The Journal of International Advanced Otology

J Int Adv Otol 2022; 18(4): 315-319 • DOI: 10.5152/iao.2022.21498

Original Article

A Unified Classification of Middle Ear and Petrous Bone Congenital Cholesteatomas

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Cite this article as: Honnurappa V, Mahajan N, Kumar Vijayendra V, Redleaf M. A unified classification of middle ear and petrous bone congenital cholesteatomas. *J Int Adv Otol.* 2022;18(4):315-319.

BACKGROUND: The aim of this study was to classify congenital cholesteatoma along an entire spectrum of involvement ranging from the middle ear to petrous apex.

METHODS: A total of 131 patients (85 adults and 46 children) underwent operations for congenital cholesteatoma over the duration of 27 years.

RESULTS: For most cases, middle ear mucosa was normal, the first ossicle eroded by the mass was the stapes, and the mastoid air cell system was well-pneumatized on intraoperative and radiographic views. Totally 34% of patients presented with facial nerve weakness and 45% of these cholesteatomas arose from the supralabyrinthine area (32.8%) and from the petrous apex (12.2%).

CONCLUSION: In this unified classification system, the otologist sees congenital cholesteatoma as a continuum, with facial nerve involvement and anacusis as part of the picture. This system of congenital cholesteatoma accommodates the supralabyrinthine and petrous bone locations of the disease.

KEYWORDS: Congenital cholesteatoma, facial nerve, petrous apex, middle ear mucosa, mastoid air cell

INTRODUCTION

Congenital cholesteatomas are found behind an intact tympanic membrane and are epidermal cysts arising from embryologic remnants of keratinizing squamous epithelium which persists in the temporal bone throughout gestation to birth.[1,2] The classic features of congenital cholesteatoma are no prior history of otorrhea or surgical procedure or tympanic membrane perforation, normal pars tensa and pars flaccida, and a white mass behind the intact membrane.[1] Some reports state that middle ear congenital cholesteatomas arise from the anterior-superior quadrant of the mesotympanum.[1-4] However, there is substantial contradictory evidence that these arise predominantly from the posterior quadrant.[5-7]

Classification systems for congenital cholesteatoma have focused on the distribution of cholesteatoma within the mesotympanum, its extension through the additus ad antrum, with possible erosion of the ossicles.[8,9] These classifications neither include supralabyrinthine extensions nor facial nerve involvement. Petrous bone cholesteatoma is described as a separate entity.[2,10]

Our experience at a tertiary otology referral center has shown that when history, physical exam, radiography, and intraoperative findings are considered together, congenital cholesteatoma can be considered as a continuum from the middle ear up to petrous apex involvement. In this study, we therefore present a unified classification system of congenital cholesteatoma. This analysis accommodates the supralabyrinthine and petrous bone instances of the disease and the frequent involvement of the facial nerve in patients who travel for medical evaluation.

MATERIALS AND METHODS

This study was reviewed by the ethics committee of Vijaya E.N.T. Care Centre (Approval no: 2020-05) and received approval with protocol number #2020-05. All subjects in this study underwent pre-operative counseling and gave informed consent to the medical and surgical care which they elected to follow.

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Received: August 15, 2021 • **Accepted:** October 22, 2021
Available online at www.advancedotology.org

Identification Criteria for Congenital Cholesteatoma

The identification of congenital cholesteatoma is based on clinical examination, radiological findings, and operative findings. Presenting symptoms range from none at all to an itching sensation, a blocked sensation, a hearing loss, and facial nerve weakness in 34% of patients. Physical exam finds a normal tympanic membrane or a bulging cholesteatoma sac that has lateralized the malleus (Figure 1). Not occasionally there is a facial nerve palsy. Myringotomy can be useful to confirm the diagnosis in cases associated with otitis media with effusion which complicates the diagnosis. High-resolution computed tomography (CT) (Figure 2) of the temporal bone can show the extent of involvement, ossicular status, and surrounding bone involvement. On magnetic resonance imaging (MRI), lesion appears to be hypo- or iso-intense on T1, does not enhance with contrast, and is hyperintense on T2, and diffusion restriction is present on non-echo planer diffusion-weighted sequence (Figure 3). Surgical findings are normal pars tensa and pars flaccida, lateralized tympanic membrane (TM) and chorda tympani and malleus handle, normal middle ear mucosa and promontory tympanic plexus, and well-pneumatized air cell system (Figure 4).

A Unified Congenital Cholesteatoma Classification System

Type 1: Small intact sac confined to middle ear but not involving ossicles and mastoid

Type 2: Lesion involving the whole middle ear without extending into mastoid with ossicular destruction—2a: without facial nerve palsy, 2b: with facial nerve palsy

Type 3: Lesion extending into mastoid—3a: without facial nerve palsy, 3b: with facial nerve palsy

Type 4: Lesion arising from the supralabyrinthine area and extending into middle ear and internal auditory meatus—4a: without facial nerve palsy, 4b: with facial nerve palsy

Type 5: Lesion arising from the petrous apex with anacusis—5a: without facial nerve palsy, 5b: with facial nerve palsy.

RESULTS AND ANALYSIS

Subjects

From 1992 to 2019, 131 patients underwent operations for congenital cholesteatoma (Table 1). These subjects presented serially to our center and were not part of a multi-institution or government registry. Of them, 53 were males and 78 females and 85 were adults and 46 were children below the age of 15 years. Forty-five patients of the 131 (34%) had facial nerve weakness, 10 of these were children. The youngest patient with facial nerve involvement was 7 months old and the oldest was 54 years old. Forty-three underwent nerve decompression — which involved decompression of the facial nerve bony canal proximal and distal to the site of cholesteatoma involvement. Another 3 patients (age: 7 months, 12 months, and 54 years) had nerve grafting because of complete erosion of the facial nerve in the horizontal segment and second genus. All 147 presenting patients were offered operations and 16 refused.

Distribution of Cholesteatoma Sites

In this series, the location of congenital cholesteatomas could be determined from the combined information obtained from physical examination, radiological evaluation, and intraoperative findings (Table 2). Classification types 2b, 3b, 4b, and 5b tally those 45/131 patients (34%) who presented with facial weakness (of note, some of the patients with supralabyrinthine/petrous apex involvement had previously been misdiagnosed as having "Bell's Palsy" because of facial weakness without an obvious tumor in the middle ear space). Classification types 4a, 4b, 5a, and 5b tally those 59/131 patients (45%) whose cholesteatomas arise from the supralabyrinthine area and extend into the middle ear and internal auditory meatus (type 4) (43/131) or arise from the petrous apex (type 5) (16/131).

Intraoperative view of the middle ear found the mucosa to be normal in most of the cases and that the first ossicle to be eroded by the mass was the stapes in most of the cases. The tympanic membrane and the chorda tympani were often lateralized. Intraoperative and radiographic views of the mastoid air cell system were found to be well-pneumatization almost in all cases.

DISCUSSION

Our unified classification of congenital cholesteatoma is based on 131 patients operated on over the duration of 27 years. The tympanic membrane was intact, the middle ear mucosa was normal, the stapes was the first ossicle to be eroded by the disease, and the mastoids were well-pneumatized. Many of these had epithelial rests located in the supralabyrinthine temporal bone or in the petrous apex (Figure 2b-d). Both adults and children had facial nerve involvement upon presentation.

Derlaki et al[1] reviewed the original reports of congenital cholesteatoma and reported their own 10 cases. They reported the identification of the origin of congenital cholesteatoma as developmental rests which failed to involute. Their findings agreed with this series that the stapes is the first ossicle to become eroded. Their classification system stops short at the extension of the middle cholesteatoma through the aditus ad antrum. Cawthorne[10] reviewed the evolving understanding of congenital cholesteatoma in 1963 and presented a series of 13 cases. These authors support the finding that the tympanic membrane is intact in congenital cholesteatoma, that petrous involvement can affect facial nerve and auditory function, and that the mastoid is well-pneumatized in these cases. Levenson et al[2] also reviewed the issue of the origins of the retained epithelial tissue and reported 20 cases of congenital cholesteatoma. One-fourth of their patients had otologic malformations. The middle ear mucosa of their series was normal in 14/20 patients and 17/20 had pneumatized

MAIN POINTS

- This study presents a unified classification scheme of congenital cholesteatoma location from middle ear cleft to petrous apex.
- In this system, congenital cholesteatoma is a continuum with anacusis and facial nerve weakness accompanying the more medial sites.
- One clinical takeaway is that facial nerve weakness with a normal tympanic membrane and middle ear space should encourage the search for petrous apex involvement.

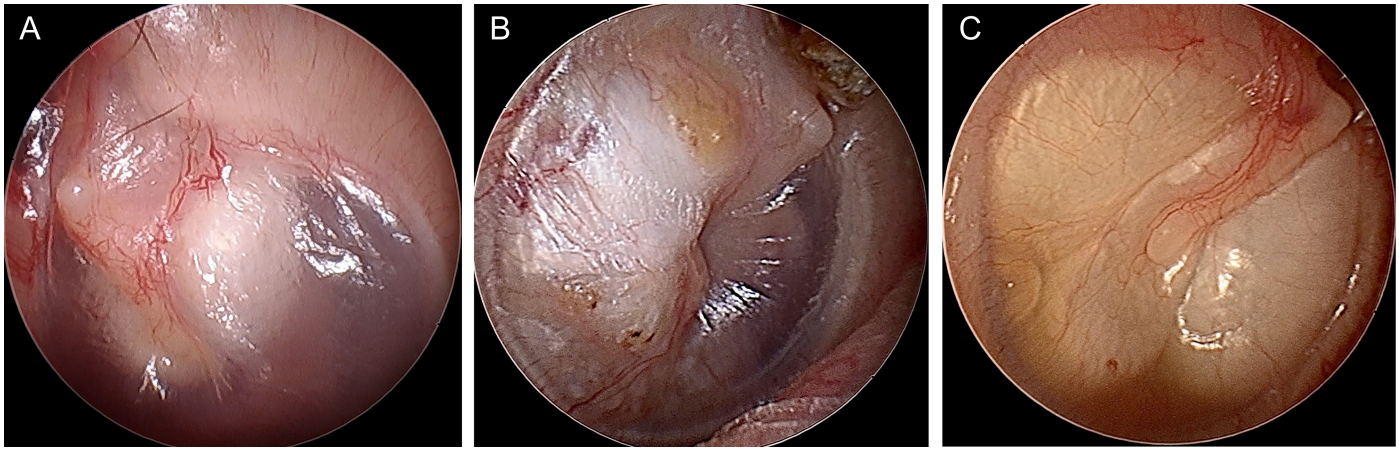

Figure 1. a-c. (a) White mass behind an intact TM (left ear), (b) white mass in posterior part pushing TM laterally (right ear), and (c) white mass filling entire middle ear (right ear).

mastoids. The presence of otitis media complicated the diagnosis in some cases.

The presentation of supralabyrinthine and petrous apex congenital cholesteatomas can be misleading, even those isolated sites may be more common than previously suspected (45% in our study); the middle ear is clear on examination and the patient may have unexplained facial palsy misdiagnosed as "Bell's." Our study found facial nerve weakness or palsy in 34% of subjects. Others have found facial nerve involvement in supralabyrinthine/petrous apex congenital cholesteatomas ranging from 51% to 95%.[11,12] Involvement of the otic capsule with anacusis was also common both for these studies and ours — but this may be a result of patients' self-selection since they would only seek medical evaluation if they had a discernible

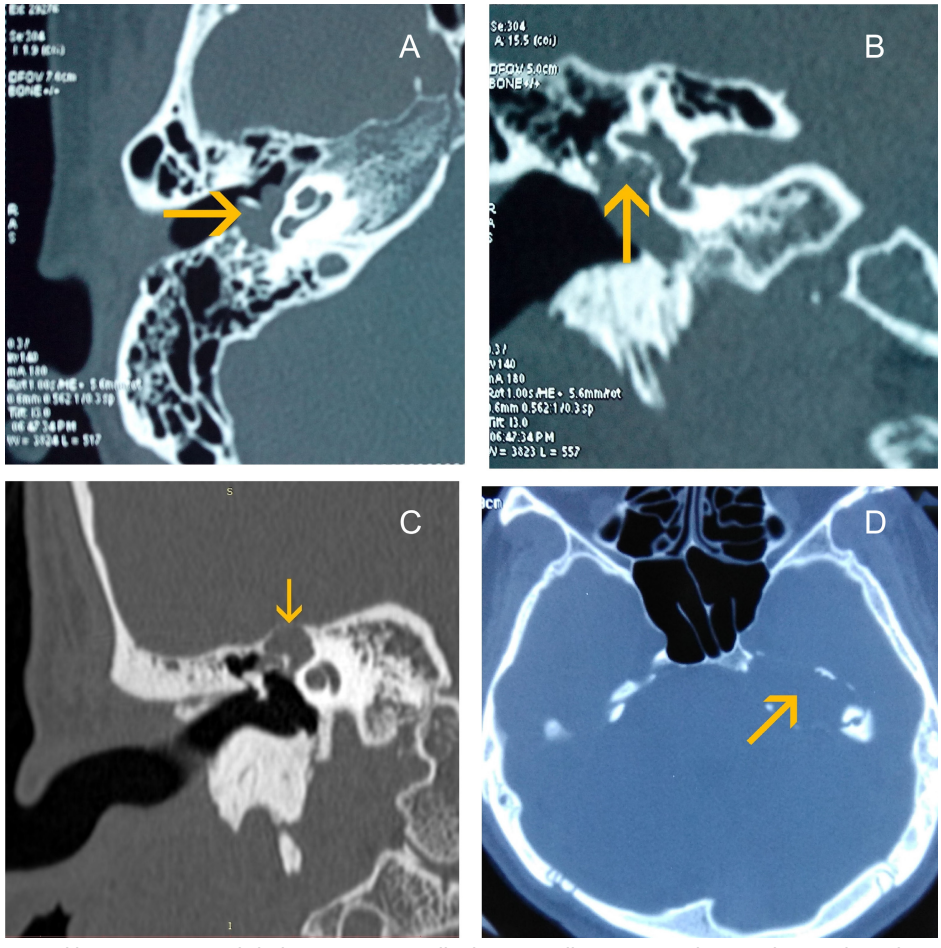

Figure 2. a-d. CT of the temporal bone in congenital cholesteatoma typically shows a well-pneumatized mastoid — (a) shows lateralization of the TM, (b) can show stapes erosion, (c) can involve the supralabyrinthine area including the geniculate ganglion, and (d) can involve the petrous apex. CT, computed tomography.

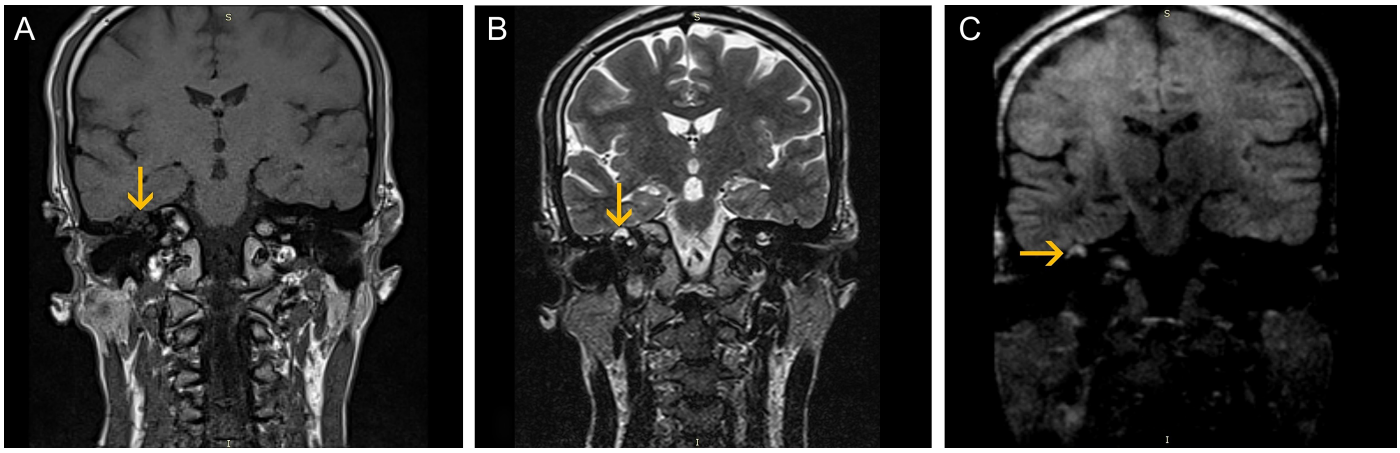

Figure 3. a-c. Non-echo planar MRI shows (a) hypointense mass on T1(arrow), (b) hyperintense mass on T2 (arrow), and (c) diffusion restriction of mass (arrow) on diffusion-weighted images. MRI, magnetic resonance imaging.

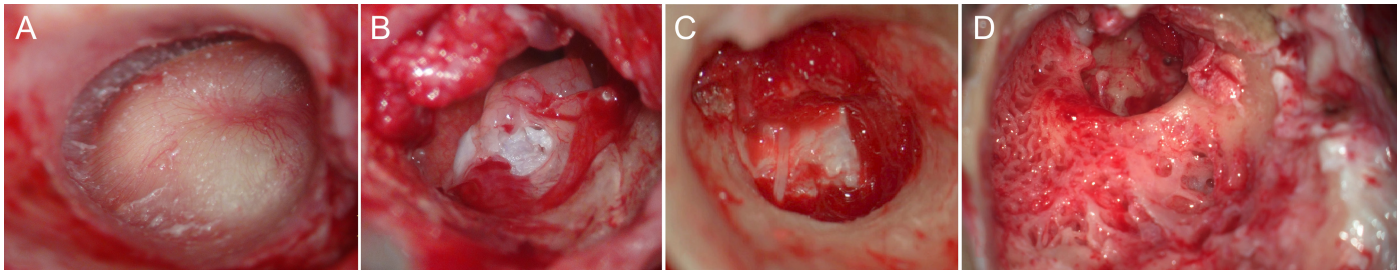

Figure 4. a-d. Intraoperative anatomical findings in congenital cholesteatoma typically are (a) an intact TM (left ear), (b) lateralized malleus handle (left ear), (c) right chorda tympani (right ear), and (d) a well-pneumatized mastoid air cell system (right ear).

symptom. An additional note should be made to clarify our reluctance to use the term "epidermoid" in our study: We are focused here on congenital cholesteatomas of the temporal bone, while the often confusing term "epidermoid" should refer to intra-dural masses, mostly in the cerebellopontine angle.

Our study has some limitations. We wanted to include only patients with as much descriptive information as possible, which dictated that only those who underwent operations are tallied. Therefore, the 16 patients who refused operation were not tallied here. In addition, the 34% with facial involvement may be overrepresented since these patients are more likely to be motivated to undergo operation for the chance to restore some facial function.

Conceptually, the petrous bone cholesteatomas have arisen from the same process as the middle ear cholesteatomas and are included here as part of a classification continuum. This is not to say that supralabyrinthine and petrous apex cholesteatomas are physically continuous or contiguous with the middle ear space. Some specialists may object that a unified classification system is not appropriate because they feel that the middle ear and the supralabyrinthine cholesteatomas should be considered differently, even though both are congenital.

Otologists/neurotologists may have additional concerns with this classification. It consolidates congenital cholesteatoma locations — from any location or size in the middle ear — to the petrous apex.

Table 1. Distribution of 131 Congenital Cholesteatomas by Age

Age Group (years)	No. of Cases
1-10	18
11-20	33
21-30	38
30-40	23
40-50	15
50-60	04
Total	131

Table 2. Distribution of 131 Congenital Cholesteatomas by Classification Type

Unified Congenital Cholesteatoma Classification Type	No. of Cases
1	11
2a	21
2b	07
3a	19
3b	14
4a	29
4b	14
5a	06
5b	10
Total	131

However, patients are necessarily excluded from the analysis, who did not have symptoms which caused them to seek evaluation or did not agree to the imaging and operations which are used to confirm the cholesteatoma site. We simply cannot know where the tumors were in patients who did not come to us or did not pursue the evaluation. The corollary of this issue is that only the symptomatic patients would come to us, especially in the cases of petrous apex cholesteatomas, which are discovered only when the otic capsule is violated. Hence the high proportion of patients presented with facial nerve dysfunction or anacusis. These symptoms of labyrinthine erosion, so common in our series, draw a distinction with the classic scheme of Derlacki et al[1]. In sum, for these reasons, neither our patient roster is complete nor could the patient roster in previous studies be.

The above limitations notwithstanding this study's strengths are the numbers of patients, the multi-modality and detailed evaluations of the cholesteatomas' locations, and the years and depth of experience in characterizing these cholesteatomas.

CONCLUSION

In this study, we present a unified classification scheme of congenital cholesteatoma. The numbers of patients and the modalities of evaluations of each patient definitively establish the locations of their masses. This classification system encourages the otologist to see congenital cholesteatoma as a continuum and see facial nerve involvement and anacusis as part of the picture. In addition, this series indicates that any facial nerve weakness with a normal tympanic membrane and middle ear space should undergo radiological investigations in the form of high resolution computed tomography (HRCT) and MRI sequences including non-echo-planer diffusion-weighted images to look for petrous apex involvement.

Ethics Committee Approval: Ethical committee approval was received from the Ethics Committee of Vijaya E.N.T. Care Centre (Approval no: 2020-05).

Informed Consent: Written informed consent was obtained from all participants who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – V.H.; Design – V.H.; Supervision – M.R.; Materials – N.M., V.K.V.; Data Collection and/or Processing – N.M., V.K.V.; Analysis and/or Interpretation – V.H., N.M.; Literature Review – N.M., M.R.; Writing Manuscript – V.H., N.M., V.K.V., M.R.

Acknowledgments: The authors would like to thank Dr. Agadurappa Mahadevaiah for the guidance and mentorship.

Declaration of Interests: The authors have no conflict of interest to declare.

Funding: The authors declared that this study has received no financial support.

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