric cysts are rare benign cystic lesions of the central nervous system. Although more commonly encountered in the regions of the thoracic and cervical spine, intracranial neurenteric cysts should be included in the differential diagnosis of an intracranial cystic mass. Imaging features can help distinguish lesions, but definitive diagnosis relies on histopathologic analysis. Previous case reports have found that they are typically symptomatic, although with advances and increases in imaging, neurenteric cysts are being diagnosed more frequently, and may even be detected prior to symptoms. The differential for intracranial cystic lesions is large, although there are typical features and important distinctions to be aware of. Accurate diagnosis of neurenteric cyst on imaging is important to allow appropriate surgical planning, which can reduce the risk of recurrence.

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