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Congenital Cholesteatoma with Mastoid and Temporal Epidural Abscess in a Patient with Congenital Aural Atresia and Grade II Microtia: A Case Report

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Abstract: Congenital cholesteatoma arising in an ear with congenital aural atresia is a rare entity, and diagnosis is difficult because the atretic plate prevents otoscopic assessment of the tympanic membrane. A 17-year-old male with left grade II microtia and congenital aural atresia presented with worsening left-sided headache, fever, severe mixed hearing loss, and recurrent purulent discharge from a left retroauricular fistula. Temporal computed tomography and contrast-enhanced magnetic resonance imaging showed chronic otomastoiditis with an extensive cholesteatoma eroding the temporal bone, destroying the tegmen tympani, and forming a mastoid abscess extending into the left temporal epidural space. A combined procedure was performed, comprising craniotomy with abscess evacuation and pericranial dural grafting by neurosurgery together with subtotal petrosectomy and abdominal fat obliteration by otolaryngology. Intraoperatively the ossicular chain was fused and no continuity was found between the cholesteatoma matrix and external canal skin, supporting a congenital origin. Culture grew Proteus mirabilis. Symptoms resolved initially, but retroauricular discharge recurred five months postoperatively, after which the patient was lost to follow-up. Congenital aural atresia conceals cholesteatoma from clinical examination, so cross-sectional imaging carries the diagnosis. Disease of this extent warrants structured long-term surveillance.

Keywords: Congenital Cholesteatoma, Congenital Aural Atresia, Microtia, Mastoid Abscess, Epidural Abscess

INTRODUCTION

Congenital aural atresia occurs in approximately 1 per 10,000 to 20,000 live births, is usually unilateral, and affects males more often than females (Aggarwal & Rayamajhi, 2020; Kelley & Scholes, 2007). It commonly accompanies microtia, and the two anomalies share a developmental origin in the first and second branchial arches. Despite this frequency, the

genetic basis of non-syndromic microtia and congenital aural atresia remains poorly defined, with forty candidate genes described and no consistent inheritance pattern established in almost half of reported pedigrees (Govindan et al., 2025).

Congenital cholesteatoma is a keratinising squamous epithelial cyst that arises within the temporal bone without a preceding perforation or otologic procedure. The diagnostic standard remains that proposed by Levenson et al. (1986), which requires a psearly white mass medial to an intact tympanic membrane, a normal pars tensa and pars flaccida, and no history of otorrhoea, tympanic membrane perforation, or previous otologic surgery (Levenson et al., 1986; Tabook et al., 2015). Congenital cholesteatoma is often silent until expansion damages adjacent structures, at which point it may cause otorrhoea, otalgia, hearing loss, middle ear effusion, balance disturbance, facial nerve palsy, retroauricular fistula, or symptoms of intracranial extension (Sitompul & Handoko, 2023).

The coexistence of these two conditions is uncommon, and the reported frequency has shifted. Older series and reviews cite a figure of 4 to 7 per cent of atresia cases (Aggarwal & Rayamajhi, 2020; Kelley & Scholes, 2007; Sohal et al., 2013). A systematic review by Chan et al. (2023) pooling 238 patients with congenital aural atresia and 473 with congenital canal stenosis reported a considerably lower incidence of 1.7 per cent in atresia, against 43.0 per cent in stenosis. This distinction matters clinically, because the two anomalies carry very different risks: a narrow but patent canal traps squamous debris and generates external canal cholesteatoma, whereas a bony atretic plate does not (Jamarun et al., 2025; Mazita et al., 2011).

The diagnostic problem in an atretic ear is structural. None of the first three Levenson criteria can be assessed, because there is no visible tympanic membrane to inspect and no canal through which otorrhoea could ever have occurred. Diagnosis therefore rests on cross-sectional imaging and on findings at surgery rather than on clinical examination. Patients consequently present late, and several published cases describe advanced disease at first diagnosis, including acute mastoiditis with extension into the infratemporal fossa, mastoid abscess as the sole presenting sign, and brain abscess complicating a hidden canal cholesteatoma (Abdel-Aziz, 2012; Jamarun et al., 2025; Tabook et al., 2015). Delayed recognition allows the disease to reach structures where surgical clearance becomes hazardous and where complication rates rise (Popescu et al., 2024).

Intracranial extension in this setting is described only sporadically, and no standard management pathway exists for the combination of extensive congenital cholesteatoma, mastoid abscess, and epidural collection in an atretic ear. Reported cases have generally been managed by mastoidectomy alone, and the role of subtotal petrosectomy with concurrent neurosurgical drainage has not been examined in this specific anatomical context.

This report describes a 17-year-old male with congenital aural atresia and grade II microtia in whom congenital cholesteatoma presented with mastoid abscess and temporal epidural abscess, managed by simultaneous craniotomy and subtotal petrosectomy. The report addresses three questions: how a congenital origin can be defended when Levenson's criteria cannot be applied, what surgical strategy is appropriate when the disease has already breached the tegmen, and how the risk of recurrence should be monitored once the mastoid has been obliterated.

METHOD

Study design. This is a descriptive, retrospective, single-patient case report conducted at the Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Udayana University / Prof. Dr. I.G.N.G. Ngoerah General Hospital, Denpasar, Bali, a tertiary referral centre for the eastern Indonesian region.

Data sources. Clinical data were extracted retrospectively from the hospital medical record covering the period from the first outpatient visit in June 2023 to the last recorded

contact in November 2024. Extracted items comprised history and physical examination, pure tone audiometry and tympanometry, laboratory results, non-contrast high-resolution temporal bone computed tomography, contrast-enhanced magnetic resonance imaging, operative notes from both the otology and neurosurgery teams, histopathology, microbiological culture with antibiotic sensitivity, and outpatient follow-up records.

Definitions and instruments. Auricular deformity was graded using the Marx classification. Hearing thresholds were reported as pure tone averages for air and bone conduction, with an air-bone gap of more than 10 dB and a bone conduction threshold above 25 dB defining a mixed loss. A congenital origin of the cholesteatoma was assessed against the criteria of Levenson et al. (1986), supplemented by imaging and intraoperative findings where those criteria could not be applied.

Reporting standard. The report follows the CARE (CAse REport) guideline and its 13-item checklist (Gagnier et al., 2014). A completed checklist is available from the corresponding author. Clinical events are presented chronologically and summarised in Table 1.

Ethics and consent. Written informed consent for the publication of clinical data and accompanying images was obtained from the patient and is held by the corresponding author. Under institutional policy, review by the research ethics committee is not required for a single de-identified case report. All direct identifiers were removed from the text and from the figures before submission. No statistical analysis was performed.

RESULTS AND DISCUSSION

Case presentation

A 17-year-old male was referred to the outpatient otorhinolaryngology clinic with discharge from behind the left ear that had begun in April 2023 (Figure 1). The discharge was yellowish and thick, resembling pus. It was accompanied by left-sided headache and high fever. The headache was initially intermittent, but during the two weeks preceding presentation it became severe and persistent. He reported that hearing deteriorated further after the headache began. There were no complaints of spinning sensation, dizziness, tinnitus, seizures, weakness of the upper or lower limbs, or reduced consciousness. He also denied any sensation of heaviness on one side of the face.



Figure 1. Clinical photograph of the patient showing the left retroauricular fistula

He reported that since childhood the left ear had felt full, that the left ear canal was smaller than the right, and that hearing on the left was poorer than on the right. A small nodule had been noticed behind the left auricle one year earlier and had enlarged progressively. He had been treated by an otorhinolaryngologist and had undergone evacuation of pus on three occasions in the outpatient clinic, without resolution of symptoms. He denied any history of recurrent rhinitis, nasal obstruction, morning sneezing, diabetes mellitus, hypertension, cardiac disease, or other metabolic disease. He acknowledged a history of cigarette smoking and alcohol consumption.

On physical examination the patient appeared moderately ill and was fully conscious (Glasgow Coma Scale E4V5M6), with blood pressure 110/80 mmHg, pulse 88 beats per minute, respiratory rate 21 breaths per minute, and axillary temperature 36.7 °C. He was afebrile and free of headache at the time of examination. Examination of the nose and throat was within normal limits. The right ear showed no abnormality, with no tragus tenderness, a wide canal free of discharge, and an intact, non-hyperaemic tympanic membrane (Figure 2A). On the left, the auricle was smaller than the right and consistent with Marx grade II microtia. There was no tragus tenderness. The external auditory canal was narrow and ended blindly at approximately 1 cm depth against a bony atretic plate, so the tympanic membrane could not be visualised (Figure 2B). A fistula was present in the left retroauricular area, discharging pus, with no oedema and minimal surrounding tenderness.



Figure 2. Otoendoscopic appearance of the right ear showing an intact tympanic membrane (A) and of the left ear showing a narrow, blind-ending external auditory canal terminating in a bony atretic plate, with the tympanic membrane not visualised (B)

Pure tone audiometry showed normal hearing in the right ear and severe mixed hearing loss in the left ear, with an air conduction pure tone average of 80 dB, a bone conduction average of 43 dB, and an air-bone gap of approximately 37 dB (Figure 3). Tympanometry showed a type A curve on the right and a type B curve on the left (Figure 4).

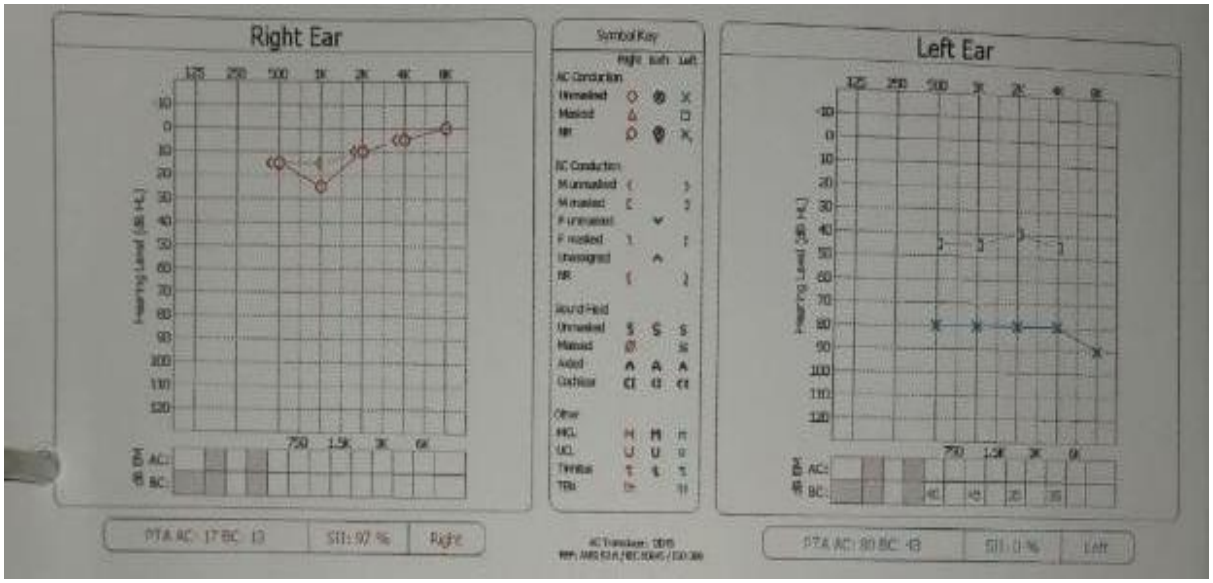


Figure 3. Pure tone audiogram showing normal hearing on the right and severe mixed hearing loss on the left

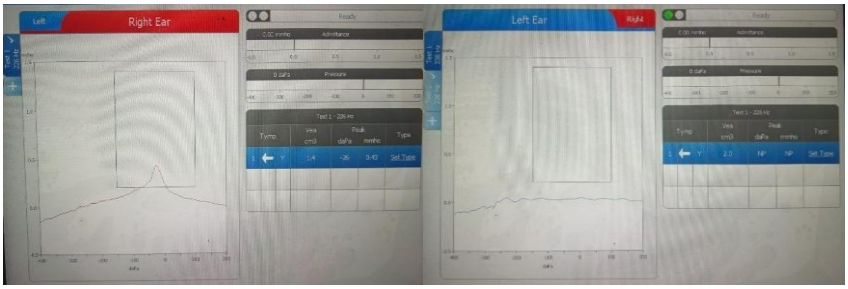


Figure 4. Tympanogram showing a type A curve on the right ear and a type B curve on the left ear

Laboratory investigation showed no elevation of the white blood cell count, but C-reactive protein was markedly raised at 114 mg/L against a reference range below 5 mg/L. Non-contrast high-resolution computed tomography of the temporal bone demonstrated left chronic otomastoiditis with cholesteatoma occupying the left Prussak's space and extending into the epitympanum, mesotympanum, hypotympanum, aditus ad antrum, and left mastoid air cells. The lesion eroded the squamous and petrous portions of the left temporal bone, destroyed the left tegmen tympani, reached the medial aspect of the atretic left external auditory canal, and was accompanied by an abscess component extending into the epidural space of the left temporal region (Figure 5).

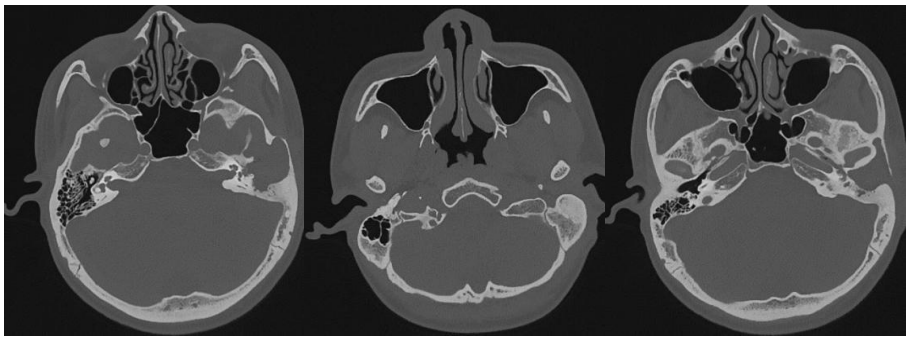


Figure 5. Non-contrast high-resolution computed tomography of the temporal bone

Contrast-enhanced magnetic resonance imaging revealed a thick-walled supratentorial extra-axial cystic lesion in the left temporal region extending to the left mastoid, consistent with an epidural abscess (Figure 6). The working diagnosis was left chronic suppurative otitis media of the atticoantral type, left mastoid abscess with intracranial extension in the form of a temporal epidural abscess, left congenital aural atresia, left severe mixed hearing loss, and left Marx grade II microtia. Left subtotal petrosectomy was planned. The patient was referred to neurosurgery for a joint operative plan and to anaesthesia, where he was classified as ASA III on the basis of active intracranial infection.

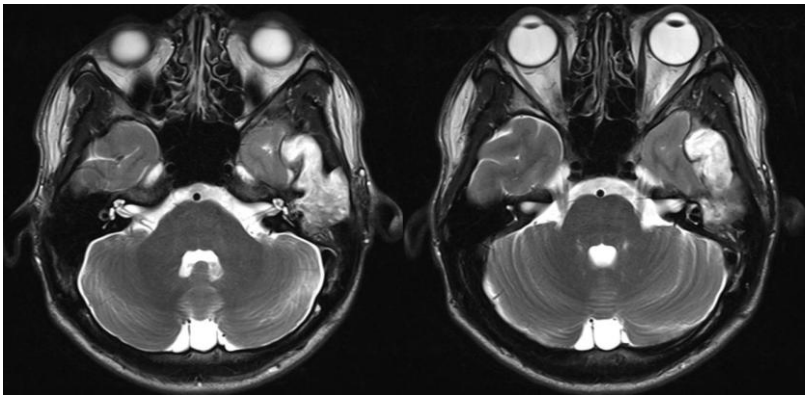


Figure 6. Contrast-enhanced magnetic resonance imaging showing the left temporal epidural abscess

A joint operation with neurosurgery was performed on 4 June 2024. The neurosurgical team performed a craniotomy with evacuation of the epidural abscess and placement of a pericranial graft over the dural defect at the tegmen superior to the antrum. The otology team performed a left subtotal petrosectomy. Operative findings were congenital cholesteatoma filling the tympanic cavity, fusion of the tympanic cavity contents including the ossicular chain, an exposed sigmoid sinus covered by granulation tissue, and atresia of the external acoustic canal, which was naturally closed. No continuity was identified between the cholesteatoma matrix and the skin of the external auditory canal. The mastoid cavity was obliterated with an abdominal fat graft (Figure 7).



Figure 7. Intraoperative finding of cholesteatoma occupying the tympanic cavity

Histopathological examination of specimens taken from the mastoid cavity and the left temporal base confirmed cholesteatoma. Culture of the wound bed swab and of the pus grew Proteus mirabilis, a gram-negative organism of the Enterobacteriaceae group.

Routine review was carried out to two months postoperatively. At the outpatient visit on 15 August 2024 the incision had healed well, with no discharge from the ear canal or the incision site. Two-weekly evaluation was advised but was not attended. The patient returned to the clinic on 8 November 2024, five months after surgery, again complaining of discharge from behind the ear. Revision imaging and a second-look procedure were advised. He did not attend any further appointment and was lost to follow-up, as he resides in Banyuwangi, East Java, and declined to return to Bali for continued care. Postoperative audiometry and postoperative magnetic resonance imaging were therefore never obtained.

Table 1. Chronology of clinical events, interventions, and outcomes

Time point	Event	Findings and action
April 2023	Onset of symptoms	Purulent discharge from left retroauricular region, left headache, fever
April 2023 to May 2024	Three outpatient drainage procedures	Evacuation of retroauricular pus by an otorhinolaryngologist, no resolution
May 2024, two weeks before referral	Symptom escalation	Headache became severe and persistent, hearing deteriorated further
June 2023 onward, first tertiary visit	Assessment	Marx grade II microtia, aural atresia with bony plate at 1 cm, retroauricular fistula with pus, CRP 114 mg/L
Same admission	Audiology	Right normal, left severe mixed loss (AC 80 dB, BC 43 dB), tympanogram type B on the left
Same admission	Imaging	HRCT: cholesteatoma filling middle ear and mastoid, tegmen tympani destroyed. MRI: left temporal epidural abscess

4 June 2024	Combined surgery	Craniotomy, abscess evacuation and pericranial dural graft, plus left subtotal petrosectomy with fat obliteration
Postoperative	Pathology and microbiology	Cholesteatoma confirmed, Proteus mirabilis isolated
15 August 2024, 2 months	Follow-up	Wound healed, no discharge, headache resolved
8 November 2024, 5 months	Recurrence	Retroauricular discharge recurred, revision imaging and second look advised
After November 2024	Outcome	Lost to follow-up, patient resides in Banyuwangi and declined further travel

Discussion

Cholesteatoma is an epithelial cyst containing keratinised desquamated epithelium, found in the ear canal, tympanic cavity, and mastoid. Distinguishing a congenital from an acquired lesion determines both the operative plan and the counselling given to the patient. In an ear with a bony atretic plate this distinction cannot be made by examination, because the first three Levenson criteria all depend on inspecting a tympanic membrane that is not accessible, and because otorrhoea through the canal is anatomically impossible. Diagnosis therefore has to be constructed from imaging, operative findings, and history.

Four observations supported a congenital origin in this patient. First, computed tomography and inspection at surgery confirmed a complete bony atretic plate, leaving no epithelial route from the external canal into the middle ear. Second, no continuity was found between the cholesteatoma matrix and the skin of the external auditory canal at operation, which is the single most direct argument against an external canal cholesteatoma that had migrated medially. Third, the ossicular chain was fused to the contents of the tympanic cavity, indicating a middle ear developmental anomaly of the same embryological period. Fourth, the three previous procedures were subcutaneous drainages of a retroauricular abscess that did not expose the mastoid cavity, so they do not constitute otologic surgery in the sense intended by Levenson's fourth criterion.

The principal competing diagnosis is external auditory canal cholesteatoma, and the distinction between atresia and stenosis is what settles it. Chan et al. (2023) found cholesteatoma in 1.7 per cent of ears with congenital aural atresia but in 43.0 per cent of ears with congenital canal stenosis, and Jamarun et al. (2025) described an extensive canal cholesteatoma complicated by brain abscess in a child whose canal measured 4 mm or less. A narrow but patent canal accumulates squamous debris, whereas a bony plate does not. Because this patient had complete atresia rather than stenosis, and because the matrix was not continuous with canal skin, an acquired canal cholesteatoma is unlikely. Mazita et al. (2011), reviewing a mixed cohort of congenital canal anomalies, found cholesteatoma in 43.9 per cent of cases, of which two presented with postauricular discharge from a mastoid abscess, the same presentation seen here.

The audiometric findings deserve comment because they were not purely conductive. The conductive component is explained by the atretic plate, the fused ossicular chain, and the mass of keratin debris occupying the tympanic cavity. The bone conduction threshold of 43 dB, however, indicates a sensorineural component that requires a separate explanation. Two mechanisms are plausible. Coexisting inner ear dysplasia is common in this population. Park et al. (2019) found inner ear anomalies in 23.0 per cent of 61 patients with unilateral congenital aural atresia and microtia, most often lateral semicircular canal dysplasia at 16.4 per cent, and more frequently in ears with lower Jahrsdoerfer scores. Alternatively, the sensorineural component may be secondary to serous labyrinthitis or labyrinthine erosion from adjacent suppuration. High-resolution computed tomography did not report labyrinthine erosion, and dedicated inner ear assessment was not undertaken, so the two mechanisms could not be separated in this patient. Abell et al. (2025) showed in 186 children that hearing in aural atresia

is worse than in canal stenosis and does not improve over time, which supports a conservative prognosis for the affected ear irrespective of mechanism.

The surgery was performed jointly with neurosurgery because of the intracranial epidural abscess. The neurosurgical team carried out a craniotomy with evacuation and placed a pericranial graft as a dural substitute in the tegmen area above the antrum, which indicates that the dura in contact with the abscess was infected and had to be excised and replaced. The otology team performed a left subtotal petrosectomy with obliteration using fat harvested from the abdominal subcutaneous tissue. Three considerations favoured subtotal petrosectomy over a canal wall down mastoidectomy. The disease filled the entire mastoid and had eroded the tegmen antri, so exposure had to extend beyond what a standard mastoidectomy provides. The external canal was already closed by the atretic plate, which meant that blind sac closure, ordinarily an additional step, was anatomically pre-existing. There was no serviceable hearing in the affected ear to preserve. Grinblat et al. (2020), reporting 107 subtotal petrosectomies with blind sac closure and autologous fat obliteration at a mean follow-up of 7.1 years, recorded no cases of recurrent or entrapped cholesteatoma, which supports the technique in ears with extensive disease. As in the case reported by Aggarwal and Rayamajhi (2020), the primary aim of surgery was clearance of disease rather than hearing rehabilitation.

Postoperatively the patient improved, with closure of the surgical wound, resolution of headache, and no discharge from the incision. Discharge nevertheless recurred five months after surgery. This outcome was foreseeable from the extent of the original disease. Volgger et al. (2020), reviewing 110 revision procedures, identified extension into the tympanic sinus or towards the mastoid as a predilection factor for residual disease. Handoko et al. (2026), synthesising 23 systematic reviews covering 12,819 patients, reported that recurrence rises from 7 per cent at 12 months to 39 per cent at 15 years, that mastoid obliteration reduces recidivism with odds ratios between 0.33 and 0.45, and that planned second-look surgery is protective. Large cholesteatomas may leave microscopic residual matrix that is not visible at the primary operation, and obliteration with fat, while reducing recurrence overall, conceals any residual disease from direct inspection.

That concealment is precisely why imaging surveillance matters after an obliterative procedure. Non-echo planar diffusion-weighted magnetic resonance imaging is the appropriate modality: in a meta-analysis of 32 diagnostic studies, Amoodi et al. (2022) reported pooled sensitivity of 92.2 per cent and specificity of 91.7 per cent for residual or recurrent cholesteatoma, and concluded that it is a reasonable non-invasive alternative to second-look surgery. Popescu et al. (2024), reviewing 580 patients over two decades, likewise placed diffusion-weighted imaging at the centre of postoperative monitoring. In this patient, the recurrence of discharge at five months should have prompted such a study, but it was not obtained.

The reason it was not obtained is worth stating plainly rather than treating as an incidental loss. The patient lives in Banyuwangi, East Java, and declined to travel to Bali again. The journey involves a ferry crossing and a full day of travel for each appointment. In an archipelagic health system, a surveillance protocol that assumes repeated attendance at the operating centre will fail for a predictable subgroup of patients. Two practical measures follow: counselling about the recurrence risk and the surveillance schedule should occur before discharge rather than at the point of recurrence, and shared care should be arranged with a centre nearer the patient's residence, with imaging performed locally and reviewed remotely.

The presentation described here shares features with several published cases. Sitompul and Handoko (2023) reported congenital cholesteatoma in unilateral congenital aural atresia with grade I microtia, and Aggarwal and Rayamajhi (2020) reported a 16-year-old with congenital aural atresia, grade III microtia, facial palsy, and postauricular swelling, in whom the tympanic membrane likewise could not be assessed. Both emphasised that congenital

cholesteatoma should be suspected in this anatomical setting and that preoperative temporal bone imaging is needed to define the extent of disease before deciding on treatment. Abdel-Aziz (2012) described extension into the infratemporal fossa with acute mastoiditis, Tabook et al. (2015) described mastoid abscess as the only presenting sign, and Haritha et al. (2025) described coexistent congenital cholesteatoma with a dehiscent facial nerve. The present case extends this group by adding intracranial epidural abscess, which none of these reports encountered.

The isolation of Proteus mirabilis is consistent with the microbiological pattern of chronic suppurative otitis media. Mohamed Ali et al. (2022), culturing middle ear discharge from 225 patients, found gram-negative organisms in 77.3 per cent of monomicrobial isolates, with Proteus species among the more frequent. The culture in this patient was taken from a wound bed swab and from pus, and surface swabs may reflect colonisation rather than the organisms within the cholesteatoma sac, so the result should be interpreted with that limitation in mind.

Several limitations apply. This is a single case, so no inference about frequency or about the comparative merit of surgical techniques can be drawn from it. A Jahrsdoerfer score was not calculated, no genetic evaluation was performed, and family history of cholesteatoma was not elicited, although Bonnard et al. (2023) have shown that first-degree relatives of surgically treated patients carry an elevated risk. Postoperative audiometry, non-echo planar diffusion-weighted imaging, and any assessment beyond five months are absent because the patient did not return, and the true long-term outcome is therefore unknown.

CONCLUSIONS

A 17-year-old male with congenital aural atresia and grade II microtia developed congenital cholesteatoma complicated by mastoid abscess and temporal epidural abscess, treated by simultaneous craniotomy with dural grafting and subtotal petrosectomy with fat obliteration. The case supports three points. First, congenital aural atresia removes the anatomical basis for Levenson's criteria, so a congenital origin must be argued from imaging and from operative findings, in this instance from a complete bony atretic plate, absence of continuity between matrix and canal skin, and a fused ossicular chain. Second, when disease has filled the mastoid and breached the tegmen, subtotal petrosectomy with obliteration provides the exposure required for clearance, and the pre-existing atretic canal makes blind sac closure anatomically straightforward. Third, obliteration hides residual disease from inspection, so scheduled non-echo planar diffusion-weighted magnetic resonance imaging should be planned from the outset rather than arranged after symptoms recur. The recurrence and subsequent loss to follow-up in this patient illustrate that a surveillance plan requiring repeated travel to a distant referral centre is unlikely to be completed, and that shared care closer to the patient's residence should be arranged before discharge.

Declarations

Consent for publication

Written informed consent for publication of this case report and the accompanying figures was obtained from the patient and signed in hard copy. All direct identifiers have been removed from the text and from the figures.

Ethics approval

Under institutional policy, review by the research ethics committee is not required for a single de-identified case report, and no approval or exemption letter was issued for this report. The work was conducted in accordance with the Declaration of Helsinki.

Conflict of interest. The authors declare no conflict of interest.

Funding. This case report received no funding from any source.

Author contributions

Both authors contributed to patient management, data collection, drafting, and critical revision of the manuscript. Both authors read and approved the final version.

Data availability

The de-identified data supporting this report are available from the corresponding author on reasonable request.

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