

Otosclerosis as a Neoplasm of the Outer Layer of the Otic Capsule

Leslie Michaels M.D. and Sava Soucek M.D.

The Royal National Throat, Nose and Ear Hospital, Gray's Inn Road, London, England

We studied stained step sections embedded in celloidin of 54 temporal bones from patients with otosclerosis at the Royal National Throat, Nose and Ear Hospital, London and the House Ear Institute, Los Angeles, California, in order to investigate the histopathologic changes in that disease of the otic capsule (OC).

We will consider here firstly the normal structure of the otic capsule and then the changes of the pathological condition of otosclerosis. Finally we will depict the appearances to be found in otosclerosis which indicate to us that this disease is a neoplasm. For convenience we will illustrate the features of the cochlear portion of OC otosclerosis only. The vestibular arrangements are similar, but structured in a more complex fashion and so more difficult to display clearly.

Fig. 1. Normal Otic Capsule

The outer and inner layers are delineated by a thin naturally occurring pale-staining line. The outer layer is composed of normal osteons, (each a central Haversian canal surrounded by osteocytes). Note the regular shape and size of the osteons. The inner layer is packed with globuli ossei (not discernible here)



Fig. 2. Otosclerotic plaques

Two plaques are visible in this photo (A and B), but there are probably more in the whole otic capsule. Each plaque is well-defined and shows a similar structure, with a basophilic zone on its cochlear side, which we have identified as an "invasive front" because it is composed of osteoblasts, as compared with the osteocytes that make up the rest..

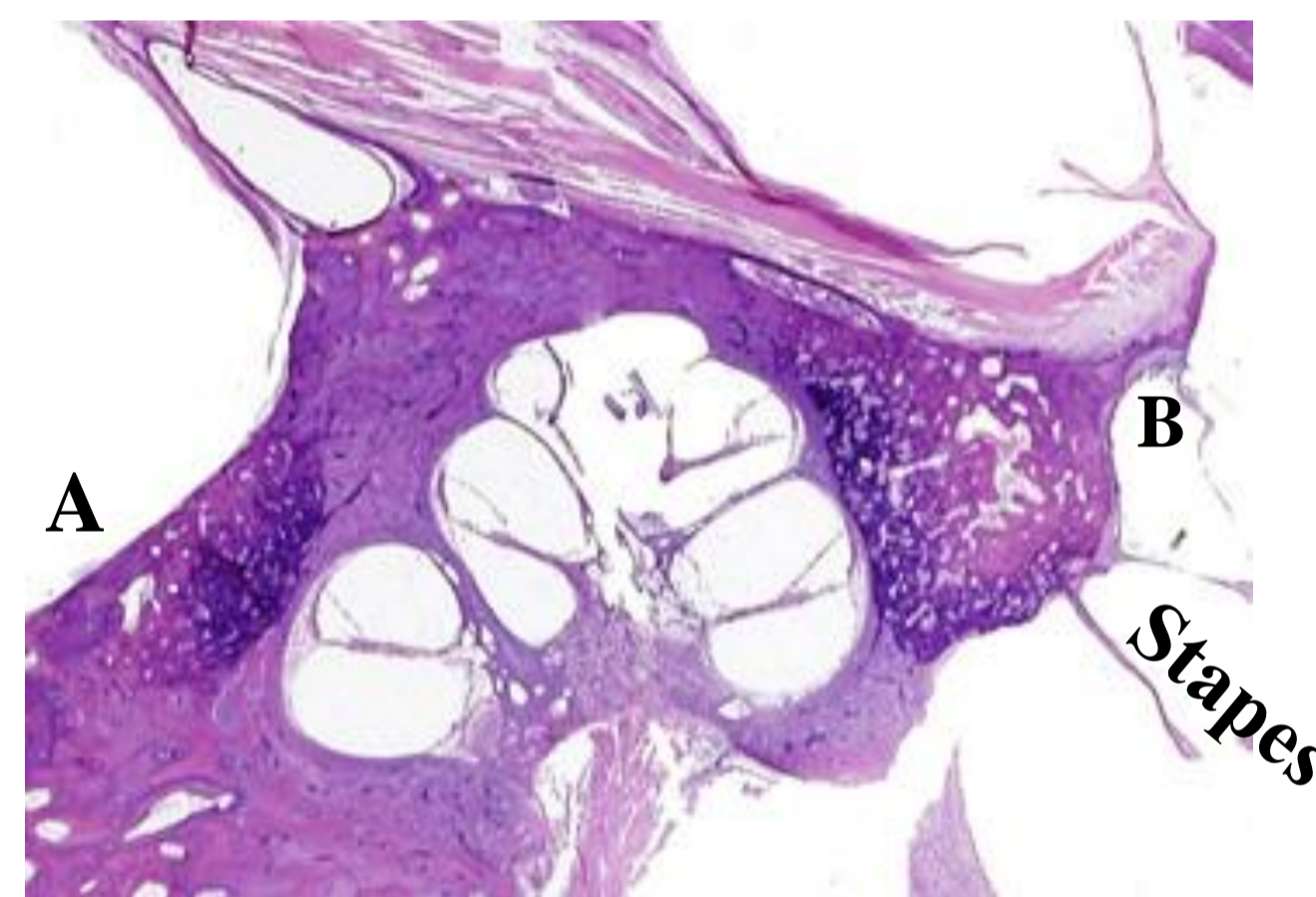


Fig. 3. Right-hand plaque (B) from Fig. 2, enlarged

We now see that the plaque is packed with Haversian canals and that these are particularly numerous in the basophilic invasive front. Otosclerosis is smoothly lined up against the the posterior cochlear wall in the vicinity of the tensor tympani muscle. In contrast the cochleo-vestibular surface of the invasive front is highly irregular with invasion of the stapediovestibular joint, uneven bulging at the vestibular surface and even invasive outgrowths into cochlear wall. Note increasing differentiation away from invasive front, the Haversian canals showing a large area of marked dilatation (otospongiosis often seen radiologically).

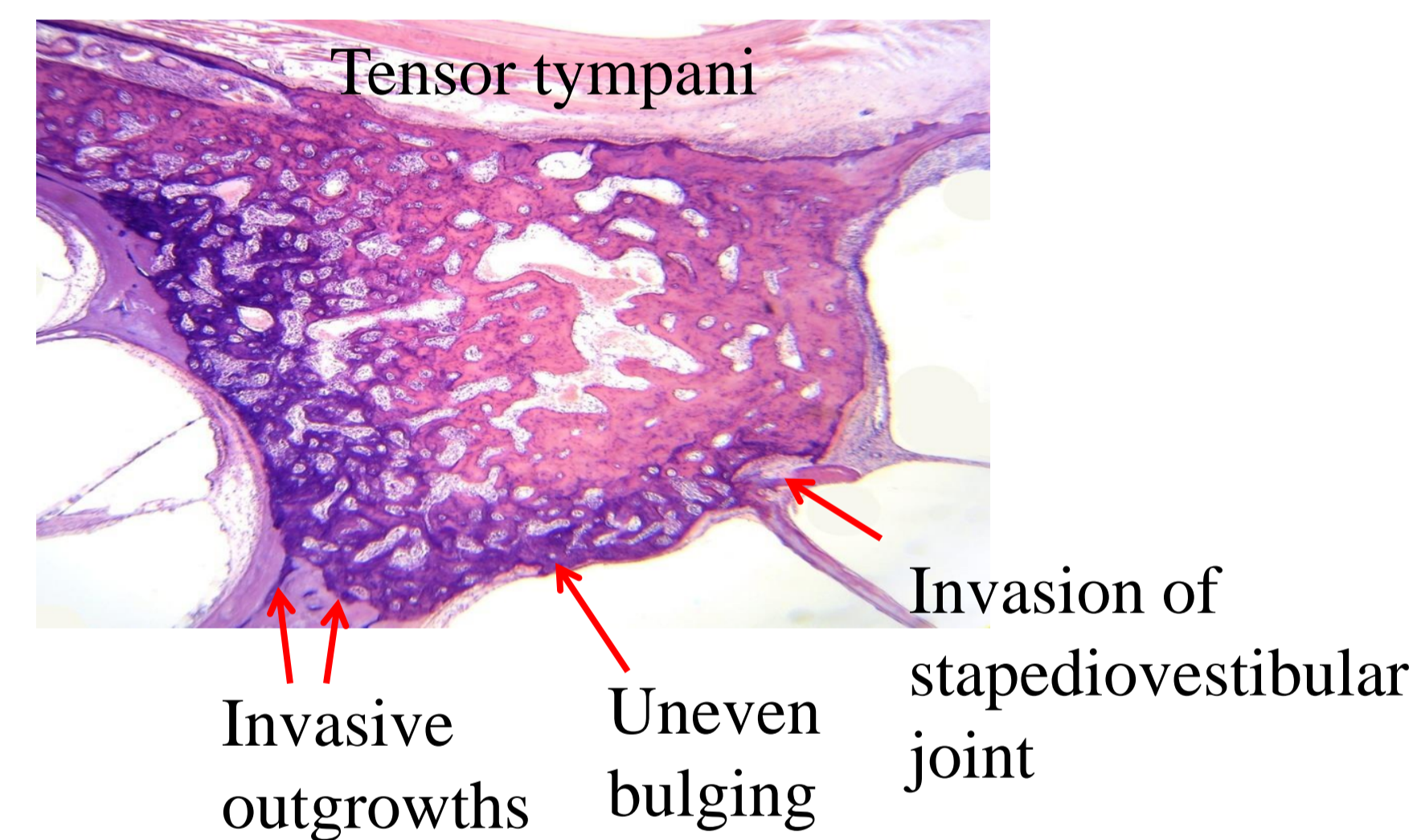


Fig. 4. Invasive front region of otosclerotic plaque, further enlarged

The outlines of the osteons, comprising Haversian canals and surrounding bony tissue, are identified. Both elements are highly distorted

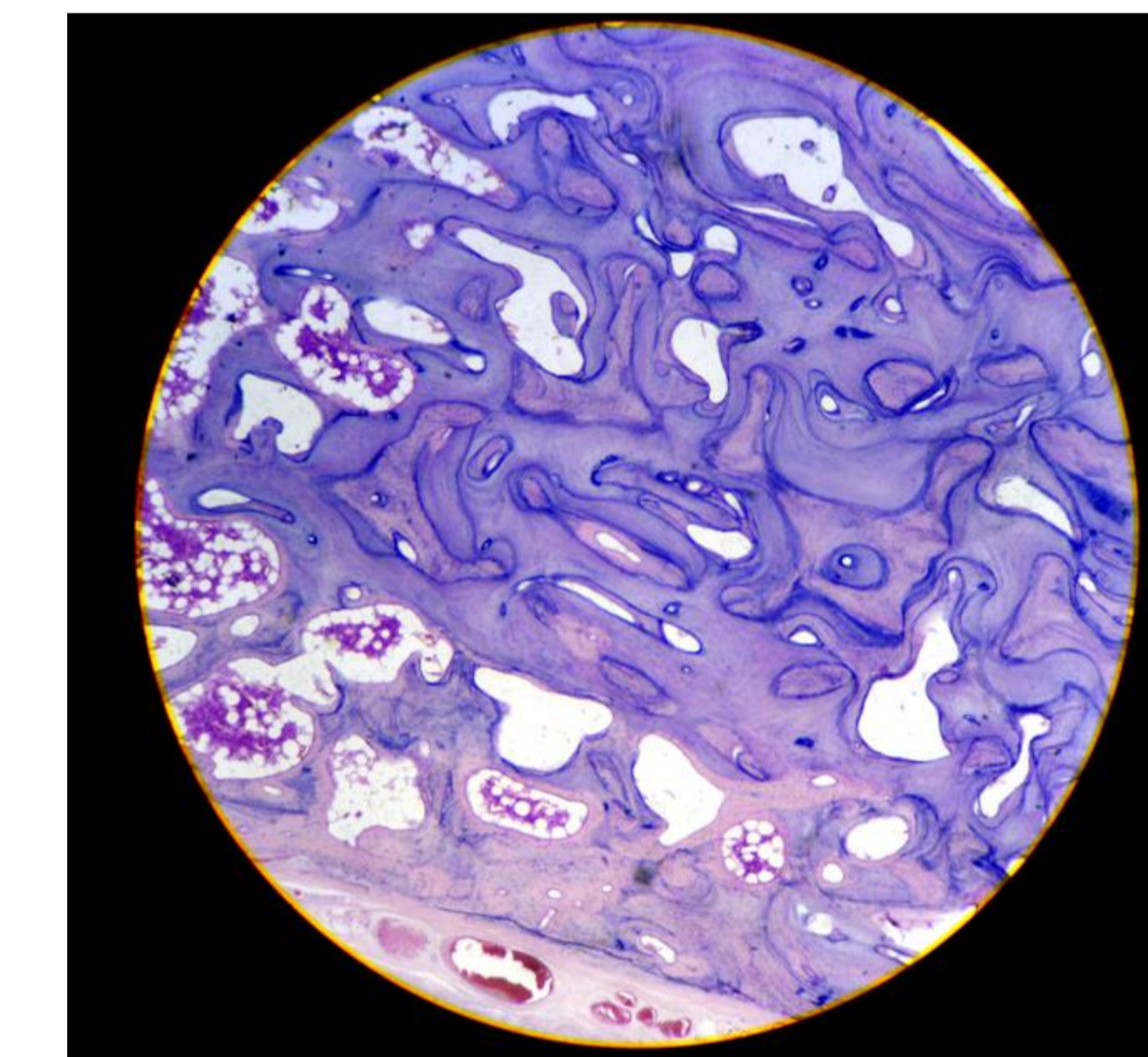
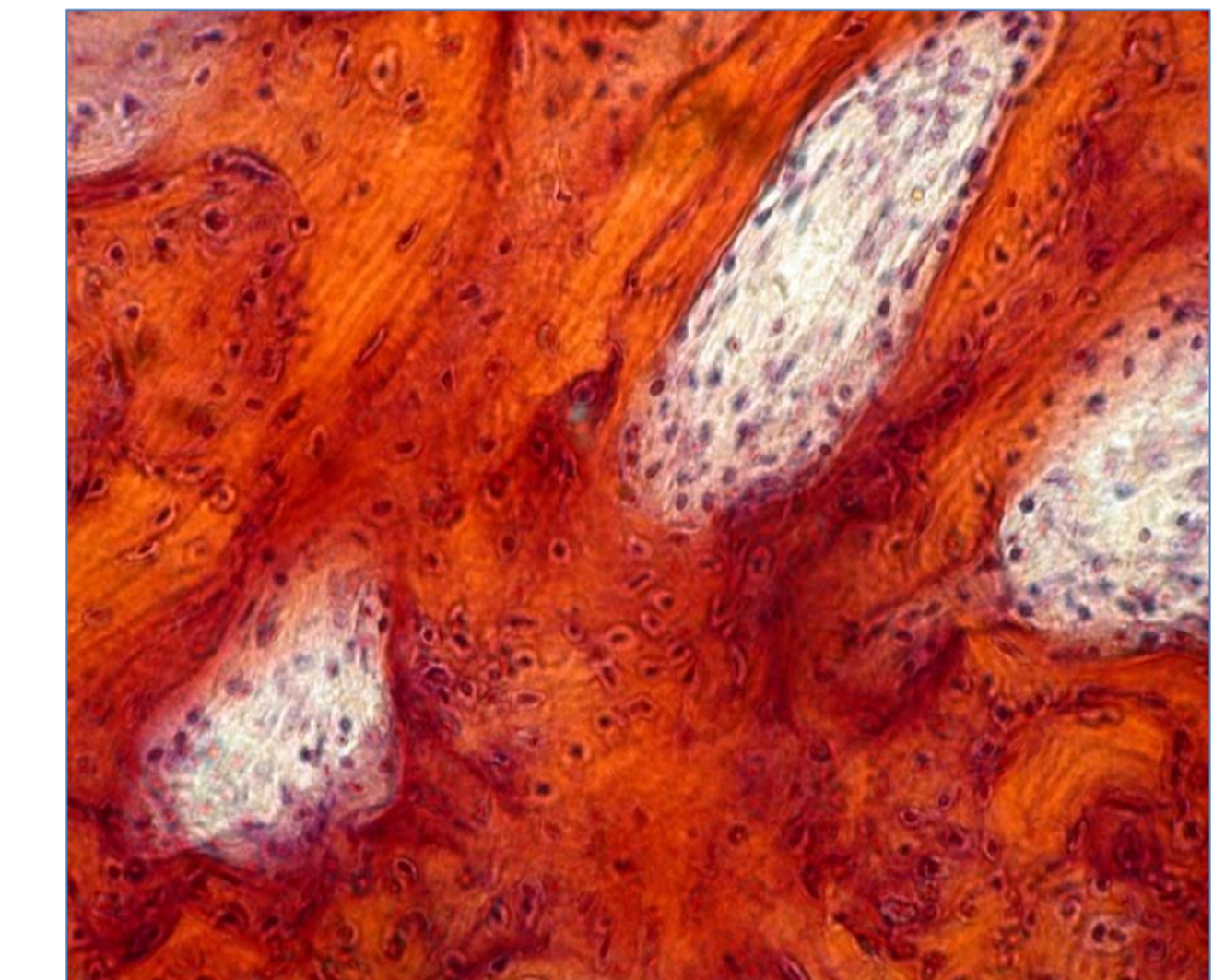


Fig. 5. High-powered view of intermediate region of otosclerotic plaque, photographed in polarised light (crossed polaroid)

Part of an abnormal osteon, showing abnormal Haversian canals surrounded by osteocytes and birefringent bands of collagen



Further Reading

Michaels L, Soucek S. Origin and growth of otosclerosis Acta Oto-Laryngologica, 2011; 131: 460-468

Michaels L, Soucek S. Atypical mature bone in the otosclerotic otic capsule as the differentiated bone of an invasive osseous neoplasm. Acta Oto-Laryngologica. 2014;134;2;118-123

Acknowledment

We would like to thank Dr. Fred Linthicum, Director of the Temporal Bone Pathology Department, House Ear Institute, Los Angeles, California, for many years of hospitality in his department and the invaluable tuition in ear pathology that accompanied it.

Features of Otosclerosis Suggesting that it is an Invading Neoplasm

Multiple otosclerotic plaques in the same OC (Fig. 2)

Each plaque has a similar morphology: smooth line-up against periosteum of outer layer of OC

indicating possible origin there (Figs. 2 & 3); basophilic zone in each plaque, in cochlear OC

always on the cochlear side of plaque, with histology of osteoblasts and multiple Haversian canals

suggesting invasive front (Figs. 2 & 3); invasive fronts with fine and blunt invasive outgrowths

(Figs. 3 & 6); clear evidence of invasion of cochlea, vestibule, stapediovestibular joint and

footplate (Figs. 3 & 6).

Fig. 6. Otosclerotic invasive front has destroyed footplate of stapes

Note irregular outgrowths

