Otosclerosis is a primary osteodystrophy of the otic capsule, and a cause of progressive conductive hearing loss in adults. Severe cases of otosclerosis can result in a combination of sensorineural and conductive hearing loss. Otosclerosis can be categorized on the basis of the extent of involvement into fenestral and retrofenestral types, and on the phase of disease, into spongiotic (active) or sclerotic (inactive). The otic capsule just anterior to the oval window is the typical site of manifestation. Disease limited to this area is referred to as fenestral otosclerosis and is most commonly lucent on CT due to resorption of the enchondral bone during the spongiotic (active) phase. As the disease progresses to the inactive or sclerotic phase, these lesions undergo remineralization and can become indistinguishable on CT from the normal dense otic capsule. Notably, otosclerotic foci are usually larger in volume than the bone they replace, causing thickening of the affected otic capsule.

Diagnosis of otosclerosis is classically based on history, physical examination, and audiometric testing. High-resolution CT is the technique of choice to confirm the diagnosis and evaluate alternate diagnoses or coexisting diseases and for preoperative anatomic assessment. The aforementioned variable disease activity and the presence of sclerotic foci mimicking normal bone can make diagnosis of otosclerosis by CT challenging.

The purpose of this study was to assess the qualitative and quantitative differences in otic capsule contour and thickness just

ABSTRACT

BACKGROUND AND PURPOSE: Otosclerosis is commonly identified on CT as a focus of hypodensity in the otic capsule anterior to the oval window. However, otosclerosis can have a sclerotic phase approximating the density of normal bone, making diagnosis challenging. This study assesses differences in otic capsule contour and thickness anterolateral to the anterior margin of the oval window in patients with otosclerosis compared with individuals with normal hearing.

MATERIALS AND METHODS: Axial CT of 104 ears with clinically diagnosed otosclerosis and 108 consecutive ears of audiometrically normal individuals were retrospectively reviewed. Two radiologists independently evaluated the pattern of otosclerosis, otic capsule contour, and bone thickness on standardized axial images at the level of the oval window and cochleariform process. Measurements were made from the posterior-lateral margin of the cochlea to the apex of the otic capsule convex contour just anterolateral to the anterior margin of the oval window. In the absence of a convex contour, the sulcus between the oval window and the cochleariform process was identified, and measurement to the depth of the sulcus was used. Receiver operating characteristic analysis determined the best cutoff value of otic capsule thickness.

RESULTS: Mean otic capsule thickness (± 2 SDs) was 3.08 (0.93) mm and 1.82 (0.31) mm in patients with otosclerosis and individuals with normal hearing, respectively (P < .001), with excellent interobserver agreement. Otic capsule thickness of >2.3 mm had 96.2% sensitivity, 100% specificity, 100% positive predictive value, and 96.4% negative predictive value for otosclerosis. A bulging/convex contour of the otic capsule had 68.3% sensitivity, 98.1% specificity, 97.3% positive predictive value, and 76.3% negative predictive value.

CONCLUSIONS: Patients with otosclerosis have significantly thicker bone abutting the oval window than individuals with normal hearing.
FIG 1. Qualitative description of the otic capsule contour relative to an imaginary line drawn from the anterior margin of the oval window to the cochleariform process. A, Normal temporal bone with a concave contour. B, Lucent phase of fenestral and retrofenestral otosclerosis with a flattened contour. C, Sclerotic phase of fenestral otosclerosis with a bulging or convex contour.
value of the otic capsule thickness that had the best combination of sensitivity and specificity for differentiating patients with otosclerosis from individuals with normal hearing. Interobserver reliability was evaluated using the Pearson product-moment correlation coefficient.

RESULTS

Study Groups
The enrolled population characteristics, CT modalities, and types of otosclerosis are shown in Table 1.

Qualitative Assessment of the Otic Capsule
Otic capsule contour in otosclerosis and normal hearing patients is shown in Table 2. Bulging or convex contour had 68.3% sensitivity, 98.1% specificity, 97.3% positive predictive value, and 76.3% negative predictive value (Table 3).

Quantitative Assessment of the Otic Capsule
Patients with otosclerosis had significantly thicker otic capsules near the oval window ($P < .001$) measured from the posterolateral margin of the cochlea lumen closest to the middle ear (junc-

Table 1: Characteristics of the study groups

| Patient Characteristics | Otosclerosis | Normal Hearing |
|-------------------------|--------------|----------------|
| No. of Patients         | 58           | 54             |
| No. of Ears             | 104          | 108            |
| Sex                     |              |                |
| Male (%)                | 21 (36.2)    | 14 (25.9)      |
| Female (%)              | 37 (63.8)    | 40 (74.1)      |
| $P$ value               | .31          |                |
| Mean age (yr)           | 49.7         | 38.5           |
| Age (SD) (yr)           | 13.9         | 13.7           |
| $P$ value               | <.001        |                |
| CT modalities           |              |                |
| MDCT (%)                | 35 (60.3)    | 44 (81.5)      |
| CBCT (%)                | 23 (39.7)    | 10 (18.5)      |
| $P$ value               | .02          |                |
| Otosclerosis involvement (%) |            |                |
| Fenestral only          | 71 (68.3)    | –              |
| Fenestral and retrofenestral | 31 (29.8)   | –              |
| Isolated round window   | 2 (1.9)      | –              |
| Otosclerosis phase of disease (%) |          |                |
| Sclerotic disease       | 25 (24.0)    | –              |
| Mixed sclerotic lucent disease | 34 (32.7)  | –              |
| Lucent disease          | 45 (43.3)    | –              |

Table 2: Otic capsule contour

| Contour                  | Otosclerosis | Normal Hearing | Total |
|--------------------------|--------------|----------------|-------|
| Bulging/convex           | 71           | 2              | 73    |
| Flattened/concave        | 33           | 106            | 139   |
| Total                    | 104          | 108            | 212   |

Two-tailed $P$ value < .001
Table 3: Otic capsule contour and thickness for diagnosis of otosclerosis

| Diagnostic Performance                  | Bulging/Convex Contoura | Otic Capsule Thickness >2.3 mm<sup>b</sup> |
|----------------------------------------|-------------------------|------------------------------------------|
| Sensitivity (%)                         | 68.3                    | 96.2                                     |
| Specificity (%)                        | 98.1                    | 100                                      |
| Positive predictive value (%)          | 97.3                    | 100                                      |
| Negative predictive value (%)          | 76.3                    | 96.4                                     |

<sup>a</sup>Bulging/convex contour across an imaginary line between the anterior margin of the oval window and the cochleiform process.

<sup>b</sup>Otic capsule thickness measured from the posterolateral margin of the cochlea closest to the middle ear (junction of the basal and middle turns) to the most convex contour.

Table 4: Mean otic capsule thickness in millimeters (2 SDs)

| Study Groups | Radiologist 1 | Radiologist 2 | Overall |
|--------------|---------------|---------------|---------|
| Otosclerosis (n = 104) | 3.09 (0.92) | 3.06 (0.97) | 3.08 (0.93) |
| Normal hearing (n = 108)  | 1.87 (0.32) | 1.78 (0.33) | 1.82 (0.31) |

Table 5: Overall mean otic capsule thickness in millimeters (2 SDs) and percentage of patients with a bulging/convex contour by phase of otosclerosis

| Phase of Otosclerosis | Mean Thickness | Bulging/Convex Contour (%) | Flattened/Concave Contour (%) |
|-----------------------|----------------|---------------------------|-------------------------------|
| Sclerotic (n = 25)    | 3.02 (0.82)    | 64.0                      | 36.0                          |
| Mixed (n = 34)        | 3.20 (1.08)    | 70.6                      | 29.4                          |
| Lucent (n = 45)       | 3.06 (0.82)    | 68.9                      | 31.1                          |
| P value               | .26<sup>a</sup> | .86<sup>b</sup>          |                               |

<sup>a</sup>P value calculated with a 1-way ANOVA test.

<sup>b</sup>P value calculated with a 2 × 3 table χ<sup>2</sup> test.

### DISCUSSION

The otic capsule is composed of an inner layer of endosteum, a middle layer of persistent primary enchondral bone, and an outer layer of periosteum. Normally, the otic capsule does not undergo postdevelopmental remodeling. However, during the active or spongiotic phase of otosclerosis, the dense middle layer of enchondral bone is resorbed and replaced by spongy vascular bone, resulting in lower density on CT. During the inactive phase, the affected areas of otosclerosis undergo new bone formation, thus mimicking the density of normal bone on CT. Our study hypothesis is based on the pathophysiology that otosclerotic foci undergo continuous resorption and remodeling, eventually resulting in production of more mature bone, often larger than the original affected area, thus leading to focal thickening of the otic capsule. Thus, our study included all consecutive patients with clinically diagnosed otosclerosis regardless of the phase of disease. Because the otic capsule adjacent to the anterior margin of the oval window is expected to enlarge with remodeling, this bone should be thicker in patients with otosclerosis compared with individuals with normal hearing regardless of the phase of disease. Our study demonstrated that there was no significant difference between the phase of otosclerosis and the thickness of the otic capsule contour. In addition, there was no significant difference between the phase of otosclerosis and the presence of a bulging/convex contour.

We found that a bulging or convex contour of the otic capsule across an imaginary line drawn from the anterior margin of the oval window to the cochleiform process had 98.1% specificity and 97.3% positive predictive value. The high specificity suggests that individuals with normal hearing are unlikely to have a bulging or convex contour and that this technique is a good diagnostic tool to rule in disease. Although predictive values are influenced by prevalence, this high positive predictive value can imply that patients with a bulging or convex contour have a high probability of otosclerosis. This finding is consistent with the bone remodeling, deposition, and thickening of the otic capsule seen in otosclerosis on histology. In addition, this finding is compatible with the clinical symptoms of conductive hearing loss as the normal sulcus immediately anterolateral to the oval window is lost, resulting in stapes fixation and impedance of sound.

For quantitative assessment, otosclerosis resulted in a significantly thicker otic capsule near the oval window than in individuals with normal hearing (P < .001). Otic capsule thickness of >2.3 mm of the most convex contour provided the best trade-off between sensitivity and specificity and is also >3 SDs from the average otic capsule thickness in individuals with normal hearing. This finding is consistent with the bone remodeling, deposition, and thickening of the otic capsule seen in otosclerosis on histology. In addition, this finding is compatible with the clinical symptoms of conductive hearing loss as the normal sulcus immediately anterolateral to the oval window is lost, resulting in stapes fixation and impedance of sound.

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immediately surrounding the fissula ante fenestram, which is one of the last parts of the otic capsule to ossify.\textsuperscript{15} Small pericochlear lucencies can be seen in children,\textsuperscript{16,17} but these would not be expected to affect the otic capsule contour or the overall thickness of the otic capsule. Because our patients were adults, we would not have expected differences related to the age of the patients. Most interesting, 1.9\% of patients with otosclerosis had isolated round window lesions without associated disease anterior to the oval window, a higher percentage than reported in prior literature.\textsuperscript{18} Although imaging may not be necessary in the diagnosis of patients who present with characteristic clinical findings and typical audiometric test findings,\textsuperscript{9} imaging is helpful for the diagnosis in cases of sensorineural or mixed hearing loss, evaluating other differential diagnoses or coexisting diseases, and preoperative anatomic assessment.\textsuperscript{2,3,6,8} Most of our patients underwent imaging for preoperative evaluation before stapes prosthesis and cochlear implant insertion for fenestral and retrofenestral otosclerosis, respectively.

There was a statistically significant difference between patients with otosclerosis who underwent CBCT versus MDCT in patients with normal hearing. This was expected because there is a trend at our institution to perform more CBCT for patients with conductive hearing loss. MDCT is preferred at our institution for the entities commonly seen in the normal-hearing population, including facial paralysis and vertigo. We would not expect a difference in terms of measurement based on the 2 modalities.

Prior literature has suggested that MDCT and CBCT are limited in cases of tiny foci of $<1\, \text{mm}$, superficial foci, inactive disease, and density variation of $<200\, \text{HUY}$.\textsuperscript{19–22} In addition, CBCT has been found to have a 0\% sensitivity for detection of otosclerosis in the sclerotic phase; however, detection was based on a visual grading system with no standardized measurement techniques used.\textsuperscript{19} As in our study, Swartz et al\textsuperscript{17} described the presence of a bony excrescence or a bulging contour of the otic capsule anterior to the oval window as a helpful finding in otosclerosis; however, the interval improvement in MDCT technology now enables reproducible quantitative measurements in standardized planes, improving accuracy and reproducibility. A recent systematic review suggests that high-resolution CT has a low sensitivity of 58\%, high specificity of 95\%, and a high positive predictive value of 92\% but is limited in submillimeter disease, retrofenestral disease, and dense sclerotic lesions.\textsuperscript{24} Our study improves the sensitivity and specificity for the diagnosis of otosclerosis using a standardized plane and clearly defined landmarks for quantitative assessment of the otic capsule, regardless of phase of disease.

A limitation of this study was that there was no histologic confirmation of disease as a definitive diagnosis of the degree of disease activity. However, our patients met clinical features sufficient to confirm otosclerosis. In addition, the characterization of the phase of disease was subjective on the basis of perceptual differences in otic capsule density. While there are currently no quantitative measures of CT density to define the different phases of disease, our study shows that the thickness of the otic capsule and the presence of a bulging contour are independent of the phase of disease. In addition, our patients with otosclerosis had undergone either an MDCT or CBCT but not both; thus, we could not compare the 2 modalities. Additional study may provide this information to support decision-making for the choice of CT imaging modalities. In addition, submillimeter foci of otosclerosis may be too small to result in a contour bulge or significant thickening of the otic capsule; however, these lesions may be less likely to result in conductive hearing loss. Another limitation is that this study was performed at 1 institution. While our study showed excellent interobserver reliability, further studies at other institutions may help confirm reproducibility. Finally, other osteodystrophies could produce a bulging contour, though entities such as Paget disease would not be expected to involve only the area anterior to the oval window.

**CONCLUSIONS**

Using a standardized axial plane parallel to the lateral semicircular canal, a bulging or convex contour of the otic capsule relative to a line drawn between the anterior margin of the oval window and the cochleariform process occurred with high specificity and positive predictive value in patients with otosclerosis. The otic capsule along the anterior margin of the oval window at the level of the cochleariform process is significantly thicker in patients with otosclerosis compared with individuals with normal hearing. Use of a quantitative assessment of the otic capsule may help the radiologist accurately diagnose otosclerosis.

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