

Use of Cone Beam Computed Tomography in the Diagnosis of Otosclerosis

Ilsón Sepúlveda^{a*}, Thomas Schmidt^b, Enrique Platín^c

^a Radiology Department; Otorhinolaryngology and Head & Neck Surgery Service, General Hospital of Concepción, Cone Beam Concepción, Advanced Craniofacial Imaging Center. Concepción, Chile

^b Otorhinolaryngology Head and Neck Surgery Service, General Hospital of Concepción. University of Concepción School of Medicine, Concepción, Chile

^c School of Dentistry North Carolina University, Chapel Hill, NC, USA

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Corresponding Author:

Sepúlveda. I*

Professor; Radiologist

Email: isepulvedaguilar (at) gmail (dot) com

Schmidt. T

Associate Professor

Platín. E

Clinical Professor

Abstract

Otosclerosis is a unique autosomal dominant focal osteodystrophic disease specific to the otic capsule causing stapedial fixation and conductive hearing loss. Currently, radiology plays a crucial role in its diagnosis with Cone Beam Computed Tomography (CBCT) emerging as a powerful tool in the field of otolaryngology. CBCT is a three dimensional technique that uses lower radiation, has fewer artifacts and offers higher resolution when compared to multislice Computed Tomography (CT). It is considered to be an excellent imaging modality for radiological exploration of the ear.

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1. Introduction

Radiological imaging is needed to determine the presence and extent of disease, treatment planning, monitor disease progression, and assess treatment efficacy (Cakli et al., 2012) High-resolution CT (HRCT) is the imaging method of choice in the evaluation of structural disorders affecting the human temporal bone in cases of conductive and mixed hearing loss with normal tympanic membranes (Liktov et al., 2014; Purohit et al., 2014)

Conventional CT imaging includes multidetector CT (commonly used to acquire 16–64 slices with some machines capable of acquiring up to 320 slices simultaneously), helical progression, and sometimes-dual energy subtraction. CT produces images in slices of the anatomical region in question. Voxels can be non-isotropic (not identical in all planes) compromising the accuracy of measurements. (Cakli et al., 2012)

CBCT is a relatively new radiological technique. It was initially developed for angiography, but it has found multiple applications in Oral and Maxillofacial Radiology (OMFR). The technique is characterized by fast image acquisition with relatively high resolution, lower radiation exposure to patients, and fewer artifacts when compared to conventional CT (Sepulveda et al., 2014).

During a CBCT examination, the single scanner and the detector rotate around the patient's head acquiring from as little as 200 basis images and up to 1,800 depending on the design of the unit. Slice thickness of CBCT varies from 0.075 to 0.5 mm. The digital data of the multi-angle projections are processed by algorithms that reconstruct the volume of the target voxels (Révész et al., 2016; Dahmani-Caussea et al., 2011) This imaging technique has higher resolution than the conventional CT, with a spatial resolution up to 0.08 mm, capable of detecting small structures useful in the preoperative and intraoperative

evaluation of the temporal bone structures. CBCT offers fast imaging of the region of interest making it easier to examine children and claustrophobic adults (Teymoortash et al., 2011; Hiraumi et al., 2016).

Analysis of the dosimetry overwhelmingly shows that the use of CBCT uses considerable lower radiation doses when compared with MSCT. CBCT has an effective radiation dose of 55.2–80 usv (micro Sieverts) for the examination of one temporal bone, compared to 3,600 for the 16-slice CT, and 4,800 usv for the 4-slice CT (Teymoortash et al., 2011) This is made possible by the virtue of the beam geometry, type of emission (pulsed rather than continuous exposure over the entire rotation) as well as more efficient detection (Faccioli et al., 2009).

CBCT has other advantages including the cost of installation and maintenance, installation is more straightforward including the smaller volume and lower heat emission requires less space and less onerous safety measures. Also, machines can be made mobile for use in the operating room (Dahmani-Caussea et al., 2011)

Otosclerosis

Otosclerosis was first described in 1735 by Valsalva as ankyloses of the stapes to the margins of the oval window (Assaly et al., 2013). Otosclerosis, also called ‘otospongiosis’ is a unique autosomal dominant focal osteodystrophic disease specific to the otic capsule, with a 20% to 40% penetrance (Salomone et al., 2008). It consists of one or several circumscribed foci of new, softer, and more vascular bone that replace the avascular endochondrial bone of the adult causing stapedial fixation and conductive hearing loss. (Purohit et al., 2014; Assaly et al., 2013; Lee et al., 2009). In 70 to 90% of cases the disease is bilateral and is infrequent and difficult to diagnose in infancy (Min et al., 2010)

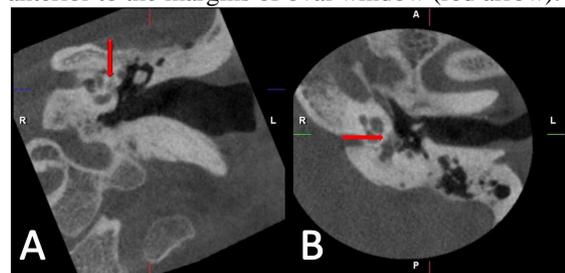
The disease is more common in women than men occurring bilaterally in 85 % of patients, typically presenting in the 2nd- 4th decades of life with conductive hearing loss (CHL), sensorineural hearing loss (SNHL) or mixed hearing loss (MHL) and/or tinnitus. (Liktov et al., 2014; Purohit et al., 2014). Among the Caucasian white adults, the estimated prevalence is approximately 0.3–0.4 % (Révész et al., 2016). Histologic otosclerosis without clinical symptoms is much more common than has been reported between 8–11 % in a large postmortem study (Liktov et al., 2014)

Histopathologically, the active lesions (otospongiosis) can be recognized by their spongy

structure and immature osseous tissue, by the size of the marrow spaces that contain a very cellular connective tissue with numerous osteoclastic giant cells, and most importantly by an increased and dilated vascularity. Inactive lesions (otosclerosis) are characterized by solid compact mosaic-like osseous tissue that contains few and small marrow spaces as well as infrequent and small blood vessels. (Assaly et al., 2013)

Otosclerosis is categorized into two types, fenestral and retrofenestral/ cochlear. The more common fenestral type of otosclerosis involves the lateral wall of the bony labyrinth (Purohit et al., 2014). Fenestral otosclerosis is characterized by aberrant bone apposition in the region of the embryonic fissula ante fenestram, which tends towards the annular ligament (ligament fixation) and also the stapes footplate (stapes ankylosis) producing conductive hearing loss (Purohit et al., 2014; Révész et al., 2016; Pont et al., 2015)

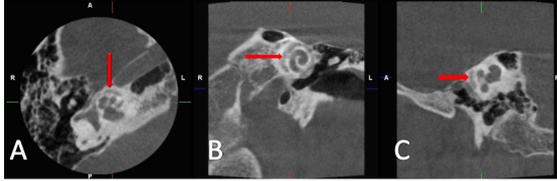
Figure 1: 46 -year-old male patient with clinical history of conductive hearing loss diagnosed with Fenestral Otosclerosis. CBCT of temporal bone (A) coronal (B) axial images show hypodense foci anterior to the margins of oval window (red arrow).



2. Experimental

Retrofenestral or cochlear otosclerosis is much less common; however, it is nearly always associated with fenestral otosclerosis (Purohit et al., 2014). It can cause pure sensorineural hearing loss (SNHL) due to fine architectural changes in the modiolar and in the pericochlear bone (both apical and basal) resulting in decreased elasticity of the spiral ligament (Révész et al., 2016). Several pro-inflammatory cytokines (TNF-alpha, TGF-beta, RANK, RANKL, BMP) may play a potential role in the pathogenesis of otosclerosis-associated sensorineural hearing loss (Liktov et al., 2014)

Figure 2: 24-year-old male patient with clinical history of sensorineural hearing loss diagnosed with Cochlear Otosclerosis. CBCT of temporal bone (A) axial (B) coronal (C) oblique sagittal images show a pericochlearhypodense double ring (red arrow).



examination and audiological tests (such as conductive hearing loss, absent stapedial reflexes, Carhart's notch, and type A tympanogram) (Lee et al., 2009; Min et al., 2010)

Confirmation of the diagnosis requires surgical middle ear exploration (Jin-Young Min). HRCT of the temporal bone is the modality of choice for the preoperative evaluation of otosclerosis (Lee et al., 2009) Sensitivity levels of HRCT scans has been reported to be between 70.5–84.5 % in patients with stapes fixation (Révész et al., 2016). Histomorphologically, active otosclerosis (otospongiosis) is diagnosed in the presence of hypodense or radiolucent foci of demineralization in the otic capsule. Otospongiosis is the early (active) phase of otosclerotic lesions. As the disease progresses, it shifts to the late (inactive) phase of otosclerosis with new spongy bone formation (Assaly et al., 2013; Lee et al., 2009).

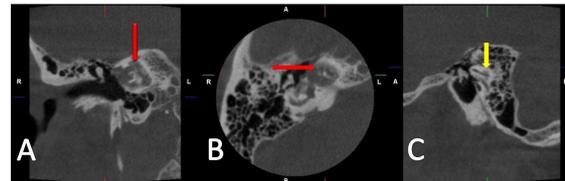
On CT, spongiotic foci approach soft-tissues in attenuation, pericochlearhypodense double ring (cochlear otosclerosis) or anterior to the margins of oval window (fenestralotosclerosis). As the disease progresses, these changes can be spread into any portion of the bony labyrinth including lateral walls of the internal auditory canal (Assaly et al., 2013). The imaging differentials of fenestralotosclerosis are few. The cochlear cleft is a small non-osseous space in the otic capsule in the region of the fissula ante fenestram, commonly seen in children. An inexperienced reader may mistake a cochlear cleft for a demineralized focus in the region of the fissula ante fenestram (Purohit et al., 2014)

The otospongiosis can be classified radiologically into 4 grades, according to the extension of the lesion. Grade 1 is further classified into 1a and 1b; 1a: isolated lesion of the footplate, which is identified as thick (above 0.6) and hypodense. Grade 1b area of pre-stapedialhypodensity below or equal to 1 mm, without extension to the anterior portion of the middle cochlear turn. Grade 2: hypodense lesion with a diameter above 1 mm, without contact with the cochlear lumen. Grade 3:

hypodense pre-stapedial focus in contact with the cochlear lumen. Grade 4 is divided into 4a and 4b as well. Grade 4a: plaques around the cochlea, and 4b plaques around the lumen of the semicircular canals and the vestibule (Pont et al., 2015)

The differential diagnosis is made with otitis media with effusion, congenital fixation of the stapes, disarticulation of the middle ear bones, tympanosclerosis, osteogenesis imperfecta, Paget's disease and congenital cholesteatoma (Salomone et al., 2008)

Figure 3: 60-year-old male patient with clinical history of deep sensorineural hearing loss diagnosed with Cochlear Otosclerosis. CBCT of temporal bone (A) axial (B) coronal (C) oblique sagittal images show a pericochlearhypodense double ring (red arrow) and hypodense foci around the lumen of the lateral semicircular canal (yellow arrow)



The treatment may be medical (anti-enzyme or anti-bone remodeling drugs) or surgical. The treatment of fenestralotosclerosis is primarily surgical with stapedectomy and stapes prosthesis insertion. Patients with cochlear otosclerosis are usually treated medically using fluorides that may limit the growth of active otosclerotic foci and thereby prevent progression of SNHL. Patients with bilateral profound SNHL may derive significant benefit from Cochlear Implant. Personal sound amplification devices (PSAD) are a further option, particularly in patients with surgical contraindications. (Purohit et al. 2014; Salomone et al., 2008)

2. Discussion

M. Dahmani-Causse, et al., assessed the morphologic concordance between CBCT and Multislice Helical on stapes and footplate assessment and measurement of footplate thickness, and quantitatively in terms of dosimetry of 12 temporal bones from fresh human cadavers of unknown clinical history. They concluded that the exploration of the stapes, incudostapedial joint, anterior stapediovestibular joint and footplate were qualitatively more precise on CBCT, and footplate thickness showed less overestimation than on MSCT. CBCT delivered 22 times less radiation than MSCT.

Tsung-Lun Lee, et al., looked at 24 auditory systems of patients with HRCT who had been clinically, surgically and pathologically confirmed with otosclerosis. HRCT was positive in 46% of the clinically, surgically and pathologically confirmed cases. The investigators concluded that HRCT was high in specificity (100%) but low in sensitivity (46%) for the diagnosis of otosclerosis that when compared with the Western literature, may stem from a greater percentage of inactive otosclerosis.

Balázs Liktör, et al., analyzed radiological imaging of histologically active foci of otosclerosis (n = 21). CBCT demonstrated in all of the cases a sensitivity of 100 %. However, CBCT was unable to detect histologically inactive otosclerosis (n = 11) sensitivity = 0 %. They determined that temporal bone CBCT is a useful imaging method in the preoperative evaluation of histologically active fenestral otosclerosis.

Conclusion

When compared with multi detector CT, CBCT is an excellent alternative for the diagnosis of active otosclerosis. It results in lower radiation to patients, fewer artifacts, and equal or higher resolution depending on the unit used.

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