

OTOSCLEROSIS

- ▣ Otosclerosis, more aptly called otospongiosis, is a primary disease of the bony labyrinth
 - ▣ one or more foci of irregularly laid spongy bone replace part of normally dense enchondral layer of bony otic capsule
 - ▣ Most often, otosclerotic focus involves the stapes region leading to stapes fixation and conductive deafness
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- ❑ The exact cause of otosclerosis is not known
- ❑ Anatomical basis.
 - Bony labyrinth is made of enchondral bone which is subject to little change in life
 - There are areas of cartilage rests in this hard bone which due to certain nonspecific factors are activated to form a new spongy bone
 - Fissula ante fenestram-lying in front of the oval window — the site of predilection for stapedial type of otospongiosis.

- ▣ Heredity.
 - About 50% of otosclerotics have positive family history; rest are sporadic
 - Autosomal dominant trait with incomplete penetrance and a variable expressivity.
- ▣ Race
 - White races are affected more than black Americans. It is common in Indians but rare among Chinese and Japanese.

▣ Sex

- Females are affected twice as often as males but in India, otosclerosis seems to predominate in males.

▣ Age of onset

- Hearing loss usually starts between 20 and 30 years of age and is rare before 10 and after 40 years

- ▣ Effect of other factors
 - Hearing loss due to otosclerosis may be initiated or made worse by pregnancy.
 - Deafness may increase during menopause, after an accident or a major operation.
 - The disease may be associated with osteogenesis imperfecta with history of multiple fractures
 - The triad of symptoms of osteogenesis imperfecta, otosclerosis and blue sclera is called van der Hoeve syndrome

- ▣ Viral infection
 - Electron microscopic and immunohistochemical studies have shown RNA related to measles virus

TYPES OF OTOSCLEROSIS

▣ Stapedial otosclerosis

- Stapedial otosclerosis causing stapes fixation and conductive deafness is the most common variety.
- lesion starts just in front of the oval window in an area called “fissula ante fenestram
- This is the site of predilection (anterior focus)
- Lesion may start behind the oval window (posterior focus)
- Around the margin of the stapes footplate (circumferential)
- In the footplate but annular ligament being free (biscuit type)
- it may completely obliterate the oval window niche (obliterative type)

Cochlear otosclerosis

- ▣ Cochlear otosclerosis involves region of round window or other areas in the otic capsule
- ▣ It may cause sensorineural hearing loss probably due to liberation of toxic materials into the inner ear fluid

Histologic otosclerosis

- ▣ This type of otosclerosis remains asymptomatic and causes neither conductive nor sensorineural hearing loss.

PATHOLOGY

- ▣ Grossly, otosclerotic lesion appears chalky white, greyish or yellow.
- ▣ Sometimes, it is red in colour due to increased vascularity, in which case, the otosclerotic focus is active and rapidly progressive.

Microscopically, spongy bone appears in the normally dense enchondral layer of otic capsule

- ▣ In immature active lesions, there are numerous marrow and vascular spaces with plenty of osteoblasts and osteoclasts and a lot of cement substance which stains blue (blue mantles) with haematoxylin-eosin stain
- ▣ Mature foci show less vascularity and laying of more bone and more of fibrillar substance than cementum, and is stained red

THANK YOU