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Congenital cholesteatoma assessment based on staging and classification criteria for middle ear cholesteatoma proposed by the Japan Otological Society

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ABSTRACT

Objective: We aimed to assess the clinical characteristics of extent patterns in congenital cholesteatoma, based on the Japan Otological Society (JOS) staging system.

Methods: This was a retrospective chart review that included 80 ears of 80 patients with congenital cholesteatoma who underwent primary surgery at a tertiary academic medical center. The main characteristics and outcomes reviewed were sex, age, clinical background, surgical method, and stage classification according to two staging classifications: the criteria advocated by JOS and Potsic staging system.

Results: The age at the time of surgery ranged from 1 to 35 years (average 8.4 years), and there were 54 men and 26 women. According to the JOS staging system, 12 ears were Stage Ia (15%), 7 ears were Stage Ib (9%), 1 ear was Stage Ic (1%), 59 ears were Stage II (74%), and 1 ear was Stage III (1%). In the study of postoperative residual recurrence, there were 4 cases after the primary operation and 3 cases after the staged operation. All 3 ears with residual disease after planned surgery were cholesteatomas that extended to all the tympanomastoid space.

Conclusion: We consider the JOS staging system to be more suitable, in terms of anatomical classification and surgical procedure selection for comparison between Europe, the United States, and Asia. Specifically, it was advantageous that the PTAM classification and the S classification are associated with surgical procedure selection and postoperative course.

1. Introduction

Because of the widespread use of school medical examinations and the development of optical instruments and diagnostic imaging, more congenital cholesteatoma cases are detected at an early stage. However, the pathology remains unclear, and there is no unified view of the choice of surgical method. Moreover, the risk of residual disease is high. To date, because there is no uniform standard with which to assess the extent of congenital cholesteatoma in Japan, Potsic stage classification of congenital cholesteatoma [1], which relatively accurately reflects clinical results, has been used. However, it is challenging to compare postoperative results due to the different locations of cholesteatomas in

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the tympanic cavity. In 2017, the JOS proposed a pathologic classification for middle ear congenital cholesteatoma [2] (the JOS staging system). We used this staging system to examine cases in which the first surgery was performed in our department. In addition, the differences between this staging system and the Potsic staging system were assessed.

2. Materials and methods

We examined 80 ears of 80 patients (males, 54; females, 26) who were diagnosed with congenital cholesteatoma of the middle ear and had undergone initial surgery at Jikei University Hospital, Tokyo, Japan, between 1993 and 2013. We only extracted the details of cases that had been followed up for at least 3 years postoperatively. If the staged surgery was chosen, the follow-up period was considered to have begun after the second surgery. The age at the time of surgery ranged from 1 to 35 years (average, 8.4 years; ≤5 years, 31 ears; 6–10 years, 34 ears; 11–15 years, 6 ears; and ≥16 years, 9 ears). The institutional review board of Jikei Medical University approved this study protocol and waived informed consent for the retrospective medical records review (approval number: 29208 (8824)).

Middle ear congenital cholesteatoma was defined, based on the diagnostic criteria of the JOS staging system, as a cholesteatoma that did not have continuity with the eardrum and ear canal and congenitally occurred in the middle ear cavity. The inclusion criteria were as follows: (1) no abnormal findings, including perforations and depressions, in the eardrum; (2) no continuity between the cholesteatoma and eardrum; (3) exclusion of petrous bone cholesteatoma. A history of otitis media was not considered a reason for study exclusion.

The adopted surgical procedure was based on a microscopic transcanal approach and whether or not a mastoidectomy was also performed. An endoscope was used in combination when necessary. Endoscopes have been used since at least 1996. Most of the procedures were performed by two chief professors, and the others were performed by a trained surgeon under a professor's supervision.

The surgical procedure was based on the International Otology Outcome Group's international consensus on the categorization of tympanomastoid surgery [3] and was classified as no mastoidectomy (Mx), only scutum removed (M2a), the combination of canal wall preserved and scutum removed (M1a+2a), or whole canal wall removed (M2c).

Planned surgery was selected for cholesteatoma expansion cases. In the staged procedure, the planned operation was performed approximately 1 year after the primary operation. Regarding the selection of a staged procedure, planned surgery was used, in principle, in cases where the cholesteatoma epithelium had entered the fine mastoid air cell system of the mastoid bone and in cases where it had progressed to the front of the tympanic cavity or the mastoid cavity. We also confirmed the presence of residual epithelium endoscopically, and we selected a staged procedure if there was a possibility of residual epithelium during the operation.

We analyzed the location of the cholesteatoma in the tympanic cavity from the site where its white mass was seen as an intraoperative finding. We described it according to the eardrum quadrant: anterosuperior (ASQ) anteroinferior, (AIQ), posterosuperior (PSQ), and posteroinferior (PIQ). The tympanomastoid space is divided into four sections: the protympanum (P), tympanic cavity (T), attic (A), and mastoid (M). They are used in the PTAM classification, which represents the pathological extent of the cholesteatoma. The JOS staging system for congenital cholesteatoma, however, was used to determine the anatomical classification. In brief, the JOS staging system is as follows: Stage I is defined as located inside the tympanic cavity. Stages Ia, Ib, and Ic are confined to the ASQ or AIQ, PSQ and/or PIQ, and both sides of the tympanic cavity, respectively. Stage II is a cholesteatoma involving two or more PTAM system sites. Stage III is a cholesteatoma with extracranial complications or intratemporal pathology. Stage IV is a case with intracranial complications.

We also examined the extent of the stapes lesion in accordance with the S classification. S0 has no stapes involvement. S1 and S2 have stapes involvement with or without the superstructure destruction, respectively. An S3 case has indistinguishable stapes footplate involvement, and SN is where the stapes is invisible at surgery. The S classification is regarded as a factor related to surgical procedure selection, with the postoperative course as a subclassification.

We also compared the results between the JOS staging system and the Potsic staging system (Stage I–IV) regarding the residual epithelium after the primary and planned surgery and the remaining lesion at the planned surgery. The Potsic staging system is as follows: Stages I and II are cases without ossicular involvement and mastoid extension of a single quadrant and multiple quadrants, respectively. Stage III has ossicular involvement, but no mastoid extension. Stage IV cases have mastoid extension.

3. Results

3.1. Relationship between staging and extension

According to intraoperative findings, location of the cholesteatoma in the PSQ was the most common (52/80 ears, 65%), followed by the PIQ (29/80 ears, 36.3%), ASQ (27/80 ears, 33.8%), and AIQ (16/80 ears, 20.0%). The average age of first diagnosis of congenital cholesteatoma was 5.1 years in ASQ cases and 7.7 years in PSQ cases. More cases located in the ASQ were found in younger patients.

Using the PTAM classification, 20/80 ears (25.0%) were classified as T, 24/80 ears (30.0%) as TA, 1/80 ears (1.3%) as AM, 14/80 ears (17.5%) as TAM, 6/80 ears (7.5%) as PTA, and 15/80 ears (18.8%) as PTAM. These cases were classified by the JOS staging system. Stage Ia was identified in 12/80 ears (15.0%), Stage Ib in 7/80 ears (8.8%), Stage Ic in 1/80 ears (1.3%), Stage II in 59/80 ears (73.8%), and Stage III in 1/80 ears (1.3%). Thus, Stage II accounted for most cases. The only Stage III case was PTAM, and the other PTAM cases were classified as Stage II. Thus, in principle,

Table 1. Distribution of cases by JOS staging system.

stage	One-stage tympanoplasty	Planned staged tymapanplasty	Total
I a	12		12 (15.0%)
I b	6		
	1	7 (8.8%)	
I c		1	1 (1.2%)
II	19	40	59 (73.8%)
III		1	1 (1.2%)
total	37 (46.3%)	43 (53.8%)	80 (100%)

Table 2. S staging by JOS staging system (Tables 1–6).

stage	I a	I b	I c	II	III	Total
S0	9			1		10
S1	2			17		19
S2	1	7	1	35		45
S3				6		6

the classification of PTAM was subdivided into Stage II and Stage III (Table 1).

When the stapes lesions were analyzed according to the 'S' subclassification, 10/80 ears (12.5%) were classified as S0, 19/80 ears (23.8%) as S1, 45/80 ears (56.3%) as S2, and 6/80 ears as S3 (7.5%). Regarding each JOS stage, Stage Ia was present in nine S0 ears, two S1 ears, and one S2 ear. Stage II was present in one S0 ear, 17 S1 ears, 35 S2 ears, and six S3 ears. All Stage Ib, Ic, and III cases were S2. In relation to the surgical procedure, the results were as follows: S0, 10/80 ears (12.5%; Mx 10 ears); S1, 19/80 ears (23.8%; 2 ears for Mx, 1 ear for M2a, and 16 ears for M1a+M2a); S2, 45/80 ears (56.3%; 12 ears for Mx, 8 ears for M2a, 23 ears for M1a+M2a, and 2 ears for M2c); and S3, 6/80 ears (7.5%; 1 ear for Mx and 5 ears for M1a+M2a). (Table 2)

Using the Potsic staging system, 10/80 ears (12.5%) were Stage I, 1/80 ears (1.3%) was Stage II, 39/80 ears (48.8%) were Stage III, and 30/80 ears (37.5%) were Stage IV.

3.2. Relationship between extension and surgical procedure

Mx was performed in 25 ears (31.3%), M2a in 9 ears (11.3%), M1a+2a in 44 ears (55.0%), and M2c in 2 ears (2.5%). When M2c was chosen, obliteration was performed concurrently.

In examining the relationship between extension according to the JOS staging system and the choice of the surgical procedure, all 12 Stage Ia ears were Mx, and the single Stage Ic case and single Stage III case were both M1a+2a. Of the 7 ears in Stage Ib, 5 (71.4%) were Mx, 1 (14.3%) was M1a+2a, and 1 (14.3%) was M2c. Of the 59 ears in Stage II, 8 (13.6%) were Mx, 9 (15.3%) were M2a, 41 (69.5%) were M1a+2a, and 1 ear (1.7%) was M2c (Table 3).

Of the 80 subjects, one-stage tympanoplasty was performed in 37 ears (46.3%), and planned surgery (staged tympanoplasty) was performed in 43 ears (53.8%).

We analyzed the relationship between extension, according to the JOS staging system, and the choice of surgery. Primary surgery (one-stage tympanoplasty) was performed for all 12 Stage Ia cases, and planned surgery was performed for 1 case of Stage Ic and 1 case of Stage III. In Stage Ib cases, 6/7 ears (85.7%) were selected for primary surgery. In the 59 ears with Stage II, 19 ears (32.2%) were chosen for primary surgery, and the remainder underwent planned surgery. No fixed trend was observed in surgical procedure selection (Table 4).

Regarding the relationship between the PTAM classification and the surgical procedure selection, in T cases (20 ears), 17 ears (85.0%) were Mx, 2 ears (10.0%) were M1a+M2a, and 1 ear (5.0%) was M2c. In TA cases (24 ears), 3 ears (12.5%) were Mx, 6 ears (25.0%) were M2a, and 15 ears (62.5%) were M1a+M2a. The only AM case was M1a+M2a. In TAM cases (14 ears), 3 ears (21.4%) were Mx, 1 ear (7.1%) was M2a, 9 ears (64.3%) were M1a+M2a, and 1 ear (7.1%) was M2c. In PTA cases (6 ears), 2 ears (33.3%) were Mx, 1 ear (16.7%) was M2a, and 3 ears (50.0%) were M1a+M2a. In PTAM cases (15 ears), 1 ear (6.7%) was M2a and 14 ears (93.3%) were M1a+M2a (Table 5).

In examining the relationship between the staging of the Potsic staging system and the surgical procedure selection, 10 ears of Stage I and 1 ear of Stage II were Mx. In the 39 Stage III ears, 10 ears (25.6%) were Mx, 7 ears (17.9%) were M2a, 21 ears (53.8%) were M1a+M2a, and 1 ear (2.6%) was M2c. Staged surgery was not selected for any Stage I (0/10) and or Stage II (0/1) cases, but it was performed in 43.6% (17/39) of Stage III ears and 86.7% (26/30) of Stage IV ears, as per the Potsic staging system.

3.3. Relationship between postoperative residual and extension

Postoperative residual disease occurred in 4 primary operations and 3 staged operations. According to the JOS staging system, cases of residual disease after primary surgery were found in 1 ear in Stage Ib and 3 ears in Stage II. Residual disease after planned surgery was found in 2 ears in Stage II and 1 ear in Stage III.

Additionally, an evaluation of the residual, as per the PTAM classification, showed residual disease after primary surgery in 1 ear for T, 2 ears for TA, and 1 ear for PTA. All 3 ears with residual disease after planned surgery were PTAM (Table 6).

As per the Potsic staging system, all 4 ears with residual disease after primary surgery were Stage III, and all 3 ears with residual disease after planned surgery were Stage IV.

4. Discussion

4.1. Relationship between staging and extension

Compared with congenital cholesteatoma reports in Europe and the United States [4–7], Japanese patients had a higher incidence of congenital cholesteatoma in the PSQ [8–11]. This correlates with our study's results, where congenital cholesteatoma in the middle ear were most frequent in the PSQ (53/80 ears, 66.3%). This further supports the idea that there is a difference in the origin of cholesteatoma between European and Japanese populations.

Table 3. JOS staging system and surgical method.

stage	Mx	M2a	M1a+2a	M2c	Total
I a	12				12(15.0%)
I b	5		1	1	7 (8.8%)
I c			1		1 (1.2%)
II	8	9	41	1	59 (73.8%)
III			1		1 (1.2%)
Total	25 (31.3%)	9 (11.3%)	44 (55.0%)	2 (2.5%)	80 (100%)

Table 4. JOS staging system and selection of planned staged tympanoplasty.

stage	One-stage tympanoplasty	Planned staged tymapanplasty	Total
I a	12		12 (15.0%)
I b	6	1	7 (8.8%)
I c		1	1 (1.2%)
II	19	40	59 (73.8%)
III		1	1 (1.2%)
total	37 (46.3%)	43 (53.8%)	80 (100%)

In a United States report [12], the average age at diagnosis of congenital cholesteatoma of the middle ear was 5–7 years, but in our study, the average age was slightly higher (8.4 years). When only the age at diagnosis was examined, ASQ cases were diagnosed at an average age of 5.1 years and PSQ cases at 7.7 years. ASQ cases were detected more often in younger patients. Another report from the United States [13] concluded that the age at diagnosis was lower than in Japan because many cases were detected early by pediatricians during school screening.

In all cases where the cholesteatoma was located anterior to the tympanic cavity, primary surgery and Mx were selected, and no postoperative residual disease was observed. In contrast, in certain cases where cholesteatomas were located posterior to the tympanic cavity, some of which required mastoidectomy, staged surgery was chosen in some instances, and residual disease was observed in 1 case.

We believe that the JOS staging system, which is subdivided according to the location of the cholesteatoma in the tympanic cavity, is beneficial both epidemiologically and clinically. According to the JOS staging system, the highest proportion of cases were Stage II (59/80, 73.8%). The original lesion may have been located posteriorly in many cases; thus, more cholesteatomas extended to the superior tympanic cavity and further to the mastoid cavity.

In the analysis according to the Potsic staging system, there was only 1 case of Stage II, and most cases were Stages III and IV. When the cholesteatoma was in the posterior upper quadrant, the ossicles in this region may often have been destroyed. We believe that this subdivided classification is more suitable than the conventional Potsic staging system in terms of classifying disease progress.

4.2. Relationship between extension and surgical procedure

According to the JOS staging system, we removed the cholesteatoma by Mx in 17/20 ears (73.9%) in Stage I and 8/59 ears (13.6%) in Stage II. In both the single- and multi-quadrant cases, we were able to remove the cholesteatoma without mastoidectomy when the mass was small. Removing the cholesteatoma without a mastoidectomy was more difficult when there was extension to the supratubal recess in ASQ cases, extension to the periarticular incudostapedial joint in PSQ cases, and extension to the tympanic sinus in PIQ cases.

We mainly performed microscopic surgery and used an endoscope to remove any residual cholesteatoma. If it appeared that the cholesteatoma would be difficult to remove completely, due to progression into the attic or mastoid cavity, we decided on a more aggressive approach and performed a mastoidectomy.

In the PTAM subclassification, patients who underwent M1a+2a were as follows: 2/20 ears (10.0%) in T, 3/6 ears (50.0%) in PTA, 15/24 ears (62.5%) in TA, 9/14 ears (64.3%) in TAM, 14/15 ears (93.3%) in PTAM, and 1/1 ear in AM. Thus, the PTAM classification was extremely effective in selecting surgical procedures based on the extent of cholesteatoma progression.

According to the JOS staging system, all cases of Stage Ia (12 ears) could be removed by Mx. However, it was often difficult to cope with any progression to the supratubal recess. In the PTA cases which did not extend into the mastoid cavity but progressed into the supratubal recess, 4/6 ears (66.7%) required a mastoidectomy. In many of these cases, we had to open the anterior tympanic space, confirm it by endoscopy through the mastoid side, and remove it.

Table 5. PTAM staging system and surgical method.

stage	Mx	M2a	M1a+2a	M2c	Total
T	17		2	1	20 (25%)
TA	3	6	15		24 (30.0%)
AM			1		1 (1.2%)
TAM	3	1	9	1	14 (17.5%)
PTA	2	1	3		6 (7.5%)
PTAM		1	14		15 (18.8%)
total	25 (31.3%)	9 (11.3%)	44 (55.0%)	2 (2.5%)	80 (100%)

Table 6. Distribution of recurrences by PTAM staging system.

	Total number (ear)	One-stage (ear)	Residual after One-stage tympanoplasty (ear)	Planned staged (ear)	Residual after Planned staged tympanoplasty (ear)	Residual at Planned staged tympanoplasty (ear)
T	20	18	1 (5.6%)	2	0	1 (50%)
TA	24	12	2ear (8.3%)	12	0	3 (25%)
TAM	14	3	0	11	0	0
PTA	6	3	1 (33.3%)	3	0	0
AM	1	1	0	0	0	0
PTAM	15	0	0	15	3 (20%)	8 (53.3%)

In general, there are many congenital cholesteatoma cases in which the growth of mastoid cells is favorable. For this reason, there are often cases where it is difficult to remove the cholesteatoma entirely because the cholesteatoma epithelium has spread into the fine mastoid air cell system of the mastoid bone. In our study, the average age at surgery was 8.4 years; therefore, to preserve the patient's quality of life, we have performed canal wall-preserved mastoidectomy. Congenital cholesteatoma is likely to re-aerate the mastoid cavity postoperatively, even in cases that have spread into the mastoid cavity. Therefore, we carefully evaluate the indications for whole canal wall removal. In our study, M2c was performed in only 2 cases, where the growth of the mastoid cavity was extremely poor in 1 case and the cholesteatoma had spread into the mastoid cavity in the other case.

In subclassification S (Table 2), the destruction of the stapes superstructure was recognized in 51/80 ears (63.8%). The stapes is present in the posterior tympanic cavity. The facial nerve is present around the stapes, and there is also a tympanic sinus, which is a blind spot under a microscopic transcanal approach. Thus, operating on this site may not be feasible with a transcanal approach and may require mastoidectomy. In our study, Stage Ia was mostly S0 (9/12 ears, 75%) and did not require mastoidectomy. However, Stage Ib and Stage Ic were all S2, and in some of these cases, mastoidectomy had to be selected. As the S classification progressed to include the other stages, the proportion of cases requiring M1a+2a increased. Thus, it was clear that S subclassification was a critical factor in the surgical choice.

On the other hand, cases in which a small cholesteatoma confined to one quadrant resulted in ossicular destruction, and cases in which extension into the mastoid cavity resulted in ossicular destruction, were categorized as Potsic Stage III. Although there is a case where Mx is possible even if there is ossicular destruction, it was considered unlikely to be reflected by the Potsic staging system. Thus, the JOS staging system is considered to be more suitable than the Potsic staging system for analyzing surgical selection.

In both the JOS staging system and the Potsic staging system, as the Stage progressed, the proportion of cases selected for staged surgery tended to increase. However, there was no certain tendency, even when examining the subclassification.

4.3. Relationship between postoperative residual disease and extension

Congenital cholesteatoma has a low level of inflammation and, therefore, fewer mucosal lesions. Mastoid honeycomb growth is considered better than in cases of acquired cholesteatoma, and reaeration after mastoidectomy is also good. Therefore, cases of recurrent cholesteatoma are considered relatively benign, whereas residual cholesteatoma is considered a significant problem.

In our study, for all patients who had residual disease after one-stage surgery, residual cholesteatoma was present in 4/37 ears (10.8%). We examined each case based on the JOS staging system. In the first case, M2c was performed for Stage Ib, and residual cholesteatoma was found in the facial recess. In the second case, M1a+2a was performed for Stage II (PTA), and residual cholesteatoma was found in the anterosuperior part (area) of the tympanic cavity. In the third case, M2a was performed for Stage II (TA), and residual cholesteatoma was observed at the aditus to the mastoid antrum. In the last case, Mx was performed for Stage II (TA), and residual cholesteatoma was found on the back of the eardrum in the anterior tympani.

There was no certain tendency in the staging of the JOS staging system and the PTAM subclassification. Instead, it was thought that residual cholesteatoma occurred in the blind spot of microscopic surgery. Thus, the incidence of residual cholesteatoma needs to be monitored and may decrease with the development of endoscopic surgical techniques, or a clear tendency will appear in the JOS staging system if the number of examined residual cases increases.

All residuals after staged surgery were cholesteatomas. According to the JOS staging system, 2 ears were Stage II, 1 ear was Stage III, and all Stage II and III cases were PTAM cases. All of the residual disease sites were inside the mastoid cavity, and one was in the lateral semicircular canal. More advanced cases tended to cause residual cholesteatoma after staged surgery.

There were 12/43 ears (27.9%) in which cholesteatoma remained after a staged procedure. We excluded and examined one of these cases, which was regarded as a special case in which M2c was performed for Stage Ib because the mastoid growth was very poor. All other cases were Stage II, as per the JOS staging system, and in the PTAM subclassification, 3 ears were TA, 1 ear was TAM, and 8 ears were PTAM cases. It was suggested that cases with an advanced PTAM stage might affect the cholesteatoma remains from a staged procedure.

Thus, it is useful to evaluate the residual cholesteatoma after staged surgery by PTAM subclassification.

According to the Potsic staging system, there was no residual cholesteatoma in Stages I and II, and there was a tendency for the rate of residual cholesteatoma to be higher in Stages III and IV. There was no clear correlation between the residual rate and staging. However, as previously pointed out, the Potsic staging system accurately reflected the clinical results in our study.

5. Conclusion

The Potsic staging system is considered simple and easy to use, and it accurately reflected our clinical results. However, the JOS staging system is considered more suitable in terms of the anatomical staging system and surgical procedure selection for comparisons between Europe, the United States, and Asia. Specifically, it was advantageous that the PTAM classification, which is pathological and anatomical, and the S classification, which is associated with surgical procedure selection and postoperative course, were positioned as subclassifications.

Declaration of Competing Interest

The authors have no conflicts of interest directly relevant to the content of this article.

References

[1] Potsic WP, Samadi DS, Marsh RR, Wetmore RF. A staging system for congenital cholesteatoma. Arch Otolaryngol Head Neck Surg 2002;128(9):1009–12.

[2] Tono T, Sakagami M, Kojima H, Yamamoto Y, Matsuda K, Komori M, et al. Staging and classification criteria for middle ear cholesteatoma proposed by the Japan Otological Society. Auris Nasus Larynx 2017;44:135–40.

[3] Matthew Y, Adrian J, Paul M, John P, Bruce B, Tetsuya T, et al. International otology outcome group and the international consensus on the categorization of tympanomastoid surgery. J Int Adv Otol 2018;14(2):216–26.

[4] Bennet M, Warren F, Jackson GC, Kaylie D. Congenital cholesteatoma: theories, facts, and 53 patients. Otolaryngol Clin N Am 2006;39(6):1081–94.

[5] Lazard DS, Roger G, Denoyelle F, Chauvin P, Garabedian EN. Congenital cholesteatoma: risk factors for residual disease and retraction pockets–a report on 117 cases. Laryngoscope 2007;117(4):634–7.

[6] Cohen MA, Kuroiwa MA, Berkowitz RG. Acquired cholesteatoma in children following congenital cholesteatoma surgery. Int J Pediatr Otorhinolaryngol 2011;75(1):43–8.

[7] Stapleton AL, EgloŠ AM, Yellon RF. Congenital cholesteatoma: predictors for residual disease and hearing outcomes. Arch Otolaryngol Head Neck Surg 2012;138(3):280–5.

[8] Kashiwamura M, Fukuda S, Chida E, Matsumura M. Locations of congenital cholesteatoma in the middle ear in Japanese patients. Am J Otolaryngol 2005;26(6):372–6.

[9] Hidaka H, Yamaguchi T, Miyazaki H, Nomura K, Kobayashi T. Congenital cholesteatoma in predominantly found in the posterior-superior quadrant in the Asian population: systematic review and meta-analysis. Otol Neurotol 2013;34(4):630–8.

[10] Yamatodani T, Mizuta K, Hosokawa K, Takizawa Y, Sugiyama K, Nakaishi H, et al. Congenital middle ear cholesteatoma: experience from 26 surgical surgical cases. Ann Otol Rhinol Laryngol 2010;122(5):316–21.

[11] Takagi T, Gyo K, Hakuba N, Hyodo J, Hato N. Clinical features, presenting symptons, and surgical results of congenital cholesteatoma based on Potsic's staging system. Acta Oto-Laryngol 2014;134(5):462–7.

[12] Kazahaya K, Potsic WP. Congenital cholesteatoma. Curr Opin Otolaryngol Head Neck Surg 2004;12(5):398–403.

[13] Rizer FM, Luxford WM. The management of congenital cholesteatoma: surgical results of 42 cases. Laryngoscope 1988;98(3):254–6.