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Case-Based Review

# Primary Mastoid Cholesteatoma: A Case Report and Review of the Literature

**Sarah King[1], Mackenzie O'Connor[2], Arianna Winchester[2], Michael Bartellas[2], J. Thomas Roland Jr[2]**

[1]Department of Otolaryngology-Head and Neck Surgery. University of Kansas School of Medicine, Kansas City, Kansas, USA
[2]Department of Otolaryngology-Head and Neck Surgery, New York University Grossman School of Medicine, New York, New York, USA

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Congenital cholesteatomas are defined as a collection of epithelium in the absence of prior surgery or pathologic retraction. They are most commonly found in the middle ear and are thought to arise from a residual epithelial rest present since birth; however, a small number of cases present with disease isolated to the mastoid bone. One such case and a review of prior reports are presented. A 29-year-old male with a 2-year history of headaches and no otologic surgery was found to have a destructive mass centered in the right mastoid bone, invading the jugular foramen and posterior fossa without middle ear disease. He underwent a mastoidectomy with complete resection of the cholesteatoma. A literature review identified 21 prior cases of isolated mastoid congenital cholesteatoma. Congenital cholesteatoma should be considered in the differential diagnosis when patients present with postauricular pain or headaches. Mastoidectomy is considered the treatment of choice.

**KEYWORDS:** Cholesteatoma, headaches, mastoid, mastoidectomy, temporal bone

## INTRODUCTION

Cholesteatomas are slow-growing lesions arising from abnormal collections of keratinizing squamous epithelium and containing keratin debris.[1] They can be congenital or acquired, with congenital cholesteatomas accounting for only 1%-5% of reported cases.[1] Congenital cholesteatoma is a collection of epithelium and debris in the absence of prior surgery or pathologic retraction and can arise from various temporal bone sites, including the petrous apex, cerebellopontine angle, middle ear, mastoid process, and external auditory canal.[2,3] The most common site is the mesotympanum, and the most rare is the mastoid process.[2,4] Though the exact etiology is unknown, middle ear congenital cholesteatoma has been linked to the presence of congenital epithelial rests in the mesotympanum, and temporal bone disease to the migration of epithelial tissue via the Seessel epipharyngeal pouch into the petrous air cells.[5] Potsic et al[6] proposed a staging system for the disease based on location and extent of disease in reference to tympanic membrane retraction, which is often used to predict recidivism. The most advanced Potsic stage IV is assigned if mastoid disease is present but refers to disease extending from the middle ear. The aim of this study was to report a rare case of a congenital cholesteatoma of mastoid origin with no involvement of the middle ear and review previous reports in the literature.

## METHODS

Case information is reported below. Informed consent was obtained from the patient. A comprehensive literature review was conducted to gather relevant studies on the presentation, management, and outcomes of primary mastoid cholesteatoma. Databases searched included PubMed, Google Scholar, and Embase, focusing on articles published in English. Keywords such as "primary mastoid cholesteatoma," "congenital cholesteatoma," and "mastoidectomy" were used to identify pertinent studies. Articles were selected based on their relevance to isolated mastoid cholesteatomas, excluding cases involving middle ear pathology or acquired disease.

## CASE REPORT

A 29-year-old male presented to the institution following an incidental finding of a right mastoid mass. The patient's medical history was significant for recent trauma sustained during a physical altercation, which prompted temporal bone computed tomography (CT) approximately 2 months prior. The scan did not show any temporal bone trauma but did reveal a mastoid mass, and

**Corresponding author**: Mackenzie O'Connor, e-mail: Mackenzie.o'connor@Nyulangone.org

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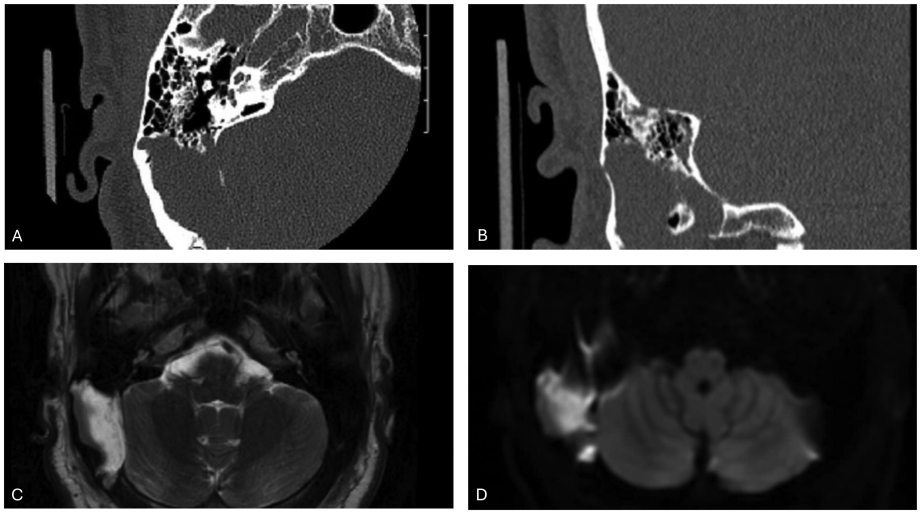


**Figure 1.** Preoperative axial (A) and coronal (B) non-contrast CT of the temporal bone, axial T2-weighted magnetic resonance imaging (C), and diffusion weighted imaging (D) demonstrating large right destructive lesion abutting the posterior fossa and eroding the sigmoid plate.

there was no history of otorrhea, hearing loss, trauma, or previous otologic surgery.

The patient reported a 1.5-year history of intermittent, throbbing headaches, primarily localized to the occipital region. He also described episodic dizziness, which was exacerbated by positional changes, such as standing or head turning, and was occasionally accompanied by vomiting. Notably, the patient was incarcerated, potentially limiting his access to consistent medical care and delaying the evaluation of these symptoms.

On physical examination, the patient appeared alert and oriented. Otoscopic examination revealed normal-appearing tympanic membranes bilaterally, with no visible signs of infection or perforation. All cranial nerves were intact. Audiometric testing indicated normal hearing bilaterally. No abnormal findings were noted in the neurological examination, including nystagmus or abnormal Romberg test.

Temporal bone CT and thin-cut magnetic resonance imaging showed a well-circumscribed mass measuring 6 × 4 × 4 cm, centered within the right posterior mastoid air cells with inner and outer table bony dehiscence (Figure 1). The mass extended anteroinferiorly, invading the right jugular foramen and right posterior fossa, abutting the lateral margin of the right cerebellum with a mild mass effect. There was no involvement of the middle ear cavity, external auditory canal, or petrous apex. Vascular imaging revealed narrowing of the right lateral transverse and sigmoid sinus without intraluminal obstruction.

The patient underwent surgical intervention via canal wall up mastoidectomy and removal of disease. Intraoperatively, a very large cholesteatoma involving the entire posterolateral mastoid was removed; however, the middle ear was free of disease. There was bony erosion and dehiscence over the exposed sigmoid sinus. The sigmoid sinus was compressed but intact. The dura was exposed and preserved (Figure 2). The mastoid cavity was packed with an abdominal fat graft to fill the space. Histopathological examination of the excised tissue confirmed primary cholesteatoma. The patient's postoperative course was uncomplicated, and postoperative audiogram confirmed no change in hearing.

## MAIN POINTS

- Congenital cholesteatoma originating from the mastoid without involvement of the middle ear is rare but possible.
- Isolated mastoid cholesteatoma may present as headache, vertigo, pain, or without symptoms and should be considered in the differential for temporal bone masses.

## DISCUSSION

Congenital cholesteatomas (CC) are rare epidermoid cysts thought to originate from residual epidermoid tissue that persists during embryonic development or migration of epithelial tissue directly through a residual embryonal pouch.[1,5,7] While CCs typically originate in the middle ear, they have also been reported to arise in other parts of the temporal bone such as the petrous apex, the external auditory canal, and in extremely rare cases, the mastoid process. Isolated mastoid cortex cholesteatomas are even less common, with only 21 documented cases in the literature.[4] See Table 1 for all reviewed literature.

Derlecki and Clemis first described primary mastoid cholesteatoma in 1965, defining it as a cholesteatoma with the following criteria: 1) exhibit all the characteristics of a CC (normal tympanic membrane, no history of infection or otorrhea, and no prior otologic surgery) and 2) demonstrate no involvement of the middle ear, attic, or aditus.[8,12] This case met these criteria, with the cholesteatoma confined to the mastoid with erosion due to expansion and without middle ear involvement.

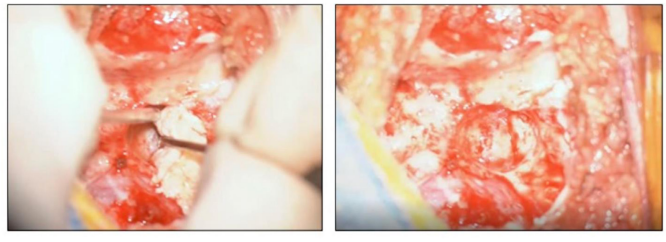

**Figure 2.** Intraoperative photos showing the main specimen before (left) and after (right) removal off of pre-sigmoid dura.

**Table 1.** Literature review for cases of primary mastoid cholesteatoma.

| | | | | Structures Involved | | | | |
|---|---|---|---|---|---|---|---|---|
| | # Patients | Age | Symptoms | SS | Posterior EAC | FN | PF | Management |
| Derlacki et al (1965)[8] | 1 | 24 | Postauricular pain, swelling | n/a | Eroded | n/a | n/a | Atticotomy, mastoidectomy |
| Luntz et al (1997)[4] | 1 | 54 | Neck pain, imbalance | Dehisced | Intact | n/a | n/a | CWU mastoidectomy |
| Cüreoglu et al (2000)[9] | 1 | 70 | Neck pain, postauricular swelling | n/a | Eroded | n/a | n/a | Modified radical mastoidectomy |
| Mevio et al (2002)[2] | 1 | 36 | Positional vertigo | Dehisced, compressed | n/a | n/a | Exposed | Modified radical mastoidectomy (retrolabyrinthine) |
| Warren et al (2007)[10] | 3 | 30 | None | Dehisced | n/a | Exposed | Exposed | CWU mastoidectomy |
| | | 28 | Otalgia | Dehisced | n/a | n/a | n/a | Unavailable |
| | | 13 | Neck mass | n/a | Eroded | Exposed | Intact | Modified radical mastoidectomy, neck dissection, parotidectomy |
| Hidaka et al (2010)[11] | 1 | 65 | Postauricular pain, swelling | Eroded | Intact | n/a | Exposed | CWU mastoidectomy, adjacent abscess drainage |
| Gianuzzi et al (2011)[12] | 3 | 71 | Postauricular/neck pain | Dehisced | Intact | Exposed | Intact | CWU mastoidectomy, fat graft |
| | | 77 | Dizziness | Dehisced | Intact | Exposed | Exposed | CWU mastoidectomy, fat graft |
| | | 60 | None | Dehisced | Intact | Exposed | Exposed | CWU mastoidectomy, fat graft |
| Cvorovic et al (2011)[13] | 1 | 29 | Postauricular pain, tinnitus, mixed hearing loss | Dehisced | Intact | Exposed | Exposed | Modified radical mastoidectomy |
| Nagato et al (2012)[14] | 1 | 10 | EAC stenosis | Intact | Eroded | n/a | Intact | CWU mastoidectomy |
| Koltsidopoulos et al (2012)[15] | 1 | 52 | Otorrhea | Dehisced, compressed | Eroded | Exposed | n/a | CWU mastoidectomy |
| Hong et al (2014)[16] | 1 | 59 | Otorrhea | Intact | Eroded | n/a | n/a | CWU mastoidectomy |
| Sepehri et al (2018)[17] | 1 | 87 | None | Intact | Intact | Intact | Intact | Surveillance |
| Annalisa et al (2021)[18] | 1 | 58 | Postauricular/neck pain | Dehisced | n/a | Exposed | Exposed | CWU mastoidectomy |
| Richards et al (2022)[19] | 3 | 34 | Headaches, dizziness | Intact | Intact | Intact | Exposed | CWU mastoidectomy |
| | | 73 | Postauricular pain/swelling | Dehisced | Intact | Exposed | Exposed | CWU mastoidectomy, fat graft |
| | | 48 | Headache | n/a | n/a | n/a | n/a | Surveillance |
| Mckenna et al (2023)[20] | 1 | 14 | Otorrhea | n/a | Eroded | Exposed | n/a | CWU mastoidectomy |

SS: sigmoid sinus, FN: facial nerve, PF: posterior fossa. Canal wall up mastoidectomy External auditory canal.

The etiology of congenital cholesteatomas remains a topic of debate, with the most widely accepted theories suggesting they stem from embryonic epidermoid remnants in the middle ear.[21] Another "Implantation Theory" proposes that during the fontanel closure process, squamous epithelium trapped within the tympanomastoid suture line could generate mastoid CC.[10,16,21] Other theories, such as those suggesting acquired causes (e.g., infection or trauma), do not fit as well in this case, particularly given the patient's lack of a relevant medical otologic history.

Primary mastoid CCs are often asymptomatic in their early stages, which often leads to delayed diagnosis. In the 21 reported cases reviewed here, it was found that the average age of presentation for mastoid CCs is 47 years, with a range of 13-87 years. This patient's case underscores the silent progression typical of these lesions, as his symptoms (dizziness, occipital headaches, and fatigue) only emerged after the cholesteatoma reached a considerable size, causing local erosion and mass effect on major blood vessels and the posterior fossa. Prior reports indicate that symptoms such as pain, dizziness, and swelling typically arise when the lesion has expanded enough to affect nearby structures, such as the periosteum, cranial nerves, or cerebellum.[19] The patient's dizziness, likely due to compression of the adjacent cerebellum, aligns with reported cases where symptoms are secondary to structural impact.[7]

Management of primary mastoid cholesteatomas is challenging due to their rarity and potential for significant bony erosion. Surgical removal is essential to prevent complications like intracranial extension, hearing loss, and cranial nerve damage.[1,7,12] For less extensive cholesteatomas confined to the mastoid, cortical mastoidectomy,

which preserves the canal wall, is often preferred to maintain ear structure and optimize hearing outcomes.[16] However, when disease extent or anatomical challenges warrant greater visibility, canal wall down mastoidectomy is employed.[1] Canal wall down offers superior exposure and a lower recurrence rate by creating a common cavity, though it can lead to a larger residual space that may require ongoing maintenance and could impact hearing.[1] Current management approaches continue to emphasize individualized treatment plans, weighing factors such as lesion size, patient anatomy, and the likelihood of recurrence.

## CONCLUSIONS

Mastoid cholesteatomas without involvement of the middle ear are rare, and only 21 cases are reported in the literature. This case adds to this body of work and provides a review of prior cases for context. Congenital cholesteatoma should be considered in the differential diagnosis when patients present with otherwise unexplained pain or swelling localized to the mastoid process or dizziness. Mastoidectomy is considered the treatment of choice.

**Data Availability Statement:** The data that support the findings of this study are available on request from the corresponding author.

**Informed Consent:** Verbal informed consent was obtained from the patient who agreed to take part in the study.

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**Declaration of Interests:** The authors have no conflicts of interest to declare.

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