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Case Report

A pearl in the ear: Intracranial complications of pediatric cholesteatomas

Krystyne Basa [a, b], Jessica R. Levi [a, b], Erin Field [c], Robert C. O'Reilly [c, *]

[a] Boston Medical Center, 1 Boston Medical Pl, Boston, MA 02118, United States
[b] Boston University School of Medicine, 72 E. Concord St, Boston, MA 02118, United States
[c] Nemours/Alfred I. duPont Hospital for Children, 1600 Rockland Rd, Wilmington, DE 19803, United States

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ABSTRACT

A nine-year-old male had a cholesteatoma of the mastoid and middle ear found incidentally after myringotomy tube placement. Associated asymptomatic sigmoid plate dehiscence with sinus invasion or thrombosis and ossicular chain destruction complicated his case. He had canal wall down tympanomastoidectomy and was followed for 4.5 years. Disease recurrence necessitated revision. Our case highlights an unusual clinical presentation, possible complications, and the aggressive quality of a benign lesion common in the pediatric population. To our knowledge, this is the first report of an asymptomatic lateral sinus obstruction secondary to an invasive cholesteatoma in this population.

1. Introduction

Cholesteatomas are keratinized lesions found within the external ear canal, middle ear, mastoid and petrous bone. The annual worldwide incidence of cholesteatomas in the pediatric population is three per 100,000 [1]. There are two types: congenital and acquired [1–4]. Congenital cholesteatomas are pearly white, usually asymptomatic, masses found in the anterior superior quadrant of the tympanic membrane [2]. Acquired cholesteatomas present with tympanic membrane abnormalities, typically a retraction pocket, associated otorrhea, and recurrent otitis media [2].

Regardless of origin, cholesteatomas are typically slow growing and can be destructive. Pediatric cholesteatomas are considered more aggressive than those found in adults [3,4]. Presenting symptoms can include conductive hearing loss, facial nerve weakness, and imbalance [5].

Here, we present a case of an extensive right-sided cholesteatoma extending into the middle ear with associated ossicular chain destruction, sigmoid plate dehiscence, and sigmoid sinus extension. To our knowledge, this is the first pediatric case of an asymptomatic sigmoid sinus obstruction secondary to an invasive cholesteatoma.

2. Case report

A nine-year-old male with a history of recurrent bilateral otitis media presented to our clinic for evaluation of a known right-sided cholesteatoma. At an outside hospital, the child underwent bilateral myringotomy and tympanostomy tube placement. At the postoperative visit, the child had an abnormal right tympanic membrane. Computed tomography showed the middle ear was completely opacified with soft tissue, concerning for a cholesteatoma. There was erosion of the mastoid air cells as well as much of the bone over the sigmoid. He subsequently was referred to our institution.

On presentation to our institution, his symptoms included intermittent otorrhea from his right tube. He denied vertigo and facial weakness. On audiologic evaluation, he had a right-sided conductive hearing loss. Physical exam was notable for a smoothly marginated area of debris adjacent to the right tympanic membrane, which was completely opaque. A tympanostomy tube was visible and in place; however, there were no visible middle-ear landmarks. We obtained an MRI (Figs. 1 and 2) to determine the extent of the disease and to rule-out an erosive process beginning

* Corresponding author. Nemours/Alfred I. duPont Hospital for Children, PO Box 269, Wilmington, DE 19899, United States.
E-mail address: roreilly@nemours.org (R.C. O'Reilly).

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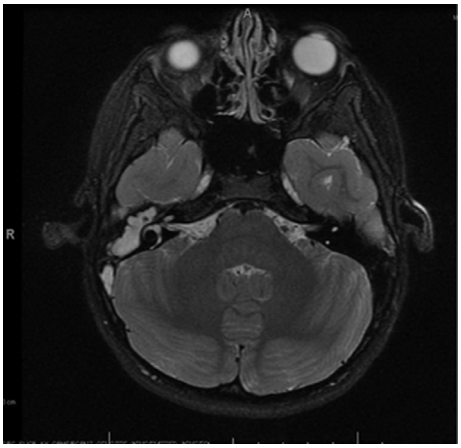

Fig. 1. T2-weighted axial non-contrast MRI with hyperintense right-sided mass.

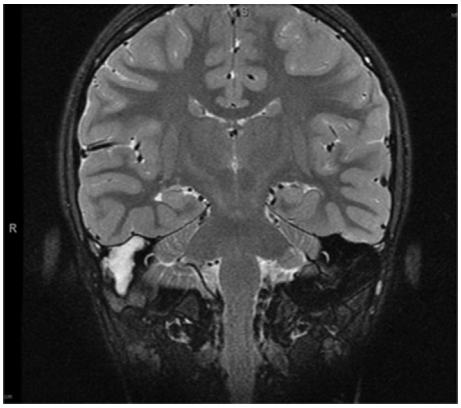

Fig. 2. T2-weighted coronal MRI with contrast showing right-sided hyperintense mass.

from the posterior fossa since there was evidence of sigmoid plate erosion on CT.

Initial MRI/MRV showed a soft-tissue mass in right middle ear and within the mastoid air cells. This mass was T2 hyperintense, and it measured approximately 2.4 × 0.8 × 2.5 cm. There was also noted extension into the medial aspect of the right external auditory canal and the anterosuperior inner ear structures. There were similar soft-tissue findings in the distal right transverse and proximal sigmoid sinuses and a notable filling defect in this region on MRV, but there was no complete occlusion of the sinuses. This most likely reflected cholesteatoma extension into the venous sinuses. Interestingly, there was no clear connection between the middle ear mass and the venous sinus mass present on imaging.

He subsequently was referred to neurosurgery due to concerns about the extension of the mass into the dural sinuses. Since our patient's presentation was unconventional, a biopsy was performed first. Pathology came back as a right-sided cholesteatoma and a tympanomastoidectomy was scheduled for a later date.

2.1. Surgical technique

The patient's first procedure was a right middle ear exploration to obtain a tissue biopsy with removal of the incus. The patient had a large vascular granuloma at the 6-o'clock position, which was removed. The tympanic membrane was completely featureless and thickened. The previously inserted tympanostomy tube was seen in the anterior inferior quadrant. A tympanomeatal flap was elevated, and the middle ear space was entered inferiorly. In the middle ear space, there was balloon mucosa, and a cholesteatoma sac filled the middle ear cleft. Biopsies were taken of the middle ear mucosa over the promontory. The chorda tympani was wrapped entirely in the cholesteatoma sac. There was erosion of the lenticular process of the incus and head of the malleus and lack of the superstructure of the stapes. The chorda tympani and body of the incus were subsequently removed during this procedure. The chorda tympani was sent for specimen with the biopsied mucosa.

The postoperative pathology report was consistent with a cholesteatoma. Therefore, tympanomastoidectomy with neurosurgery available was planned for a later date.

For the second procedure, removal of the cholesteatoma, a canal wall down (CWD) tympanomastoidectomy approach was chosen. The tegmen and the dura posterior to the sigmoid sinus were carefully skeletonized, and the mastoid air cells were completely dissected to expose the cholesteatoma sac. The sac was entered and the matrix was carefully removed. Matrix found in the mastoid was carefully removed as well, along with the mastoid mucosa. The canal wall was then taken down, and all visible cholesteatoma was removed from the middle ear cleft.

Intraoperatively, there was destruction of the mastoid air cells with extension into the subarcuate space and a largely dehiscent sigmoid, measuring up to 2.5 cm in length. The sigmoid was rubbery to palpation, thought to be thrombosis within the dural sinus; however, there was no obvious direct extension of the cholesteatoma into the sigmoid itself. In addition, the middle ear space and mastoid were noted to be acutely infected. At this point, the neurosurgeon came in to visualize the sigmoid. Both he and the senior otolaryngologist mutually agreed that resection of the sigmoid would not be prudent, as the patient was asymptomatic. In addition, complete removal of the diseased area would have required a large resection.

A fascial graft was taken, and a meatoplasty was performed. The superior aspect of the remaining tympanic membrane was resected, and the fascial graft was placed to create a “cavum minor.” Gelfoam (Pfizer, New York, NY) squares were used as support, and the remainder of the fascial graft was used to line the mastoid cavity. Surgiflo (Ethicon, Somerville, NJ) was used to fill the mastoid cavity in the middle ear space. The meatoplasty was then sutured opened. Postauricular wound closure was achieved in three layers.

2.2. Results

The patient was followed by Otolaryngology and Neurosurgery for 4.5 years after his initial CWD tympanomastoidectomy. A planned MRI was obtained 3.5 months after surgery, which showed stable soft tissue/filling defect of the sigmoid. Nine months after the procedure, he began to complain of short-term memory problems, intermittent headaches, visual disturbances, and imbalance. Neurosurgery continued to follow him for these complaints and diagnosed him with migraines. However, given the history of sigmoid sinus thrombosis, an MRI/MRV (Figs. 3 and 4) was obtained nine months after his CWD tympanomastoidectomy. Clinically, he had developed some narrowing of the meatoplasty and adhesions over the mastoid bowl. His images showed growth of a recurrent mastoid cholesteatoma but stable sigmoid findings with no interval growth intracranially. Because of evidence of disease recurrence, he underwent a revision of the right-sided meatoplasty and cleaning of the mastoid bowl 17 months after his initial tympanomastoidectomy. During the time of the revision surgery, cholesteatoma was found filling the mastoid. There was also notable new bone growth over the sigmoid sinus. All visible cholesteatoma again was removed. Serial MRI/MRVs were continued to monitor for