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# Ruptured Congenital Cholesteatoma Presenting as Atticoantral Chronic Suppurative Otitis Media in a 4-Year-Old: A Diagnostic Challenge

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**A B S T R A C T**

Chronic suppurative otitis media (CSOM) of the atticoantral type is a distinct otologic pathology driven by cholesteatoma, characterized by enzymatic bone erosion and a high risk of intracranial complications. While typically acquired through retraction pockets, a subset of these cases represents congenital cholesteatomas (CC) that have silently expanded and ruptured the tympanic membrane. This presentation mimics acquired disease, creating a diagnostic blind spot. We report the case of a 4-year-old male presenting with a one-year history of persistent, foul-smelling otorrhea and otalgia in the right ear. Despite a history of recurrent cold, there was no prior trauma or otologic surgery. Physical examination revealed an attic perforation with purulent discharge. Computed tomography scan (CT-scan) identified a soft tissue mass filling the epitympanum and mesotympanum with significant erosion of the scutum and ossicles. The patient underwent a modified radical mastoidectomy with type II tympanoplasty. Intraoperative findings revealed an extensive cholesteatoma sac consistent with a congenital origin that had secondarily ruptured. In conclusion, congenital cholesteatoma must be considered in the differential diagnosis of pediatric CSOM, even in the presence of a perforation. This ruptured presentation highlights the necessity of early HRCT imaging over conventional radiography. The canal wall down approach remains a critical strategy for eradicating extensive pediatric disease to prevent recidivism.

## 1. Introduction

Chronic suppurative otitis media (CSOM) represents a pervasive and debilitating pathology that continues to pose a significant public health challenge worldwide. It is a condition that transcends mere localized infection, often resulting in profound physical, social, and psychological sequelae for the affected individual.[1] The burden of this disease is disproportionately borne by developing nations, where factors such as limited access to healthcare, malnutrition, and high population density contribute to its persistence. In the specific context of Southeast Asia, the prevalence of CSOM is alarmingly high, estimated at approximately 7.8% according to data from the World Health Organization. Globally, the disease affects between 65 to 330 million individuals, making it a leading cause of preventable hearing loss.[2]

The clinical landscape of CSOM is classically stratified into two distinct entities based on the behavior of the disease and the associated risks: the

tubotympanic type, often termed benign or safe, and the atticoantral type, designated as malignant or dangerous.[3] This binary classification is not merely semantic but dictates the urgency of therapeutic intervention. The benign type is typically confined to the mucosa of the middle ear, characterized by a central perforation and mucopurulent discharge. In stark contrast, the atticoantral or malignant type is defined by the presence of cholesteatoma. This variant accounts for nearly 95.7% of all dangerous CSOM cases. Despite the nomenclature malignant, cholesteatoma is histologically benign; however, its clinical behavior is aggressively destructive. It acts as a bone-eroding process within the temporal bone, driven by a cystic accumulation of keratinizing squamous epithelium in the middle ear cleft, where it does not natively belong.[4]

The danger posed by atticoantral disease lies in the unique capacity of cholesteatoma to destroy bone. This destruction is mediated through a dual mechanism involving mechanical pressure necrosis and enzymatic osteolysis.[5] As the cholesteatoma sac expands due to the accumulation of desquamated keratin debris, it exerts pressure on the surrounding bony structures, compromising their blood supply. Concurrently, the associated chronic inflammatory response triggers the release of osteolytic enzymes and cytokines that actively resorb bone. This relentless expansion threatens critical anatomical structures housed within the temporal bone, including the ossicular chain, the facial nerve canal, the bony labyrinth, and the tegmen tympani, which separates the ear from the brain. Consequently, untreated atticoantral disease can lead to catastrophic intratemporal and intracranial complications, such as facial paralysis, labyrinthitis, meningitis, and brain abscess. The microbiological environment of CSOM further complicates its management. The etiology is multifactorial, involving a complex interplay of aerobic and anaerobic bacteria, often organized within biofilms that confer resistance to standard antimicrobial therapies. Pathogens such as *Staphylococcus aureus* (including Methicillin-resistant strains or MRSA), *Pseudomonas aeruginosa*, and *Proteus* species are frequently isolated from the purulent discharge. The infected cholesteatoma sac is particularly recalcitrant to medical therapy because the keratin mass lacks a blood supply, preventing systemic antibiotics from reaching effective concentrations within the core of the lesion. Topical antibiotics, while useful for superficial infection control, fail to eradicate the underlying pathology.[6]

Children are disproportionately affected by otitis media, a susceptibility largely attributed to the immature anatomy and physiology of the pediatric ear.[7] In infants and young children, the Eustachian tube is shorter, wider, and oriented more horizontally—approximately 10 degrees relative to the skull base—compared to the 45-degree angulation seen in adults. This anatomical configuration, combined with an immature immune system, predisposes children to the reflux of nasopharyngeal pathogens into the middle ear, leading to recurrent acute otitis media (AOM) and persistent effusion. Within the pediatric population, the origin of cholesteatoma is a subject of significant academic debate. The majority of cases are classified as acquired, arising from retraction pockets in the pars flaccida or posterior superior quadrant of the tympanic membrane due to chronic Eustachian tube dysfunction (the invagination theory). Alternatively, acquired cholesteatoma may develop secondary to the migration of squamous epithelium through a pre-existing tympanic membrane perforation. However, a distinct and clinically critical subset of pediatric cases is congenital in origin. Congenital cholesteatoma (CC) arises not from infection or retraction, but from aberrant embryonic epithelial cell rests that become trapped within the temporal bone during fetal development. These epidermoid formations typically reside in the anterior epitympanum or posterior mesotympanum. The classic diagnostic criteria for congenital cholesteatoma, proposed by Levenson,

require the presence of a white mass behind an intact, normal tympanic membrane in a patient with no history of otorrhea, perforation, or prior otologic surgery.[8]

While the classic presentation of congenital cholesteatoma involves an intact drum, the natural history of the disease is one of progressive expansion. This growth creates a diagnostic paradox that often confounds clinicians. As the congenital cyst enlarges—estimated to grow at a rate of approximately 1 mm per year—it eventually impinges upon the tympanic membrane. If left undiagnosed, the cyst may erode through the drum, resulting in a spontaneous rupture. Once the congenital cholesteatoma ruptures, the clinical picture undergoes a dramatic transformation. The child, who may have been previously asymptomatic, suddenly presents with otorrhea and a visible perforation. At this stage, the condition mimics acquired atticoantral CSOM indistinguishably. The defining intact drum criterion is lost, leading many clinicians to misdiagnose the condition as an acquired disease secondary to chronic infection. This distinction is not merely academic; congenital cholesteatomas often have different growth patterns and extensions compared to acquired retraction pockets. The ruptured presentation represents a silent killer scenario where the pathology has advanced significantly—eroding ossicles and expanding into the mastoid—before the first symptom appears. This delayed presentation is a major factor contributing to the extensive disease often seen in young children.[9]

The limitations of clinical examination in these complex pediatric cases necessitate the use of advanced imaging modalities. Otoscopy and otomicroscopy, while essential for identifying the perforation and the presence of keratin debris, provide limited information regarding the extent of the disease within the pneumatized spaces of the temporal bone. Conventional radiography, such as the Schuller view, has historically been used but offers low sensitivity. It typically reveals non-specific findings such as mastoid sclerosis or hypopneumatization, failing to delineate the soft tissue mass or subtle bony erosions. Computed tomography (CT) scan of the temporal bone has emerged as the gold standard for preoperative assessment. CT-scan offers superior spatial resolution, allowing for the precise visualization of the cholesteatoma mass and its destructive effects on the bony architecture. Key radiological markers of cholesteatoma on CT-scan include the presence of a soft tissue mass with sharp margins, blunting or erosion of the scutum (the lateral epitympanic wall), and destruction of the ossicular chain and tegmen tympani. CT-scan is indispensable for surgical planning, as it alerts the surgeon to anatomical hazards such as a dehiscent facial nerve, a low-lying dura, or a fistula of the lateral semicircular canal.

The management of pediatric cholesteatoma is primarily surgical, with the dual objectives of eradicating the disease to create a dry, safe ear and preserving or reconstructing hearing function. The two main surgical approaches are canal wall up (CWU) and canal wall down (CWD) mastoidectomy. The choice between these techniques remains a point of contention in pediatric otology.[10] CWU mastoidectomy preserves the posterior bony canal wall, maintaining near-normal anatomy and avoiding the maintenance issues associated with a mastoid cavity. However, this approach is associated with a higher risk of residual and recurrent disease (recidivism), particularly in children who often have aggressive disease and persistent Eustachian tube dysfunction. Conversely, CWD mastoidectomy involves lowering the posterior canal wall to exteriorize the mastoid air cells into the external auditory canal. While this creates a cavity that requires lifelong cleaning, it significantly reduces the rate of recurrence and is often considered the safer option for extensive or recurrent cholesteatoma. In cases of ruptured congenital cholesteatoma, where the disease may be extensive due to years of silent growth, a Modified Radical Mastoidectomy (a type of CWD procedure) combined with tympanoplasty is frequently

required to ensure complete removal of the matrix and prevent intracranial complications.

Against this background of diagnostic complexity and surgical challenge, this case report aims to highlight the clinical entity of ruptured congenital cholesteatoma presenting as atticoantral CSOM in a preschool-aged child. The novelty of this study lies in its detailed documentation of the diagnostic blind spot where congenital disease mimics acquired pathology following tympanic membrane rupture. We aim to demonstrate the critical importance of looking beyond the perforation to identify the congenital origin of the disease, utilizing CT-scan to map the silent destruction that occurred prior to presentation. Furthermore, this report seeks to validate the efficacy of the Modified Radical Mastoidectomy (Canal Wall Down) approach with simultaneous ossicular reconstruction (Type II Tympanoplasty) as a definitive management strategy for extensive pediatric cholesteatoma. By analyzing the correlation between the clinical presentation, radiological findings, and intraoperative reality, this study provides valuable insights for otologists managing dangerous ear disease in the pediatric population, emphasizing that a perforated drum does not exclude a congenital etiology.

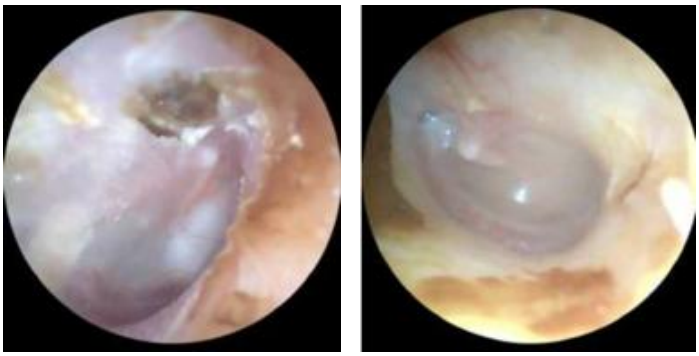
## 2. Case Presentation

Written informed consent was obtained from the patient's legal guardian (parents) for the publication of this case report and any accompanying images. The identity of the patient has been anonymized to protect privacy.

A 4-year-old male child presented to the Otorhinolaryngology outpatient clinic at a tertiary referral center (Prof. Dr. I.G.N.G. Ngoerah General Hospital, Denpasar). The patient was brought by his family with a chief complaint of persistent discharge from the right ear for approximately one year. The discharge was described as yellowish-white, foul-smelling, and intermittent. The patient also reported pain (otalgia) deep within the right ear canal.

The medical history revealed frequent episodes of upper respiratory tract infection (common cold) characterized by clear rhinorrhea, without significant cough or fever. Crucially, there was no history of trauma to the right ear, no history of facial asymmetry, no vertigo, and no severe headaches associated with nausea or vomiting. The patient had no history of prior otologic surgery. This absence of prior intervention or trauma is a significant historical marker when considering the etiology of the cholesteatoma.

Upon clinical admission, the patient was observed to be in good general condition and fully alert, presenting as compos mentis. A focused otorhinolaryngological examination was subsequently performed (Figure 1). Inspection of the affected right ear revealed a normal-appearing auricle with no elicitable tenderness over the tragus or the retroauricular region, effectively ruling out acute mastoiditis at presentation. The external auditory canal was patent; however, it contained a significant amount of mucopurulent secretion. Following careful suction clearance, otoscopic visualization demonstrated a pathological perforation specifically localized to the attic (pars flaccida) region of the tympanic membrane. Crucially, whitish keratin debris was visible extending through the perforation, a clinical finding strongly indicative of an underlying cholesteatoma matrix. Conversely, examination of the contralateral left ear demonstrated normal anatomy, characterized by a patent canal free of secretions and an intact tympanic membrane with a preserved light reflex. The remainder of the head and neck examination, including anterior rhinoscopy and oropharyngeal assessment, was unremarkable, and there was no palpable cervical lymphadenopathy.



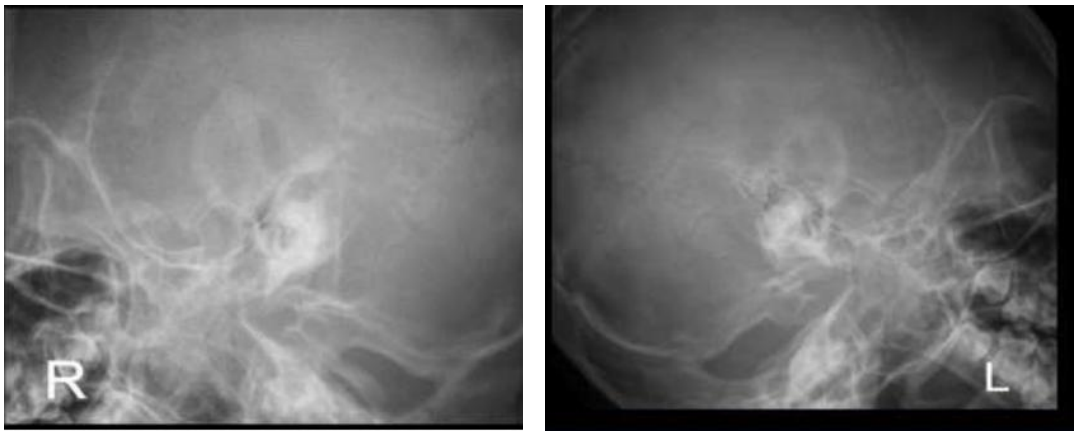
(A) (B)

Figure 1. Otoscopic examination of the tympanic membrane. (A) right ear (pathologic); (B) left ear (normal).

The pre-operative functional assessment of the auditory system presented specific challenges inherent to the pediatric demographic. Due to the patient's young age (4 years) and limited ability to cooperate with subjective testing instructions, standard qualitative assessments using tuning forks—such as the Rinne and Weber tests—were precluded. Consequently, objective measures were prioritized to evaluate the middle ear status. Tympanometry was successfully performed, yielding a Type A tympanogram bilaterally. In the context of the affected right ear, which harbored a known attic perforation, this finding is clinically paradoxical; a Type A curve in this pathological ear likely suggests that the cholesteatoma mass or associated polypoid tissue was obstructing the perforation or filling the middle ear space in a manner that mimicked normal compliance, or potentially indicated a false-positive seal during the procedure.

To ensure the patient's physiological fitness for general anesthesia and to mitigate intraoperative risks, a comprehensive panel of laboratory investigations was conducted. The hematological profile demonstrated a Hemoglobin level of 12.7 g/dL, a Leukocyte count of 5,250/µL, and a Hematocrit of 36.60%, all of which fell within the normative reference ranges for the patient's age group. Furthermore, the platelet count was recorded at 191,000/µL. Given the vascular nature of the temporal bone and the potential for bleeding during granulation tissue removal, the coagulation profile was scrutinized; the Prothrombin Time (PPT) was 10.3 seconds, and the Activated Partial Thromboplastin Time (APTT) was 28.5 seconds. These values confirmed an intact coagulation cascade, permitting the surgical team to proceed with the planned otologic intervention without the requirement for preoperative transfusion or hematological correction.

The radiological investigation followed a stepwise algorithm, beginning with conventional 2D radiography before advancing to 3D tomographic mapping. Initial imaging utilized the Schuller view radiography performed during the first month of clinical assessment. This modality provided a gross overview of the mastoid pneumatization. The findings revealed significant hypopneumatization of the right mastoid with diffuse clouding of the mastoid air cells and marked sclerosis of the periantral triangle. While these findings were consistent with chronic inflammatory disease, the Schuller view failed to visualize a definitive cholesteatoma mass or delineate the specific extent of bony erosion. The left mastoid similarly exhibited clouding and sclerotic lines, leading to an initial radiological impression of bilateral chronic mastoiditis, with dominant pathology on the right side.




(A) (B)

Figure 2. Schuller views radiography. (A) right ear; (B) left ear.

Recognizing the limitations of plain radiography in defining complex cholesteatomatous anatomy, a computed tomography (CT) scan of the temporal bone was performed approximately one month later to provide a detailed roadmap for surgical planning. The CT scan unveiled a soft tissue density consistent with chronic otomastoiditis and cholesteatoma extensively occupying the right epitympanum, mesotympanum, and hypotympanum. The disease process was not confined to the middle ear cleft but extended posteriorly into the aditus ad antrum and laterally into Prussak's space (Table 1). Crucially, the scan demonstrated aggressive osteolytic activity, a hallmark of dangerous CSOM. There was definitive bony erosion of the scutum (the lateral wall of the epitympanum) and the tegmen tympani, the thin bony plate separating the middle ear from the middle cranial fossa. Furthermore, the ossicular chain, specifically the malleus and incus, showed signs of erosion. These findings confirmed the diagnosis of atticoantral disease and highlighted the imperative for surgical intervention to prevent intracranial complications. Secondary incidental findings included bilateral maxillary sinusitis and hypertrophy of the left inferior nasal turbinate.

Following the confirmation of the diagnosis and the completion of pre-operative optimization, the patient underwent definitive surgical management in the fifth month following initial presentation (Table 2a). The procedure was conducted under general anesthesia utilizing a hypotensive technique to minimize intraoperative bleeding and improve visualization of the microsurgical field. The patient was classified as ASA I physical status, indicating a healthy patient with no systemic disease.

The surgery commenced with a retroauricular approach, chosen to provide wide access to the mastoid cortex and the middle ear. A curvilinear incision was made through the skin and subcutaneous tissue approximately 3 mm posterior to the retroauricular sulcus. Hemostasis was achieved via infiltration with epinephrine (1:200,000). The periosteum was elevated to expose the mastoid cortex, and a self-retaining retractor was placed. The surgical landmark of Macewen's triangle (the suprameatal triangle) was identified to guide the initial cortical mastoidectomy.

**Table 1. Summary of Clinical, Laboratory, and Radiological Findings on Admission**

| PARAMETER / CATEGORY | DETAILED CLINICAL FINDINGS |
|---|---|
| **1. PATIENT PROFILE & HISTORY** | |
| **Demographics** | 4-year-old Male |
| **Chief Complaint** | Persistent **Right Ear Otorrhea** (1 year duration).<br>Characteristics: Yellowish-white, foul-smelling, intermittent. |
| **Associated Symptoms** | Right Otalgia (deep ear pain). Recurrent upper respiratory infections (common cold). |
| **Relevant Negatives** | **Negative** for: Trauma, Prior Otologic Surgery, Facial Asymmetry, Vertigo, Nausea/Vomiting. |
| **2. PHYSICAL EXAMINATION (OTOLARYNGOLOGY)** | |
| **Right Ear (Pathological)** | **External Canal:** Patent, mucopurulent secretions present.<br>**Tympanic Membrane:** **Attic Perforation** visualized.<br>**Pathology:** Whitish keratin debris (Cholesteatoma) visible through perforation. |
| **Left Ear (Healthy)** | **External Canal:** Patent, no secretions.<br>**Tympanic Membrane:** **Intact**, positive light reflex, pearly grey appearance. |
| **General & Neck** | Compos Mentis (Alert). No retroauricular pain/swelling. No palpable cervical lymphadenopathy. |
| **3. AUDIOLOGICAL & LABORATORY ASSESSMENT** | |
| **Audiometry (Tympanometry)** | **Type A** (Bilateral).<br>*Note: Paradoxical finding in the right ear likely due to mass effect sealing the middle ear space.* |
| **Hematology Panel** | Hemoglobin: 12.7 g/dL<br>Leukocytes: 5,250 /µL<br>Platelets: 191,000 /µL<br>Hematocrit: 36.60% |
| **Coagulation Profile** | Prothrombin Time (PPT): 10.3 seconds<br>APTT: 28.5 seconds<br>*Status: Within normal limits for surgery.* |
| **4. DIAGNOSTIC IMAGING (RADIOLOGY)** | |
| **Conventional X-Ray (Schuller)** | **Right Mastoid:** Hypopneumatization, clouding of air cells, sclerosis of periantral triangle. |
| **CT Scan (Temporal Bone)** | **Mass Location:** Soft tissue density in Epitympanum, Mesotympanum, Hypotympanum, Aditus ad Antrum, and Prussak's space.<br>**Bone Erosion:** **Erosion of Scutum** (Lateral Epitympanic Wall), **Erosion of Tegmen Tympani**, and **Erosion of Ossicles** (Malleus/Incus).<br>**Diagnosis:** Atticoantral CSOM with Cholesteatoma. |

Upon drilling the mastoid cortex and entering the antrum, the true extent of the pathology was revealed. A massive cholesteatoma sac was found filling the entire mastoid cavity and extending anteriorly into the epitympanum. The cholesteatoma had caused significant destruction of the surrounding bony architecture. Given the extensive nature of the disease and the erosion of the posterior canal wall, the

decision was made to perform a canal wall down (CWD) procedure. The posterior bony canal wall was lowered to the level of the facial ridge, thereby exteriorizing the mastoid air cells and converting the mastoid and middle ear into a common cavity. This maneuver is essential in extensive pediatric cholesteatoma to ensure complete removal of the matrix and to minimize the risk of residual disease. The cholesteatoma matrix was meticulously dissected and removed from the mastoid, antrum, and tympanic cavity. Inspection of the ossicular chain confirmed the preoperative CT findings: both the malleus and incus were significantly eroded by the disease process. Following the eradication of the pathology, reconstructive steps were initiated. A Type II Tympanoplasty was performed to restore the sound conducting mechanism, utilizing the remaining ossicular remnants or a graft to bridge the gap to the stapes. This procedure was combined with a Modified Radical Mastoidectomy to create a safe, dry, and self-cleaning cavity. To ensure adequate aeration of the mastoid bowl and allow for easy postoperative cleaning, a wide meatoplasty was performed. The surgical site was then packed with antibiotic-impregnated gauze, and a pressure dressing was applied.

Intraoperative tissue samples were submitted for histopathological examination, which subsequently confirmed the morphological features of cholesteatoma, characterized by keratinizing squamous epithelium. The post-operative course was uncomplicated (Table 2b). The patient recovered well from anesthesia with no immediate sequelae. Crucially, facial nerve function was assessed immediately post-operatively and found to be intact, with no evidence of facial asymmetry or paresis. The patient was monitored in the ward and discharged on the second post-operative day. Discharge medications included oral antibiotics (Cefixime 50 mg every 12 hours) to prevent secondary infection and analgesics (Paracetamol) for pain control. Follow-up care was conducted to ensure proper wound healing and graft uptake. At the first follow-up visit, one week post-surgery, the patient was asymptomatic with no subjective complaints. Two weeks post-surgery, the retroauricular sutures were removed, and the ear canal tampon was extracted. Assessment at this stage revealed a well-healed retroauricular wound and a viable graft. By the third post-operative week, the ear canal was noted to be clean, and the surgical cavity was dry, indicating a successful initial anatomical outcome.

**Table 2a. Diagnostic Profile and Initial Medical Management**

| CLINICAL PHASE | DETAILS OF MANAGEMENT & FINDINGS |
|---|---|
| **1. DIAGNOSIS HIERARCHY** | |
| **Working Diagnosis** | Pre-operative Chronic Suppurative Otitis Media (CSOM) - Atticoantral Type (Dangerous) Dextra. |
| **Radiological Diagnosis** | Chronic otomastoiditis with cholesteatoma filling epitympanum, mesotympanum, and hypotympanum (CT Scan Findings). |
| **Final Diagnosis** | Post-operative **Malignant CSOM Dextra with Ruptured Congenital Cholesteatoma** (Post-Tympanoplasty Type II & Modified Radical Mastoidectomy). |
| **2. INITIAL MEDICAL MANAGEMENT (CONSERVATIVE)** | |
| **Regimen** | • **Antibiotic (Systemic):** Erythromycin 112 mg PO every 6 hours.<br>• **Antibiotic (Topical):** Otozambon ear drops (Polymyxin B/Neomycin), 2 drops every 12 hours.<br>• **Decongestant:** Pseudoephedrine HCl 15 mg + Triprolidine HCl 2.5 mg every 24 hours.<br>• **Analgesic:** Paracetamol 150 mg every 8 hours (prn pain). |
| **Result** | Therapy continued for 2 weeks. Failed . Symptoms persisted, prompting surgical evaluation. |

**Table 2b. Surgical Intervention, Follow-up, and Outcomes**

| CLINICAL PHASE | DETAILS OF MANAGEMENT & FINDINGS |
|---|---|
| **3. SURGICAL INTERVENTION (DEFINITIVE)** | |
| **Procedure Details** | **Anesthesia:** General (Hypotensive technique).<br>**Incision:** Retroauricular approach. |
| **Surgical Techniques** | Canal Wall Down **Modified Radical Mastoidectomy:** Posterior canal wall lowered to exteriorize extensive disease.<br>Reconstruction **Type II Tympanoplasty:** Reconstruction of the ossicular chain mechanism.<br>**Meatoplasty:** Wide meatoplasty performed to ensure cavity aeration. |
| **Intraoperative Findings** | Massive cholesteatoma sac filling the mastoid and epitympanum. Erosion of malleus and incus. Perforation in the attic region. |
| **4. POST-OPERATIVE COURSE & OUTCOME** | |
| **Post-op Medication** | **Inpatient:** Ceftriaxone 400 mg IV q12h, Pseudoephedrine 15 mg PO q8h.<br>**Discharge:** Cefixime 50 mg PO q12h, Pseudoephedrine 15 mg q8h, Paracetamol 150 mg q8h. |
| **Follow-up Timeline** | • **Day 2 (Discharge):** Good condition, no facial palsy, drain removed.<br>• **Week 1:** No subjective complaints.<br>• **Week 2:** Suture and tampon removal. Wound well-healed.<br>• **Week 3:** Ear canal clean, mastoid cavity dry, graft intact. |
| **Final Outcome** | Success Anatomical success achieved with a dry, safe ear. Functional preservation of the facial nerve (House-Brackmann Grade I). Histopathology confirmed Cholesteatoma. |

## 3. Discussion

Our study presents a compelling diagnostic conundrum that challenges the traditional binary classification of chronic suppurative otitis media (CSOM). In the landscape of pediatric otology, the distinction between acquired and congenital cholesteatoma is not merely semantic but carries profound implications for pathophysiology and surgical planning. Classically, congenital cholesteatoma is defined by the criteria established by Levenson, which necessitate the presence of a white mass behind an intact, normal tympanic membrane in a patient with no history of otorrhea, prior perforations, or otologic surgical procedures. Under strict adherence to these criteria, the patient in this report—presenting with a year-long history of purulent otorrhea and a visible attic perforation—would predominantly be classified as having primary acquired cholesteatoma.[11]

However, a deeper analysis of the clinical timeline and intraoperative findings suggests a more complex etiology: a ruptured congenital cholesteatoma. The natural history of congenital cholesteatoma is one of insidious, silent expansion. Originating from aberrant embryonic epithelial rests trapped within the temporal bone—typically in the anterior epitympanum or posterior mesotympanum—these cysts grow slowly, with an estimated radial expansion of approximately 1 mm per year.[12] In the early stages, this growth is asymptomatic and hidden behind the tympanic membrane. As the cyst enlarges within the confined spaces of the middle ear, it inevitably exerts pressure on surrounding structures. In this patient, the absence of a history of recurrent acute otitis media prior to the onset of discharge is a critical historical marker. It supports the hypothesis that the cholesteatoma was not the sequela of chronic Eustachian tube dysfunction and retraction pocket

formation (the mechanism of acquired disease), but rather a primary mass that expanded until it mechanically eroded through the pars flaccida of the tympanic membrane.[13]

Once this rupture occurs, the sterile environment of the congenital cyst is breached, allowing the ingress of pathogens from the external auditory canal. This event transforms the clinical picture into one mimicking acquired atticoantral CSOM, characterized by secondary bacterial infection and foul-smelling otorrhea.[14] This masked presentation represents a significant diagnostic blind spot. The clinician seeing the patient at this stage observes only the perforation and discharge, often failing to recognize the congenital origin of the disease. This case underscores the necessity for otologists to maintain a high index of suspicion for congenital etiologies even in the presence of perforation, particularly in young children with extensive disease but no antecedent history of ear trauma or chronic effusions.

The aggressive nature of the disease observed in this 4-year-old patient—characterized by extensive involvement of the epitympanum, mesotympanum, and mastoid with significant ossicular erosion—highlights the vulnerability of the pediatric temporal bone. Cholesteatoma in children is frequently more aggressive than in adults, a phenomenon attributed to greater cellular proliferation rates and an intense inflammatory response.[15] The underlying pathophysiology involves a dual mechanism of bone destruction: pressure necrosis exerted by the expanding keratin sac and enzymatic osteolysis mediated by osteoclasts and inflammatory cytokines recruited by the superadded infection (Figure 3).

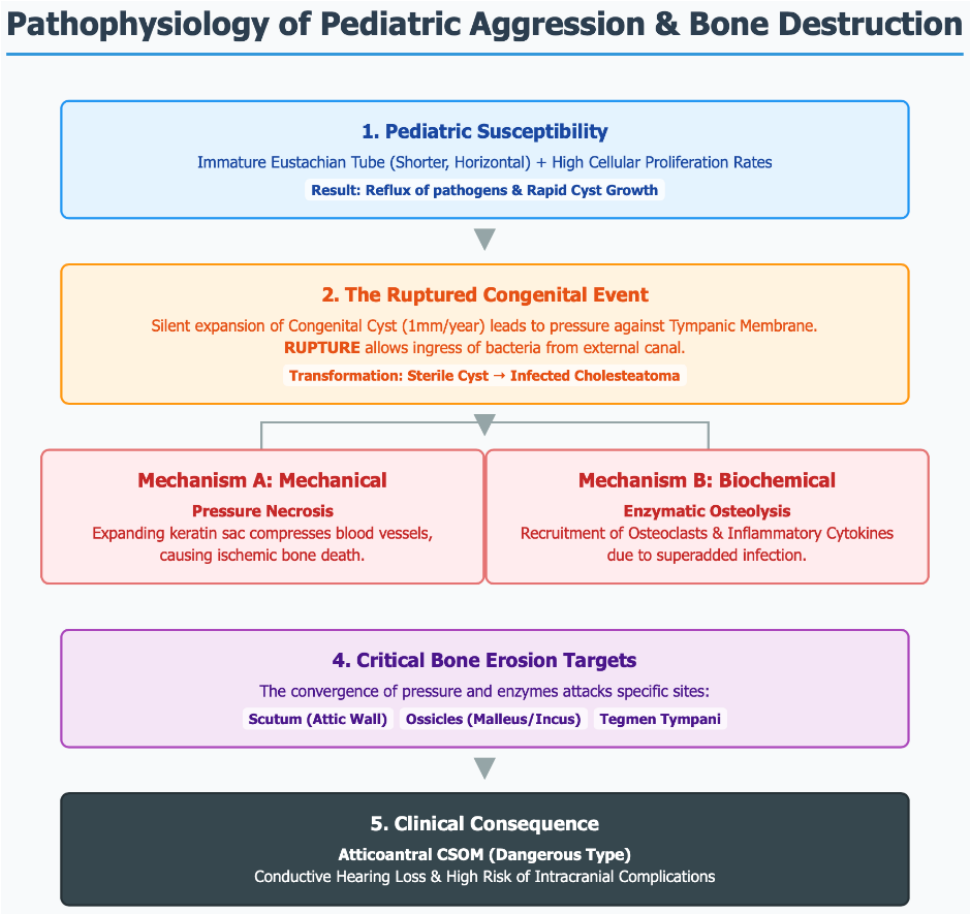



Figure 3. Pathophysiology of pediatric aggression and bone destruction.

In this case, the cholesteatoma occupied critical anatomical sub-sites including Prussak's space and the aditus ad antrum. The erosion of the scutum (the lateral wall of the epitympanum) observed on imaging is pathognomonic for attic cholesteatoma.[16] This bony erosion is not merely a radiological finding but a testament to the biochemical aggressiveness of the disease. The destruction of the ossicular chain, specifically the malleus and incus, resulted in a conductive hearing loss, which is the most common functional sequela in CSOM, affecting over 90% of patients. The disruption of the ossicular chain in this patient necessitated the intraoperative decision to perform a Type II tympanoplasty, bridging the gap between the tympanic membrane graft and the stapes to restore sound transmission. Furthermore, the pediatric Eustachian tube plays a pivotal role in the disease process. In children under the age of seven, the tube is shorter, wider, and more horizontally oriented (approximately 10 degrees) compared to the adult angulation. This immature anatomy predisposes the middle ear to the reflux of nasopharyngeal pathogens and hinders the clearance of secretions. In the context of a ruptured congenital cholesteatoma, this dysfunction exacerbates the secondary infection, creating a vicious cycle of inflammation and otorrhea that is often refractory to medical management.[17]

This case report provides a stark illustration of the limitations of conventional 2D radiography in the management of complex otologic pathology. The initial investigation utilizing Schuller view radiography revealed only hypopneumatization and sclerosis of the mastoid air cells. While sclerosis is a hallmark of chronic mastoiditis, indicating a long-standing lack of aeration, it is a non-specific finding.[18] The Schuller view failed to visualize the cholesteatoma mass itself or to delineate the critical bony erosions that were present. Relying solely on such findings can lead to a dangerous underestimation of the disease extent, potentially resulting in inadequate surgical planning or delayed intervention.

In contrast, the computed tomography (CT) scan served as the definitive diagnostic roadmap. The scan revealed a soft tissue density filling the epitympanum and mesotympanum with extension into the aditus ad antrum, correlating perfectly with the intraoperative findings. More importantly, the CT scan identified the erosion of the scutum and the tegmen tympani. Tegmen erosion is a critical preoperative warning sign, alerting the surgeon to the risk of dural exposure and intracranial extension. The superior spatial resolution of HRCT allows for the assessment of hidden areas such as the sinus tympani and facial recess, which are frequent sites of residual disease. The literature supports the assertion that HRCT is the gold standard for preoperative assessment in cholesteatoma. It offers high sensitivity for detecting ossicular erosion and labyrinthine fistulas, the latter being a complication found in 5-10% of cases. In this patient, the specific finding of scutum erosion on CT was highly predictive of the attic retraction and cholesteatoma sac found during surgery. Therefore, this study advocates for the early utilization of HRCT in any pediatric patient presenting with persistent otorrhea and attic pathology, as conventional radiography is insufficient for safe surgical decision-making.

The management of pediatric cholesteatoma is a subject of ongoing debate, primarily centered on the choice between canal wall up (CWU) and canal wall down (CWD) mastoidectomy techniques.[19] The primary goal of cholesteatoma surgery is threefold: to eradicate the disease, to create a dry and safe ear, and to preserve or restore hearing function. In this case, the surgical team opted for a Modified Radical Mastoidectomy, a CWD procedure. The rationale for this aggressive approach was multifactorial. Firstly, the intraoperative findings revealed a massive cholesteatoma filling the mastoid cavity and epitympanum with significant erosion of the posterior canal wall structures. In the presence of such extensive disease, attempting to preserve the posterior canal wall (CWU) carries a significantly higher risk of

recidivism. Recidivism rates in pediatric CWU procedures have been reported to be as high as 30-50%, often necessitating revision surgery. Children are particularly prone to recurrent retraction pockets due to persistent negative middle ear pressure and aggressive epithelial growth.

By performing a CWD procedure—specifically lowering the facial ridge and converting the mastoid and middle ear into a common cavity—the surgeon eliminates the bony scaffold that supports the formation of new retraction pockets. This technique exteriorizes the disease, allowing for direct surveillance of the cavity during follow-up. While CWD procedures result in an anatomical alteration requiring lifelong cavity care (cleaning of debris), they provide the highest assurance of a safe ear, which is the priority in preventing life-threatening intracranial complications. The procedure was combined with a Type II Tympanoplasty to address the conductive hearing loss caused by the erosion of the malleus and incus. This demonstrates that the radical removal of disease does not preclude functional rehabilitation. The successful uptake of the graft and the creation of a dry, epithelialized cavity in the post-operative period validate the efficacy of this approach.[20]

The management of this patient highlights the importance of a comprehensive treatment strategy that extends beyond the operating room. The preoperative medical management, involving systemic erythromycin and topical antibiotics, failed to resolve the otorrhea. This failure is consistent with the nature of cholesteatoma, where the avascular keratin mass acts as a sanctuary for bacteria (often within biofilms), rendering systemic antibiotics ineffective. This resistance to medical therapy should serve as a red flag for clinicians, prompting early imaging and surgical consultation rather than prolonged courses of ineffective antibiotics. Furthermore, the postoperative care in CWD patients is critical. The creation of a mastoid bowl requires regular toilet to remove desquamated epithelium and cerumen to prevent infection. The wide meatoplasty performed in this case was an essential step to ensure adequate aeration of the cavity, which facilitates healing and simplifies future cleaning.

In the context of the Indonesian healthcare system and similar developing regions, where late presentation of CSOM is common due to socioeconomic factors and limited access to specialized care, this case serves as an educational paradigm. It emphasizes that dangerous ear disease can manifest in young children without the classic history of chronic ear infections. Future research should focus on the genetic and environmental factors contributing to the high prevalence of severe CSOM in Southeast Asia and evaluate the long-term quality of life outcomes in pediatric patients undergoing CWD versus CWU procedures in this specific demographic.

## 4. Conclusion

This case, a 4-year-old male presenting with atticoantral chronic suppurative otitis media, provides a critical insight into the diagnostic and therapeutic challenges of pediatric otology. We conclude that this patient likely suffered from a ruptured congenital cholesteatoma, a clinical entity where a silently expanding congenital cyst erodes the tympanic membrane, thereby mimicking the presentation of acquired disease. This case serves as a stark reminder to clinicians that the presence of a tympanic perforation does not exclude a congenital etiology, particularly in young children with no prior history of otologic trauma or chronic effusion.

From a diagnostic perspective, this report establishes the indispensability of CT-scan over conventional radiography. The Schuller view failed to capture the severity of the pathology, whereas the CT-scan accurately delineated the soft tissue extent and, crucially, the erosion of the scutum and tegmen tympani. We strongly recommend that CT-scan be considered the standard of care for any pediatric

patient presenting with persistent, antibiotic-resistant otorrhea and attic pathology to ensure accurate surgical planning and risk stratification. Therapeutically, this study validates the Modified Radical Mastoidectomy (Canal Wall Down) with Type II Tympanoplasty as a safe and effective management strategy for extensive pediatric cholesteatoma. While the preservation of anatomy is an ideal, the aggressive nature of pediatric disease and the high risk of recidivism often necessitate a CWD approach to ensure the complete eradication of the cholesteatoma matrix. The successful outcome in this case—a dry, safe ear with a reconstructed sound-conducting mechanism—demonstrates that radical disease clearance can be achieved without sacrificing the potential for functional hearing rehabilitation. Ultimately, early detection, precise imaging, and definitive surgical intervention remain the cornerstones of preventing the devastating complications of this silent destroyer of the temporal bone.

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