The Journal of Laryngology and Otology
April 1991, Vol. **105**, pp. 261–267

Mode of growth of acquired cholesteatoma

MEHER D. WELLS, M.D., F.R.C.S., L. MICHAELS, M.D., F.R.C.Path. (London)

Abstract
A histopathological study of acquired cholesteatoma in four temporal bones from two adults and one child is presented. The findings suggest that the cholesteatoma originated from the retraction pockets of the tympanic membrane and there was active growth of the squamous epithelium of the retraction pockets, which may be enhanced in the presence of otitis media.

Introduction

It is well recognized that some acquired cholesteatomas arise from retraction pockets of the tympanic membrane. It is not clear as to how the cholesteatoma is derived from them, how it grows subsequently or what is the relationship to otitis media. The purpose of this report was to make a histopathological study of acquired cholesteatoma in relation to its origin and mode of growth.

Method

Temporal bones were removed at post mortem. The microslice technique (Michaels *et al.*, 1983), was used in each case. The microslices were embedded in paraffin, sectioned in the vertical plane (6 μm) in Case 1 and horizontal planes in Cases 2 and 3. All the sections were stained with haemotoxylin and eosin.

Case 1

A 60-year-old male who had bilateral chronic otitis media with cholesteatoma had had a left radical mastoidectomy performed many years previously. There was persistent otorrhoea from the right side at the time of death. An audiogram showed a 25 dB conductive hearing loss on the right and a 75 dB conductive hearing loss on the left. The cause of death was unrelated to the otitis media. Both temporal bones were removed at post mortem.

Histological appearance (right temporal bone)

There was a postero-superior retraction pocket of the pars flaccida which formed a sac extending within the middle ear to the facial nerve. Tongues of squamous epithelium extended from the deeper part of the sac into the middle ear. There were keratin pearls in the wall of the sac. There was also chronic inflammation and cholesterol clefts in the middle ear and mastoid cells. In addition to the cholesteatoma sac there was another sac of the retraction pocket in the attic. The malleus, incus and stapes were present. (Figs.1–3).

Left temporal bone

A radical mastoidectomy had been performed. The tympanic membrane, malleus and incus had been destroyed. The superstructure of the stapes had been destroyed by disease. The footplate of the stapes was intact. Postero-superiorly there was thickening of squamous epithelial growth and keratin pearl formation. There was sub-epithelial fibrosis of the remaining lining. The bony facial ridge was deficient. There was no active inflammation (Figs. 4–6).

Case 2

An 81-year-old female died suddenly on admission to hospital. The right temporal bone was removed at post mortem.

Histological appearance

There was a perforation of the tympanic membrane and a large cholesteatoma sac occupied the whole of the middle ear. The mucosal layer of the tympanic membrane was replaced by squamous epithelium which formed the anterior wall of the cholesteatoma sac. There was no transition from normal cuboidal middle ear mucosal epithelium to squamous epithelium of the cholesteatoma sac. The squamous epithelium near the anterior crus of the stapes which was destroyed at the lower third, was more cellular than other areas of the sac which may denote increased activity and possible growth at this site. There was also a further cholesteatoma sac in the attic which in serial sections was not connected with middle ear cholesteatoma sac (Figs. 7–9).

Case 3

An 11-year-old child had bilateral grommets inserted

From: Department of Histopathology, University College and Middlesex School of Medicine, Institute of Laryngology and Otology, London.
Accepted for publication: 31 December 1990.

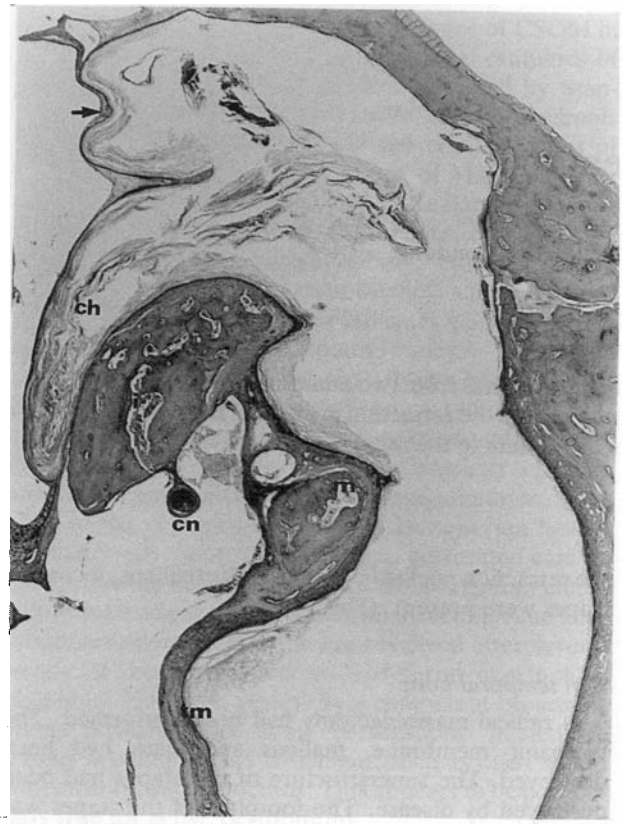


FIG. 1

Case 1. Vertical section of right temporal bone showing retraction pocket formation cholesteatoma sac (ch), incus (i), malleus (m), tympanic membrane (tm), chorda tympani nerve (cn), and keratin pearl (arrow).

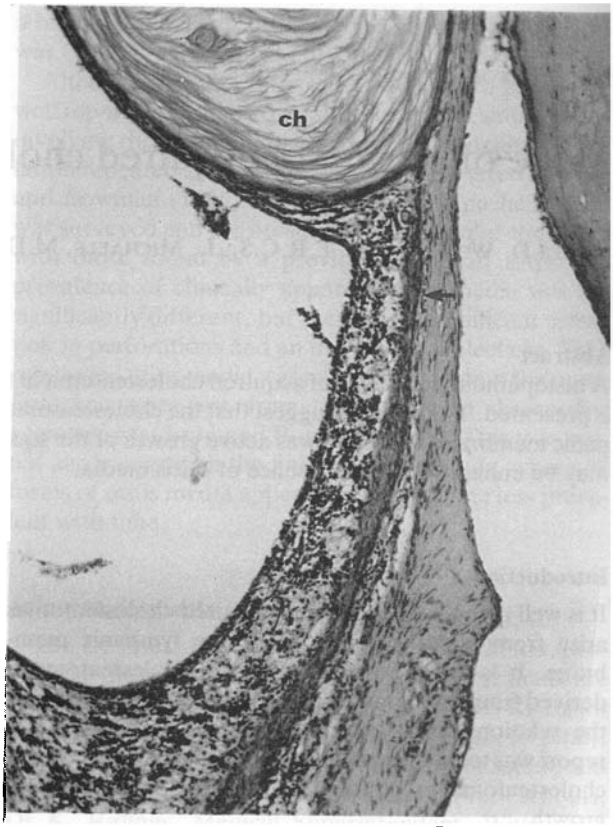


FIG. 2

Case 1. Down growth of squamous epithelium (arrow) from cholesteatoma sac (ch).

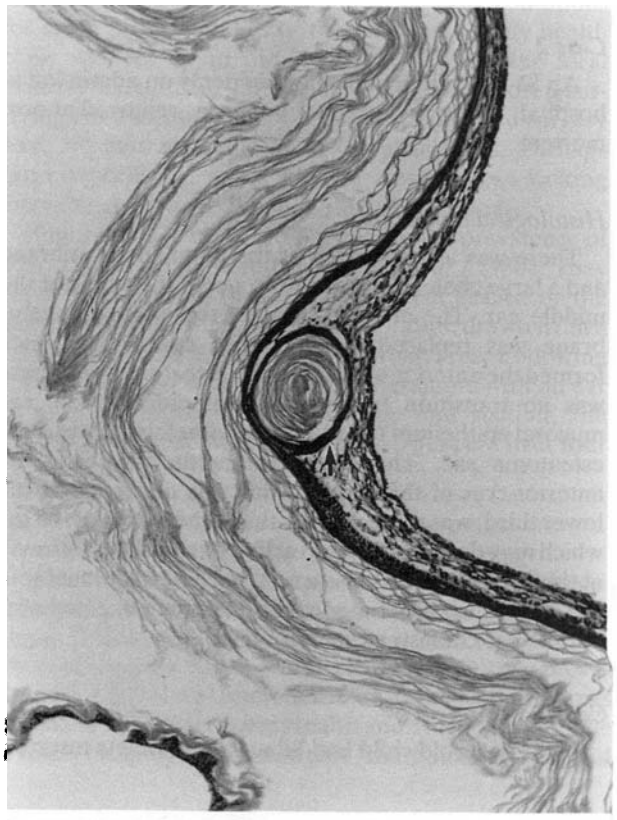


FIG.3

Case 1. Keratin pearl in the tympanic membrane (arrow).

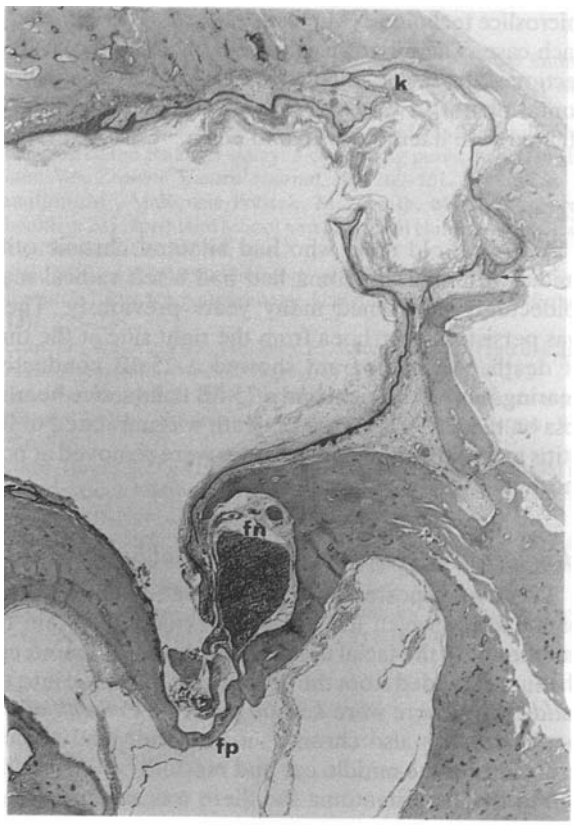


FIG. 4

Case 1. Vertical section of left temporal bone showing thickened squamous epithelium in the covering mastoidectomy cavity (arrow), keratin pearl (k), facial nerve (fn) and footplate of the stapes (fp).

https://doi.org/10.1017/S0022215100115567 Published online by Cambridge University Press

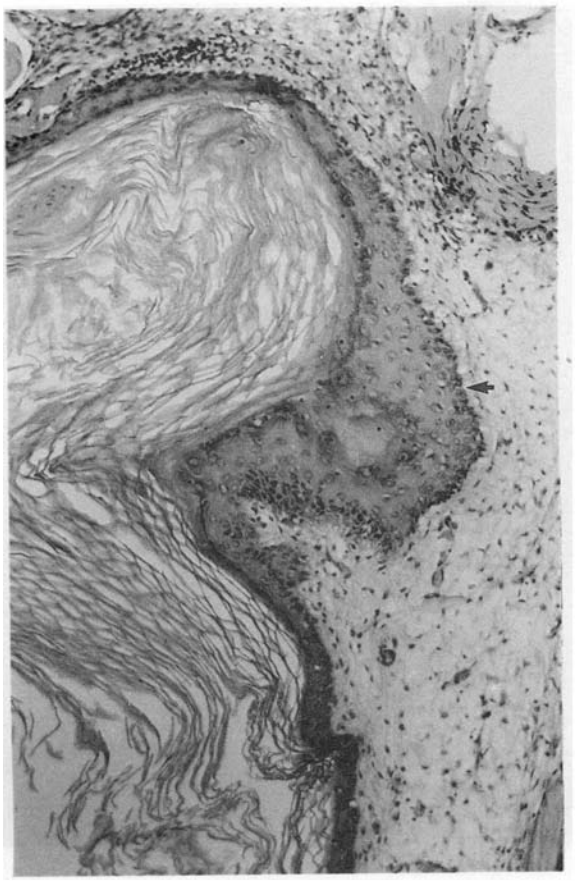


FIG. 5
Case 1. Showing thickened epithelium (arrow seen in Fig. 4).

for otitis media with effusion. After extrusion of the grommets there was a persistent conductive hearing loss on the right side. A right exploratory tympanotomy was planned. Soon after the incision was made the patient had a cardiac arrest and died. Post mortem examination showed status asthmaticus. The right temporal bone was removed at post mortem.

Histological appearance

The surgical defect of the tympanotomy in the tympanic membrane was seen. There was haemorrhage in the connective tissue layer of the pars tensa. A cholesteatoma sac posterior to the malleus was present. It was open towards the tympanic membrane. There were extensions of the squamous epithelium of the sac in the mastoid. There were intra-epithelial pearls in the sac adjacent to the incus. The malleus and incus showed a moth-eaten surface erosion. The superstructure of the stapes was destroyed except for part of the post crus. There was dehiscence of the bony facial canal. The chorda tympani and facial nerve were normal. There was also cholesterol granuloma with cholesterol clefts in the mastoid cells. Cholesteatoma involved the mastoid air cells. The anterior part of the middle ear was free of disease. There was respiratory ciliated epithelial lining in the anterior part of the mesotympanum (Figs. 10 & 11).

Discussion

Cholesteatoma is defined as the presence of squamous epithelium in the middle ear cleft. Its growth associated with infection can cause destruction of surrounding structures and if untreated may also cause death.

For almost a century the origin of acquired cholesteatoma has been debated and its mode of growth has remained unclear. The two most popular concepts are that it arises from metaplasia or from migration of squamous epithelium from the tympanic membrane into the middle ear; a congenital basis for acquired cholesteatoma has also been suggested.

Wendt (1873) proposed that squamous metaplasia of the epithelium of the middle ear cleft can lead to cholesteatoma. Squamous epithelium in the middle ear mucosa has been demonstrated by Palva *et al.*, (1968) and Sadé *et al.* (1982) who suggested that it may be the origin of acquired cholesteatoma. We were unable to demonstrate areas of transition of the normal middle ear epithelium into squamous epithelium or cholesteatoma in the four temporal bones presented in this study.

The concept of migration of squamous epithelium into the middle ear firstly as retraction pockets of the tympanic membrane and then forming cholesteatoma is widely accepted and has clinical and histological support. Friedmann (1955) in his elegant study of experimentally induced otitis media in guinea pigs, frequently found migration or extension of stratified squamous epithelium from the external auditory meatus or the tympanic membrane into the bullae of the guinea pig. The squamous epithelium formed small implantation cysts and thick lamella of keratinized epithelial cells which when detached and mixed with pus produced the characteristic picture of aural cholesteatoma. In a histopathological study of retraction pockets of human temporal bones, Wells and Michaels (1983) have suggested that persistent retraction pockets are formed secondary to chronic inflammation and fibrosis in the middle ear with loss of the connective tissue layer of the tympanic membrane. Poor aeration of the middle ear may be an important factor in retraction pocket formation. In many pockets also there are stratified squamous lined sacs and squamous epithelial 'pearls' originating from the retraction pockets and these structures may penetrate into the middle ear. It seems possible that it is these epithelial ingrowths rather than the main retraction pocket itself which goes on to cholesteatoma formation. Our study of 1983 has in fact amplified the migration theory of early workers, Habermann (1888), Bezold and Siebenmann (1908), Wittmaack (1933) and Tumarkin (1961). Histopathology of the four temporal bones with cholesteatoma in our three cases indicates that the origin of acquired cholesteatoma is from the squamous epithelium of the tympanic membrane. The isolated squamous epithelial cyst in the attic in Case 1 could have been derived from the retraction pocket in a similar way to that described in our 1983 study.

It is known that some retraction pockets remain inactive or self cleansing indefinitely while some may grow to produce active disease in the middle ear with destruction of surrounding structures and other complications. In our four temporal bones the growth of the cholesteatoma sac seems to take place as tongues of squamous epithelium from retraction pockets in a fashion which is almost like a neoplasm. The squamous epithelium is thick at the site of the growth. Ruedi (1963) has shown in

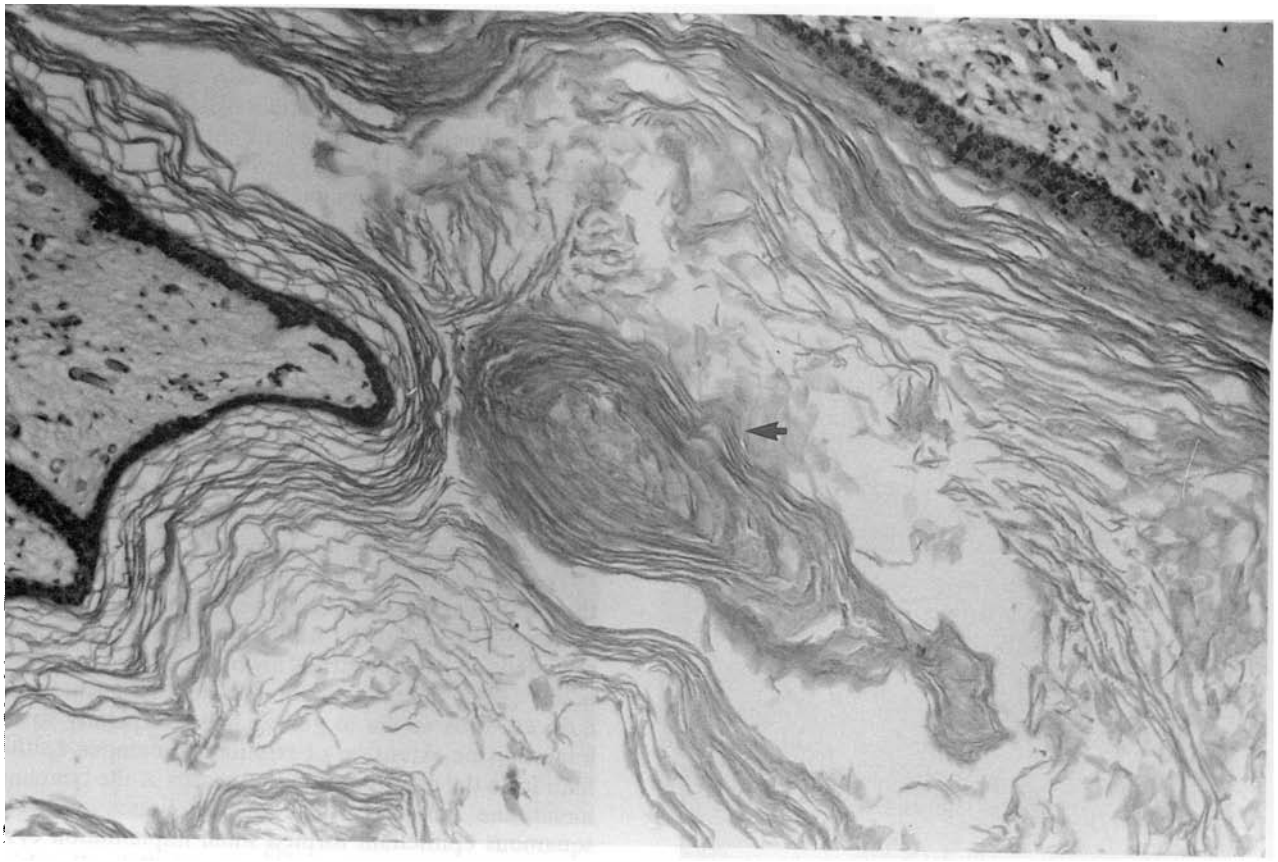


FIG. 6
Case 1. Showing keratin pearl (arrow) seen in Fig. 4.

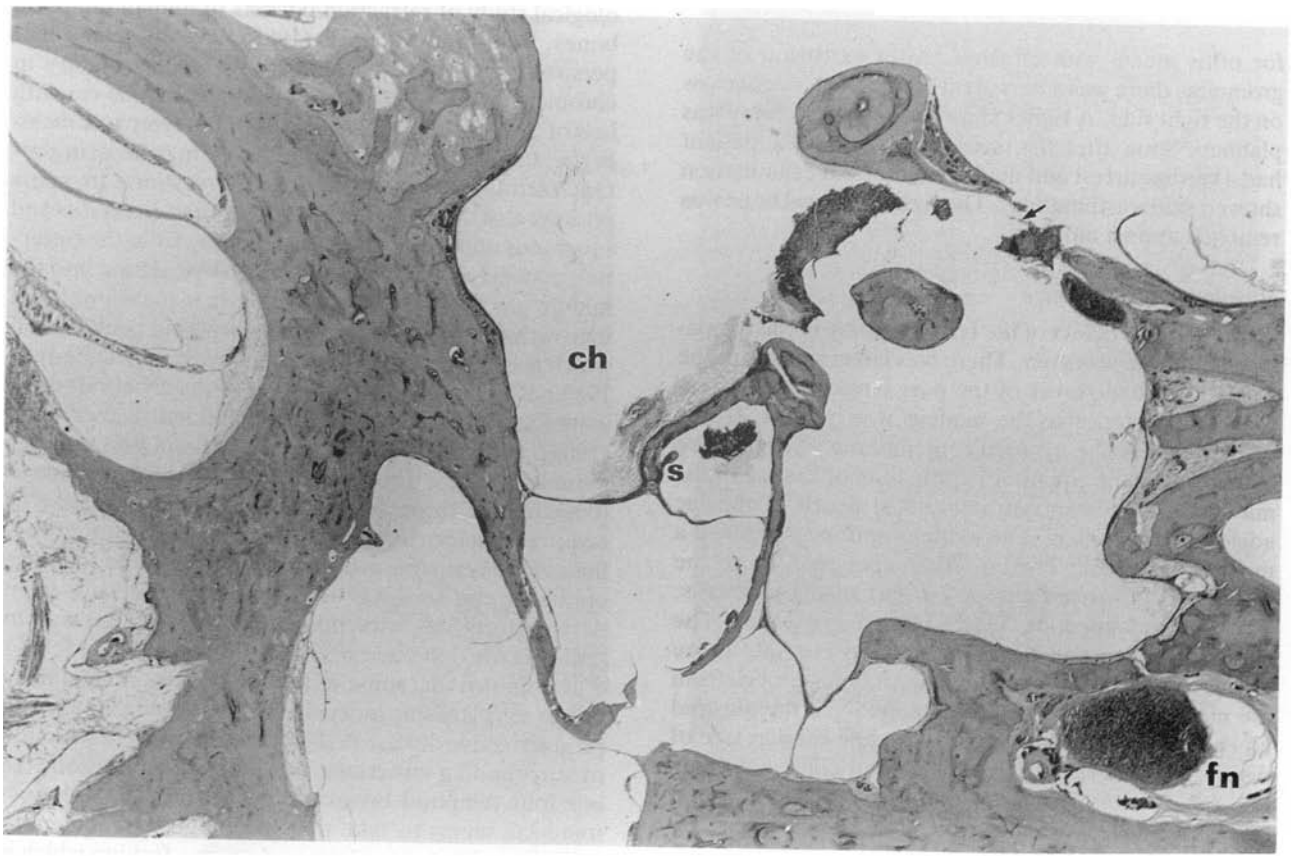
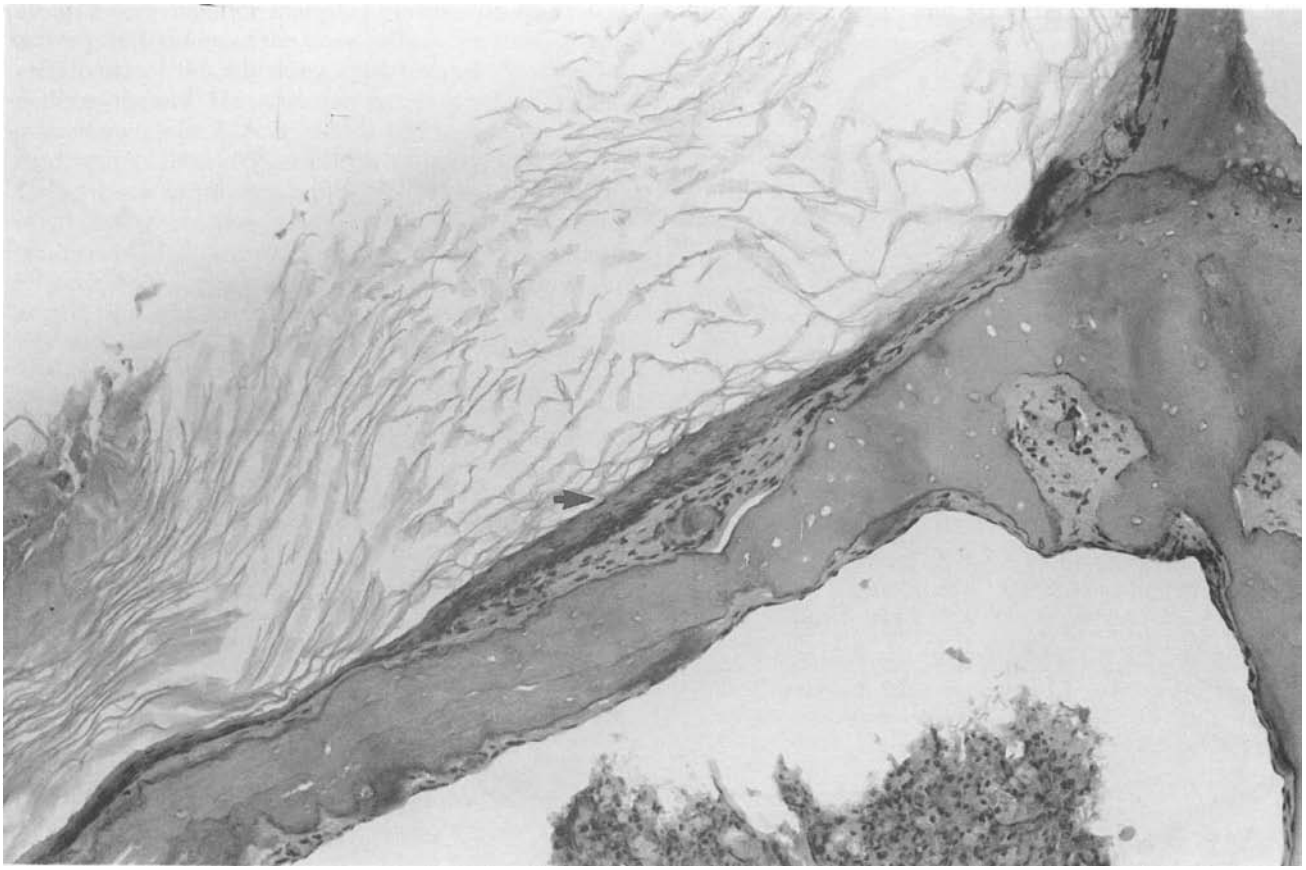


FIG. 7
Case 1. Horizontal mid modular section of right temporal bone showing perforation of the tympanic membrane (arrow), cholesteatoma sac (ch), stapes (s), partly destroyed and lined by cholesteatoma sac and facial nerve (fn).

https://doi.org/10.1017/S0022215100115567 Published online by Cambridge University Press



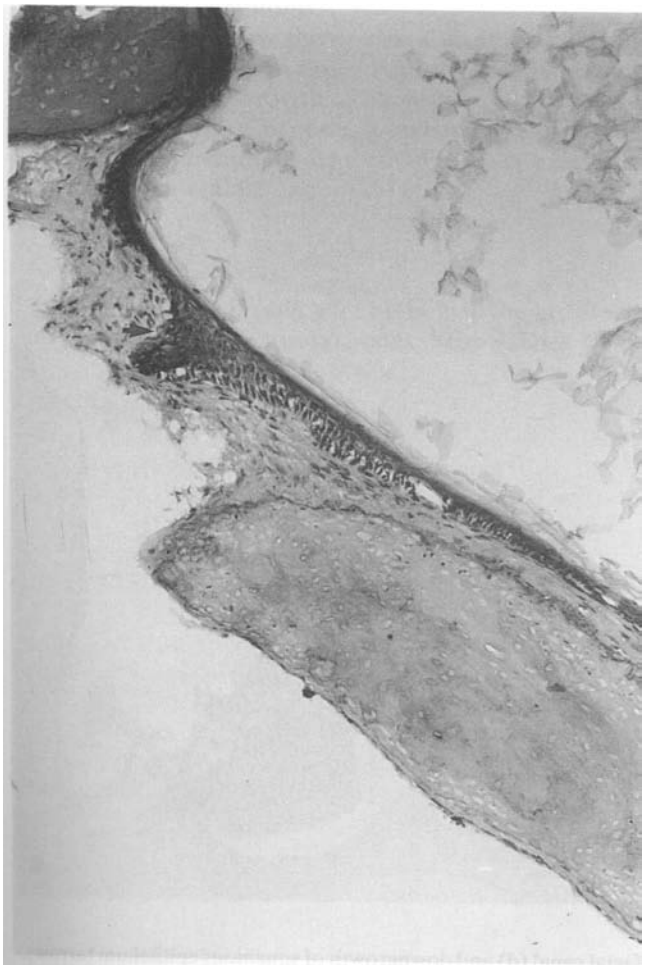


FIG. 8 & 9

Case 2. Showing thickened squamous epithelium on the anterior crus of the stapes (arrows). Higher magnification of part of Fig. 7.

https://doi.org/10.1017/S0022215100115567 Published online by Cambridge University Press

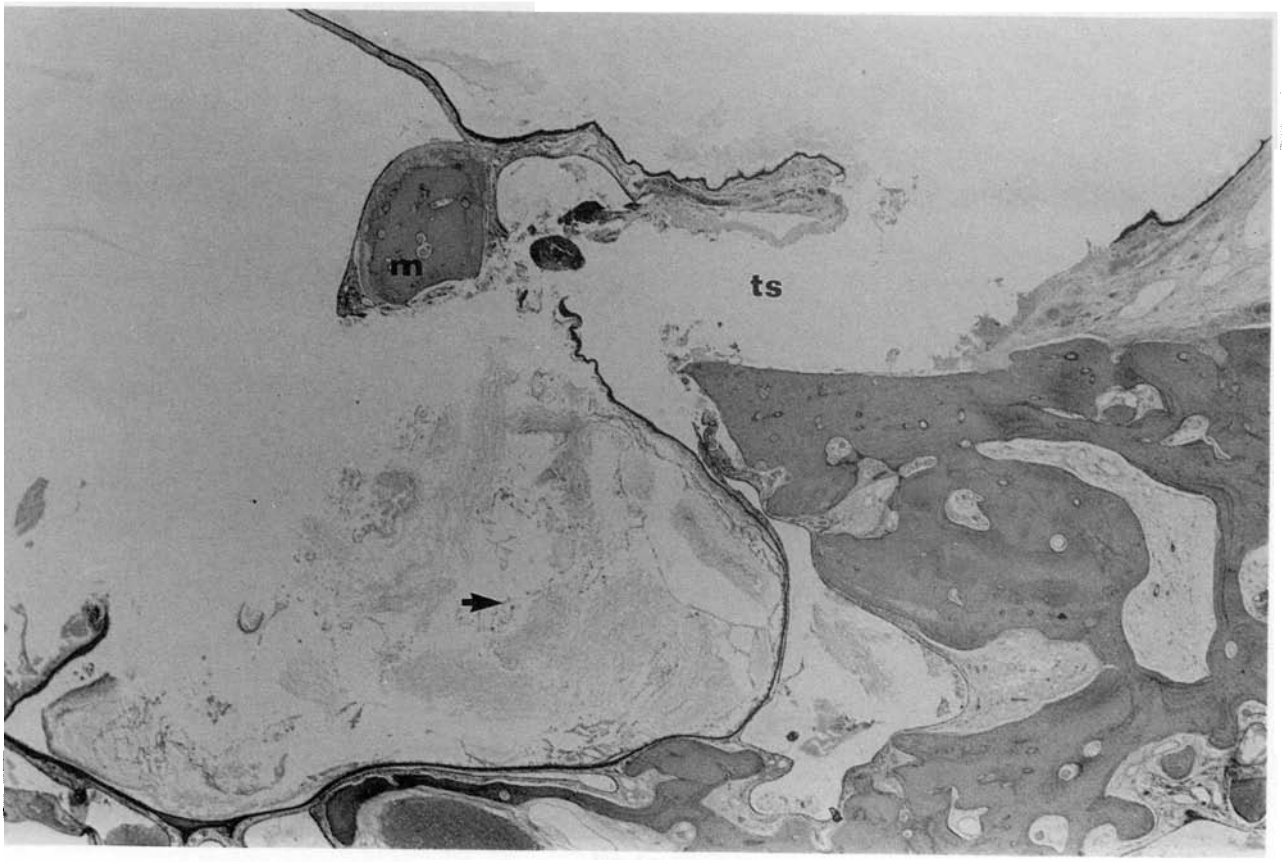


FIG. 10

Case 3. Horizontal section of right temporal bone showing cholesteatoma sac (arrow) behind an intact tympanic membrane, tympanotomy site (ts) and malleus (m).

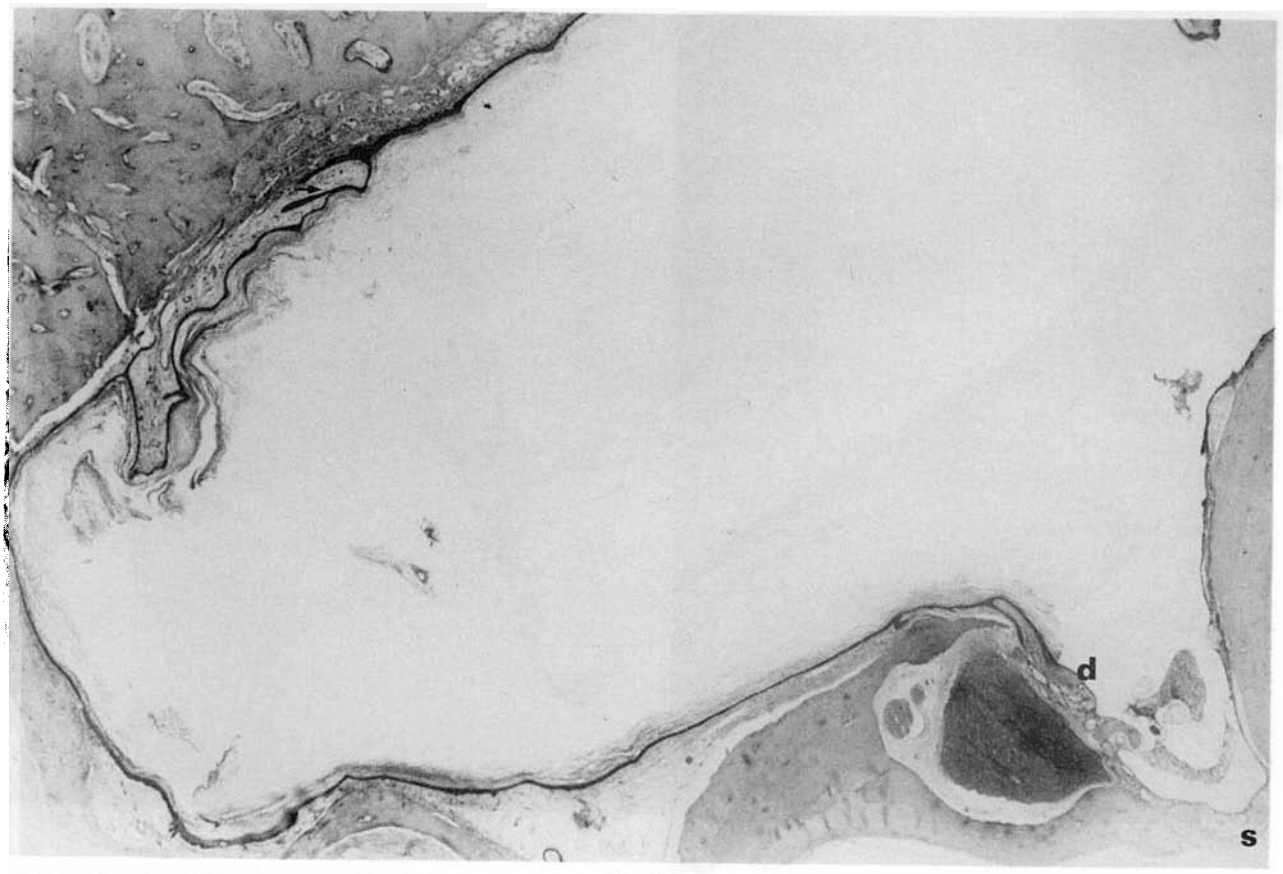


Fig. 11

Case 3. Showing the footplate of the stapes (s) and dehiscence of the bony facial canal (d) and downgrowth of squamous epithelium (arrow).

https://doi.org/10.1017/S0022215100115567 Published online by Cambridge University Press

an area of a superior marginal perforation there was active proliferation of the basal cells in the stratum germinativum of the adjoining epidermis of the external auditory meatus. He suggested that presence of inflammation increased hyperkeratosis and acanthosis of the epidermis of the external auditory meatus that adjoins the tympanic membrane superiorly. In all his cholesteatoma cases there was associated chronic otitis media, which is also confirmed in our study. The presence of inflammation appears to be an important factor in the growth of cholesteatoma.

There is evidence to suggest that residual cholesteatoma after mastoidectomy is less aggressive in its growth and activity. Sheehy *et al.*, in 1977, in an analysis of 1,024 primary cases of mastoid surgery for cholesteatoma found residual cholesteatoma in 380 cases. The incidence of residual disease was higher than in adults. It was most frequently seen in the middle ear than in the epitympanum and least frequently in the mastoid. Sheehy *et al.* also state that they had not seen a report of a life-threatening complication from residual cholesteatoma (residual or primary) as it ceases to grow when it comes into contact with soft tissue such as the dura or facial nerve. It is likely that inflammation enhances the growth of cholesteatoma. In residual disease fewer complications may be seen because the infection and inflammation is largely eliminated which may reduce the activity of the squamous epithelium. In Case 1, a left radical mastoidectomy was performed many years previously, the bony facial bridge was deficient and there was residual cholesteatoma present in the mastoid cavity. Except for radical mastoid surgery, this patient had no other aural symptoms.

This study supports the origin of acquired cholesteatoma from retraction pockets; inflammation is an important factor for the growth of cholesteatoma. There are specific sites in the wall of the sac where active growth of squamous epithelium in the form of tongues extends the disease in the middle ear.

Acknowledgements

We thank Tony Frohlich for his help in the histological preparation of the temporal bones, Miss S. Dick for typing the manuscript and Mr Peter Schwartz for his help with the references.

Key words: Cholesteatoma

References

Bezold, F. R., Siebenmann, F. (1908) Textbook of Otology. Chicago Medical Book Co., Hammond Press, W. B. Conkey Company: Chicago, Illinois. p. 201–209.

Friedmann, I. (1955) The comparative pathology of otitis media—experimental and human. II. The histopathology of experimental otitis with particular references to experimental cholesteatoma. *Journal of Laryngology and Otology,* **69:** 588–601.

Habermann, J. (1888) Zur enstehung des cholesteatomas des mittelohres (Cysten in der schleimhaut der paukenhohle, atrophie der nerven in der schnecke). *Archiv für Ohrenheilkunde,* **27:** 42–50.

Michaels, L., Wells, M., Frohlich, A. (1983) A new technique for the study of temporal bone pathology. *Clinical Otology,* **8:** 77–85.

Palva, T., Palva, A., Darmmert, K. (1968) Middle ear mucosa and chronic ear disease. *Archives of Otolaryngology,* **87:** 21–29.

Ruedi, L. (1963) Acquired cholesteatoma. *Archives of Otolaryngology,* **78:** 252–261.

Sade, B., Babiacki, A., Pinkus, G. (1982) The metaplastic and congenital origin of cholesteatoma. Cholesteatoma and mastoid surgery procedures. 2nd International Conference, Tel Aviv, Israel, March 22–27, 1981. Kugler Publications: Amsterdam. p. 305–309.

Sheehy, J. L., Brackmann, D. E., Graham, M. S. (1977) Cholesteatoma surgery: residual and current disease—a review of 1,024 cases. *Annals of Otology Rhinology and Laryngology,* **86:** 451–462.

Tumarkin, A. (1961) Pre-epidermosis. *Journal of Laryngology, and Otology,* **75:** 487–500.

Wells, M., Michaels, L. (1983) Role of retraction pockets in cholesteatoma formation. *Clinical Otolaryngology,* **8:** 39–45.

Wendt, A. (1873) Uber das verhalten des gehorsorgans und des nasenrachenraum bei varid. *Archiv für Ohrenheilkunde,* **7:** 85–87.

Wittmaack, K. (1933) Wie entsteht ein genuines cholesteatom? *Archiv für Ohrenheilkunde,* **137:** 306–331.

Address for correspondence:
Mrs M. D. Wells, M.D., F.R.C.S.,
Consultant ENT Surgeon,
The William Harvey Hospital,
Kennington Road,
Willesborough,
Kent TN24 0LZ.