Otosclerosis

Diagnosis, Evaluation, Pathology, Surgical Techniques, and Outcomes

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Introduction

Worldwide, the number of patients suffering from otosclerosis has declined considerably. Although this is a cause for rejoicing, it places the modern-day otologist in a unique predicament. It leaves him or her with very little surgical experience in dealing with otosclerosis. The patient who has undergone surgery for otosclerosis many years ago and now faces problems as a result of that surgery are among the many challenges that the otologist of today will need to face. The modern-day otologist will need to overcome his or her own learning curve before being able to perform the surgery reliably and deliver consistently good results.

There are still quite a few nuances that need to be learned if the otologist is to successfully and effectively treat patients with otosclerosis. Our book strives to help today's otologist achieve that goal. The references are limited to just a few. The atlas section on the surgical procedures is provided by internationally acknowledged experts. Their experience and techniques have been gleaned through years of performing this amazingly demanding procedure and consistently delivering excellent results. Each beautiful picture and illustration is worth a thousand words. Their surgical videos are priceless.

We hope that the current trends continue where otosclerosis will just be a faded memory in the history of illnesses that affect humanity. But until then, all otologists will still need to be familiar with all the dimensions that the treatment of otosclerosis brings.

We thank the contributors for sharing their valuable experience, their extraordinarily beautiful illustrations, and their videos. We appreciate their unselfishness and their graciousness. It is their hope, as well as ours, that all who read this book will benefit from it and as a result their patients will benefit from it too.

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PART I

Basic Science

CHAPTER 1

Pathology of Otosclerosis

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INTRODUCTION

Otosclerosis is primarily a disease of localized bone remodeling.[1] It is thought to affect only the bony capsule of the middle and inner ear selectively. Otosclerosis is a process occurring in two phases: (1) active phase as characterized by bone resorption (spongiosis), and (2) phase of remission characterized by bone deposition (sclerosis).

It is a disease affecting enchondral bone of the otic capsule characterized by disordered resorption and deposition of bone.

An otosclerotic focus consists of areas of bone resorption, new bone formation, vascular proliferation, and a connective tissue stroma.

AGE OF ONSET

It can range from 10 to 48 years of age, however, the mean age of onset is commonly the age of 30.[2]

DeJuan[3] in his study reported the onset of clinical otosclerosis to be 28% between the ages of 18 and 21 years, 40% between 21 and 30, and 22% between 31 and 40.[4]

PREVALENCE

The exact incidence remains unclear and next to impossible to determine.

However, in one report autopsy studies conducted by Konigsmark and Gorlin[5] revealed an incidence of 5% to 18% of the general population.

Jahn and Vernick[6] report that 10% of Caucasians have histologic otosclerosis, but only 1% of these develop clinical manifestations of otosclerosis.

Many authors have noted that the incidence of patients suffering from otosclerosis has declined steeply in recent times.

RACE

There appears to be a definite racial predisposition. Caucasians are more predisposed than Africans. Asians too are far less affected than Caucasians, and the prevalence in American Indians seems to be extremely low across the continent.[2]

GENDER

Shambaugh[7] noted a female preponderance in his study. Otosclerosis is not a genetically sex-linked characteristic disease. Thus, a ratio of 1:1 would have been expected.

Hueb et al[8] reported a higher incidence of bilateral otosclerosis in women than men. This prompted them to believe that this would more likely cause women to seek medical advice than men. This in turn could likely explain the gender disparity. On the other hand, endocrinological factors predisposing to the appearance of otosclerotic foci is a possibility to be considered.

OTOSCLEROSIS AND PREGNANCY

There are many reports that associate the onset of hearing loss caused by otosclerosis and the onset of pregnancy. Shambaugh[9] found that in an analysis of 475 female patients suffering from otosclerosis that 50% suffered from hearing impairment with the onset of pregnancy. Although he and many authors note a correlation between the onset of hearing loss following pregnancy, they are not clear on why or how this occurs. As mentioned earlier, endocrinologic factors have been suspected for this occurrence.

TYPES OF OTOSCLEROSIS

1. Histologic otosclerosis
2. Fenestral otosclerosis
3. Cochlear otosclerosis
4. Malignant otosclerosis
5. Far advanced otosclerosis

Histologic Otosclerosis (Figures 1–1 and 1–2)

Histologic otosclerosis is a finding on microscopic examination of temporal bones. The location of the otosclerotic changes is such that the patient suffers no symptoms related to the otosclerosic changes. Therefore, it generally does not involve the stapes bone, the stapediovestibular joint, or the cochlear endosteum. It is therefore asymptomatic.

Fenestral Otosclerosis

This refers to an otosclerotic lesion that involves the stapes bone and/or the stapediovestibular joint. This in turn causes a conductive hearing loss. This is

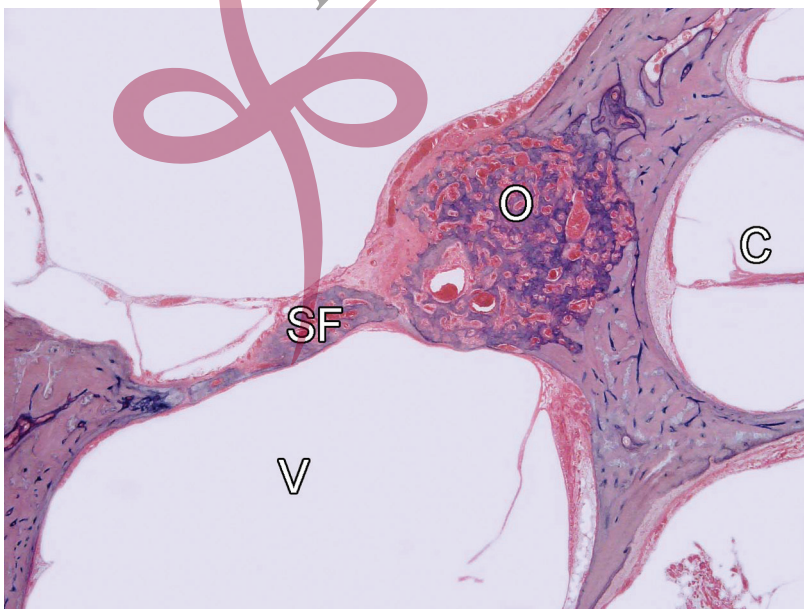

Figure 1–1. Section of human temporal bone demonstrating "histologic otosclerotic" focus. O = "histologic" otosclerotic focus; SF = stapedial footplate; C = cochlea; V = vestibule. Courtesy of University of Minnesota temporal bone collection. Kindly contributed by Dr. Cureoglu and Dr. Paparella.

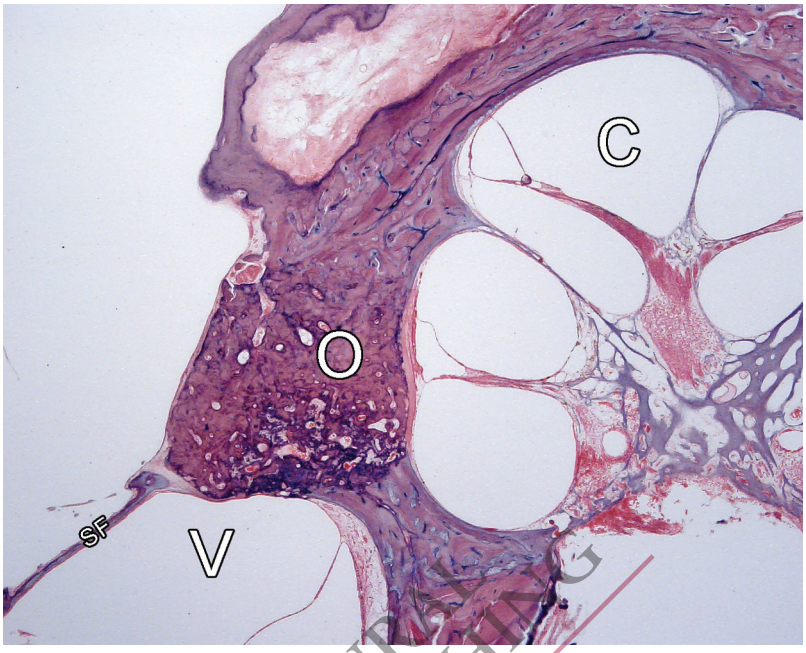

Figure 1–2. Magnified view of histologic otosclerosis. O = otosclerotic focus; V = vestibule; SF = stapedial footplate. Courtesy of University of Minnesota temporal bone collection. Kindly contributed by Dr. Cureoglu and Dr. Paparella.

the most common site of involvement representing 81 to 95% of cases.[10]

Cochlear Otosclerosis (Figure 1–3)

Cochlear otosclerosis is a term used in cases in which the otosclerotic lesion invades the cochlear endosteum and is usually reserved for the occurrence of pure sensorineural hearing loss due to otosclerosis without any conductive component.

"Malignant" (Obliterative) Otosclerosis

This is defined as severely active otosclerosis involving both oval and round windows and most of the bony labyrinth and is manifested initially by mixed hearing loss, which then relentlessly progresses to severe profound sensorineural hearing loss. Both windows are obliterated by the otosclerotic focus. Lamellar new bone is seen in the inner ear of such patients.

Far Advanced Otosclerosis (FAO)

Far advanced otosclerosis is defined as no measurable air or bone conduction or air conduction no better than 95 dB and bone conduction at 55 dB to 60 dB at one frequency only.

A negative Rinne's test result with a 256 Hz magnesium tuning fork is the best way to separate a FAO sensorineural hearing loss from sensorineural hearing losses of other causes.

SITES OF INVOLVEMENT OF THE TEMPORAL BONE BY OTOSCLEROSIS IN ORDER OF FREQUENCY

1. Commonly seen anterior to the oval window
2. The round window niche (Figures 1–4 and 1–5)
3. Posterior to the round window
4. Posterior wall of the internal auditory canal

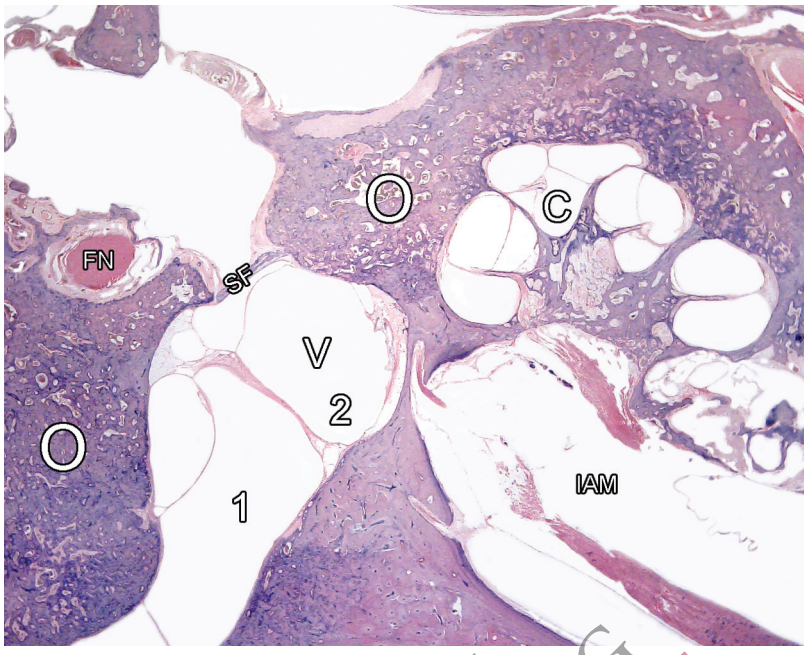

Figure 1–3. Histopathology section of human temporal bone demonstrating cochlear otosclerosis. O = otosclerotic focus; C = cochlea deformed by the otosclerosis; V = vestibule; 2 = saccule; 1 = utricle; SF = stapes footplate; FN = facial nerve. Courtesy of University of Minnesota temporal bone collection. Kindly contributed by Dr. Cureoglu and Dr. Paparella.

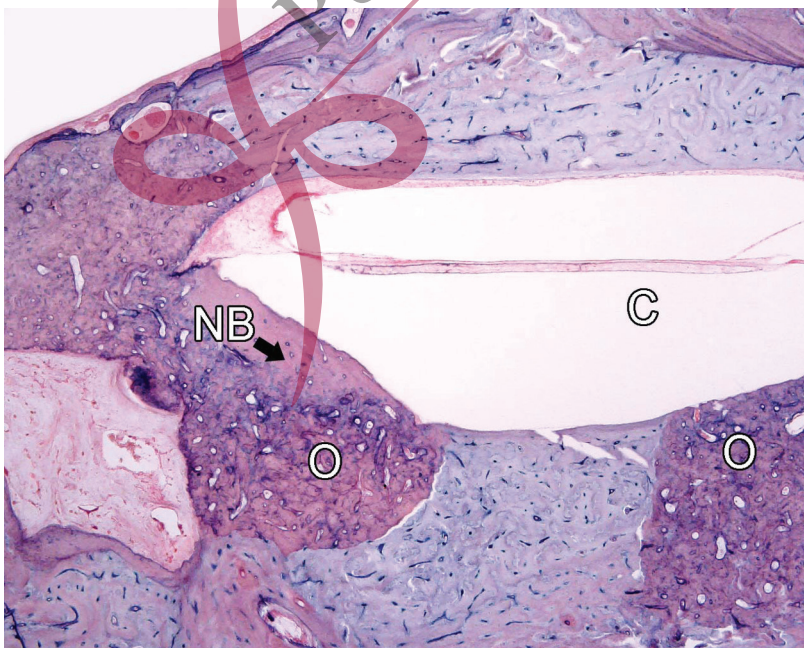

Figure 1–4. HP section demonstrating otosclerotic focus obliterating the round window. O = otosclerotic focus; C = cochlea; NB = new bone formation. Courtesy of University of Minnesota temporal bone collection. Kindly contributed by Dr. Cureoglu and Dr. Paparella.

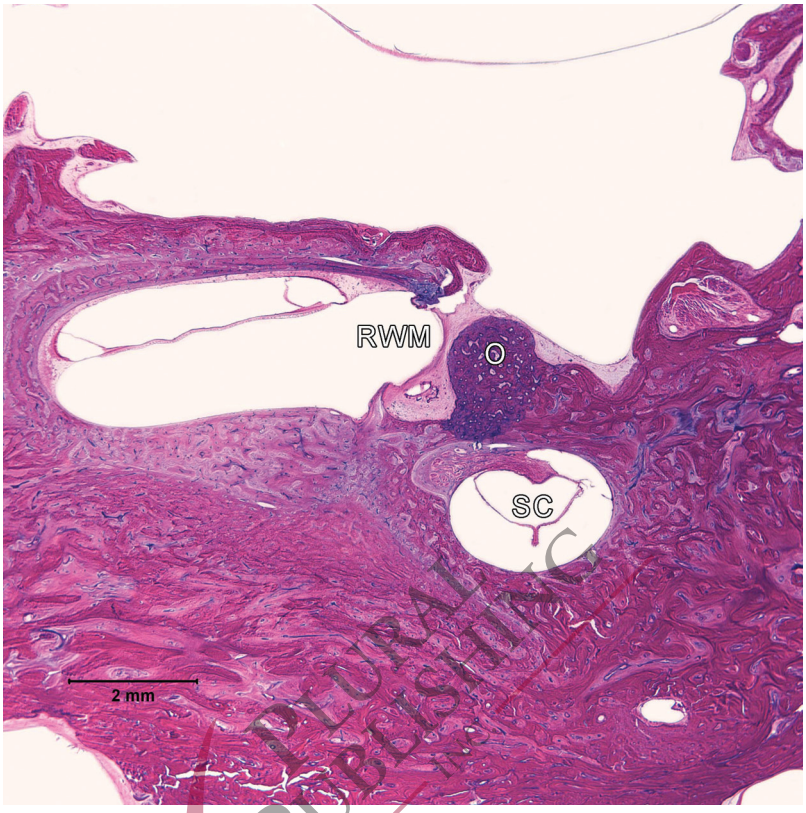

Figure 1–5. HP section demonstrating otosclerotic focus near round window niche. O = otosclerotic focus; RWM = round window membrane; SC = Semicircular canal. Courtesy of University of Minnesota temporal bone collection. Kindly contributed by Dr. Cureoglu and Dr. Paparella.

5. Around the cochlear aqueduct
6. Semicircular canals
7. Totally within the stapedial footplate

HISTOPATHOLOGY OF OTOSCLEROSIS

The otic capsule itself undergoes very little remodeling. The otic capsule contains small regions of immature cartilaginous tissue called the "globuli interossei." This may be the loci of the earliest lesions of otosclerosis.

Otosclerosis is characterized by the following:

1. Bone resorption
2. New bone formation
3. Vascular proliferation
4. Connective tissue stroma.

The very **first** stage of otosclerosis is resorption of enchondral bone around blood vessels.

Second: This results in enlargement of perivascular spaces. Vascular spaces become wider. This is the initial stage characterized by diffuse or patchy demineralization that coincides with preotosclerotic lesions.

Third: This is then followed by deposition of immature (woven) bone.

Fourth: Resorption and deposition of immature bone occurs continuously within an otosclerotic focus with production of more mature (lamellar) bone.