SAGE Open Medical Case Reports
Volume 9: 1–5

DOI: 10.1177/2050313X211067913
journals.sagepub.com/home/sco
SAGE

Neurenteric cyst of the temporal bone: A case report

Timothy Fan[1], Ezer Benaim[1], Jorge Lee Diaz[2] and Charles MacDonald[1]

Abstract

Neurenteric cysts are rare, developmental malformations mainly found in the spinal canal. The authors report on a 29-year-old woman who presented with congenital left-sided hearing loss and a 9 days history of left ear pain and facial weakness (House–Brackmann IV). Radiological examination revealed a complex 7 mm cystic structure involving the petrous and mastoid portions of the left temporal bone. Intraoperatively, a red-brown multicystic mass was discovered and excised completely. Histologic findings showed simple columnar epithelium with goblet cells and bilayers of submucosal musculature, resembling normal intestinal linings. Post resection, the patient regained nearly full facial nerve capabilities (House–Brackmann II) with complete resolution of pain, but no changes to her hearing. The objectives of this case report are to describe the clinical, radiologic, intraoperative, and pathologic characteristics of an extradural temporal neurenteric cyst with sensorineural deficits to contribute to the growing knowledge regarding this rare entity.

Keywords

Neurenteric cyst, temporal bone, temporal cyst, congenital hearing loss, facial paralysis

Date received: 18 September 2021; accepted: 2 December 2021

Introduction

Neurenteric cysts, also known as enterogenous cysts or endodermal cysts, are a rare developmental malformation hypothesized to originate from the persistence of the neurenteric canal between the notochordal plate and the endoderm.[1,2] The majority of neurenteric cysts arise at the cervicothoracic intraspinal region, with a small fraction occurring intracranially, and even rarer occurrences elsewhere.[1,3] It is difficult to assess the true frequency of neurenteric cysts, due to its sparsity in literature, however, one publication estimated the incidence rate of neurenteric cysts in the central nervous system (CNS) to be 0.01%.[4]

While rare, neurenteric cysts may induce headache, seizures, and cranial nerve palsies due to mass effect.[1] In addition, neurenteric cysts may hemorrhage internally, leading to rapid destabilization of patient's health.[1] Infrequently, neurenteric cysts may eject content externally, inducing aseptic meningitis, hydrocephalus, or brain abscess.[1] Albeit uncommonly seen, neurenteric cysts have also been associated with spinal dysraphism, and gastrointestinal (GI), renal, cardiac, and dermal abnormalities.[5,6] Publications of extradural neurenteric cysts originating from the temporal bone are limited in literature,[7] with no prior reports of cases contributing to or exacerbating ipsilateral congenital inner ear malformations, resulting in hearing loss. The authors aim to describe the clinical, radiologic, intraoperative, and pathologic characteristics of a temporal neurenteric cyst to contribute to the current understandings of this rare entity.

Case report

A 29-year-old woman with a lifelong history of unevaluated left-sided deafness was referred to our clinic with left ear and neck pain and left facial weakness for the past 9 days. Physical exam noted normal otoscopy with tender and swollen posterior

[1]Department of Otolaryngology—Head and Neck Surgery, University of Tennessee Health Science Center, Memphis, TN, USA
[2]Department of Radiology, LeBonheur Children's Hospital, Memphis, TN, USA

Corresponding Author:
Timothy Fan, Department of Otolaryngology—Head and Neck Surgery, University of Tennessee Health Science Center, 910 Madison Avenue, Suite 423, Memphis, TN 38163, USA.
Email: tfan3@uthsc.edu

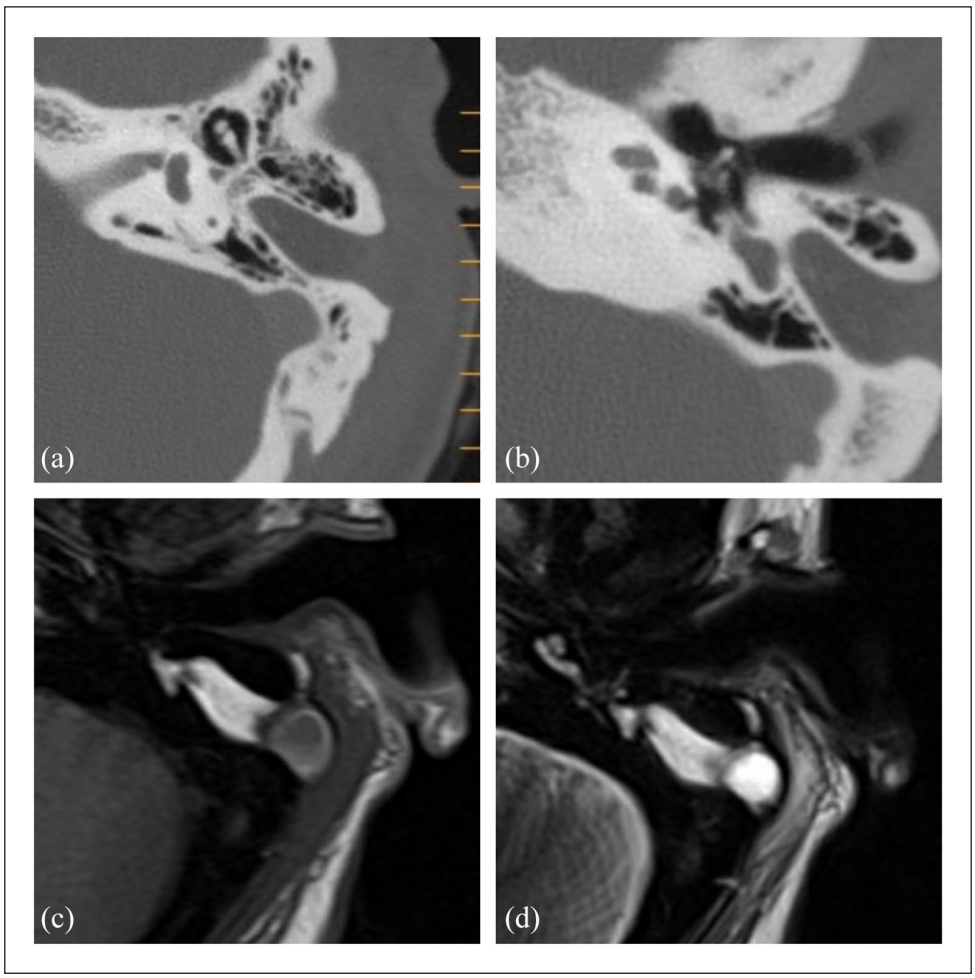

Figure 1. (a) Axial CT of the head with no lateral semicircular canal present. (b) Axial CT of the head showing a hypoplastic cochlea. (c) Axial T1-weighted MRI of the head without gadolinium revealing a mastoid lesion with bright signal. (d) Axial T2-weighted MRI of the head with bright signals from both components of a mastoid lesion.

auricle without erythema, and left hemifacial paresis (House–Brackmann IV). She was afebrile, and laboratory studies were inconsistent with infection. Audiometry demonstrated profound sensorineural hearing loss on the left side with normal hearing on the right side. She has no known family history of early hearing loss.

Fine-cut bone-window computed tomography (CT) scans revealed a 7 mm left-sided complex cystic mass with well-defined bony defect peripherally extending posteriorly to the margin of the mastoid and medially to the posterior genu of the facial nerve canal. There was no lateral semicircular canal (Figure 1(a)) and the cochlea appeared hypoplastic (Figure 1(b)). Magnetic resonance imaging (MRI) examination showed a cystic mass involving the left temporal bone, extending into the soft tissues below the mastoid tip. The cystic structure was non-enhancing on T1 with gadolinium and bright on T1 without gadolinium (Figure 1(c)) and T2 (Figure 1(d)), suggesting internal fluid with protein content. Fine needle aspiration near the sternocleidomastoid muscle yielded 30 mL of beige cloudy fluid with macrophages, calcifications, and a few epithelial cells.

The patient underwent tumor resection through a combined trans-mastoid and transcervical approach, extending the incision from the temporal bone down to the superior sternocleidomastoid muscle. On gross inspection, the tumor was red-brown in color with multicystic compartments filled with gelatinous fluid (Figure 2a). Distinct thick bony shell, possibly due to long-term bony remodeling, was noted peripherally (Figure 2b). The mass was intimately associated with the descending portion of the facial nerve and was removed in its entirety.

Histologically, the cystic structure was lined by simple columnar epithelium with goblet cells. Microscopic examination of the cystic wall revealed chronic inflammation with histiocytes, focal calcifications, and giant cell reaction. A trichrome stain highlighted fibrosis and muscular tissue, and desmin immunoreactive smooth muscles were seen in two distinct submucosal layers, resembling the muscularis mucosa and muscularis propria (Figure 3). The histology was consistent with a neurenteric cyst.

Our patient experienced immediate and significant improvement in pain post-surgery and was discharged home

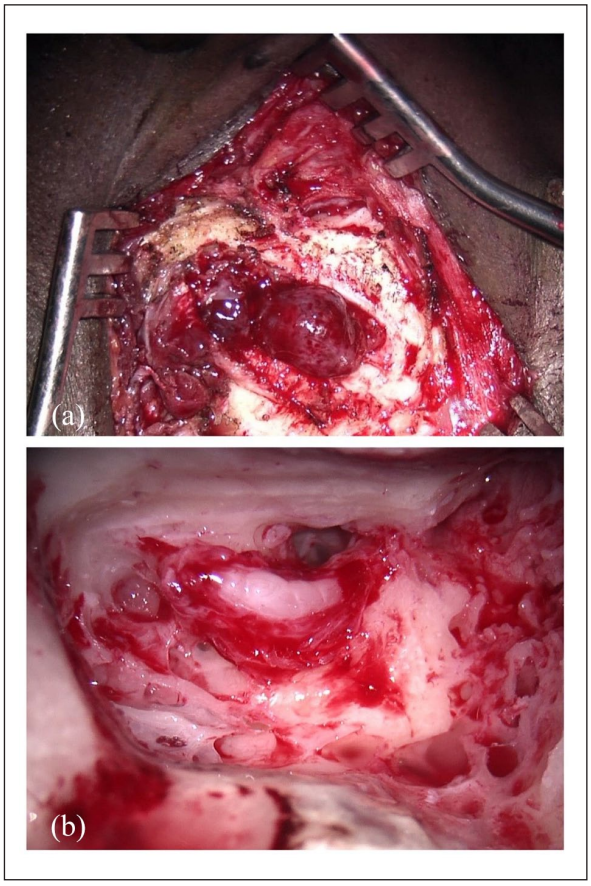

Figure 2. (a) Pre-excision view of the mastoid cystic mass. (b) Post-excision view of the remaining bony shell and underlying facial nerve after complete resection of the mastoid cystic mass.

on the same day of surgery. Follow-up in clinic, approximately 1 week later, showed continuation in her improved facial nerve function (House–Brackmann II) with minimal pain, but no changes from her baseline hearing capacity. No complications or signs of recurrence were found at the 2 months follow-up visit. The patient declined postoperative imaging studies and was lost to additional follow-up afterward.

Discussion

Neurenteric cysts are rare congenital lesions mostly found in intraspinal or intracranial locations.[8–11] The existence of extradural neurenteric cysts may be explained by communication between intra- and extradural spaces through dysraphic spinal columns.[12] It is proposed that neurenteric cyst embryological formation progresses through complex endodermal and ectodermal interactions, and abnormal interlayer adhesions may give rise to locations anywhere between the gut and skin, including the temporal bone.[13,14]

Our case is unique for the neurenteric cyst was not only extradural but it appeared to originate from the temporal bone with possible contributions to the patient's congenital hypoplastic cochlea and absent lateral semicircular canal,

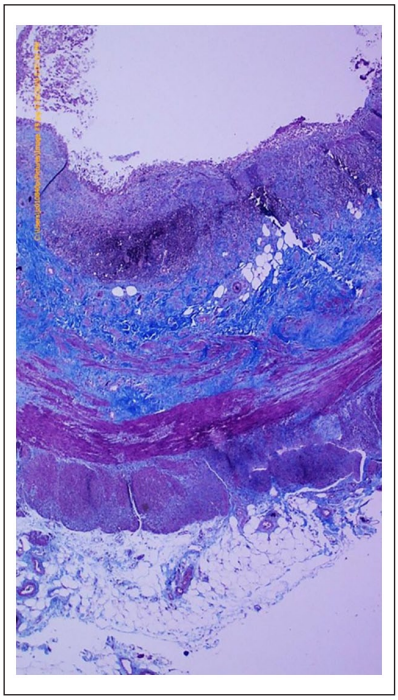

Figure 3. Low-powered (x40) view of simple columnar epithelium with goblet cells and two submucosal muscular layers (trichrome stain).

which is absent in literature. Eynon-Lewis et al.[15] reported on an extradural neurenteric cyst, causing unilateral hearing loss, similar to our patient, however, the neurenteric cyst in their case report was located at the cerebellopontine angle, a relatively common location for neurenteric cysts.[16] While the neurenteric cyst reported by Eynon-Lewis et al.[15] had extensive internal auditory canal and petrous bone destruction, no cases of mastoid and inner ear remodeling due to neurenteric cysts were previously reported. Albeit no conclusive connections can be drawn between the mastoid cyst and our patient's congenital hearing loss, the chronic cystic growth, as evidenced by its thick bony shell, most likely obliterated the inner ear space and induced her anomalies. Future reporting of similar cases is necessary to delineate possible embryological correlations.

CT is suitable for inspecting bony changes, while MRI is ideal for visualizing neural anatomy for suspected neurenteric cysts.[3] On MRI examinations, neurenteric cysts are commonly round, homogeneous, lobulated, isointense or hyperintense on T1-weighted, and hyperintense on T2-weighted images.[2,17] The intensity of MRI fluctuates depending on the concentration of protein and/or possible bleeding in the neurenteric cystic content.[17] Our patient had heterogeneous signaling on MRI, which is atypical for neurenteric cysts.[2] In addition, the observed thick bony capsule on CT is indicative of the neurenteric cyst's ability to induce long-term bony turnover. The cystic content can range from gelatinous, xanthochromic, to oily fluid with or without small crystals and calcifications.[1] As a result of its origin, neurenteric cysts

contain cuboidal to columnar GI or respiratory epithelium, with possible cilia and goblet cells.[1]

Plausible differential diagnoses for our patient's cystic mass in the mastoid includes epidermoid and dermoid cysts, and schwannoma. An epidermoid cyst could have similar characteristics on CT and MRI, with T1 shortening (hyperintense signal) secondary to proteinaceous material and/or prior bleeding, but the T1 shortening would have restricted diffusion, contrasting a neurenteric cyst. Dermoid cyst can have the same CT appearance, which was the original presumed diagnosis, but on MRI, the T1 shortening would be secondary to the fat content and should be suppressed with a fat-suppression sequence. Since there was no suppression of the T1 signal, this excludes this entity. For schwannoma with cystic components, the enhancement would be more pronounced with possible sparing of the cystic components, and the localization should follow the cranial nerve (CN) VII or VIII distributions. Most differential diagnoses are easily distinguished through histologic examinations.

The optimal treatment for neurenteric cysts is complete excision.[3] Although it was evident that our patient had an uncommon temporal bone entity based on presentation and imaging characteristics, forming differentials and surgical planning were challenging without biopsies. As a result, it was critical that both total resection and appropriate histologic specimens were obtained in the first surgery. Neurenteric cysts naturally fluctuate through cycles of rupturing and refilling, and recurrence risks increase with partial excisions, biopsy, and aspiration of cystic content.[1] Chavda et al.[18] demonstrated that eight patients with partial excision of neurenteric cysts had 37% recurrence rates 30 years post operations. While generally indolent in nature, neurenteric cysts may induce acute symptomatic presentations through mass effect, hemorrhage, and/or infarctions, which may explain our patient's abrupt presentation.[1] In rare instances, intracranial neurenteric cysts have shown capability of malignant transformation into adenocarcinoma, and long-term follow-up MRI is recommended for detection of both recurrence and transformation risks.[12]

Conclusion

We report on a temporal neurenteric cyst with marked peripheral mastoid bone remodeling, unique communication with posterior genu of the facial nerve, and hypoplasia of the cochlea and lateral semicircular canal. This case demonstrates that prompt total resection may be curative for patients with extensive extradural neurenteric cysts. It is our hope that this case report will contribute to raising awareness of neurenteric cysts and assist in navigating the differential diagnoses of temporal bone tumors, especially with the involvement of the mastoid and inner ear.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval

Ethical approval to report this case was obtained from the University of Tennessee Health Science Center (21-08230-NHSR).

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

IRB approval

21-08230-NHSR (University of Tennessee Institutional Review Board)

Permission

Patient written informed consent was obtained for this case report

ORCID iDs

Timothy Fan https://orcid.org/0000-0001-8542-7086

Ezer Benaim https://orcid.org/0000-0001-8190-0091

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