



Case Report

Fenestration of intracranial neurenteric cyst: A case report

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ABSTRACT

Background: Neurenteric cysts are rare congenital lesions of endodermal origin which result from the failure of the neurenteric canal to close during embryogenesis. The majority of neurenteric cysts occur in the spinal cord, though in rare instances can occur intracranially, typically in the posterior fossa anterior to the pontomedullary junction (80%) or in the supratentorial region adjacent to the frontal lobes (20%).

Case Description: We present the case of a 75-year-old woman with an extra-axial cystic lesion centered in the premedullary cistern causing brainstem compression. The lesion was later histopathologically confirmed to be a neurenteric cyst. She presented initially with a 4-month history of worsening headache, dizziness, and unsteady gait. We performed a left retrosigmoid craniotomy for cyst fenestration/biopsy with the aid of operating microscope and stealth neuronavigation. Following the procedure, the patient recovered without complications or residual deficits.

Conclusion: This case illustrates the successful fenestration of an intracranial neurenteric cyst with good clinical outcome. We present the pre- and post-operative imaging findings, a technical video of the procedure, histopathological confirmation, and a brief review of the relevant clinical literature on the topic.

Keywords: Cerebellopontine angle, Fenestration, Intracranial neurenteric cyst, Premedullary cistern, Retrosigmoid craniotomy

INTRODUCTION

Neurenteric (endodermal) cysts are uncommon, benign lesions of the central neuraxis that typically occur intraspinally because of aberrant embryological development of the notochord.^[1,4,8,14] These lesions typically present either incidentally on neuroimaging or with typical symptoms of mass effects, such as headache and dizziness. In addition, lesions in this region have the potential to cause mass effect, compression of cranial nerves, and hydrocephalus, thus highlighting the importance of surgical resection rather than a “watch and wait” strategy.^[3,10] Given the rarity of these lesions and their relatively indistinct neuroimaging findings, they are often misdiagnosed prior to resection.^[1] Here, we present a case of a neurenteric cyst in the premedullary cistern, the associated pre- and post-operative imaging findings, histopathology images of the cyst, and a video demonstration of cyst fenestration after a left retrosigmoid craniotomy.

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CASE REPORT

History and examination

A 75-year-old woman presented with a 4-month history of worsening headaches along with progressive weakness, dizziness, and lightheadedness mainly when going from supine or sitting to standing. On neurological examination, the patient was noted to have mild resting tremor in the left hand and horizontal diplopia when attempting to look to the left, along with impaired convergence. The patient was also noted to have gait unsteadiness and poor balance.

Pre-operative neuroimaging

Computed tomography (CT) of the head without contrast showed a mildly hyperdense large extra-axial mass at the upper aspect of the premedullary cistern extending mildly to the inferior left cerebellopontine angle with compression of the brainstem [Figure 1a]. The mass was closely associated with the adjacent vertebral arteries and basilar artery. Posterior fossa cisterns were noted to be crowded by the mass. No evidence of osseous skull base involvement/destruction was observed.

Magnetic resonance imaging (MRI) with/without contrast of the brain showed a T2-fluid-attenuated inversion recovery (FLAIR), well-circumscribed, hyperintense mass in the left cerebellopontine angle and premedullary cistern [Figure 1b]. There was local mass effect on the brainstem without associated edema. The sagittal T1-FLAIR image showed isointense mass [Figure 1c].

Operative details

A left retrosigmoid craniotomy was performed for neuroenteric cyst fenestration/biopsy with the use of

operating microscope and use of Medtronic Stealth neuronavigation. A yellowish cyst was identified which was opened using an arachnoid knife and yellowish fluid immediately returned [Video 1]. This was copiously irrigated. A small section of the cyst was removed using scissors and sent for histopathological evaluation.

Histopathological findings

Microscopic examination of the tissue sections demonstrated a benign fibromembranous cystic lesion lined by pseudostratified, ciliated columnar epithelium, and providing a histopathological diagnosis of neurenteric cyst. [Figure 2] depicts the photomicrographs of hematoxylin and eosin (H and E) stained tissue sections.

Postoperative imaging and course

Postoperatively, the patient recovered well, and post-operative CT of the head without contrast showed no residual mass [Figure 3]. Headache and dizziness resolved post-operatively and patient was symptom-free at 5-month follow-up. However, the patient was diagnosed with Parkinson's disease on a neurology consultation, likely responsible for her resting tremor noted on initial presentation.

DISCUSSION

Clinical characteristics

A neurenteric cyst is a central nervous system lesion of endodermal origin which most frequently occurs in the spine. Intracranial neurenteric cysts have been estimated to represent about 17.9% of occurrences and of these, the vast majority occur in the posterior fossa. Rare cases of neurenteric cyst have been noted to occur in the brainstem,

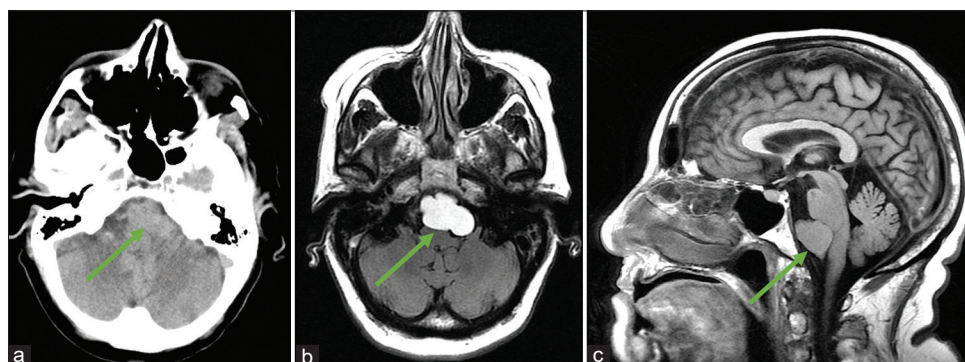


Figure 1: Pre-operative neuroimaging. (a) Axial computed tomography-head image showing mildly hyperdense large extraaxial mass at the upper aspect of the premedullary cistern extending mildly to the inferior left cerebellopontine angle with compression of the brainstem. (b) Axial magnetic resonance imaging (MRI)-brain T2- fluid-attenuated inversion recovery (FLAIR) image showing hyperintense left premedullary cistern and cerebellopontine angle mass. (c) Sagittal MRI-brain T1-FLAIR image showing isointense premedullary cistern mass. Arrow points to the left premedullary cistern mass later histopathologically confirmed to be a neurenteric cyst.

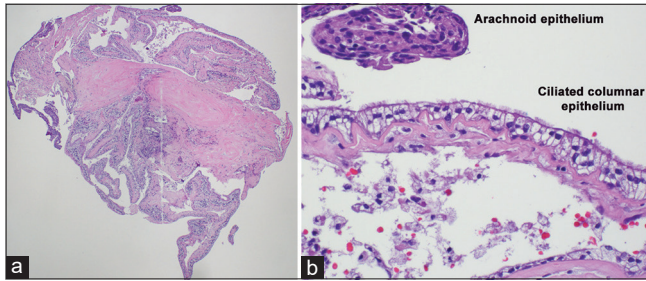


Figure 2: Microscopic examination of the cyst lining reveals a pseudostratified ciliated columnar epithelium. (a) Low-magnification hematoxylin and eosin (H and E) staining showing fibromembranous tissue lined by columnar epithelium. Imaged at $\times 4$ magnification. (b) High-magnification H and E staining showing pseudostratified ciliated columnar epithelium and adjacent arachnoid epithelium. Imaged at $\times 40$ magnification.

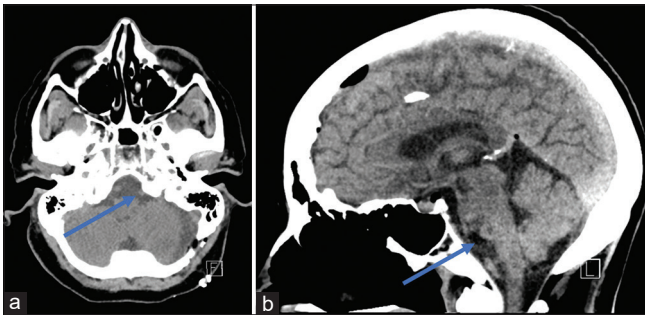


Figure 3: Post-operative neuroimaging. Computed tomography-head images, axial (a) and sagittal (b), showing empty spaces (arrows) at neurenteric cyst location after fenestration.

fourth ventricle, and supratentorial region.^[1] Even though the precise etiology of neurenteric cyst formation remains largely unknown, the widely accepted theory is that it arises from aberrant embryologic development during notochord formation.^[3,8,12] Most patients present with signs and symptoms of local mass effect such as cranial nerve deficits, signs of aseptic meningitis due to irritation from local spilling of cystic material, and myelopathy in the case of intraspinal lesions.^[7,10,14] It is likely that our patient's horizontal diplopia was the result of such cranial nerve compression.

Imaging

The radiological characteristics of neurenteric cysts are heterogeneous. CT of neurenteric cysts typically reveal hypodense lesions, but can also appear isodense or, more rarely, hyperdense. CT-head in our case revealed a mildly hyperdense lesion. In addition, our patient's cyst did not show internal enhancement, which is consistent with the literature.^[7,11]

On MRI-brain, a classic neurenteric cyst usually presents as a well-circumscribed cystic mass which is enhancing and slightly iso- or hyperintense on T1-weighted sequences

and slightly more hyperintense on T2-weighted-FLAIR sequences with mild restriction diffusion noted on diffusion-weighted imaging secondary to its high proteinaceous content.^[7,11,12] The majority of neurenteric cysts are small, measuring < 2 cm.^[12] However, there are rare reported cases of larger-sized supratentorial neurenteric cysts with intracystic calcifications.^[13]

Surgical management

The standard of care for patients with a symptomatic neurenteric cyst is complete surgical excision. However, complete excision can be difficult if the cyst is adherent to important local structures, as was the case with our patient's cyst which was adherent to the nearby vertebral and basilar artery. Although difficult, it is important to achieve as neurenteric cysts have an 11.9–37% recurrence rate during a span of 4 months to 14 years, thus resulting in the recommended 10-year follow-up period.^[1] Furthermore, while complete surgical excision is the standard of care and was indicated in our patient, this might not necessarily always be the case. Waqas *et al.* reported a case of a 41-year-old man with a prepontine epidermoid cyst who was treated with steroids and antibiotics, and symptoms resolved with radiologic regression of the cyst.^[15] This indicates that in certain situations, such as when a patient may have significant risk of surgical complications and is clinically stable, a “watch and wait” approach with close clinical and radiologic follow-up may prove fruitful despite the rarity of spontaneous regression. However, more data on such non-progressive and minimally symptomatic cases are needed.

In the present case report, the cyst was fenestrated using a left retrosigmoid approach. This is the primary means employed to gain access to the cerebellopontine angle - an anatomically complex site with many critical structures. Various vascular and neoplastic lesions that are approached in this manner include vestibular schwannomas, meningiomas, and aneurysms of nearby arteries.^[5,9]

Histopathology

Three major histopathologic sub-types of neurenteric cysts have been noted in literature: Types A, B, and C.^[2,6,8,12] Type A most closely resembles respiratory or gastrointestinal epithelium and contains a pseudostratified layer of ciliated columnar or cuboidal epithelium situated on a basement membrane and connective tissue. Type B cysts contain simple, non-ciliated mucin-producing epithelium superior to/underlaid by more connective tissue and can contain smooth muscle, glandular, and lymphoid tissue, as well as nerve ganglia. Type C cysts are essentially type B cysts with additional glial cells. The cyst presented in our case can be classified as a type A cyst because of the presence of pseudostratified ciliated columnar epithelium [Figure 2].

CONCLUSION

Neurenteric cysts are a rare, benign lesion of the central nervous system, typically occurring in the spine and less commonly intracranially. In the present report, we describe the case of a 75-year-old woman with a symptomatic lesion centered in the premedullary cistern causing brainstem compression. Fenestration of the intracranial cyst through retrosigmoid craniotomy resulted in good post-operative outcome. The associated imaging findings, operative video detailing fenestration of the intracranial neuroenteric cyst, and histopathologic findings are presented, along with relevant literature on the topic.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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