DEFINITION

1. **Otosclerosis** is a disease which is localised to the bone of the middle and inner ear and in which areas of lamellar bone are destroyed, being replaced by bone of greater thickness, cellularity and vascularity. It is not a generalised disease of bone. Three stages in the condition are described -

   1.1. an active stage in which the bone structure is replaced by woven bone.

   1.2. an inactive stage in which the bone becomes highly mineralised and thickened.

   1.3. an intermediate stage where the processes of softening and thickening are both occurring.

   Any one focus may contain areas at different stages of activity.

2. The condition is sometimes referred to as **otospongiosis**.

CLINICAL MANIFESTATIONS

3. The clinical manifestations depend upon the location and extent of the lesions and otosclerotic foci may be present without causing symptoms.

4. Symptomatic otosclerosis typically presents with bilateral, gradually increasing, hearing loss which starts insidiously between the ages of 20 and 50 years. Unilateral otosclerosis occurs in about 15% of cases and the condition may remain unilateral or may eventually affect the other ear.

5. Speech is often heard better in the presence of background noise, this phenomenon being referred to as **paracusis**, and speech discrimination is usually good.

6. Pain is not a feature although some patients complain of discomfort.

7. Tinnitus is common and may be the presenting symptom but may disappear as the disease progresses.

8. Attacks of vertigo are uncommon and, when they occur, are usually transient.

9. In the majority of cases, the hearing loss is mainly conductive - ie produced by damage to the middle ear - although sensorineural hearing loss - produced by damage to the inner ear - may be identifiable to a variable degree. Most patients who present with conductive loss will eventually develop a sensorineural impairment greater than that produced by ageing alone.

10. The bone conduction audiogram may show reductions of sensitivity which are greatest around the sound frequency of 2000 Hz. This configuration is called the **Carhart notch** and is characteristic of fixation of the stapes in the middle ear.

11. The course of otosclerosis is variable but the hearing loss is usually progressive, occasionally rapidly so. Progress may be steady or intermittent and may become arrested naturally.

12. Suitable hearing aids usually give useful results and although operative treatment is well established it may be contra-indicated in particular cases. It is recognised that, in about 2% of cases, some degree of sensorineural hearing loss (which may even be total) may occur after operation (however competently performed), even many years later.

AETIOLOGY

13. The familial pattern of otosclerosis has long been recognised and various studies have shown that the condition is a genetically determined disease, transmission being as a simple autosomal dominant with variable "penetrance" or manifestation. Although the existence of the condition in a particular family may be denied this is usually because of lack of diagnosis or even manifestations in other members since the genetic determiner may be "carried" without leading to manifestation.

14. Otosclerosis is not caused by infection or inflammation, metabolic disorders, vascular disease, other ear disorders such as Ménière's disease and otitis media, trauma or exposure to loud and/or prolonged noise.

15. Once established, the course of otosclerosis may be accelerated by periods of endocrine activity such as puberty, pregnancy and lactation or thyrotoxicosis. It may also be accelerated during the healing process following fracture of major bones. Otherwise, environmental factors do not have any effect on the course of otosclerosis.

CONCLUSION

16. Otosclerosis is a genetically-determined localised disease of the bone of the middle and inner ears. Its onset is unrelated to environmental factors or other diseases although its course may be adversely affected by certain factors which have been listed at paragraph 15 above.

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