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Temporal Bone Histopathology Case of the Month: Otosclerosis without stapes fixation

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Introduction

Otosclerosis is an autosomal-dominant inherited disease with incomplete penetrance. Although it causes hearing loss in only 0.3 to 0.4% of the population, it has been reported to be present in post mortem temporal bones of 2.5 to 10% of the white population.¹ Furthermore, 10% of the population with both otosclerosis and hearing loss can develop a sensorineural hearing loss when the cochlea becomes involved.²

The pattern of otosclerosis is variable. Otosclerosis most commonly originates in the bone anterior to the oval window involving the fissula ante fenestrum and causes hearing loss as it begins to invade the annular ligament of the stapes footplate. On histopathology it can also be found originating from both anterior and posterior to the annular ligament causing a bipolar stapes fixation. It can also originate from the stapes footplate itself creating a thickened footplate with well delineated margins. When the lesions flows across the ligament and onto the footplate, it can cause both fixation and complete obliteration of the footplate. Finally, if the disease grows towards the cochlea and invades the endosteum, it can cause an isolated sensorineural hearing loss or mixed hearing loss.² In this paper, we will describe a case of otosclerosis incidentally noted without any involvement of the stapes, illustrating its prevalence in the asymptomatic population.

Case Report

44-year-old-man presented with right-sided tinnitus and progressive hearing loss over eight years and eventual facial numbness and palsy over his ipsilateral face. Preoperative audiogram revealed no hearing on the right side and a pure tone average of 13 dB on the left side without an air-bone gap and speech discrimination scores of 92% at 40 dB. Acoustic reflexes were not conducted. A pantopague myelogram revealed 3.5 to 4 cm acoustic neuroma on the right. He subsequently underwent retrosigmoid resection of this tumor but did not survive the recovery for unknown reasons. On postmortem histologic slides he was

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incidentally noted to have an otosclerotic focus around his anterior oval window without stapes fixation in both ears.

Histopathology, left ear

A small (1.5 mm) otosclerotic lesion occupies the anterior oval window niche without fixing the stapedial footplate. (Figs 1&2) The cochlear contents are normal except for some loss of stria vascularis volume not sufficient to affect thresholds. The spiral ganglion count is 30,850.

Discussion

Histologically, otosclerosis can be broken into two different stages of disease, an otospongiotic and an otosclerotic stage. The otospongiotic stage is characterized by replacement of normal bone architecture by “woven bone” that appears to be disorganized small lamellae in which there is an equal amount of connective tissue containing fibrocytes, small blood vessels, and osteoclasts which rarely are found at the edge of the lesion. The otosclerotic stage is composed of more dense disorganized bone with much less connective tissue than the spongiotic phase. These two stages often coexist. There may be multi centric foci of both types of lesion.³ This patient is an example of a typical otosclerotic focus adjacent to the fissula ante fenestrum without stapes or cochlear involvement.

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Figure 1. Otosclerotic (vs. spongiotic) focus (black arrow) in the anterior oval window niche without involvement of the stapes footplate (black arrowheads) (Hematoxylin and Eosin [H&E] $\times 20$)

Figure 2.

Transition (black arrowheads) between normal bone and the otosclerotic focus. Note the small irregular bone lamellae and small connective tissue spaces and absence of osteoclasts. The lacunae in the otosclerotic lesion are filled with viable osteocytes whereas the lacunae in the adjacent normal bone are empty. (H&E $\times 200$)