

Case Report

Imaging in Otosclerosis: Behind the Echoes of the Cochlea

Pramod Vijayasarathy* 

Department of Radiodiagnosis, Kempegowda Institute of Medical Sciences Hospital and Research Centre, Bangalore, India

Abstract

Otosclerosis is a primary bone disorder of the otic capsule characterized by abnormal bone remodeling, leading to progressive conductive or mixed hearing loss, often accompanied by tinnitus. It affects approximately 0.3-0.4% of Caucasians, with lower prevalence in Asian and African populations, and accounts for 5-10% of adult-onset conductive hearing loss worldwide. Beyond auditory dysfunction, patients may present with associated symptoms such as tinnitus, vertigo, and, less commonly, rhinitis-like complaints due to Eustachian tube dysfunction, impacting quality of life. We present two cases—one fenestral and one cochlear otosclerosis—illustrating the clinical spectrum. HRCT remains the gold standard, while MRI assists in assessing cochlear involvement and excluding mimics. Advanced modalities, including ultra-high-resolution CT and photon-counting detector CT, further enhance lesion characterization. Given its global burden and potential for misdiagnosis, this study underscores the need for heightened awareness, timely imaging, and close collaboration between radiologists and otolaryngologists to optimize outcomes.

Keywords

Otosclerosis, Otospongiosis, HRCT Temporal Bone, MRI Temporal Bone, Hearing Loss, Tinnitus, Cochlea

1. Introduction

Otosclerosis is a hereditary bone dysplasia or focal osseous dyscrasia primarily affecting the endochondral bone of the otic capsule within the temporal bone [1-4]. It is characterized by abnormal resorption of mature dense bone by osteoclasts, replaced by spongy, vascular woven bone [3]. In the early active phase (*otospongiosis*), lesions appear demineralized and vascular, while in the inactive phase they become dense and sclerotic [2, 3, 5, 6]. The most frequent site is the *fissula ante fenestram*, located anterior to the oval window [1, 3]. Progressive hearing loss and tinnitus in otosclerosis impose a significant psychosocial and functional burden, often impairing communication, reducing social interaction, and affecting work productivity. These symptoms can also

negatively impact quality of life, contributing to emotional distress, anxiety, and depression in affected individuals [7]. Here, we present two illustrative cases of otosclerosis followed by a comprehensive discussion of its imaging features, differential diagnosis, and treatment considerations.

2. Methodology

High-resolution CT (HRCT) of the temporal bone was performed using a 16 slice GE brightspeed multidetector CT scanner with thin collimation. Images were acquired at 120 kV and 200-250 mA with a slice thickness of 0.5 mm and a

*Corresponding author: pram.med29@gmail.com (Pramod Vijayasarathy)



high-resolution bone reconstruction algorithm. Axial images were obtained parallel to the infra-orbito-meatal line, followed by multiplanar reformats (coronal and sagittal) to assess the otic capsule, oval window, round window niche, cochlea, and semicircular canals.

3. Case Presentation

Case 1:

A 42-year-old female presented with a two-year history of progressive bilateral conductive hearing loss, with pulsatile tinnitus more pronounced in the right ear. Audiometry confirmed a conductive hearing loss with an air-bone gap. Otoscopic examination was unremarkable.

HRCT Findings: Temporal bone thin coronal and axial sections (0.625 mm thickness) showed small foci of demineralization in the form of hypodense foci anterior to the oval window at the fissula ante fenestram. There was thickening and fixation of the stapes footplate on the right side (Figure 1).

Case 2:

A 38-year-old male presented with a one-year history of progressive bilateral mixed hearing loss (conductive and sensorineural), pulsatile tinnitus, and recurrent episodes of vertigo. Audiometry confirmed mixed hearing loss with an air-bone gap. Otoscopic examination revealed a reddish area behind the tympanic membrane interpreted as the *Schwartz sign*, a clinical marker of active otospongiosis due to vascular spongiotic lesions.

HRCT Findings: Temporal bone thin coronal and axial sections revealed diffuse demineralization around and behind the cochlea in a spiral fashion, termed the “halo sign” (Figure 2).

4. Figures

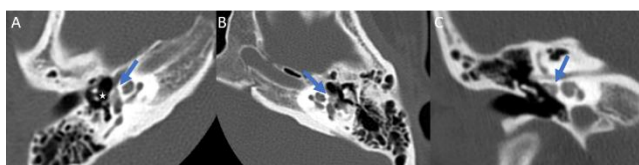


Figure 1. Fenestral Otosclerosis.

- 1) **Figure 1 (A):** Axial HRCT of the right temporal bone shows characteristic hypodense (lucent) foci anterior to the oval window at the fissula ante fenestram (blue arrow) indicating bone demineralization. The stapes footplate is thickened and fixed, consistent with stapes fixation (star).
- 2) **Figure 1 (B):** Axial HRCT of the left temporal bone shows characteristic hypodense (lucent) foci anterior to the oval window at the fissula ante fenestram (blue arrow) indicating bone demineralization.
- 3) **Figure 1 (C):** Coronal HRCT of the right temporal bone shows

characteristic hypodense (lucent) foci anterior to the oval window at the fissula ante fenestram (blue arrow) indicating bone demineralization.

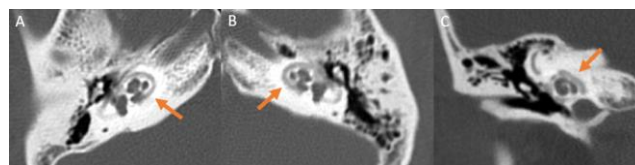


Figure 2. Cochlear Otosclerosis.

- 1) **Figure 2 (A):** Axial HRCT of the right temporal bone reveals diffuse hypodense demineralization around the cochlea, known as the “halo sign” (orange arrow).
- 2) **Figure 2 (B):** Axial HRCT of the left temporal bone reveals diffuse hypodense demineralization around the cochlea (orange arrow).
- 3) **Figure 2 (C):** Coronal HRCT of the right temporal bone reveals diffuse hypodense demineralization around the cochlea (orange arrow).

5. Discussion

5.1. Prevalence and Demographics

Otosclerosis is a major cause of acquired hearing loss, affecting an estimated 15 million people in the United States [3]. The prevalence of clinical otosclerosis is 0.3-0.4% in Caucasians, but only 0.5% in Asians and 0.1% in African-Americans [2-4]. Histological otosclerosis is more common, reported in up to 13% of autopsy studies [3, 4]. Otosclerosis shows bilateral involvement in up to 70-80% of cases. It may be syndromically associated with *Van der Hoeve syndrome*, *osteogenesis imperfecta* (blue sclera, bone fragility), and *Paget's disease*. The disease shows a female predominance (2:1), with onset usually between the second and fifth decades [3, 6, 8].

5.2. Genetic Predisposition and Pathogenesis

Genetic predisposition plays a major role, with autosomal dominant inheritance and incomplete penetrance described [1]. A positive family history is seen in ~60% of cases [4]. To date, eight loci (OTSC1-5, 7, 8, 10) have been mapped. Candidate genes include COL1A1/COL2A1 (collagen synthesis), TGF- β 1, BMP2, BMP4 (bone metabolism), RELN (neuronal migration), VDR (vitamin D regulation), and SERPINF1 [4, 11]. Osteoprotegerin (OPG) has been identified as a protective factor preventing abnormal otic capsule remodeling; OPG-deficient mice mimic otosclerosis [11]. Environmental triggers such as the measles virus may also play a role [11].

5.3. Clinical Presentation and Diagnosis

Patients usually present with *progressive conductive hearing loss*, sometimes progressing to mixed or rarely pure SNHL [2, 5-8]. Associated symptoms include *tinnitus* and less commonly *vertigo* [6, 8]. Audiometry typically shows *air-bone gap*, *Carhart notch*, and absent stapedial reflexes [2, 5, 6]. Otoscopy is often normal, though in some patients the *Schwartz sign* (promontory blush) may be visible [3, 9].

5.4. Imaging Modalities and Findings

High-Resolution CT (HRCT)

HRCT is the *gold standard* for diagnosing and staging otosclerosis [2, 5, 9].

Findings: Fenestral hypodensities at the fissula ante fenestram, footplate thickening, and the “*double-ring/halo sign*” around the cochlea in retrofenestral disease [2, 9].

Sensitivity/Specificity: Reported sensitivity ranges 34-95%, with an average pooled sensitivity ~58% and specificity ~95% [1, 2, 6]. Positive predictive value is >90% [2]. HRCT is most reliable for fenestral disease but less sensitive for tiny foci (<1 mm), retrofenestral lesions, and inactive dense foci [1, 2].

Limitations: Vulnerable to slice thickness >1 mm; hypodense vascular connective tissue can mimic disease leading to false positives [2].

Bone density quantification studies have shown significantly reduced Hounsfield units in otosclerosis compared to normal otic capsule [1, 2].

Magnetic Resonance Imaging (MRI)

MRI is less sensitive than HRCT but can identify active disease and exclude mimics [9].

Findings: Pericochlear intermediate T1 signal, perilabyrinthine enhancement post-contrast, and curvilinear T2 hyperintensities [9].

Utility: Useful in cases of predominant SNHL/tinnitus or when HRCT is negative but suspicion remains. MRI also rules out mimics such as vestibular schwannoma, Paget's disease, or osteogenesis imperfecta [9].

Advanced Imaging Techniques

Recent advances have significantly improved diagnostic yield

Cone-beam CT (CBCT): Ultra-high spatial resolution and reduced dose; useful in preoperative assessment but limited by density calibration and artifacts [10].

Photon-counting detector CT (PCDCT): Converts photons directly to electrical signals, yielding better contrast, fewer artifacts, improved dose efficiency, and superior visualization of foci and prosthesis positioning [10, 11].

Ultra-high-resolution CT (UHRCT): Submillimeter isotropic imaging with improved detection of small fenestral foci compared with standard HRCT [10].

Comparative studies demonstrate that while HRCT remains the standard, PCDCT and UHRCT offer superior visualization of both early fenestral lesions and postoperative prostheses, and may become the new

benchmark for otosclerosis imaging [10, 11]. However, they are not yet standard of care due to cost, accessibility, and technical constraints. They are instead positioned as potential future directions.

5.5. Radiological Classification and Prognosis

The *Veillon classification* is commonly applied [1, 6]:

- 1) *Group I:* No radiological abnormality.
- 2) *Group II (Ia-II):* Minimal lesions without cochlear contact (footplate thickening, small pre-stapedial foci).
- 3) *Group III:* Foci in contact with the cochlear lumen.
- 4) *Group IV (IVa/IVb):* Extensive pericochlear or perivestibular lesions.

Prognostic implications:

Extensive (IV): Associated with poor pre/postoperative thresholds, reduced bone conduction gain, and higher complication rates [1].

Type III lesions: Despite minimal cochlear contact, patients show excellent postoperative outcomes, with >50% achieving >10 dB BC gain [1, 6].

Localized fenestral disease: Predicts favorable outcomes with air-bone gap closure after stapes surgery [6].

Variability in HRCT findings of otosclerosis arises from differences in *scanner technology* (detector type, slice thickness, reconstruction algorithms), *radiologist expertise*, and the *stage of disease* at presentation. Early otospongiotic foci may be subtle and easily overlooked, whereas advanced sclerotic changes are more conspicuous. These factors explain the wide range of reported detection rates and diagnostic accuracy in the literature [12].

5.6. Differential Diagnosis

Several pathologies can mimic otosclerosis radiologically:

- 1) *Osteogenesis imperfecta:* Diffuse otic capsule demineralization plus systemic skeletal fragility, blue sclera [6].
- 2) *Paget's disease:* “Cotton-wool” skull with generalized thickening and coarsening of trabeculae [6].
- 3) *Cholesteatoma:* Soft-tissue lesion with bony erosion of middle ear cavity [5].
- 4) *Osteonecrosis:* Patchy sclerosis and lucency not localized to fissula ante fenestram [5].
- 5) *Otosyphilis:* Aggressive destructive lesions with contrast-enhancing soft tissue [5].

5.7. Treatment

Conventional Approaches

Hearing aids: Beneficial in early/mild cases but do not halt progression [8].

Surgery: *Stapedotomy/stapedectomy* remains the gold standard for fenestral disease, with air-bone gap closure in most patients [1, 8]. *Laser-assisted stapedotomy (CO₂/KTP*

lasers) has improved safety and precision.

Advanced and Adjunctive Treatments

HRCT-assisted surgical planning: Allows preoperative prosthesis length prediction and assessment of anatomical risk factors [1].

Medical therapy:

Sodium fluoride: Previously used, but evidence remains limited [5].

Bisphosphonates: Promising in preliminary studies for stabilizing cochlear disease by inhibiting osteoclast-mediated bone resorption [10] but robust clinical trial data is still lacking to recommend them routinely.

Cochlear implantation: Reserved for profound SNHL or failed stapes surgery, with good functional outcomes in advanced retrofenestral disease [5, 9].

6. Conclusion

HRCT remains the gold standard for diagnosing and staging otosclerosis, while MRI complements by detecting cochlear involvement and excluding mimics. Recent CT innovations further enhance lesion and prosthesis visualization. Timely imaging, combined with multidisciplinary management, is critical for optimal hearing preservation and improved patient outcomes.

7. Teaching Points

- 1) *HRCT* is most sensitive for fenestral disease and essential for surgical planning.
- 2) *MRI* helps detect active disease, cochlear involvement, and rule out differential diagnoses.
- 3) *Advanced CT techniques* (CBCT, UHRCT, PCDCT) provide higher resolution and better postoperative assessment.
- 4) *Collaborative management* between radiologists and ENT specialists ensures the best outcomes.

Abbreviations

BC	Bone Conduction
BMP2/BMP4	Bone Morphogenetic Protein 2 / 4
CBCT	Cone-Beam Computed Tomography
COL1A1/COL2A1	Collagen Type I Alpha 1 / Collagen Type II Alpha 1
ENT	Ear, Nose, and Throat (Otorhinolaryngology)
HRCT	High-Resolution Computed Tomography
MRI	Magnetic Resonance Imaging
OPG	Osteoprotegerin
PCDCT	Photon-Counting Detector Computed Tomography
SNHL	Sensorineural Hearing Loss

TGF-β1

UHRCT

VDR

Transforming Growth Factor Beta 1

Ultra-High-Resolution Computed Tomography

Vitamin D Receptor

Acknowledgments

We extend our heartfelt gratitude to the staff, residents, and technicians of the Radiodiagnosis Department at Kempegowda Institute of Medical Sciences for their unwavering support. We also acknowledge the invaluable assistance provided by the Postgraduates. We sincerely thank the Otorhinolaryngology Department for their significant contributions. Lastly, we are deeply appreciative of the patients who participated in this research.

Author Contributions

Pramod Vijayasarathy is the sole author. The author read and approved the final manuscript.

Patient Consent for Publication

Written informed consent has been obtained. The images are anonymized to protect patient identity and privacy.

Data Availability Statement

Not applicable.

Conflicts of Interest

The author declares no conflicts of interest.

References

- [1] Halily S, Abdulhakeem B, Oukessou Y, Rouadi S, Abada R, Roubal M, Mahtar M. CT scan findings impact on hearing thresholds in otosclerosis: A study of 108 patients. *Ann Med Surg (Lond)*. 2022 May 2; 77: 103716. <https://doi.org/10.1016/j.amsu.2022.103716>
- [2] Kanzara T, Virk JS. Diagnostic performance of high resolution computed tomography in otosclerosis. *World J Clin Cases*. 2017 Jul 16; 5(7): 286-291. <https://doi.org/10.12998/wjcc.v5.i7.286>
- [3] Lin T Rheinboldt M. Fenestral otosclerosis. *Appl Radiol*. 2018; (11): 37-39.
- [4] El Omri M, Mouna B, Chelly S, Ghammem M, Haouas J, Meherzi A, et al. Imaging of Otosclerosis: Radio-surgical Correlation. *EJENTAS*. 2023; 2023: 171089.
- [5] NHS. Otosclerosis [Internet]. 2023 [cited 2024 May 16]. Available from: <https://www.nhs.uk/conditions/otosclerosis/>

- [6] Driscoll CLW, Carlson ML, Patel NS. Otosclerosis. In: Lalwani AK, editor. *Current Diagnosis & Treatment Otolaryngology—Head and Neck Surgery*. 4th ed. McGraw-Hill Education; 2020.
- [7] Leposavić L, Leposavić I, Jasović-Gasić M, Milovanović S, Nikolić-Balkoski G. Psychosocial aspects of acquired hearing impairment in the patients with otosclerosis. *Psychiatr Danub*. 2006 Jun; 18(1-2): 30-8. PMID: 16804497.
- [8] Purohit B, Op de Beeck K, Hermans R. Role of MRI as first-line modality in the detection of previously undiagnosed otosclerosis: a single tertiary institute experience. *Insights Imaging*. 2020 May 19; 11(1): 71.
<https://doi.org/10.1186/s13244-020-00878-3>
- [9] Cai L, Xu Y, Liu Y, Chen Q. Study on Genetic Factors of Otosclerosis: Review. *Ann Otolaryngol Rhinol*. 2022; 9(3): 1289.
- [10] Stankovic KM, McKenna MJ. Current research in otosclerosis. *Curr Opin Otolaryngol Head Neck Surg*. 2006 Oct; 14(5): 347-51. <https://doi.org/10.1097/01.moo.0000244194.97301.19>
- [11] Lazzerini F, Forli F, Capobianco S, et al. Current trends in imaging for otosclerosis and the potential role of photon-counting computed tomography. *Acta Otorhinolaryngol Ital* 2025; 45(3-Suppl. 1): S18-S28.
https://doi.org/10.14639/0392-100X-suppl.1_3-45-2025-A1335
- [12] Lagleyre S, Sorrentino T, Calmels MN, Shin YJ, Escudé B, Deguine O, Fraysse B. Reliability of high-resolution CT scan in diagnosis of otosclerosis. *Otol Neurotol*. 2009 Dec; 30(8): 1152-9. <https://doi.org/10.1097/MAO.0b013e3181c2a084>

Biography



Pramod Vijayasathya, MBBS, MD, DNB, MICR, EDIR, DICR, is a radiologist with extensive expertise in diagnostic imaging. He is currently a Senior Resident at Kempegowda Institute of Medical Sciences, Bangalore, Karnataka, India. Previously, he served as a Specialist

Radiologist at Aster Multispecialty Hospital, Bangalore, where he delivers comprehensive radiological services including MRI, CT, ultrasound, Doppler, mammography, elastography, fluoroscopy, and image-guided procedures. Dr. Pramod has contributed significantly to medical literature with multiple publications in reputed national and international journals. He has also authored book chapters, presented at national and international conferences, and contributed to academic radiology education. His clinical and research interests include cardiac imaging, fetal imaging, neuroimaging, fusion imaging, and interventional techniques. A recipient of academic and professional honors, Dr. Pramod remains committed to advancing imaging sciences and improving patient care through innovation and collaboration.

Research Field

Pramod Vijayasathya: Temporal bone imaging, cardiac imaging, neuroimaging, fetal imaging, fusion imaging, genitourinary imaging, artificial intelligence.