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Staging and classification criteria for middle ear cholesteatoma proposed by the Japan Otological Society☆

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ABSTRACT

In order to provide a basis for meaningful exchange of information among those treating cholesteatoma, the Committee on Nomenclature of the Japan Otological Society (JOS) was appointed in 2004 to create a cholesteatoma staging system as simple as possible to use in clinical practice in Japan. Following the announcement of preliminary criteria for the staging of pars flaccida (attic) cholesteatoma in 2008, we proposed the 2010 JOS staging system for two major types of retraction pocket cholesteatoma, pars flaccida and pars tensa cholesteatoma. Since then, the JOS staging system has been widely used in clinical studies of cholesteatoma in Japan, allowing standardization in reporting of surgical outcomes based on the respective stages of cholesteatoma. We have recently expanded the range of cholesteatoma by adding cholesteatoma secondary to a tensa perforation and congenital cholesteatoma as the 2015 JOS staging system for middle ear cholesteatoma. Although further revisions may be required for universal acceptance of these criteria, we hope our staging system will open the way for international consensus on staging and classification of middle ear cholesteatoma in the near future.

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1. Introduction

Middle ear cholesteatoma is characterized by a mass lesion formed by keratinizing squamous epithelium, keratin debris and varying thickness of perimatrix, with or without surrounding inflammatory reaction. Because of the multifactorial and progressive nature of the disease process, pathophysiological conditions vary considerably from case to case. Hence, the operating surgeon is required to make a subjective decision regarding the most appropriate surgical technique for the individual situation, to achieve optimal surgical goals, including disease eradiation and subsequent functional and anatomical stabilization. Classification and staging of cholesteatoma would provide a standardized assembly of a relatively homogenous group of patients, allowing rational interpretation of surgical results based on respective pathophysiological conditions of the disease process. Needless to say, proper classification criteria can only be created

☆ All authors belong to the Committee on Nomenclature of the Japan Otological Society (JOS). The criteria set has been approved by the JOS Board of Directors.

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by a consensus-based approach, preferably with the support of an academic society, and should undergo revision over time.

2. Impact of the previous proposal of criteria (the 2010 JOS staging system) on clinical studies of cholesteatoma in Japan

The Committee on Nomenclature of the Japan Otological Society (JOS) was appointed in 2004 to create a cholesteatoma staging system as simple as possible to use in clinical practice, in order to provide a basis for meaningful exchange of information among those treating cholesteatoma. Although several criteria for cholesteatoma classification/staging have been proposed worldwide [1–5], none of them are simple enough nor are they authorized by an academic society. Assuming that the tympanomastoid surgery based on the type of cholesteatoma has been discussed and cultivated differently in each country, it was hoped that a consensus-based staging system primarily for Japanese otolaryngologists could be developed to start with.

Following the announcement of preliminary criteria for the staging of pars flaccida (attic) cholesteatoma in 2008 [6], we proposed the 2010 JOS staging system [7] for two major types of retraction pocket cholesteatoma, pars flaccida and pars tensa cholesteatoma. The basic concept of this system was twofold: (1) The middle ear space was divided into four sites: protympanum (P), tympanic cavity (T), attic (A) and mastoid (M); (2) Three stages were defined: in stage I, the lesion is confined to the attic or the tympanic cavity; in stage II, the lesion extends beyond the attic or the tympanic cavity; in stage III, intratemporal and/or intracranial complications are observed. Stages II and III can be further sub-classified according to the extension of the cholesteatoma using the PTAM system. For example, if the matrix has invaded the protympanum, the attic and the mastoid antrum without causing any complications, it is described as ‘‘stage II PAM’’.

Since then, the 2010 JOS staging system has been widely used in clinical studies of cholesteatoma in Japan, allowing standardization not only in reporting of surgical outcomes but also in clinical communications between physicians and patients based on the respective stage of cholesteatoma. The number of medical articles referring to the JOS system has gradually increased in Japanese literature during the past 7 years. A search of the key words of ‘‘cholesteatoma’’ AND ‘‘staging’’ in the Japan Medical Abstracts Society web site, produced 46 original papers and 60 conference abstracts between 2008 and 2015 [8].

3. Background of the current revision of the previous criteria

In 2012, we were given the opportunity to present the JOS staging system during the 9th International Conference on cholesteatoma and ear surgery in Nagasaki, Japan (Chairman of the meeting, Prof. Haruo Takahashi) [9]. In one of the panel discussions entitled ‘‘Panel with response analyzer to build up international consensus on classification and staging of middle ear cholesteatoma (modulators: Prof. Nuri Ozgirgin and Prof. Naoaki Yanagihara)’’, this system gained favourable responses from the international audience as judged by the results of the response analyzer. However, one point to be considered in order to make this staging system internationally acceptable was a suggestion to separate the intracranial complications from the stage III elements because intracranial complications are life-threatening complications which are of great concern worldwide.

Although the 2010 JOS staging system was designed exclusively for pars flaccida and pars tensa cholesteatomas, at least two entities unrelated to a retraction pocket in origin are fairly well recognized in Japan: cholesteatoma secondary to a chronic tensa perforation and congenital cholesteatoma. The former is cholesteatoma that develops secondary to ingrowth of squamous epithelium through a pre-existing tensa perforation (so-called secondary acquired cholesteatoma). Careful observation using a microscope or an endoscope is essential to discriminate a continuous epithelial invasion from the perforation edge to the underside of the pars tensa with no direct adhesive lesion to the promontrial mucosa [10]. Although a clear distinction between this type of cholesteatoma and pars tensa cholesteatoma may be difficult in some cases, the specific staging criteria described below would help better understanding of the characteristic process of this disease entity. The latter is congenital cholesteatoma that develops behind an intact tympanic membrane. Although academic studies abroad showed that the typical location of congenital cholesteatoma is in the anterior superior quadrant of the tympanic cavity [11,12], this is not necessarily true in Japan, where the posterior superior quadrant is reportedly the most common site [13,14]. Therefore, it would be worthwhile to survey clinical data of congenital cholesteatoma nationwide using the same staging criteria, specific for this project.

The current committee to improve the criteria was convened under the Japan Otological Society in 2014 to address these issues. The original Japanese version has already been published as the 2015 JOS staging system [15] and its English version is reported here following a public comment period of 2 months. The cholesteatoma classification has been slightly modified from the Japanese version through a collaborative dialogue with members of the cholesteatoma guidelines group of the European Academy of Otology and Neurotology.

4. Proposed new set of criteria: the 2015 JOS staging system for middle ear cholesteatoma

4.1. Classification of cholesteatoma

I. Acquired cholesteatoma
  1) Retraction pocket cholesteatoma (so-called primary acquired cholesteatoma)
    a) *Pars flaccida cholesteatoma (Attic cholesteatoma)*: Cholesteatoma originating in a pars flaccida retraction pocket.

b) *Pars tensa cholesteatoma*: Cholesteatoma originating in a pars tensa retraction pocket. The range of retraction varies from a postero-superior quadrant (sinus cholesteatoma) to a partial or an entire pars tensa adhesion.

c) *Combination of pars flaccida and pars tensa cholesteatoma*: Cholesteatoma involving both the attic and pars tensa. They may develop as separate retraction pockets or as an indivisible collapse with substantial attic and posterior canal wall defects.

2) Nonretraction pocket cholesteatoma

a) *Cholesteatoma secondary to a chronic tensa perforation (so-called secondary acquired cholesteatoma)*: Cholesteatoma that develops secondary to ingrowth of squamous epithelium along the perforation edge to the underside of the pars tensa and the malleus handle without forming a well-defined retraction pocket.

b) *Transplanted cholesteatoma following trauma or otologic procedures*: Cholesteatoma that usually develops as a cyst resulting from the transplantation of squamous epithelial cells into the middle ear during traumatic or iatrogenic injuries to the tympanic membrane.

II. Congenital cholesteatoma

Cholesteatoma that develops behind an intact tympanic membrane. History of otitis media does not necessarily exclude congenital cholesteatoma but cases having previous otologic procedures should be excluded.

III. Unclassifiable cholesteatoma

Cholesteatoma is not applicable to any of the above categories. The otoscopic characteristics of cholesteatoma may be modified if the tympanic membrane undergoes secondary pathological changes following infections or otologic procedures.

4.2. Staging of cholesteatoma

4.2.1. Divisions of the middle ear space: PTAM system (Fig. 1)

In order to simplify the extent of cholesteatoma, the tympanomastoid space is divided into four sections: the protympanum (P), the tympanic cavity (T), the attic (A) and the mastoid (M). The protympanum includes the bony Eustachian tube and the supratubal recess. The tympanic cavity consists of the mesotympanum, hypotympanum and the retrotympanum including the tympanic sinus. The posterior border of the attic is the posterior end of the incus short process or the fossa incudis. The mastoid includes the antrum and the mastoid cells.

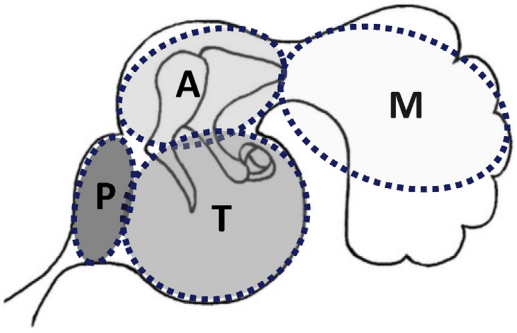

Fig. 1. Schematic drawing of divisions of the tympanomastoid space. The tympanomastoid space is divided into four sections: the protympanum (P), the tympanic cavity (T), the attic (A) and the mastoid (M) in order to represent the extent of cholesteatoma.

4.2.2. Staging system applicable to the following four types of cholesteatoma

Stage I: cholesteatoma localized in the primary site*

*The site of cholesteatoma origin, i.e. the attic (A) for a pars flaccida cholesteatoma and the tympanic cavity (T) for pars tensa colesteatoma, congenital cholesteatoma and cholesteatoma secondary to a tensa perforation.

Stage II: Cholesteatoma involving two or more sites

Stage III: Cholesteatoma with extracranial complications and/or intratemporal pathologic conditions:

Facial palsy (FP), labyrinthine fistula (LF): with conditions at risk for membranous labyrinth, labyrinthine disturbance (LD): scale out BC values for more than two speech frequencies (0.5, 1, and 2 kHz), canal wall destruction (CW): more than half the length of the bony ear canal, adhesive otitis (AO): total adhesion of the pars tensa, petrous bone or skull base destruction (PB), neck abscess (NA).

Stage IV: Cholesteatoma with intracranial complications:

Purulent meningitis, epidural abscess, subdural abscess, brain abscess, sinus thrombosis, etc.

4.3. Staging system for respective cholesteatoma types

4.3.1. Pars flaccida cholesteatoma (attic cholesteatoma)

Stage I: Cholesteatoma localized in the attic
Stage Ia: A retraction pocket with epithelial self-cleaning function
Stage Ib: A retraction pocket with persistent accumulation of keratin-debris
Stage II: Cholesteatoma involving two or more sites
Stage III: Cholesteatoma with intratemporal complications and/or pathologic conditions
Stage IV: Cholesteatoma with intracranial complications

4.3.2. Pars tensa cholesteatoma

Stage I: Cholesteatoma localized in the tympanic cavity
Stage Ia: A retraction pocket with epithelial self-cleaning function
Stage Ib: A retraction pocket with persistent accumulation of keratin-debris
Stage II: Cholesteatoma involving two or more sites
Stage III: Cholesteatoma with intratemporal complications and/or pathologic conditions
Stage IV: Cholesteatoma with intracranial complications

4.3.3. Cholesteatoma secondary to a tensa perforation

Stage I: Cholesteatoma localized in the tympanic cavity
Stage Ia: Epithelial invasion confined to the underside of the pars tensa
Stage Ib: Epithelial invasion extending to the tensor tympani tendon and the promontrial wall
Stage II: Cholesteatoma involving two or more sites
Stage III: Cholesteatoma with intratemporal complications and/or pathologic conditions
Stage IV: Cholesteatoma with intracranial complications

4.3.4. Congenital cholesteatoma

Stage I: Cholesteatoma localized in the tympanic cavity
Stage Ia: Cholesteatoma confined to the anterior half of the tympanic cavity
Stage Ib: Cholesteatoma confined to the posterior half of the tympanic cavity
Stage Ic: Cholesteatoma involving both of sides of the tympanic cavity
Stage II: Cholesteatoma involving two or more sites
Stage III: Cholesteatoma with intratemporal complications and pathologic conditions
Stage IV: Cholesteatoma with intracranial complications

4.4. Additional criteria for the evaluation of the degree of mastoid cell development and the pathological status of stapes

Two optional criteria were added to the 2010 JOS system to meet the demand of JOS members: the degree of mastoid cell development and the pathological status of the stapes. Both factors are considered to exert characteristic influences on surgical procedures and long-term outcomes. Computed tomography (CT), which is a routine preoperative examination before any tympanomastoid surgery in Japan, is used to assess the degree of mastoid cell development. The pathological status of the stapes can also be judged from high resolution CT but an accurate assessment at surgery is necessary.

4.4.1. Development of mastoid cells (Fig. 2)

MC0: almost no cell growth
MC1: cellular structures only around the mastoid antrum
MC2: well developed cellular structures
MC3: cellular structures extending to the peri-labyrinthine area

The superscript ''a'' is appended to indicate aeration in the mastoid (confirmed with preoperative CT or intraoperatively) as''MC2^{a}'' if the well-developed mastoid cells are aerated.

4.4.2. Pathological status of the stapes (Fig. 3)

S0: no stapes involvement
S1: the superstructure is surrounded by cholesteatoma and granulation
S2: the superstructure is missing but the footplate remains intact
S3: the footplate is involved and indistinguishable
SN: the stapes is not observed at surgery.

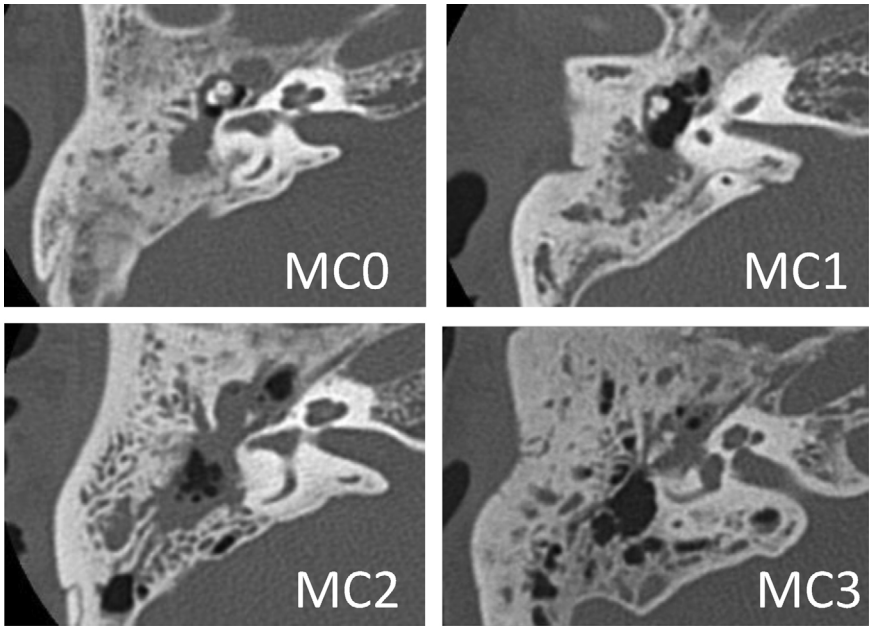

Fig. 2. Axial CT images representing degrees of mastoid cell development (MC0–3).

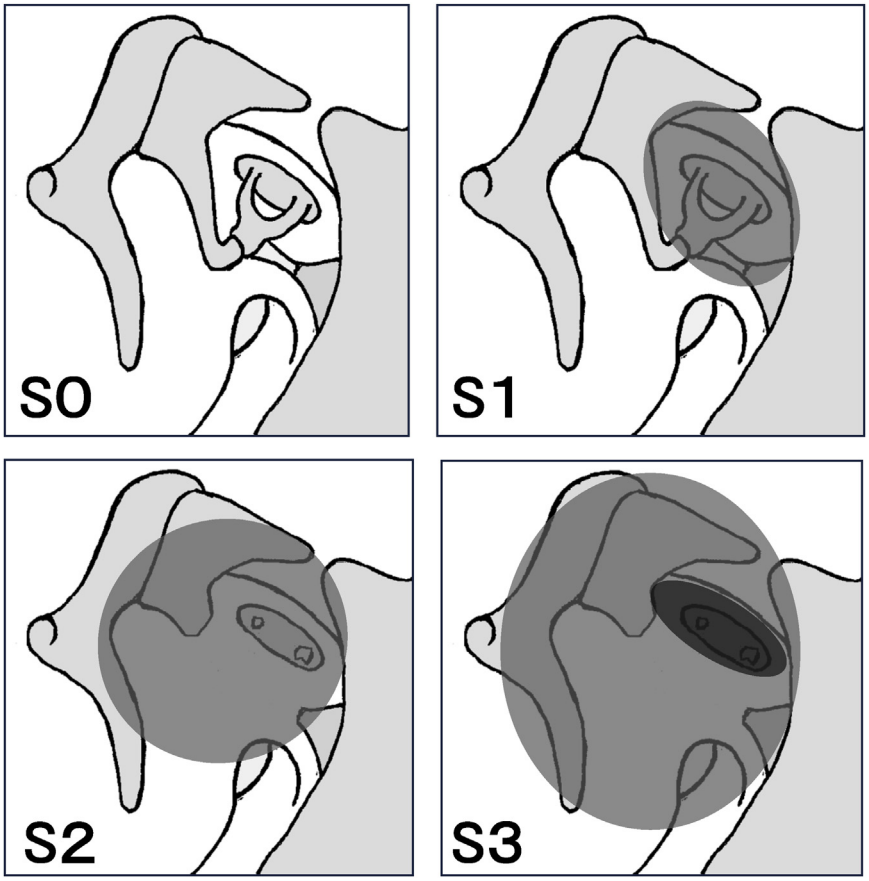

Fig. 3. Schematic drawing of criteria for pathological status of the stapes (S0–S3).

5. Conclusions

The 2015 JOS staging system for middle ear cholesteatoma has been developed over the past 7 years through a consensus-based process and in a step-by-step manner, led by the Committee on Nomenclature of the JOS. The original version of the criteria was prepared for pars flaccida cholesteatoma in 2008. The concept of the criteria fitted naturally into pars tensa cholesteatoma, leading us to propose the 2010 JOS staging system as a set of criteria for both types of retraction pocket cholesteatoma. Successful promotion of the use of this system has had a significant effect in standardizing reporting of surgical outcomes based on the respective stages of cholesteatoma. We have expanded the range of cholesteatoma by adding cholesteatoma secondary to a tensa perforation and congenital cholesteatoma as described in this article. Although further revisions may be required for universal acceptance of these criteria, we hope our staging system will open the way for international consensus on staging and classification of middle ear cholesteatoma in the near future.

Conflict of interest

The authors have no conflict of interest to declare.

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