

**Case Report**

Open Access, Volume 2

**Rapidly progressive labyrinthitis ossificans in an immunocompromised pediatric patient**Sean Holmes<sup>1\*</sup>; Katherine Babin<sup>2</sup>; Avery Bryan<sup>2</sup>; Gauri Mankekar<sup>1</sup><sup>1</sup>LSU Shreveport Dept of Otolaryngology/Head and Neck Surgery, LSU Health Shreveport, 1501 Kings Highway, Shreveport, LA, 71103, USA.<sup>2</sup>School of Medicine, LSU Health Shreveport, 1501 Kings Highway, Shreveport, LA, 71103, USA.**\*Corresponding Authors: Sean Holmes**

Otolaryngology Resident, Department of Otolaryngology/HNS, LSU Health Shreveport, 1501 Kings Highway, Shreveport, LA 71103.

Email: sholm6@lsuhsc.edu

Received: Apr 22, 2021

Accepted: May 12, 2021

Published: May 17, 2021

Archived: www.jcimcr.org

Copyright: © Holmes S (2021).

**Abstract**

In this report, we present a case of rapid otic capsule obliteration within an exceedingly short timeframe in the setting of Chronic Suppurative Otitis Media (CSOM) in an immunocompromised pediatric patient with Down Syndrome. Following maximal therapy for a right sided cholesteatoma, the patient developed a multi-drug resistant infection that cause CSOM, which within 6 weeks progressed to complete obliteration of the right cochlea and otic capsule. The possibility of congenital temporal bone microscopic dehiscence allowing infection propagation cannot be excluded. Nonetheless, this case highlights the importance of appreciating how quickly chronic middle ear disease can progress to involve the labyrinth and cause intracranial complications, even with adequate concurrent medical therapy in the form of antibiotics and surgical therapy. A greater awareness as physicians should be made on management of refractory chronic middle ear disease to better treat their potential complications, which is made apparent in this case report.

**Keywords:** Otic capsule; Chronic suppurative otitis media; Otic capsule; Tympanomastoidectomy; Multi-drug resistance.**Abbreviations:** CSOM: Chronic Suppurative Otitis Media; MDR: Multi-Drug Resistant; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; ID: Infectious Disease.**Introduction/background**

The otic capsule, described as the bony labyrinth that surrounds the membranous labyrinth of the inner ear, is composed of the cochlea, vestibule, and semi-circular canals. Partial capsular erosion can occur secondary to a variety of causes, including Chronic Suppurative Otitis Media (CSOM), meningitis, aberrant arterial supply, schwannomas, cochlear device failures, and Langerhans cell histiocytosis to name a few [1-6]. This rare case presentation of rapidly progressive capsular obliteration highlights the importance of aggressive treatment of chronic middle ear disease in children.

Chronic Suppurative Otitis Media (CSOM), once common, has become a rarity in the medical world today. The development of antibiotics along with culture driven therapy and surgical intervention has drastically reduced the development of complications of CSOM, dropping the intracranial complication rate from 2.3-4% to 0.15-0.04%. These complications include both intracranial and extracranial pathology and can include the following conditions: mastoiditis, facial nerve palsy, extratemporal abscesses, lateral sinus thrombosis, brain abscesses, cerebellar abscesses, labyrinthitis, labyrinthine fistulase, meningitis, extradural abscesses, cochlear erosion, subdural empyemas, petrositis, and ossicular erosion [7]. Although medical and sur-

gical therapies can mitigate these now rare complications, we must remain aware of the potential for these to arise, and be up to date on the knowledge of how to treat these conditions.

Suppurative labyrinthitis, a bacterial infection of the inner ear is relatively uncommon today. There is a radiological classification illustrating the four stages of suppurative labyrinthitis: 1) serous, 2) purulent, 3) fibrous and 4) osseous. The serous stage involves production of Ig rich exudate in the perilymph, which then progresses to the purulent stage of bacterial and leukocyte invasion of the perilymphatic scala-end organ necrosis. The serous and purulent stages together are considered acute labyrinthitis. The fibrous and osseous stages are together known as chronic labyrinthitis. Clinical features of acute suppurative labyrinthitis include severe vertigo with nausea, vomiting, and hearing loss. The fibrous stage is characterized by fibroblast proliferation with granulation tissue in the perilymph. This leads to the osseous stage, which describes new bone deposition in the involved labyrinth [8].

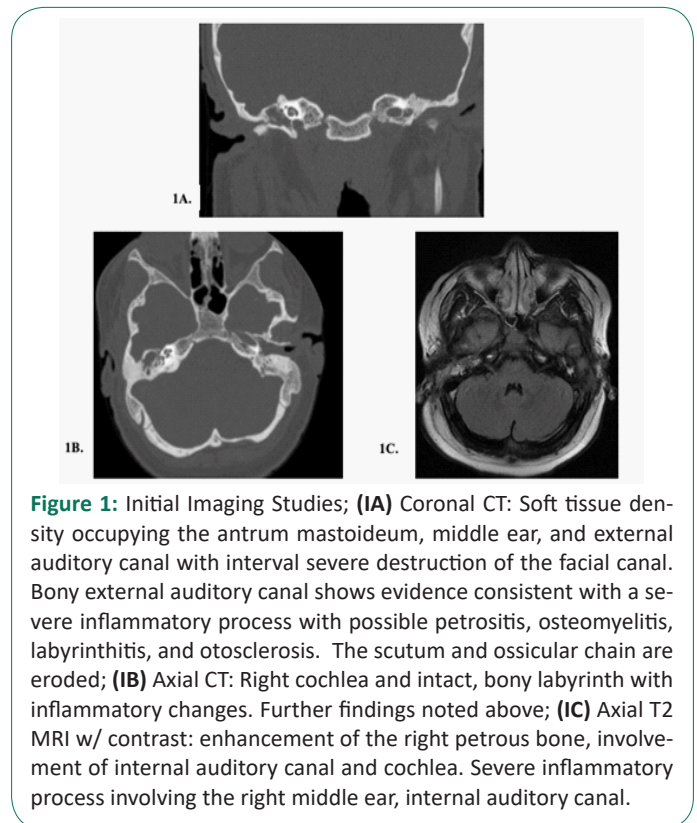
In the case of our patient presented below, he originally presented with a right sided cholesteatoma with initiation of maximal therapy. Despite medical and surgical intervention, the patient developed CSOM with a multi-drug resistant bacteria that eventually obliterated the right cochlea and otic capsule in a matter of 6 weeks. This case adequately demonstrates the importance of awareness of refractory chronic middle ear disease and its complications as physicians.

### Case presentation

A 14-year-old male with history of Down's Syndrome, Acute Myeloid Leukemia in remission, several previous ear tube placements, and bilateral mixed hearing loss presented to our facility for evaluation of chronic otitis media referred by a local otolaryngologist. Upon presentation, his parents noted bilateral otorrhea worse on the right as compared to the left, right-sided otalgia, and right-sided facial weakness. Physical examination revealed purulent otorrhea bilaterally from patent and functional ear tubes, along with a right sided grade IV House-Brackmann facial weakness. His initially presenting left sided otorrhea resolved after initial topical therapy and no further intervention was required for the left ear at the time. The left ear tube remains in place and functional at this time.

Empiric broad-spectrum IV antibiotic therapy was initiated for treatment of his right ear symptoms and physical examination, and an initial diagnostic work-up with CT and MRI was obtained which are described below.

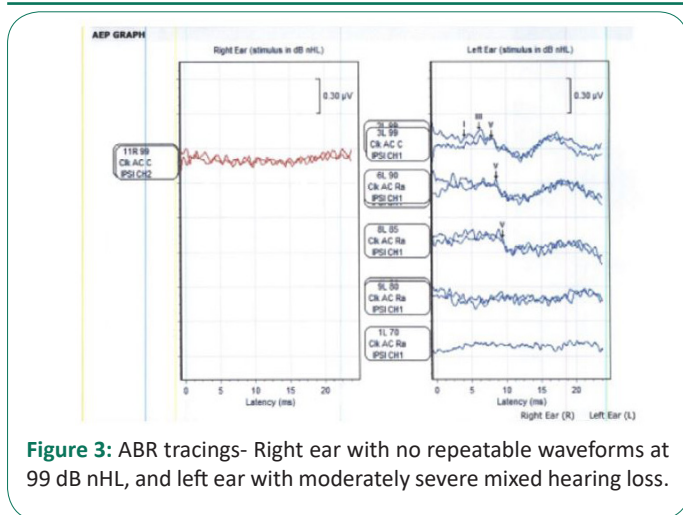
Computer Tomography (CT) showed a right-sided soft tissue density occupying the mastoid antrum, middle ear, and external auditory canal with severe destruction of facial canal and otic capsule with erosion of the cochlea and vestibule and sclerosis of the semicircular canals along with evidence of petrositis and erosion of the ossicular chain with the tegmen tympani intact (Figure 1A,B). Magnetic Resonance Imaging (MRI) revealed remarkable enhancement of the right petrous bone with involvement of the internal auditory canal including the right cochlea, as well as extension to the middle ear and external auditory canal. Erosion of the ossicular chain was observed (Figure 1C).



Following imaging, the patient underwent right canal wall-down tympanomastoidectomy with facial nerve decompression. Intraoperatively, the cholesteatoma matrix was noted to be within the epitympanum, extending into the antrum, covering the facial nerve, and without identification of the ossicles. The cholesteatoma matrix was resected, and the facial nerve was stimulated at end of case with adequate response. The patient initially did well post-operatively; however, weeks after surgery, he developed recurrent right-sided otorrhea for which further interventions were required.

The patient returned to clinic for subsequent follow up visits in 2-3 week intervals. The patient's mom reported persistent right-sided thick otorrhea, although patient continued to be afebrile. On physical examination, mucoid debris was noted in the mastoid cavity with no visualization of the tympanic membrane at all three visits. At this time, an exam under anesthesia was scheduled for further evaluation. This subsequently revealed right mastoid cavity with thickened yellow secretions and extensive granulation tissue to anterior and posterior aspects of medial mastoid cavity which was removed up to the medial aspect of the mastoid cavity as to avoid any inadvertent injuries to inner ear structures. Intra-operative cultures were obtained which revealed subsequent growth of Multi-Drug Resistant (MDR) *Escherichia Coli* (E. Coli). He completed 21 days of culture sensitivity directed IV antibiotics with cefuroxime, as per recommendations from the Infectious Disease (ID) department.

Auditory Brainstem Response (ABR) was performed after right-sided canal wall down tympanomastoidectomy, during which the patient was roughly half-way through completion of his course of IV antibiotics. Results revealed no repeatable waveforms at 99dB nHL for right ear, and moderately severe mixed hearing loss for left ear (Figure 2).



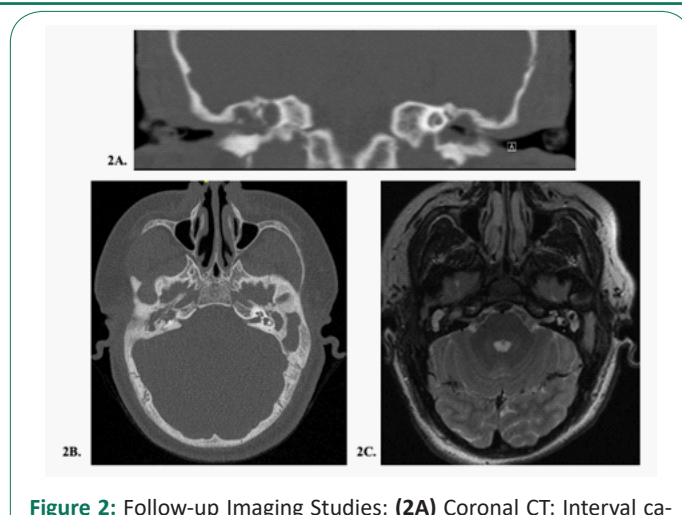
The patient clinically improved initially after culture-directed antibiotic therapy following surgical intervention. However, when he returned to clinic for a second follow-up visit despite 51 days of IV Vancomycin and Ceftriaxone, he had right sided persistent otorrhea. Cultures were obtained at this time and revealed MDR *Corynebacterium* sensitive to meropenem. The patient received 31 days of PO meropenem. At his third follow up visit, the patient had complete resolution of otorrhea and no clinical signs of otitis media.

Follow up imaging demonstrated complete obliteration of the right otic capsule. CT imaging of the right temporal bone revealed erosion of bone around jugular foramina and widening of the geniculate ganglion along with the cochlear segment of the facial canal suggesting intracranial extension consistent with complete otic capsular obliteration (Figure 3A,B). MRI revealed persistent inflammatory changes of the right temporal bone with destruction of the otic capsule including osteomyelitis and labyrinthitis with intracranial extension through the internal auditory canal with meningitis. It additionally shows involvement of the 7th and 8th cranial nerves (Figure 3C).

We continued to follow our patient monthly to monitor for recurrence of CSOM. Our patient showed no clinical symptoms of recurrence after completion of the final course of IV antibiotic therapy with Meropenem. At the third symptom free monthly follow up visit, we scheduled routine visits every three months. Our patient is one year removed from the infection and has not yet had a recurrence. He now has full resolution of facial function with right-sided House-Brackman grade I examination. Due to his history of Down's Syndrome and intermittent otorrhea due to ear tubes, methods of amplification have been difficult thus far. In the future we may consider ossicular chain reconstruction if no disease recurrence becomes evident versus bone conduction device, as this patient is a poor cochlear implant candidate due to otic capsule obliteration.

### Discussion

The bony labyrinth that surrounds the membranous labyrinth of the inner ear is otherwise known as the otic capsule. The otic capsule comprises of 3 parts: The vestibule, semicircular canals, and cochlea. Partial otic capsular invasion has been reported in literature due to aberrant internal carotid artery [1], Langerhans histiocytosis [2], facial nerve schwannomas [3], cochlear implant device failures [3,4], and chronic suppurative otitis media [5,6]. Although partial otic capsule erosion has been reported, there are no reports of complete cochlear, vestibular, and semicircular canal obliteration. We are reporting the first case of



**Figure 2:** Follow-up Imaging Studies; (2A) Coronal CT: Interval canal wall down mastoidectomy with the mastoid bowl occupied by fluid, including the middle ear and external auditory canal. Progression of the erosion of the otic capsule with communication of the vestibular cochlear system with the internal auditory canal. The ossicles are now absent. Facial canal intact. The tegmen tympani, regular foramina mastoideum, and carotid canal are intact; however, the bone surrounding the jugular foramina and appears eroded. The geniculate are ganglion and cochlear segment of the facial canal appear widened and is suspicious for intracranial extension of the inflammatory process. Bone surrounding the jugular foramina and appears eroded. Otic capsule shows decreased density throughout. Consistent with R temporal bone osteomyelitis and high suspicion of intracranial extension of inflammatory process. Additional sclerosis of semicircular canals. (2B) Axial CT: Complete otic capsule obliteration present, further findings noted above in 2A. (2C) Axial T2 MRI w/ contrast: Status-post right sided mastoidectomy with persistent inflammatory changes of the right temporal bone. Destruction of the otic capsule including osteomyelitis and labyrinthitis with intracranial extension through the internal auditory canal with meningitis also noted with involvement of the 7<sup>th</sup> and 8<sup>th</sup> cranial nerves.

complete, rapid otic capsular obliteration secondary to CSOM.

Madana et al. have defined CSOM as an insidious and chronic intractable inflammation of mucosa, submucosa with destruction of bone of the middle ear cleft characterized by persistent perforation of the tympanic membrane and recurrent otorrhea [9]. Chronic suppurative otitis media, while common in developing countries, has become a rarity in developed countries, with mortality rates falling from 35% to 5% with the introduction of antibiotics and advances in health care [9]. Complications of Chronic Suppurative Otitis Media (CSOM) include mastoiditis, facial palsy, extratemporal abscesses, lateral sinus thrombosis, brain abscess, cerebellar abscess, labyrinthitis, labyrinthine fistula, meningitis, extradural abscess, cochlear erosion, subdural empyema, petrositis, ossicular erosion [5,6,10-13]. These complications are typically insidious in nature [5], and are primarily documented in developing countries [6,14-17]. There have been few reports of complications occurring in developed countries in the last several decades including a case of petrositis and cerebellar abscess complicating chronic otitis media [18]. This resolved with a combination of oral and IV antibiotics with no hearing deficit after resolution of infection. Our patient has Trisomy 21, and is in remission from AML which further predisposes him to otitis media due to compromised immunity, mid-face hypoplasia with malformation of the eustachian tube, a shortened palate, macroglossia, and narrowing of the oropharynx and nasopharynx [19].

Two types of COSM have been described, non-cholesteatomatous/tubotympanic and cholestatomatous or atticofacial [9,14]. CSOM complications such as intracranial abscess, facial nerve palsy, meningitis, petrositis, and mastoiditis, and lateral sinus thrombophlebitis are more commonly found in the cholesteatomatous type of COSM [9]. Mostafa et al describe 422 patients with CSOM complications [5]. Of these 422 patients, 8% had cochlear erosion. Haider et al found that 66 of 279 (23.66%) of patients who underwent surgery for CSOM exhibited ossicular chain erosion [9]. Partial ossicular chain interruption was found in 69.3% of patients with cholesteatomas CSOM vs 13.9% of patients with non-cholesteatomas CSOM [5].

Our patient initially had cholesteatoma for which he underwent right canal wall down tympanomastoidectomy with facial nerve decompression, and then subsequently has resurgence of MDR bacterial infection consistent with CSOM which progressed to full blown osteomyelitis of the right temporal bone and obliteration of the vestibule, cochlea, and semicircular canals. Congenital temporal bone dehiscence, although rare, could explain the rapid invasion of infection into the inner ear leading to purulent labyrinthitis. This congenital pathway for spread of infection has been reported as a common cause of pediatric meningitis [20,21].

The difficulty in treatment for this case presentation was highlighted by the results of bacterial culture including MDR E. Coli sensitive to amikacin and ceftriaxone and MDR Corynebacterium sensitive to meropenem. This exemplifies the importance of culture of ear drainage for culture directed IV antibiotic therapy which is what ultimately led to resolution for this patient. Although treated with appropriate culture directed antibiotic therapy, our patient had refractory infection. With the antibiotic resistance crisis, developed countries could potentially see a steady increase of CSOM complications that were once nearly ameliorated with antibiotics.

## References

1. Yao W, Benjamin LC, Korzec K. Aberrant Internal Carotid Artery Causing Erosion of the Otic Capsule: An Unusual Cause of Pulsatile Tinnitus. *Otolaryngol Neck Surg* 1998; 118: 678–679.
2. Blumberg JM, Malhotra A, Wu X, et al. Langerhans Cell Histiocytosis of the Temporal Bone with Otic Capsule Involvement. *Clin Neuroradiol* 2017; 27: 163–168.
3. Loos E, Wuyts L, Puls T, et al. Cochlear erosion due to a facial nerve schwannoma. *J Int Adv Otol*. 2019; 15: 330–332.
4. Doherty JK, Linthicum FH. Cochlear endosteal erosion with focal osteomyelitis induced by cochlear implantation. *Otol Neurotol*. 2004; 25: 1029–1030.
5. Mostafa BE, El Fiky LM, El Sharnouby MM. Complications of suppurative otitis media: Still a problem in the 21st century. *ORL Published Online First*. 2009.
6. Osma U, Cureoglu S, Hosoglu S. The complications of chronic otitis media: Report of 93 cases. *J Laryngol Otol Published Online First*. 2000.
7. Sharma N, Jaiswal AA, Banerjee PK, Garg AK. Complications of Chronic Suppurative Otitis Media and Their Management: A Single Institution 12 Years Experience. *Indian J Otolaryngol Head Neck Surg*. 2015; 67: 353-360.
8. Paparella MM, Shumrick DA, Gluckman JL, Meyerhoff WL. (Eds.). *Otolaryngology: Volume II Otolology and Neurootology*. (3rd ed.). Philadelphia: W.B. Saunders Company. 1991.
9. Madana J, Yolmo D, Kalaiarasi R, et al. Microbiological profile with antibiotic sensitivity pattern of cholesteatomatous chronic suppurative otitis media among children. *Int J Pediatr Otorhinolaryngol*. 2011; 75: 1104–1108.
10. Haidar H, Sheikh R, Larem A, et al. *Hiader.pdf. Otolaryngol Open Access*. 2015; 5: 2.
11. Varshney S, Nangia A, Bist SS, et al. Ossicular Chain Status in Chronic Suppurative Otitis Media in Adults. *Indian J Otolaryngol Head Neck Surg*. 2010.
12. Smith JA, Danner CJ. Complications of Chronic Otitis Media and Cholesteatoma. *Otolaryngol. Clin. North Am*. 2006.
13. Lin YS, Lin LC, Lee FP, et al. The prevalence of chronic otitis media and its complication rates in teenagers and adult patients. *Otolaryngol - Head Neck Surg Published Online First*: 2009.
14. Wakode PT, Joshi S V., Gawarle SH. Chronic suppurative otitis media in school going children. *Indian J Otolaryngol Head Neck Surg*. 2006; 58: 152–155.
15. Migirov L, Bendet E, Kronenberg J. Cholesteatoma invasion into the internal auditory canal. *Eur Arch Oto-Rhino-Laryngology*. 2009; 266: 657–662.
16. Kangsanarak J, Fooanant S, Ruckphaopunt K, et al. Extracranial and intracranial complications of suppurative otitis media. Report of 102 cases. *J Laryngol Otol*. 1993.
17. Oberdorfer P, Kongthavonsakul K, Intachumpoo J, et al. A 14-year-old girl with tuberculous otitis media and brain abscess. *BMJ Case Rep*. 2012; 2–4.
18. Trimis G, Mostrou G, Lourida A, et al. Petrositis and cerebellar abscess complicating chronic otitis media. *J Paediatr Child Health*. 2003.
19. Mitchell R, Call E, Kelly J. Ear, nose and throat problems in children with down syndrome. *Br J Hosp Med*. 2005; 66: 504–506.
20. Harrington JW, Birck HG. Recurrent Meningitis Due to Congenital Petrous Fistula: A Case Report. *Arch Otolaryngol* 1967; 85: 572–575.
21. Kimitsuki T, Inamitsu M, Komune S, et al. Congenital malformation of the inner ear associated with recurrent meningitis. *Eur Arch Oto-Rhino-Laryngology*. 1999.