ORIGINAL ARTICLE

Conductive deafness with normal eardrum: absence of the long process of the incus

KEEHYUN PARK, YUN-HOON CHOUNG, YOU REE SHIN & SUNG PYO HONG

Department of Otolaryngology, Ajou University School of Medicine, Suwon, South Korea

Abstract

Conclusions. The most likely cause of lesions with an absence of the long process of the incus was congenital origin because many cases were associated with the stapedial anomaly. However, the possibility of a potential inflammation of this lesion could not be completely excluded, especially in cases with only an absence of the long process of the incus. Objective. To clarify whether the absence of the long process of the incus is caused by congenital anomaly or a potential inflammation. Subjects and methods. A total of 21 patients (5 bilateral cases, 16 unilateral cases) with absence of the long process of the incus were reviewed. Operative and histopathological findings of the removed incus were analyzed. Results. The 21 cases with absence of the long process of the incus were classified into 6 types. Thirteen cases showed the absence of the long process associated with the stapedial anomaly, which were all unilateral. Eight cases showed no association with the stapedial anomaly, among which five cases were bilateral. Three of eight cases not associated with the stapedial anomaly showed evidence of resorption in the incus body such as fibrosis and Haversian canal widening.

Keywords: Ossicle, congenital anomaly, inflammation, bone resorption

Introduction

The middle ear ossicles are derived from branchial arches I and II, and the footplate of the stapes is derived in part from the otic capsule. The long process of the incus is derived from branchial arch II as well as the superstructure of the stapes. Normal ossicular anatomy corresponds closely to its embryologic derivation.

Ossicular deformities may occur in a normal tympanic membrane and external auditory canal. The tympanic membrane and external auditory canal do not derive from the same embryological origins as the ossicles. As such, the two groups of malformations may exist independently of each other. Most frequently, however, they exist in combined form.

A non-progressive and 40–60 dB conductive hearing loss with a normal eardrum, without any history of trauma or infection, is highly suggestive of a congenital ossicular malformation. Among ossicular anomalies, an absence of the long process of the incus has been suggested. However, there is also a suggestion that this kind of pathology may be attributed to acute otitis media during infancy or childhood [1]. This conductive hearing loss with a normal eardrum is due to resorption of the long process of the incus by its potential inflammation [2]. The purpose of this study was to clarify whether the absence of the long process of the incus is caused by inflammation or anomaly.

Subjects and methods

The subjects comprised 21 patients who had visited Ajou University Hospital from 1994 to 2005. They were confirmed by exploratory tympanotomy as having a congenital absence of the long process of the incus without any anomalies of the tympanic membrane and external auditory canal. Patients with a previous history of head injuries or progressive hearing loss were excluded from the study.

There was no difference in distribution of sex, and the average age at diagnosis was 26.9 years (7–61 years). The average hearing threshold level in
speech frequencies before operation was 51.2 dB (41.2–62.3 dB) of the conductive type.

Malformations of the ossicles were analyzed on the basis of surgical findings. The defective portions of the removed incuses were analyzed histopathologically in 16 ears.

Results

Ossicular anomalies of 21 cases with absence of the long process of the incus were classified into the following six types. Type 1: only absence of the long process of the incus (five cases) (Figure 1A). Type 2: fibrous change of the long process of the incus (three cases) (Figure 1B). Type 3: absence of the long process of the incus without the superstructure of the stapes (six cases) (Figure 2A). Type 4: fibrous change of the long process of the incus without the anterior crus of the stapes (three cases) (Figure 2B). Type 5: absence of the long process of the incus with footplate fixation and without the anterior crus of the stapes (three cases) (Figure 3A). Type 6: fibrous change of the long process of the incus with footplate fixation of the stapes (one case) (Figure 3B). Eight cases showed only absence of the long process, among which three cases revealed a fibrous band instead of a long process. Thirteen cases showed the absence of the long process associated with the stapes anomaly, among which four cases revealed a footplate fixation. As shown in Table I, bilateralism was described according to the anomalous ossicles. Bilateral pattern was noted only in type 1 and type 2 cases, while unilateral pattern was noted in all cases associated with the stapes anomaly.

In histopathological analyses of 16 removed incuses, 3 incuses showed evidence of resorption in the body due to increased osteoclastic activity such as fibrosis (Figure 4A), fibrosis with new bone formation (Figure 4B), and Haversian canal widening (Figure 4C). These were all unilateral cases, which included one case of type 1 and two cases of type 2.

Discussion

Although congenital anomalies of the ossicles without malformations of the external ear are not common, high resolution computed tomography (CT) of the temporal bone and exploratory tympanotomy have given the otologic surgeon a greater chance of revealing them. To date, there have been many reports 

Figure 1. Operative findings and schematic presentation of absence of the long process of the incus. (A) Only absence of the long process of the incus (type 1). (B) Fibrous change of the long process of the incus (type 2). c, chorda tympani; s, stapes; arrow, fibrous band instead of incus long process; asterisk, absence of incus long process.
Figure 2. Operative findings and schematic presentation of absence of the long process of the incus associated with the stapedial anomaly (mobile footplate). (A) Absence of the long process of the incus without the superstructure of the stapes (type 3). c, chorda tympani; arrowhead, stapes footplate. (B) Fibrous change of the long process of the incus without the anterior crus of the stapes (type 4). Asterisk, absence of anterior crus of stapes; arrow, fibrous band instead of incus long process.

Figure 3. Operative findings and schematic presentation of absence of the long process of the incus associated with the stapedial anomaly (fixed footplate). (A) Absence of the long process of the incus with footplate fixation and without the anterior crus of the stapes (type 5). c, chorda tympani; asterisk, absence of stapes anterior crus. (B) Fibrous change of the long process of the incus with footplate fixation of the stapes (type 6). I, incus; s, stapes; arrow, fibrous band instead of incus long process; arrowhead, herniated facial nerve.
Bone resorption is frequently found on the surface of the incus. Inflammation or mechanical stimulation can cause the lesion of the long process of the incus [10]. The long process is vulnerable to inflammation or mechanical stimulation because of its floating structure in the middle ear cavity and poor blood supply. Also, remodeling of the incus continues throughout life, while resorption and regeneration of the bone occur repeatedly. In a few cases in our study, there was evidence of resorption of the incus body, especially in its attached portion of the long process, which showed fibrosis, new bone formation, and Haversian canal widening due to increased osteoclastic activity. It was thought that this evidence of resorption was not always related to an inflammation in the middle ear cavity during infancy or childhood. In 1958 Hough mentioned that, in some of his many cases of middle ear ossicular malformations, there was no long process of the incus, and that this condition was attributable to otitis media in infancy or childhood [1]. Iwanaga and Yamamoto [2] also reported a case of conductive hearing loss with the normal tympanic membrane due to fibrosis caused by potential inflammation of the long process of the incus. In contrast to these reports, there was a report about bilateral congenital

### Table I. Associated ossicular anomalies of cases with absence of the long process of the incus.

<table>
<thead>
<tr>
<th>Anomalous ossicle</th>
<th>Unilateral</th>
<th>Bilateral</th>
<th>Total (cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No association (8 cases)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>only absence</td>
<td>1</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>with fibrous change</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Associated with stapedial anomaly (13 cases)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>no superstructure</td>
<td>6</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>no anterior crus with fibrous change</td>
<td>3</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>footplate fixation without anterior crus</td>
<td>3</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>footplate fixation with fibrous change</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total (cases)</td>
<td>16</td>
<td>5</td>
<td>21</td>
</tr>
</tbody>
</table>

Figure 4. Light micrograph of the defective portion of the removed incuses shows bone resorption due to increased osteoclastic activity such as (A) fibrosis (asterisk), (B) fibrosis with new bone formation (arrow), and (C) widening of Haversian canal (arrowhead). (Hematoxylin and eosin stain ×100.)
absence of the long process of the incus, which was found in three successive generations [11]. In this family, the defect appeared to be an inherited trait due to either an autosomal dominant mutation or an X-linked dominant inheritance. In dominant hereditary conductive deafness through lack of incus-stapes junction, it must be assumed that a dominant genetic factor prevents the rotation and junction of the distal end of the incus with the blastema of the stapes in the sixth to the seventh fetal week, resulting in inborn conductive deafness [12].

Another interesting finding in our study was that the absence of the long process of the incus was associated with several stapedial anomalies. The upper ossicular chain is more closely related to the first arch, whereas the lower ossicular chain is related to the second arch. This is extremely important for otologists, because this is the reason why it is common to see malformations of the lower half of the ossicular chain in conjunction with stapedial anomalies. The long process of the incus and the superstructure of the stapes have the same embryological origin, and therefore the two groups of malformations may exist dependently of each other. Anatomic variations of the ossicles may differ considerably in type and extent, and in no two analyses are the malformations the same. Ultimately, it is difficult to determine why the malformation was involved only in the ossicles originating from the second arch and not in the other structure developed from the same arch. Thus, the specific factors concerning the etiogenesis of an isolated ossicular anomaly require further study.

In conclusion, the most likely cause of a lesion with an absence of the long process of the incus was congenital, because many such lesions were associated with the stapedial anomaly. However, the possibility of a potential inflammation of this lesion cannot be completely excluded, especially in cases with an absence of the long process of the incus not associated with the stapedial anomaly.

References