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The difference in congenital cholesteatoma CT findings based on the type of mass



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KEYWORDS

Congenital cholesteatoma;
Computed tomography;
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Closed type;
Open type

Abstract

Objective: A retrospective assessment of differences in congenital cholesteatoma CT findings with a focus on type of cholesteatoma mass.

Materials and methods: The medical records and CT images of 14 patients with congenital cholesteatomas in the middle ear who underwent surgery at our institution between January 2009 and July 2014 were reviewed. Cholesteatomas were classified as closed type, open type, or mixed type based on intraoperative findings. The CT findings including cholesteatoma size, location, and shape were retrospectively reviewed.

Results: Eight patients had closed type cholesteatomas, four had mixed type, and two had open type. The mean size of all cholesteatomas was 5.1 mm. None of the cholesteatoma types indicated a tendency towards a certain location. The round shape was observed more frequently in closed type cholesteatomas than in other types (closed: 5/8; mixed: 1/4; open: 0/2). Two large closed type cholesteatomas and two mixed type cholesteatomas exhibited a constricted shape. Both of the open type cholesteatomas displayed an irregular shape.

Conclusion: Small closed type congenital cholesteatomas were typically observed as round shaped lesions, but large closed type cholesteatomas and other type cholesteatomas tended to display shapes other than round.

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Cholesteatoma is a well-margined non-neoplastic lesion which usually affects the middle ear, and much less commonly, develops in the external auditory canal [1,2]. Cholesteatoma can be classified as either congenital or acquired based on pathogenesis. Middle ear congenital cholesteatoma is a relatively rare disease that accounts for approximately 2% of all middle ear cholesteatomas, and presents in children as a whitish mass behind an intact tympanic membrane [1]. Morphologically, congenital cholesteatomas can be classified as closed type (cystic type), open type (keratinizing epithelium without the formation of an epithelial cyst), or mixed type, which consists of both closed and open cholesteatoma types [3–6]. In patients with open type congenital cholesteatomas, rare abnormalities through the tympanic membrane often render early detection by otoscopy difficult [7]. Since delayed diagnosis of congenital cholesteatomas can lead to advanced lesions that require more extensive surgery, early detection is critical. Knowledge of computed tomography (CT) findings related to cholesteatoma morphology could be helpful for early detection of congenital cholesteatoma, especially for detection of open type cholesteatoma. However, CT findings based on the type of cholesteatoma mass are poorly understood. Accordingly, the purpose of this study was to assess differences in congenital cholesteatoma CT findings with a focus on the type of cholesteatoma mass.

Materials and methods

The medical records of patients with congenital cholesteatomas of the middle ear who underwent surgery at our institution between January 2009 and July 2014 were reviewed. The diagnosis of congenital cholesteatoma was made based on the criteria of Levinson et al. [8], which include:

- a whitish mass behind an intact tympanic membrane;
- absence of otorrhea or perforation of the tympanic membrane;
- no history of previous procedures, including myringotomy or insertion of grommets.

A prior history of otitis media was not considered. Cholesteatomas were classified as closed type, open type, or mixed type based on intraoperative findings. A ruptured cystic cholesteatoma with debris extruding from the cyst was classified as a mixed type cholesteatoma. Among the 21 patients who met the inclusion criteria, three patients were excluded because their respective CT images were not available for morphological assessment of the cholesteatoma due to occupation of the tympanic cavity by an advanced lesion, or accompanying otitis media. Four patients without records indicating cholesteatoma type were likewise excluded. Consequently, following application of the exclusion criteria, 14 patients were enrolled in the retrospective study. Data including age at operation, sex, CT findings (cholesteatoma size, location, and shape), and intraoperative findings (types of cholesteatoma and erosion of ossicles) were collected for each patient. The appropriate institutional ethics committee approved the design of the retrospective study, and a waiver for informed consent was obtained.

CT evaluation

Because the study design was retrospective, varieties of CT scanners were used. Ten patients underwent temporal CT in our institution, and two patients underwent CT scanning at other institutions. In our institution, high resolution CT was obtained in the axial plane with coronal reformations using the following acquisition parameters: 120 kVp, 200–300 mAs, and 0.5 mm section thickness. Acquisition parameters used at the other institutions were 120 kVp, 200–400 mAs, and 1.0 mm section thickness.

Two radiologists retrospectively reviewed the CT findings and reached a consensus. Cholesteatoma size was measured as the maximum diameter. The tympanic cavity was divided into four quadrants, including the anterior-superior (AS) quadrant, anterior-inferior (AI) quadrant, posterior-superior (PS) quadrant, and posterior-inferior (PI) quadrant. The handle of malleus was used as a separation point on CT images to divide the anterior and posterior portions, and the lower pole of the handle of malleus was used to divide the superior and posterior portions. The location of the cholesteatoma was classified into one of the four quadrants, or a combination of the quadrants. The shape of the cholesteatomas was classified as round, constricted (soft tissue density with one or two constrictions), irregular (similar to multi-lobulated appearance), or other (shape not classified in any of the above categories).

Results

The clinical and CT findings of patients with congenital cholesteatomas are summarized in Table 1. The study participants included eight males and six females with a mean age of 3.2 years (range, 1–6). Eight patients had closed type cholesteatoma, four had mixed type, and two had open type. Ossicular erosion was observed in three patients (21.4%). Destruction of the long process of the incus and stapes suprastructures was observed in a patient with a mixed type cholesteatoma. As well, the destruction of only the long process of the incus was observed in one patient with a closed type cholesteatoma, and one participant with a mixed type cholesteatoma. Ossicular anomalies were not observed in any of the patients included on the study.

CT findings

The mean size of all cholesteatomas was 5.1 mm (range, 2–10). Six patients had cholesteatomas in the AS quadrant, while three patients presented with cholesteatomas in the PS and PI quadrants, two patients had cholesteatomas in the PI quadrant, two patients had cholesteatomas in all quadrants, and one patient had cholesteatomas in both the AS and AI quadrants. None of the cholesteatoma types indicated a tendency towards a certain location. Five of the eight closed type cholesteatomas, and one of the four mixed type cholesteatomas exhibited a round shape (Fig. 1a). Two mixed cholesteatomas exhibited a constricted shape (Figs. 1b and 2), and both of the two open cholesteatomas displayed an irregular shape (Fig. 3). One closed cholesteatoma and one mixed cholesteatoma displayed soft densities that were not round, constricted, or

Table 1 Clinical and CT features of chongenital cholesteatoma.

| Patient no. | Age (years) | Sex | Intraoperative findings | | CT findings | | |
|-------------|-------------|-----|-------------------------|-------------------------------|-------------|----------|-------------|
| | | | Type of form | Ossicular erosion | Size (mm) | Location | Shape |
| 1 | 4 | M | Closed type | None | 3.9 | AS | Round |
| 2 | 2 | F | Closed type | None | 2.7 | AS | Round |
| 3 | 3 | M | Closed type | None | 4.2 | AS | Round |
| 4 | 3 | M | Closed type | None | 2.2 | AS | Round |
| 5 | 1 | M | Closed type | None | 2 | PI | Round |
| 6 | 1 | M | Closed type | None | 8.6 | PS + PI | Others |
| 7 | 3 | M | Closed type | None | 7 | All | Constricted |
| 8 | 5 | F | Closed type | Long process of incus | 10.4 | All | Constricted |
| 9 | 2 | M | Mixed type | None | 4.3 | AS | Round |
| 10 | 5 | F | Mixed type | Long process of incus | 10 | PS + PI | Constricted |
| 11 | 3 | F | Mixed type | None | 6.2 | AS + AI | Constricted |
| 12 | 6 | M | Mixed type | Long process of incus, stapes | 7.9 | PS + PI | Others |
| 13 | 5 | F | Open type | None | 4.5 | AS | Irregular |
| 14 | 2 | F | Open type | None | 4.7 | PI | Irregular |

AS: anterior-superior quadrant; AI: anterior-inferior quadrant; PS: posterior-superior quadrant; PI: posterior-inferior quadrant.

irregular, and were thus classified as other in terms of shape. According to the operative records, the mixed type cholesteatoma displaying a round shape exhibited a small opening. Ossicular erosions corresponding to the intraoperative findings were identified on the CT images, respectively.

Discussion

Although several theories exist regarding the pathogenesis of congenital cholesteatoma in the middle ear, the most widely accepted theory is that congenital cholesteatomas arise from embryonic epithelial rests [3]. Most cases of congenital cholesteatomas present as cystic masses, or closed type cholesteatomas, and open type cholesteatomas occur

less frequently [5,6,9]. However, the factors involved in the determination of whether embryonic epithelial rests develop into closed or open type cholesteatomas has not been elucidated. Several studies that focused on the differences in the clinical characteristics of closed and open type congenital cholesteatomas have been reported. More frequently, patients with closed cholesteatomas present with otorrhea or otalgia, as opposed to patients with open type cholesteatomas, who commonly experience hearing loss [7]. A closed type cholesteatoma located within the anterior portion of the middle ear can be easily removed, but an open type cholesteatoma is difficult to remove surgically, and exhibits a much higher recurrence rate than closed type cholesteatomas [4,10]. Unfortunately, studies that described the clinical characteristics of patients with

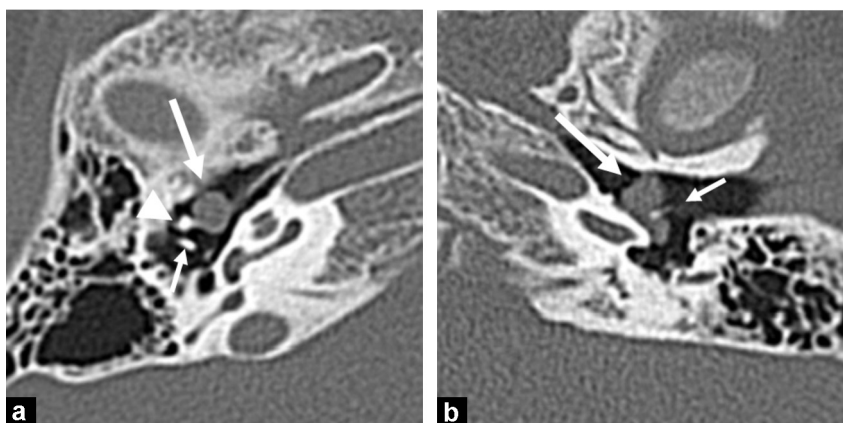


Figure 1. Closed type cholesteatoma: a: axial CT image of a 3-year-old boy (case 3) revealed a round shaped cholesteatoma (arrow) in the anterior-superior quadrant. The handle of malleus (arrowhead) and the long process of incus (small arrow) are visible; b: axial CT image of a 3-year-old boy (case 7) revealed a constricted shaped cholesteatoma occupying the anterior-superior quadrant and posterior-superior quadrant (arrow). The constriction seemed to be caused by compression from the handle of malleus (small arrow).



Figure 2. Mixed type cholesteatoma. Coronal CT image of a 3-year-old girl (case 11) revealed a constricted shape cholesteatoma. The site of the constriction (arrow) was not adjacent to ossicles (small arrow).

mixed cholesteatomas were not identified during the conduct of a literature search. In this study, we didn't include clinical presenting signs mentioned above in the inclusion criteria since the diagnosis of cholesteatoma was made using criteria, which Levenson et al. established.

A limited number of studies have evaluated differences in closed and open type cholesteatoma CT findings. Sorderberg et al. evaluated the CT findings of two patients with open type congenital cholesteatomas and five patients with closed type cholesteatomas [9]. One of the two patients with an open type cholesteatoma exhibited subtle soft tissue densities at the incudostapedial joint, while the second patient displayed no abnormal findings. On the other hand, four of five patients with closed type cholesteatomas presented with opacifications or fluid within mastoid cells. Kim et al. likewise reported that subtle soft tissue densities in the middle ear were observed more frequently in patients with open type cholesteatomas than in individuals with closed type cholesteatomas [11]. However,

neither of the prior studies included a detailed description of the morphology of the observed cholesteatomas. Iino et al. reported that mastoid cell pneumatization was worse in patients with closed type cholesteatomas than in patients with open type cholesteatomas. Likewise, Iino et al. reported that pneumatization in patients experiencing otitis media was poorer than in patients without such episodes [5]. The results of the prior study indicated that closed type cholesteatomas are closely related to middle ear inflammation, which is consistent with the clinical finding that otitis media occurs more frequently in patients with closed type cholesteatomas.

The findings of this retrospective study indicated that the shape of congenital cholesteatomas on CT images differed according to the type and size of the cholesteatoma. In our study, a round shape was observed more often in closed type cholesteatomas. In fact, all of the observed closed type cholesteatomas confined to a quadrant were round in shape, which might indicate that round shape is a typical form of early stage closed type cholesteatoma. However, the round shape was not specific to closed cholesteatomas, and was observed in the mixed cholesteatoma that displayed a small opening. A constricted shape was observed in large, closed type cholesteatomas that occupied all quadrants, as well as in mixed type cholesteatomas. The constriction in closed cholesteatomas appeared to be caused by compression from adjacent ossicles. On the other hand, the constriction in one of the mixed type cholesteatomas was not adjacent to ossicles, unlike the observations in larger closed type cholesteatomas. Consequently, the constriction may have indicated a rupture site, or a boundary between flat epithelium and the cystic mass, and could be a distinguishing feature of closed, constricted shape cholesteatomas. An irregular shape, which may be attributed to the accumulation of keratotic debris on the surface of the middle ear, was observed only in open type cholesteatomas. Although our study included only two patients with small open type cholesteatomas confined to a quadrant, considering that no other type cholesteatomas exhibited an irregular shape, the potential exists that irregular shape might be a characteristic of early stage open type cholesteatomas. However, it remains unknown whether larger open type cholesteatomas also display an irregular shape. Consequently, further

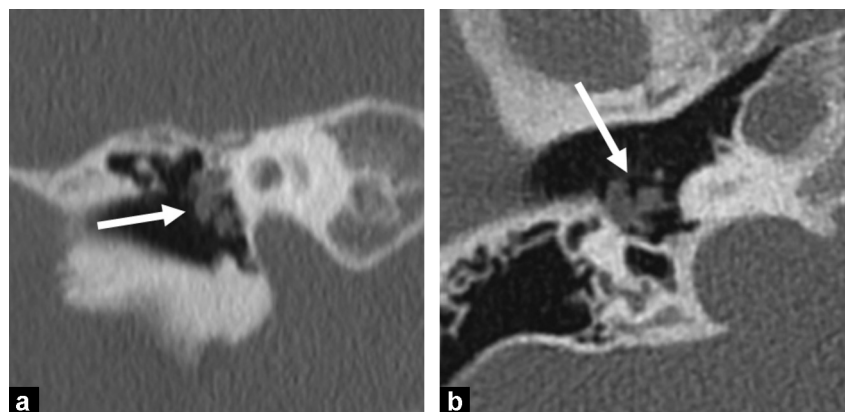


Figure 3. Open type cholesteatomas: a: coronal CT image of a 5-year-old girl (case 13) revealed an irregular shaped cholesteatoma (arrow) in the anterior-superior quadrant; b: axial CT image of a 2-year-old girl (case 14) revealed an irregular shaped cholesteatoma (arrow) in the posterior-inferior quadrant.

studies are required to confirm CT features of open type cholesteatomas.

Regarding the location of congenital cholesteatomas, Koltai et al. reported that most congenital cholesteatomas started in the AS quadrant and extended into other areas [12]. Kashiwamura et al. reported that in Japanese patients, most cholesteatomas were located in the posterior middle ear as opposed to an anterior location, whereas reports from western countries revealed that most congenital cholesteatomas originated from the anterior half of the middle ear [6]. Recently, a meta-analysis conducted by Hidaka et al. also demonstrated that the AS quadrant was less frequently involved in Asian studies compared to Western studies, and that the involvement of the PS quadrant in Asian studies was higher than association with the AS quadrant [13]. Nonetheless, the reason for this discrepancy remains unclear. A prior report established that closed type cholesteatomas arise from the anterior half of the middle ear more frequently [14]. Soderberg et al. demonstrated that all open type cholesteatomas were located in the PS quadrant, whereas Michaels reported that open type cholesteatomas were typically located in the AS quadrant [3,9]. In our study, although all of the patients were Japanese, seven of the cholesteatomas were located in the anterior half of the middle ear, which was greater than the number detected in the posterior location. When focusing on the cholesteatomas confined to one quadrant, six of the eight cholesteatomas were located in the AS quadrant, which was in accord with the results of Koltai et al.

There were several limitations to the current study, including the small number of patients owing to the rarity of this disease, and the strict inclusion criteria. In addition, various CT equipments were used due to the retrospective nature of the study, but the influence of the CT assessments was assumed insignificant. Finally, most of the patients in our study had early stage cholesteatomas confined to one or two compartments, and therefore the assessment of advanced cases was limited.

Conclusion

In conclusion, different shapes of congenital cholesteatomas could be detected on CT images according to the type or size of the mass. Small closed type cholesteatomas were typically observed as round shaped lesions, while large closed type cholesteatomas or other type cholesteatomas tended to exhibit shapes other than round. Consequently, irregular shapes might be characteristic of early stage open type cholesteatoma. Recognition of the differences in CT findings could aid in determining the type of cholesteatoma, as well as early detection of open type cholesteatomas.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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