Clinics of Surgery

Review Article ISSN: 2638-1451 | Volume 7

Otosclerosis or Otospongiosis

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Received: 21 Feb 2022
Accepted: 04 Mar 2022
Published: 11 Mar 2022
J Short Name: COS

Citation:

Hamerschimidt R, Otosclerosis or Otospongiosis. Clin Surg. V7(7): 1-4

1. Review

Otosclerosis or otospongiosis is a bone degeneration that occurs in the otic capsule, the bone structure that surrounds the cochlea and labyrinth. Is an aberrant process of bone resorption of the labyrinthine capsule followed by reparative deposition of new, immature sclerotic bone (Abdurehim, 2016) [1]. This disease most often starts at the base of the stapes, which is the smallest bone in the human body, receiving the name of fenestral otosclerosis. Over time, it can progress to the cochlea and even reach the internal auditory meatus. Therefore, it is far from being a simple "calcification" of a small ear bone, requiring correct diagnosis, long-term follow-up, and personalized treatment.

Although is a heterogeneous disorder, with variable familial manifestations in the population, it has been recognized that one that often occurs in multiple members of the same family consists of a monogenic type of inheritance (Ziff, 2016) [2]. So, it is currently considered an autosomal dominant hereditary disease. The disorder is hereditary because it is inherited from parents to children, and autosomal because the genes with this change are not those that determine the person's sex, so it is a disease that can affect both men and women, being more frequent in females in a 3:1 ratio. The dominant term, on the other hand, means that only one gene, from the father or the mother, is enough for a patient to manifest the disease.

The disease only occurs in patients with these genetic mutations, causing bone degeneration of the middle and inner ear, especially of the oval window, where the smallest of the 3 auditory ossicles, the stapes, articulates. It is established that the manifestation of the disease happens because of an unbalance between resorption and formation of new bones, as a consequence of a metabolic disorder in the endochondral layers of the bones and, therefore, a defective remodeling process. (Oliveira, 2018) [3,4] With the alteration of bone metabolism in this region, the stapes become fixed and unable to generate a sound vibration coming from the external auditory canal and tympanic membrane to the cochlea. In other words, there is no mechanical energy transferred by ossicle vibration to the cochlea. The characteristic deafness of otosclerosis patients, therefore, results from the difficulty in conducting sound through the bones (conductive deafness), as opposed to sensorineural deafness, arising from damage to the cochlea itself or the cochlear nerve. However, it is important to emphasize that, in advanced stages of the disease, the patient might show osteosclerotic extension into the cochlea and, thus, a mixed hearing loss with vestibular symptoms can be observed (Cureoglu, 2011).

The term sclerosis was formerly used concerning elderly people with different degrees of cognitive or mental deficit (at that time there was no mention of Alzheimer's or senile dementia). However, otosclerosis appears most often between 20 and 30 years of age and may affect even children. Presbycusis, on the other hand, is a hearing loss that occurs in the elderly due to damage to the cochlear hair cells.

Most patients seek help in adulthood, complaining of progressive unilateral or bilateral hearing loss, which may be accompanied by tinnitus and, more rarely, dizziness. History often reveals cases of deafness in the family, but not always. Clinical examination and appearance of the tympanic membrane on otoscopic examination

in the office are almost always normal. However, the differentiation of conductive and sensorineural deafness is made through the use of a tuning fork. It gives us important information and, associated with a normal tympanic membrane exam and typical clinical history, indicates a probable otosclerosis even before performing the necessary complementary tests.

When it comes to histopathological findings, they've shown to be fundamental to define the phases of the disease: first, the otospongiosis phase, characterized by an increase of osteoclastic activity and the development of local microvascularity; second, the transitional phase, where the spongy bones start to deposit in areas of previous bone absorption; and, finally, the otosclerotic phase, where the deposited spongy bone becomes a denser bone, usually leading to the classical symptoms. (Batson, 2017) [5].

The most important tests that usually define the diagnosis are pure tone/vocal audiometry and immitanciometry. Through them, it is possible to define the conductive hearing loss and some other parameters that suggest the stiffening of the sound transmission mechanism of the middle ear (abnormal ossification around the stapes). Audiometry confirms the conductive deafness and its degree, which were already suspected by the use of the tuning fork. In immitanciometry, two conditions are evaluated: the type of the immittance curve, usually type A or As, and the measure of the stapedial reflex, usually absent. These data are essential for making the differential diagnosis with other pathologies that also cause conductive losses, such as superior semicircular canal dehiscence or Minor's Syndrome, in which there is the presence and not the absence of stapedial reflexes.

In many cases, middle ear and mastoid computed tomography is also used to rule out other causes of deafness or even to confirm otosclerosis and assist in surgical programming. It assesses the anatomy of the oval window, relationship with the facial nerve, cochlear involvement, among others. Even though most diagnoses are made based upon history and otologic examination, radiologic confirmation of the disease might help the clinical management of the patients. Since the gold standard exam, the pathological diagnosis, is usually limited to post-operative or *post-mortem* scenarios, we must find alternatives. A high-resolution CT, for instance, has shown a sensitivity of about 90% in some studies, helping identify a cochlear endosteal involvement, which gives an important prognostic value, suggests the nature of the hearing loss (if the region is preserved, probably the sclerosis is limited to the bony labyrinth) and allows surgical planning. (Quesnel, 2018) [6].

Given the clinical condition caused by otosclerosis, there is a range of therapeutic options depending on the degree of hearing loss, evolution, patient age, presence of comorbidities, among other factors to be analyzed on a case-by-case basis. Treatment can be expectant, medication, hearing aids for individual sound amplification (ISAA), traditional surgery (stapedotomy or stapedectomy), cochlear implants, and, eventually, bone-anchored prostheses. In initial cases, with small hearing loss, especially in one ear and without much repercussion, the first treatment is expectant, that is, follow-up. As deafness evolves, the conventional hearing aid or surgery, stapedotomy, may be indicated, which has a very high success rate in experienced hands. However, even with successful surgery, long-term follow-up should be done, as the disease can continue to progress.

If surgery is indicated, stapedotomy (or stapedectomy) is the surgical treatment of choice. It is a surgery performed for decades, with some changes in technique over time and a great success rate. We currently perform this surgery in a hospital environment, under local anesthesia and sedation, or possibly general anesthesia. In addition, a cost-effectiveness study showed that a stapedectomy is a cost-effective option from a patient perspective in that it had little cost but a maximum health benefit. (Gillard, 2020) [7]. With the aid of a surgical microscope or video endoscope, through the ear canal, we verified and confirmed the stapes fixation by otosclerosis after the detachment of the tympanic membrane. We then remove part of the defective stapes, keeping the base in place, the so-called platinum. The exact measurement is made between the anvil and the stage so that the prosthesis is cut to exactly the right size. This step is extremely important. After we performed a small perforation in the platinum, using manual perforators, drills, or possibly laser, we placed the small prosthesis (6mm by 0.6mm, which after being cut is normally 4.5mm by 0.6mm) made of Teflon or titanium, to reestablish the passage of sound vibrations to the liquid inside the cochlea. The prosthesis is fitted to the second bone, the incus, and goes to the opening of the stapes footplate (Figure 1). In the immediate postoperative period, some patients may experience vertigo, which can last for a few days, more or less intense. Physical exercise, air travel, diving, and any other pressure variation should be avoided for a variable period depending on the orientation of each surgeon and each patient's recovery, usually for 30 days. The surgery has had a great success rate over the decades in which it has been performed, with satisfactory hearing improvement in most cases. It is also noteworthy that tinnitus improvement occurs in many cases after hearing improvement. However, as with all surgery, there are risks related to the procedure, including the risk of hearing deterioration. These risks must be well discussed with the surgeon before surgery.

Despite well-established surgical treatment and good results, many patients may not be operated on. Mild deafness, the coexistence of sensorineural deafness in a sufficient degree that the benefit is not adequate, associated diseases or simply the fear of surgery and its potential risks lead some patients to opt for the use of hearing aids. In otosclerosis, hearing aids work very well. With the cochlea preserved, the adaptation and functional gain promoted by the hearing aid tend to be excellent. However, we know that the disease is progressive and, over time, the increase in deafness can make patients who have used ISADs for a period prefer to try a greater gain

through surgery, which usually has a longer-lasting result. Yet, we emphasize again that surgery is not the cure for otosclerosis, as the bone deposit and resorption process can continue, depending on the genetic load and associated factors, such as pregnancy, for example, which can worsen the condition of deafness or even cause a return to hearing loss after surgery. If there is cochlear involvement and depending on the evolution of deafness in audiometry and cochlear impairment in computed tomography, drug treatment can be performed, with the use of bisphosphonates. They are medications with some controversy in the literature, but they show benefits in many cases, with alendronate being the most used, because these compounds inhibit the action of osteoclasts through inhibition of the farnesyl pyrophosphate synthase, which regulates osteoclastic function. (Gronowicz, 2014) [8]. Often patients undergo combined treatments, in which traditional surgery is associated with the use of hearing aids and medication. In some cases, it is preferred to only use hearing aids and medication, or eventually, traditional surgery and, in the future, due to important cochlear involvement, end up being indicated for cochlear implants. So, the follow-up is fundamental.

In recent years, patients with advanced degrees of otosclerosis and who have not shown satisfactory results with the treatments described above may benefit from implantable hearing aids. Among the options, we have bone-anchored hearing aids (BAHA), in the case of conductive deafness, or cochlear implants, in cases of cochlear evolution with severe or profound deafness where there is no indication for traditional surgery or satisfactory results with the hearing aids. Some studies demonstrated that BAHA comes with a significant increase in global measures of ease of communication and reduction aversiveness to sounds, which makes it a viable candidate. (Steward, 2011) [9]. Next, as an illustration, we placed a photo of a normal cochlea on computed tomography (Figure 2), and then a cochlea with a great involvement by otosclerosis and which was indicated for cochlear iplantation (Figure 3), and then the appearance on tomography of the electrodes of the cochlear implant inserted in the cochlea (Figure 4). Therefore, otosclerosis is a chronic, evolving disease that must be carefully evaluated concerning the appropriate treatment and surgical indication, always weighing the risks and benefits of any treatment instituted, with clinical, audiometric and possibly tomographic follow-up being imperative.

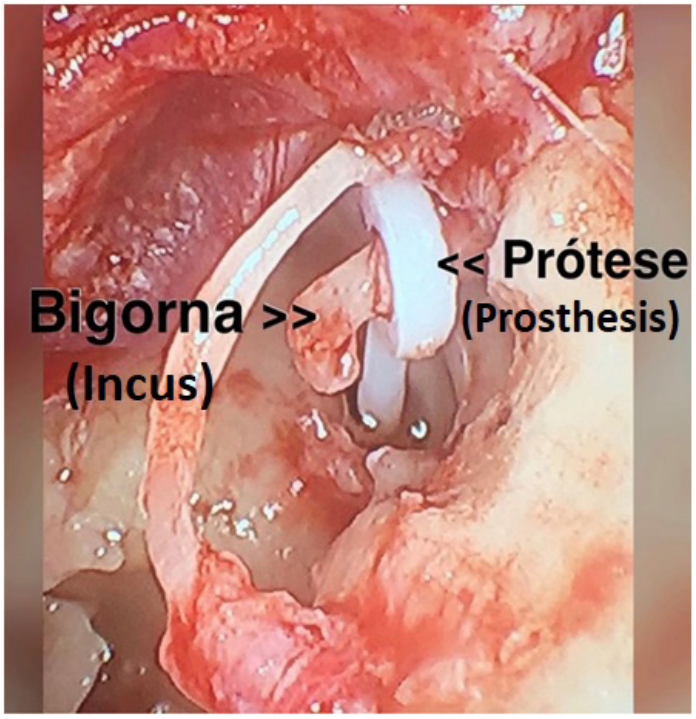

Figure 1: Photo of the teflon prosthesis fitted to the incus, after removal of the stapes, under endoscopic view.

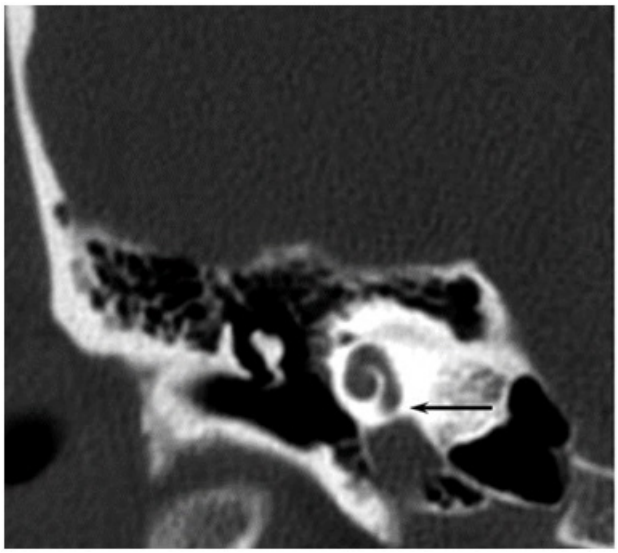

Figure 2: Computed tomography photo of a normal cochlea.

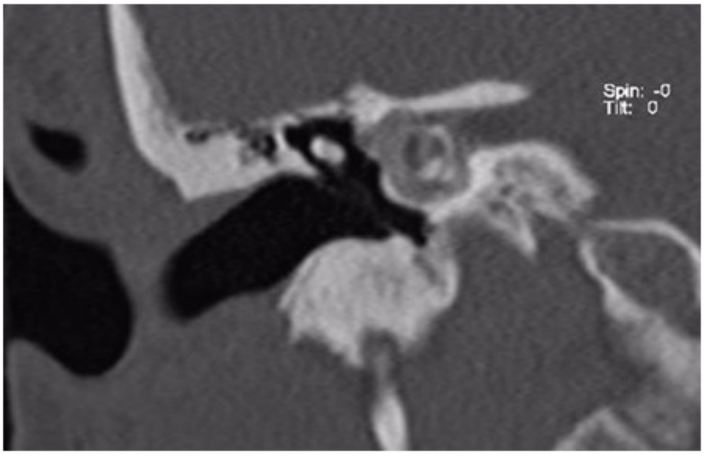

Figure 3: Photo of a cochlea with major bone degeneration caused by cochlear otosclerosis.

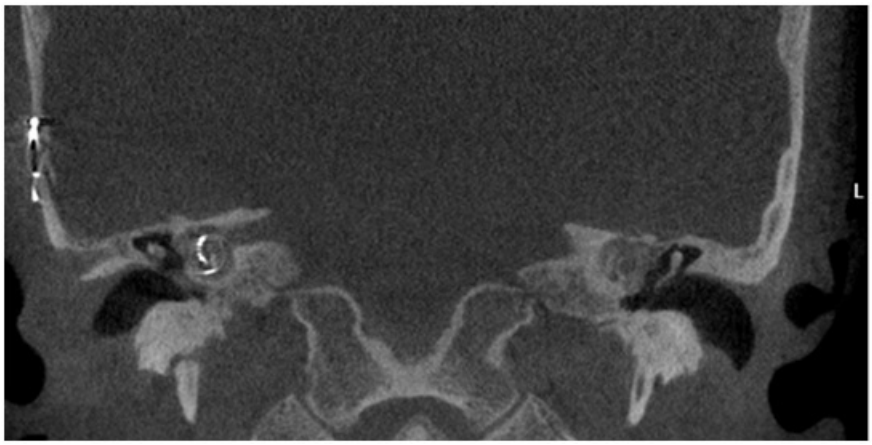

Figure 4: Photo of the electrode bundle placed inside the cochlea in a case of cochlear otosclerosis and profound deafness.

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