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Anatomical Risk Factors Identifiable on Computed Tomography in Otosclerosis

Otosklerozda Bilgisayarlı Tomografide Demonstre Edilebilen Anatomik Risk Faktörleri

Eda Cingöz[1], Mehmet Cingöz[2]

[1]University of Health Sciences Turkey, İstanbul Bağcılar Training and Research Hospital, Department of Radiology, İstanbul, Turkey
[2]University of Health Sciences Turkey, Başakşehir Çam and Sakura City Hospital, Department of Radiology, İstanbul, Turkey

Abstract

Objective: The aim of the current study was to evaluate the relationship between the dimensions of the temporal bone, high-riding jugular bulbs, and jugular bulb dehiscence in patients with otosclerosis compared with healthy controls.

Method: Two radiologists retrospectively evaluated high-resolution computed tomography images of the temporal bone from 34 patients radiologically diagnosed with otosclerosis, along with images from 34 age- and sex-matched control subjects for comparison. Measurements of temporal bone, including the length and width of the petrous bone and the angle between the midsagittal line and the petrous bone, were performed on a defined standardized slice orientation using a workstation. High-riding jugular bulbs and jugular bulb dehiscence were noted in both groups.

Results: The study cohort of 68 subjects (34 patients and 34 controls) with an average age of 48.08±11.23 years showed no significant differences in the presence of high jugular bulbs and jugular bulb dehiscence. The lengths, widths, and angles of the petrous bone were similar in both groups when analyzed bilaterally, as well as in comparisons between the affected sides of patients and the corresponding sides of the healthy controls.

Conclusion: The current study revealed that neither high-riding jugular bulb nor jugular bulb dehiscence was significantly associated with otosclerosis. Additionally, the dimensions of the petrous part of the temporal bone were not associated with the disease. Identifying the anatomical factors associated with or not associated with otosclerosis can be instrumental in clarifying its etiology and guiding future research toward a more comprehensive understanding of the disease.

Keywords: Fissula ante fenestram, jugular bulb, otosclerosis, temporal bone

Öz

Amaç: Bu çalışmanın amacı, otoskleroz hastalarında temporal kemiğin boyutları, yüksek konumlu juguler bulbus ve juguler bulbus dehisansı ile olan ilişkileri sağlıklı kontrollerle karşılaştırmalı olarak değerlendirmektir.

Yöntem: Otoskleroz tanılı 34 hasta ve yaş ve cinsiyet açısından eşleştirilmiş 34 kontrolün temporal kemiğine ait yüksek çözünürlüklü bilgisayarlı tomografi görüntülerini iki radyolog retrospektif olarak değerlendirmiştir. Temporal kemik ölçümleri, belirlenen standart kesitte bir iş istasyonu kullanılarak gerçekleştirilmiş ve petroz kemiğin uzunluğu, genişliği ve midsagittal çizgi ile petroz kemiği arasındaki açı ölçülmüştür. Her iki grupta da yüksek konumlu juguler bulbus ve juguler bulbus dehisansı not edilmiştir.

Bulgular: Ortalama yaşı 48,08±11,23 yıl olan 68 katılımcı (34 hasta ve 34 kontrol) arasında yüksek juguler bulbus ve juguler bulbus dehisansının varlığı açısından anlamlı bir fark bulunmamıştır. Petroz kemiğin uzunlukları, genişlikleri ve açıları, her iki grup arasında ve aynı zamanda hastaların etkilenen tarafları ile sağlıklı kontrollerin karşılık gelen tarafları arasında yapılan karşılaştırmalarda benzer bulunmuştur.

Sonuç: Bu çalışma, yüksek konumlu juguler bulbusun ve juguler bulbus dehisansının otoskleroz ile anlamlı bir ilişkisi olmadığını ortaya koymaktadır. Ayrıca, temporal kemiğin petroz kısmının boyutları da hastalıkla herhangi bir ilişki göstermemiştir. Otoskleroz ile ilişkili ya da ilişkisiz olan anatomik faktörlerin belirlenmesi, hastalığın etiyolojisinin açıklanmasına yardımcı olabilir ve hastalığın daha kapsamlı anlaşılması yönünde gelecekteki araştırmalara rehberlik edebilir.

Anahtar kelimeler: Fissula ante fenestram, juguler bulbus, otoskleroz, temporal kemik

Address for Correspondence: Eda Cingöz, University of Health Sciences Turkey, İstanbul Bağcılar Training and Research Hospital, Department of Radiology, İstanbul, Turkey
E-mail: edacanipek@gmail.com **ORCID:** orcid.org/0000-0003-0814-4597 **Received:** 07.05.2024 **Accepted:** 25.11.2024

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Introduction

Otosclerosis, also known as otospongiosis, is a primary osteodystrophy of the otic capsule and is a major cause of acquired hearing loss in adults. Otosclerosis is generally considered idiopathic and involves complex remodeling and subsequent fixation of the stapes bone. Such fixation significantly impairs the transmission of sound to the inner ear, potentially leading to conductive, sensorineural, or mixed types of hearing loss and results in progressive auditory impairment (1). Otosclerosis manifests in two primary forms: fenestral and retrofenestral. The fenestral type, also known as the stapedial type, primarily affects the oval window and stapes footplate. In this form, hearing loss is predominantly conductive, resulting from the thickening and fixation of stapes. On the other hand, the retrofenestral or cochlear type involves the cochlea and involves demineralization of the cochlear capsule. This typically results in sensorineural hearing loss, although the exact mechanism remains unclear. In retrofenestral otosclerosis, the prefix "retro" indicates "behind" or "deep to," rather than "posterior," referencing the medial wall of the middle ear as observed through otoscopy. Often presenting alongside fenestral involvement, these two manifestations are not separate entities; rather, they point toward a continuum, highlighting the progressive and interconnected nature of the condition (2). Retrofenestral otosclerosis is characterized by areas of demineralized spongy vascular bone within the cochlear capsule, which may further encroach upon the vestibule, semicircular canals, and internal auditory canal (Figure 1). The promontory often exhibits a pinkish hue visible through the tympanic membrane, a phenomenon known as the Schwartze sign (1,3,4). The etiology of sensorineural hearing loss in this context is hypothesized to stem from direct damage to the cochlea and spiral ligament due to lytic activity or the release of proteolytic enzymes (1,3,4). In fenestral otosclerosis, the disease process primarily targets the lateral wall of the bony labyrinth, with spongy new bone typically developing in the area of the fistula ante fenestrae. The latter is a fibrocartilaginous cleft located just anterior to the oval window, bridging the inner and middle ear (Figure 2). The pathology gradually progresses to encompass the entire footplate of the stapes, potentially extending to the cochlea. Involvement of the annular ligament induces mechanical fixation at the stapedo-vestibular joint, culminating in characteristic conductive hearing loss and an audiometric air-bone gap, often referred to as Carhart's notch (1,4).

Although the precise etiology of otosclerosis remains elusive, genetic predisposition is widely believed to contribute significantly to its development (5,6). This phenomenon is also associated with potential factors such as viral infections, disrupted bone metabolism, inflammatory and hormonal dynamics, and autoimmune responses (7,8). In addition to these factors, the relationship between anatomical characteristics and otosclerosis was evaluated. Temporal changes in bone size or shape can influence sound transmission mechanics in the middle and inner ear. Deviations in dimensions like the petrous bone length or cochlear aqueduct width may alter ossicle

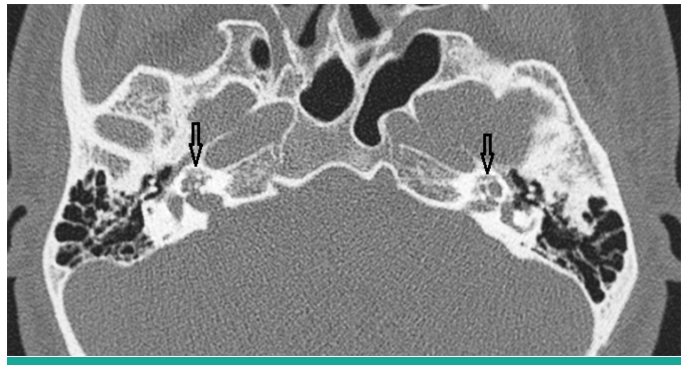

Figure 1. Temporal high-resolution computed tomography scan of a 27-year-old male patient presenting with tinnitus revealed bilateral retrofenestral otosclerosis, as indicated by black arrows

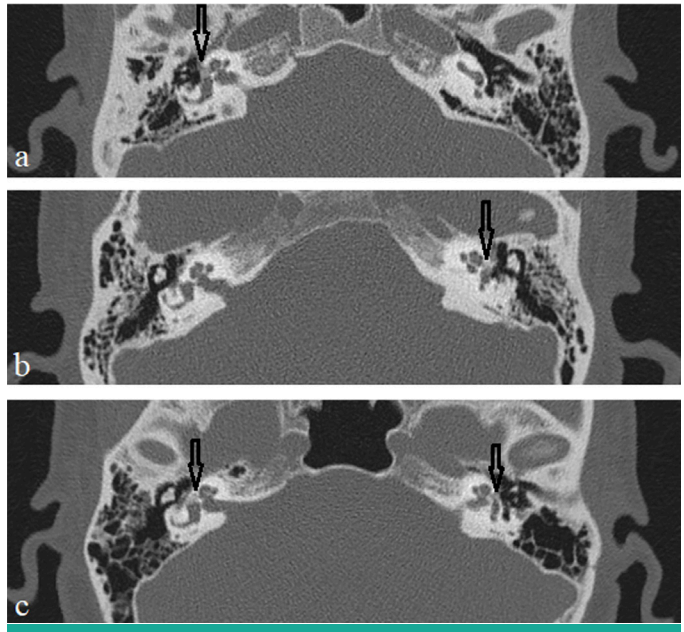

Figure 2. (a) Right fenestral otosclerosis in a 27-year-old female patient (black arrow), (b) Left fenestral otosclerosis is evident in a 27-year-old male patient (black arrow). Bilateral otosclerosis is observed in a 55-year-old female patient (black arrows)

movement, especially that of the stapes, potentially leading to fixation and stress on the stapes footplate, as seen in fenestral otosclerosis. Abnormal alignment of middle ear structures can increase mechanical stress on the stapes, promoting progressive bone remodeling. Temporal bone dimensions, particularly around the otic capsule and cochlea, may affect blood flow and metabolism, and restricted venous drainage can create hypoxia, stimulating abnormal bone remodeling in otosclerosis. Structural differences in the fissula ante fenestram due to abnormal temporal bone dimensions may also increase susceptibility to otosclerosis. A significant narrowing of the dimensions of the facial canal and cochlear aqueduct suggests that these structures are associated with otosclerosis and that anatomical variations may play a crucial role in its pathogenesis (9). The primary objective of the current study was to determine whether temporal bone dimensions are associated with the development of fenestral otosclerosis. Additionally, the relationship between otosclerosis and the presence of a high jugular bulb and jugular bulb dehiscence was evaluated.

Materials and Methods

The current retrospective study was conducted with the approval of the University of Health Sciences Turkey, İstanbul Bağcılar Training and Research Hospital, Clinical Research Ethics Committee (date: 22.12.2023/file number: 2023/12/09/086), and informed consent was waived due to the retrospective design of the study. The study included a total of 68 subjects with 34 patients radiologically diagnosed with fenestral otosclerosis aged between 41 and 55 years, alongside a control group of 34 healthy subjects matched for age and gender. The inclusion criteria for the patient cohort were a radiologic diagnosis of fenestral otosclerosis and an age of over 18 years. The exclusion criteria for both groups were the presence of retrofenestral otosclerosis, external otitis, otitis media, inner ear abnormalities, tumoral mass, history of otic surgery, semicircular canal dehiscence, and motion artifacts that could compromise diagnostic accuracy. Temporal bone high-resolution computed tomography (HRCT) images collected between January 2023 and January 2024 from subjects included in the study were retrieved from the local Picture Archiving and Communication Systems and transferred to the workstation. The data were retrospectively evaluated using the Syngo.via Siemens program. HRCT examinations were performed without the use of contrast media using a 128-detector sequenced multislice scanner (Ingenuity, Philips, Amsterdam, Netherlands). The imaging parameters were as follows: tube voltage: 120 kV; tube current: 783 mAs; slice thickness: 0.625 mm; field of view: 200; matrix: 768×768; pitch: 0.25 and rotation time: 0.42 s. The images were evaluated by two radiologists in the bone window with a window width of 4095 HU and window level of 600 HU at the workstation, enabling multiplanar reformatted images.

The axial plane was angled to align precisely with the cochlear aperture and bony orbital roof, resulting in a consistent slice orientation, as reported in previous studies in which the temporal bone was examined (Figure 3) (10). Measurements were performed by two board-certified radiologists who reached consensus. A total of 136 temporal bones from the 68 individuals were examined in the axial view that displayed the basal turn of the cochlea at its maximum length, corresponding to the defined standardized axial plane or running parallel to it (Figure 4). The following parameters were recorded: a. The length of the petrous bone from the apex to the external base at its maximum extent is aligned parallel to the primary orientation of the basal turn of the cochlea. The width of the petrous bone centered on the cochlea and perpendicular to the length of the basal turn of the cochlea at its maximum, c. The angle is formed by the length of petrous bone and the midsagittal line. The presence of high-riding jugular bulbs and jugular bulb dehiscence was also noted along with the sides affected.

Statistical Analysis

Data entry and statistical analysis were performed using SPSS for Windows version 18.0 (SPSS Inc., Chicago, IL, USA). The suitability of data for normal distribution was assessed using both visual (histograms and probability plots) and analytical methods (Shapiro-Wilk test). Numerical data were evaluated using means, standard deviations, and medians (interquartile range 1st-3rd); frequency distributions and percentages were used for summarizing categorical data. Non-normally distributed numerical data were analyzed using the Mann-Whitney U test.

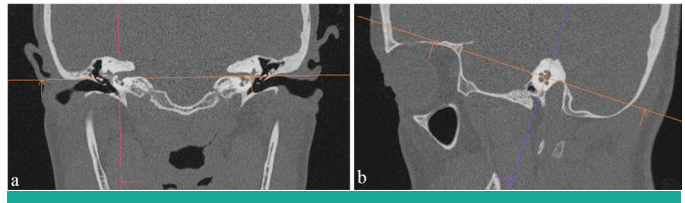

Figure 3. Observation of the bilateral scutum, Prussak's space, and ossicles in the coronal plane shifted the examination to the sagittal plane where a line connecting the cochlear aperture and the orbital roof was drawn. This line provides a guide for further axial assessments

The Pearson chi-square test was used to compare categorical data. Statistical significance was considered at $p<0.05$.

Results

The study cohort comprised 68 participants who were evenly divided into two groups: 34 patients diagnosed with fenestral otosclerosis and 34 age- and sex-matched healthy subjects. The average age of the participants was 48.08±11.23 years, with 44 (64.7%) males and 24 (35.3%) females. The distribution of otosclerosis were as follows: bilateral in 15 (44.1%) patients, left-sided in 11 (32.4%) patients, and right-sided in 8 (23.5%) patients.

High jugular bulbs were present in 55.9% of patients with fenestral otosclerosis and 50.0% of healthy individuals (p=0.627). Among patients with fenestral otosclerosis who had a high jugular bulb, 47.4% had a right jugular bulb, 21.1% had a left jugular bulb, and 31.6% had a bilateral bulb. In the healthy group with a high jugular bulb, 58.8% were found on the right, 29.4% on the left, and 11.8% were bilateral. Jugular bulb dehiscence was observed in 20.6% of patients with fenestral otosclerosis and 23.5% of healthy subjects (p=0.770). Among the patients with jugular bulb dehiscence, 85.7% had it on the right side and 14.3% on the left. In the healthy group, 75.0% of individuals presented with jugular bulb dehiscence on the right, 12.5% on the left, and 12.5% bilaterally (Table 1).

The measurements of petrous bone length, width, and angle were similar for the right and left sides in both the patient and healthy groups (p>0.05) (Table 2). A comparison of the measurements of petrous bone length, width, and angle on the affected sides of patients with the corresponding sides of the controls, as well as on both right and left sides, showed no significant differences between the patient and healthy groups (Table 3).

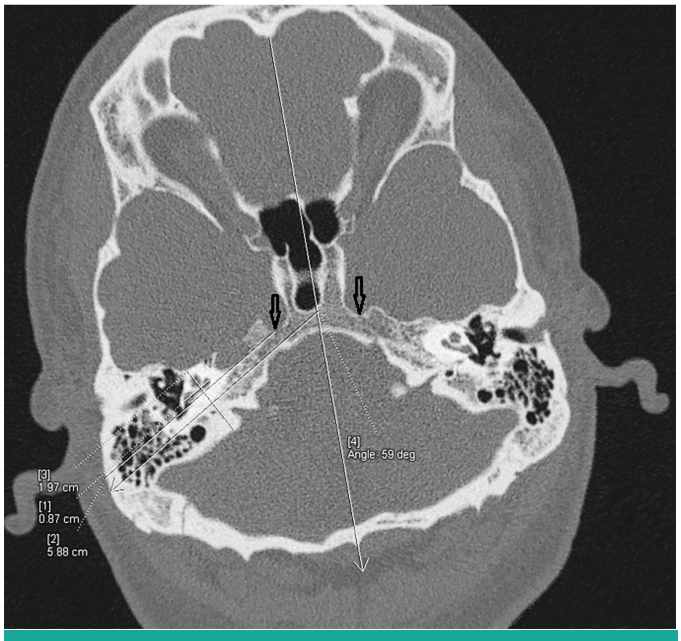

Figure 4. Measurements in the specified axial plane at the workstation using pre-optimized parameters. A line parallel to the cochlear basal turn, as indicated by [1], served as the reference for measuring the length of the petrous bone from the apex to the external base at its maximum extent, as shown in [2] (the petrous apex is indicated by a black arrow at its most medial point). Additionally, [3] the width of the petrous bone, centered on the cochlea and perpendicular to the length of the cochlea's basal turn at its maximum, and [4] the angle formed by the length of the petrous bone and the midsagittal line were measured

Discussion

Hearing loss can substantially diminish the quality of life because of challenges in adaptation and increased social isolation. One of the prevalent causes of hearing loss is otosclerosis, a disease that primarily targets the bony labyrinth of the ear. The pathological changes typically commence near the fissula ante fenestram and subsequently spread via the vascular canals to other parts of the temporal bone. Although there are no definitive data on the etiology or factors that may predispose individuals to otosclerosis, the literature is full of studies that aim to better understand the underlying mechanisms and potential risk factors associated with the development of the disease. Drabkin et al. (11) demonstrated that a *SMARCA4* mutation that causes human otosclerosis produces a

Table 1. Comparison of the prevalence of high jugular bulb and jugular bulb dehiscence between patients and healthy controls

Feature		Patient group (n=34)	Control group (n=34)	p*
		n (%)	n (%)	
High jugular bulb	Present	19 (55.9)	17 (50.0)	0.627
	Absent	15 (44.1)	17 (50.0)	
Side affected by a high jugular bulb (n=36)	Right	9 (47.4)	10 (58.8)	
	Left	4 (21.1)	5 (29.4)	-
	Bilateral	6 (31.6)	2 (11.8)	
Jugular bulb dehiscence	Present	7 (20.6)	8 (23.5)	0.770
	Absent	27 (79.4)	26 (76.5)	
Side affected by jugular bulb dehiscence (n=15)	Right	6 (85.7)	6 (75.0)	
	Left	1 (14.3)	1 (12.5)	-
	Bilateral	-	1 (12.5)	

*: Pearson's chi-square test

Table 2. Comparison of right and left side measurements of petrous bone between the patient and control groups

Measurements	Patient group (n=34)	Control group (n=34)	p*
	Mean ± SD Median (1st-3rd quartile)	Mean ± SD Median (1st-3rd quartile)	
Petrous bone length, R (mm)	63.32±5.04 63.20 (59.45-66.45)	62.89±5.34 63.25 (58.35-67.55)	0.849
Petrous bone width R (mm)	19.42±2.62 19.15 (18.07-20.75)	19.35±3.11 19.70 (17.42-21.42)	0.907
Petrous bone angle, R (°)	53.26±5.47 55.00 (49.00-57.00)	53.23±4.88 54.00 (49.75-56,25)	0.768
Petrous bone length L (mm)	62.11±4.70 61.15 (59.15-64.42)	62.44±4.84 62.50 (58.67-67.02)	0.690
Petrous bone width L (mm)	19.63±2.30 19.35 (18.17-20.40)	19.27±2.94 19.05 (16.75-20.80)	0.611
Petrous bone angle L (°)	53.38±4.41 54.00 (48.75-55.25)	52.26±4.53 52.00 (48.75-56.25)	0.844

*: Mann-Whitney U test, SD: Standard deviation, R: Right, L: Left

Table 3. Comparison of measurements of petrous bone between the affected and healthy sides

Measurements	Patient group	Control group	p*
	Mean ± SD Median (1st-3rd quartile)	Mean ± SD Median (1st-3rd quartile)	
Petrous bone length R (mm) (n=23)	62.92±5.45 61.20 (59.00-65.20)	63.25±4.96 64.00 (59.20-67.50)	0.775
Petrous bone width R (mm) (n=23)	19.41±3.03 17.60 (21.50)	19.13±3.22 18.70 (17.70-21.60)	0.886
Petrous bone angle, R (°) (n=23)	53.82±5.81 55.00 (50.00-58.00)	53.65±4.56 54.00 (50.00-56.00)	0.636
Petrous bone length L (mm) (n=26)	619.8±4.48 61.55 (60.00-64.15)	62.43±4.86 62.50 (58.75-66.85)	0.687
Petrous bone width L (mm) (n=26)	19.49±2.12 19.35 (18.17-20.32)	19.45±2.90 19.05 (17.22-20.80)	0.985
Petrous bone angle (°) (n=26)	52.60±4.48 54.00 (49.00-56.00)	52.53±4.64 53.00 (48.75-57.00)	0.979

*: Mann-Whitney U test, SD: Standard deviation, R: Right, L: Left

similar phenotype in mice, highlighting the critical role of genetic factors. Moss (7) demonstrated the relationship between measles and otosclerosis, illustrating the impact of infections on the etiology of the disease and suggested that viral exposure could influence its pathogenesis. Horner (8) suggested that hormone replacement therapies, specifically those combining estrogen and progestin, may increase the risk of developing otosclerosis and associated vestibular disorders. The same study also noted that hyperprolactinemia may similarly elevate the risk of otosclerosis, thereby highlighting hormonal influence as a significant factor in its etiology.

The relationship between otosclerosis and various anatomical factors has also been extensively studied, yielding significant findings that further our understanding of the complexities involved in the etiology of the disease. A high jugular bulb may exert pressure on middle ear structures, affecting sound transmission and potentially leading to stapes fixation and abnormal bone remodeling in otosclerosis. This condition may impair venous drainage, causing hypoxia or metabolic disturbances that could trigger or worsen otosclerosis. Additionally, disrupting the inner ear anatomy may contribute to sensorineural hearing loss, and mechanical irritation or altered fluid dynamics may lead to inflammation or immune responses, thereby influencing otosclerosis development. Gillet et al. (12) showed that alterations in the stapes footplate can be observed in otosclerosis. The stapes footplate was found to be thickened in patients when examined using only the stapes axial plane. Other studies have revealed a complex array of anatomical factors beyond just the thickening of the stapes footplate. These include significant narrowing

of the facial canal and cochlear aqueduct, among other structural abnormalities, as reported by Cakmak and Cakmak (9). These studies underscore the multifaceted nature of otosclerosis and the need for a comprehensive understanding of the impact of its anatomical features. Friedmann et al. reported no relationship between high jugular bulb and otosclerosis. Building on their findings, our research additionally demonstrated that there was no association between dehiscence of the jugular bulb and otosclerosis, identifying an additional anatomical factor that does not influence the condition (13). Additionally, the current study also investigated the anatomy of the petrous bone as a potential factor in the etiology of otosclerosis. Paetz et al. (10) previously approached the length and width of petrous bone from a developmental perspective. Building upon this, we investigated whether there were any differences in these parameters between patients with otosclerosis and healthy controls; however, we did not find any significant correlation. Lloyd et al. (14) examined the developmental aspects of petrous bone angle. Based on this study, we investigated petrous bone angle in patients with otosclerosis; however, we could not identify any statistically significant differences between the patient and control groups.

The etiology of otosclerosis is multifactorial and has not been fully elucidated. A number of factors can be implicated, including anatomical factors. In the current study, we examined the potential correlation between otosclerosis and anatomical factors, such as a high-riding jugular bulb and its dehiscence. Our study contributes to the existing literature by confirming and reinforcing previous findings of the lack of a significant relationship between otosclerosis and the high-riding status of the jugular bulb. Additionally, we confirmed the absence of any association with jugular bulb dehiscence. We also investigated the potential association between otosclerosis and temporal bone anatomy. Using established measurements from prior studies, we compared the length, width, and angle of petrous bone among cohorts of both healthy individuals and those affected by otosclerosis. We not only compared the measurements between the patient and control groups bilaterally but also the affected sides of patients with the corresponding sides of healthy controls. Nonetheless, statistical analyses revealed no significant differences between the two groups, suggesting that the anatomical dimensions of the petrous part of the temporal bone are not associated with otosclerosis.

Conclusion

Anatomical features such as the dimensions of the facial canal and cochlear aqueduct have been implicated in the etiology of otosclerosis. Previous studies have identified anatomical features, such as a high-riding jugular bulb, which are not significantly associated with the disease. In the current study, we aimed to conduct further anatomical investigations by examining the relationship between jugular bulb dehiscence and otosclerosis; however, our data indicated no significant correlation. Additionally, we performed a comprehensive analysis of the length, width, and angle of the petrous part of the temporal bone and observed no association with otosclerosis compared with healthy controls. These findings indicate that although certain anatomical structures are linked to otosclerosis, others do not contribute to its pathogenesis. Future studies should delve deeper into both the associated and non-associated anatomical factors of otosclerosis to advance our understanding of its etiology.

Ethics

Ethics Committee Approval: This study was approved by the University of Health Sciences Turkey, İstanbul Bağcılar Training and Research Hospital, Clinical Research Ethics Committee of (date: 22.12.2023/file number: 2023/12/09/086). The study was conducted in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 2000.

Informed Consent: Informed consent was waived due to the retrospective study design.

Footnotes

Authorship Contributions

Concept: E.C., Design: E.C., Data Collection or Processing: M.C. Analysis or Interpretation: E.C. Literature Research: M.C. Writing: E.C., M.C.

Conflict of Interest: No conflict of interest was declared by the authors.

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