Sudden Sensorineural Hearing Loss and Pneumolabyrinth: An Unusual Manifestation of Isolated Mastoid Cholesteatoma

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Abstract

Objectives: To describe the case of a patient with an unusual growth pattern of cholesteatoma leading to a rare complication and atypical presentation.

Methods: Case report from a tertiary Navy hospital facility. We present the case of a patient who suffered permanent profound sensorineural hearing loss and temporal vertigo as a result of an unusual growth pattern of cholesteatoma. Without manifestations of chronic ear disease, an isolated mastoid cholesteatoma eroded the posterior semicircular canal and led to a pneumolabyrinth.

Results: From the evaluation and workup for sudden sensorineural hearing loss, radiographic imaging demonstrated an erosive mass consistent with cholesteatoma. The lesion was suspected based on imaging characteristics and was confirmed on histology after resection of the whole mastoid segment.

Conclusion: In the absence of findings consistent with chronic ear disease, sudden sensorineural hearing loss is a noted presentation for cholesteatoma. The abnormal location and growth pattern for this particular cholesteatoma is presently uncharacterized and should be considered in the evaluation of patients presenting with sudden sensorineural hearing loss.

Case Presentation

A thirty year old male presented to the otolaryngology clinic for evaluation of severe right sided hearing loss, aural fullness, and tinnitus. Approximately 6 weeks earlier, he experienced symptoms attributed to a mild upper respiratory infection and he specifically recalled onset of dizziness, nausea, and aural symptoms with excessive nose blowing. At time of evaluation, vestibular symptoms had resolved yet right sided hearing loss and aural pressure persisted.

Medical and surgical history were non-contributory with the exception of previous tubal surgery at age 18 upon entering military service. As an adult, he specifically denied chronic ear infections, signs/symptoms of eustachian tube dysfunction, or head trauma. Routine occupational and recreational activities were non-suspicious.

Complete neurologic exam was normal with the exception of a tuning fork exam demonstrating a right sided sensorineural hearing loss. Bilateral microsuction showed a safe retraction of an otherwise normal right tympanic membrane that readily reduced with Valsalva. The middle ear was clear of any effusion or mass.

Audiology exam demonstrated right profound (anacusis) hearing loss, absent otoacoustic emissions, and normal tympanometry (Figure 1).

Initial Management

- Based on initial presentation, the differential diagnosis focused on inner ear pathology associated with possible idiopathic sudden sensorineural hearing loss (SSHL), perilymphatic fistula, vestibular neuritis, and/or neoplastic processes.
- Although late in presentation, initial treatment followed AAO-HNS guidelines for treatment of SSHL, with a course of intraoral steroids and radiographic evaluation.
- Initial screening non-contrasted MRI did not identify a retrocochlear lesion. However, a bright T2 signal was seen extending along the posterior fossa between the dura and posterior semicircular canal into a well pneumatized mastoid cavity.
- A subsequent temporal bone CT established a soft tissue mass centered within the mastoid air cells with dehiscence into multiple labyrinthine structures (Figure 2). A pneumolabyrinth involving the vestibule and cochlear space was identified (Figure 3). Erosion was also seen along the mastoid segment of the facial nerve with involvement of the jugular vein. The remainder of the temporal bone was well ventilated and without disease (Figure 4).
- To further define the mass, a contrasted MRI was obtained that demonstrated characteristics of cholesteatoma.
  - Hypointense on T1, hyperintense on T2
  - Nonenhancing with gadolinium
  - Restriction on diffusion weighted imaging (high signal intensity)
  - Low signal intensity on ADC mapping

Definitive Management

- Surgical management focused on identification and removal of probable cholesteatoma or other disease.
- During a tympanomastoidectomy, keratin debris was readily identified within mastoid air cells (Figure 5).
- Debridement further defined stratified squamous epithelium (malacia) eroding the posterior fossa bone and tracking along the dura. Erosion included the posterior semicircular canal, the inferior aspect of the sigmoid sinus and the jugular bulb. The mastoid segment of the facial nerve was preserved with mild adhesions but no compression (Figure 6).
- Cholesteatoma tracked along central air cell tract towards the aditus ad antrum without adherence to the incus or erosion of horizontal semicircular canal. To ensure absence of anterior epidural epimeatus the incus and head of malleus were removed. No disease extended into the epitympanum or middle ear.
- A labyrinthectomy was completed to ensure complete extirpation of disease.
- Surgical pathology later confirmed the surgical diagnosis of cholesteatoma.

Background

- Cholesteatoma is commonly defined as an expansile mass that often develops within the middle ear or mastoid, lined by keratinized stratified squamous epithelium that contains desquamated debris.
- It is classified as congenital (rare) or acquired (common).
- Acquired cholesteatoma are sub-classified into primary or secondary based on pathogenesis:
  - Primary: related to imagination theory of retraction pockets in the tympanic membrane
  - Secondary: related to inflammation theory of retraction pockets through tympanic membrane perforation from either acute or chronic otitis media, ear trauma, or it may be iatrogenic.
- Numerous theories have been put forth to attempt to explain the mechanism by which primary acquired cholesteatoma form. More recently Jäckle has offered a novel, well thought theory based on the interaction of opposing mobile mucosal surfaces of the middle ear.
- The epitympanum and mastoid cavity were typical locations for evolving tympanic membrane retraction packs.
- Patients with early cholesteatoma are either asymptomatic or develop mild symptoms of chronic otitis media (e.g., intermittent ototears, otalgia, aural pressure).
- Typical presentation includes symptoms of malodorous otitis externa, chronic recurring infections, and in some instances, aural polyps in the external auditory canal. Variable degrees of conductive hearing loss may also be present.
- Advanced development of cholesteatoma may erode the otic capsule resulting in vertigo and sensorineural hearing loss. Extension of disease may also lead to facial nerve paralysis, and intracranial infections.
- Diagnosis is typically confirmed on clinical exam using otoscopy or through surgical exploration.
- Audiologic testing typically demonstrates varying degrees of conductive hearing loss, mixed hearing loss, and very rarely sensorineural hearing loss. Tympanometry may suggest eustachian tube dysfunction.
- Although not always indicated, imaging is useful to define the extent of the disease and for pre-surgical planning.
- Non-contrasted CT will typically show a hypodense lesion within the middle ear or mastoid that may demonstrate bony resorption.
- Common characteristics on MRI include dark on T1, bright on T2, no enhancement with gadolinium, restricted on DWI (High intensity), low signal intensity on ADC mapping.
- Management goals: 1) eradicate all cholesteatoma to create safe, dry, disease free air 2) optimize hearing while minimizing the need for long term otologic care.

Discussion

- The initial presentation of our patient did not suggest cholesteatoma as the etiology for his symptoms.
- Rather than presenting with typical manifestations of early cholesteatoma, his initial presentation was a result of a late complication – labyrinthine fistula.
- With a normal middle ear exam and sudden sensorineural hearing loss, it was treated following the guidelines for management of SSNHL.
- The location of the disease was unusual for isolated common erosive lesions within the temporal bone (i.e., cholesteatoma).
- The possibility of other lesions such as an endolymphatic sac tumor or intracranial epidermoid lesion was further evaluated with CT and contrasted MRI.
- Despite the lack of clinical consistency with cholesteatoma, imaging characteristics favored this as the diagnosis.
- The unusual imaging finding of pneumolabyrinth is rarely associated with cholesteatoma and more consistent with temporal bone trauma.
- There are few management recommendations for the finding of pneumolabyrinth and almost none for pneumolabyrinth secondary to cholesteatoma.
- Hidaka et al. noted some return of hearing in patients with traumatic pneumolabyrinth isolated to the vestibule but no hearing recovery in patients with pneumolabyrinth found within the cochlea.
- With no expectation of return to functional hearing, a canal wall up mastoidectomy with labyrinthotomy was necessary to clear disease. This approach, combined with a second look procedure, was deemed a better option than exterminating the disease via a canal wall down mastoidectomy.

Conclusion

- Our patient demonstrated an atypical initial presentation for cholesteatoma as evidenced by profound SSHL and vertigo in the absence of findings consistent with eustachian tube dysfunction or chronic otitis media.
- This cholesteatoma presented in an unusual and unique location. The disease epicenter was located along the posterior fossa and extended into a well ventilated mastoid cavity. Also, the labyrinthine fistula affected the posterior semicircular canal rather than the horizontal semicircular canal.
- Management of pneumolabyrinth from cholesteatoma invasion departs from typical traumatic pneumolabyrinth. The primary focus shifts from hearing preservation to eradication of disease.

References