The Journal of Laryngology & Otology (2006), ***120***, *740–744.*

doi:10.1017/S0022215106002349
Printed in the United Kingdom
First published online 19 July 2006

Main Article

Complications of keratosis obturans

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Abstract

Three patients with extensive keratosis obturans were treated during a 12-month period. One presented with an idiopathic sensorineural hearing loss and was found to have keratosis obturans in the contralateral, asymptomatic ear. The disease process had resulted in a horizontal semicircular canal fistula in what was now, effectively, the only hearing ear. The second patient had an extensive dehiscence of the tegmen tympani. The third presented with a facial palsy. An automastoidectomy cavity was present, with circumferential skeletonization of the descending facial nerve over a length of 1.5 cm and dehiscence of the temporomandibular joint and jugular bulb. All three patients were successfully treated by surgical formalization of their automastoidectomy cavities. They appeared to represent cases of keratosis obturans rather than external auditory canal cholesteatoma, on the basis of previously published reports.

These complications and patterns of bone erosion have not previously been described in keratosis obturans. The third patient is believed to have the most extensive case of keratosis obturans yet described.

Key words: Ear, External; Keratosis; Cholesteatoma; Mastoid; Treatment Complications

Introduction

Keratosis obturans is a rare condition in which the bony portion of the ear canal becomes occluded by a plug of desquamated keratin. Whilst the condition was first described by Toynbee in 1850,[1] the term 'keratosis obturans' was first used by Wreden in 1874.[2] For many years, the terms 'keratosis obturans' and 'external auditory canal cholesteatoma' were used interchangeably. More recently, a distinction has been drawn between the two conditions on the grounds of differing clinical[3] and pathological[4,5] features.

It is generally accepted that keratosis obturans may result in some widening of the ear canal. We present three cases which demonstrate previously unreported patterns of extensive bone erosion.

Case reports

Case one

A 50-year-old woman was referred, having suffered a recent, idiopathic, sudden sensorineural hearing loss in her right ear. She had been aware of a hearing loss in the left ear for many years and wore a hearing aid on that side. Both ears were otherwise asymptomatic. She gave a history of left-sided ear infections during childhood but the ear had not discharged since the age of 18 years. There was no other significant medical history.

Examination revealed a normal looking right ear but an apparent automastoidectomy cavity on the left. This contained a hard plug of wax and keratinous debris which proved too painful to remove.

Audiometry showed a 50 to 60 dB sensorineural loss on the right, with an optimum speech discrimination score of only 32 per cent. On the left, there was a 30 dB conductive hearing loss.

A computed tomography (CT) scan of the temporal bones confirmed erosion of the posterior canal wall, with material of soft tissue density in the mastoid, and also demonstrated a large fistula of the horizontal semicircular canal (Figures 1 and 2).

A diagnosis of keratosis obturans was made. It was felt that further erosion of the labyrinthine bone would pose a significant threat to the patient's hearing, and that, despite the affected ear being effectively her only hearing ear, surgical intervention was the preferred option. At operation, there was found to be marked erosion of the posterior and superior walls of the bony ear canal, and this area was filled by densely impacted, stratified layers of squamous epithelium (Figure 3). The large fistula of the horizontal semicircular canal was confirmed and the facial nerve was found to be dehiscent in both the horizontal and upper vertical segments (Figure 4). The tympanic membrane and middle ear were normal. A modified radical mastoidectomy was performed, with particular attention being paid to performing a meatoplasty large enough to enable future cleaning of the cavity. The epithelium covering the fistula was not disturbed.

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Accepted for publication: 21 March 2006.

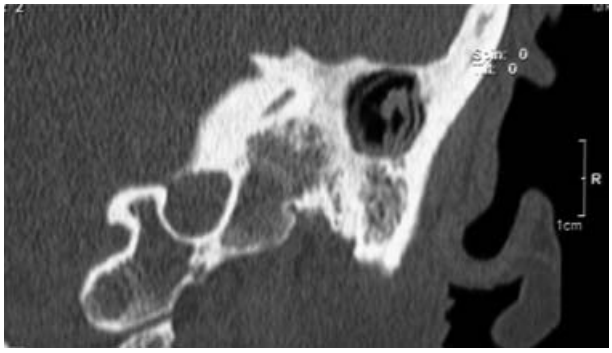

FIG. 1

Coronal computed tomography scan of patient one, showing epithelial debris in an apparently lamellar pattern within the automastoidectomy cavity.

The cavity healed rapidly, although the lining epithelium remained non-migratory. A post-operative audiogram showed an improvement of 15 dB in air conduction thresholds on the left side.

Case two

A 71-year-old woman presented with a long-standing history of a discharging left ear. There was no other medical history of note.

Examination revealed a hard crust over the pars flaccida of the tympanic membrane and superior aspect of the bony ear canal. Attempts at removing this crust were unsuccessful. A pure tone audiogram revealed a mixed hearing loss with a 10 to 30 dB air–bone gap.

A CT scan revealed marked erosion of the roof of the ear canal and outer attic wall, with apparent dehiscence of the floor of the middle cranial fossa (Figure 5). The tympanic membrane appeared thickened but the middle ear was well aerated.

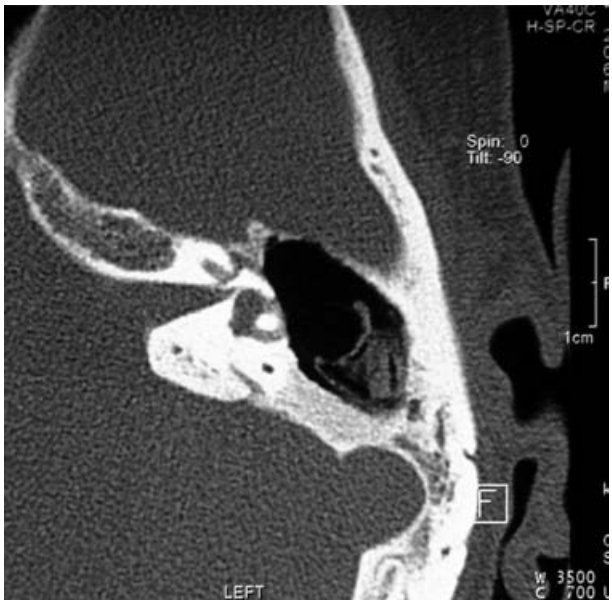

FIG. 2

Axial computed tomography scan of patient one, showing the extent of the cavity and the large horizontal semicircular canal fistula.

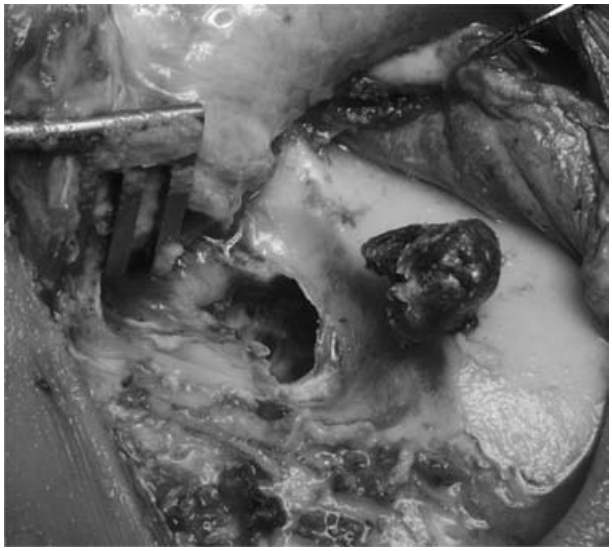

FIG. 3

The left mastoid region of patient one after limited removal of the bony cortex. The keratinous plug that was filling the cavity has been placed on the adjacent bone.

At surgery, extensive bone destruction was found, with squamous epithelium in contact with the middle fossa dura and extending back into the sino-dural angle. The tympanic membrane, middle ear and lower half of the mastoid were intact. A modified radical mastoidectomy was performed. The tegmen defect was repaired with cartilage reinforced by a middle temporal artery flap. Healing of the cavity was uneventful.

Case three

A 53-year-old man presented to our unit six weeks after suffering a rapid onset of right-sided facial weakness. He had suffered a similar episode one year previously, from which he had recovered

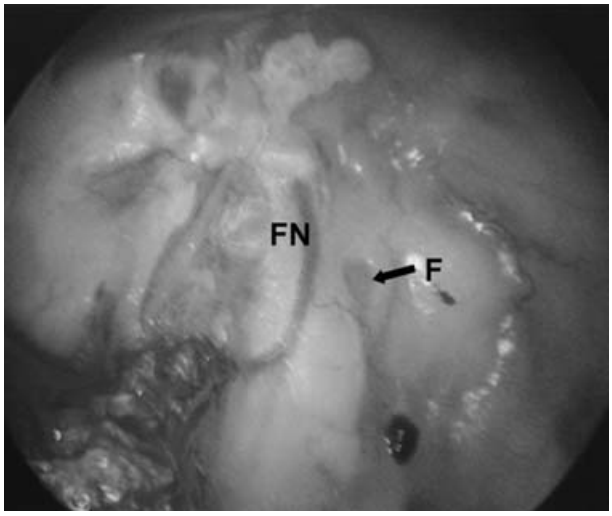

FIG. 4

Endoscopic view of the mastoid cavity of patient one. The exposed facial nerve (FN) and horizontal semicircular canal fistula (F) are clearly seen.

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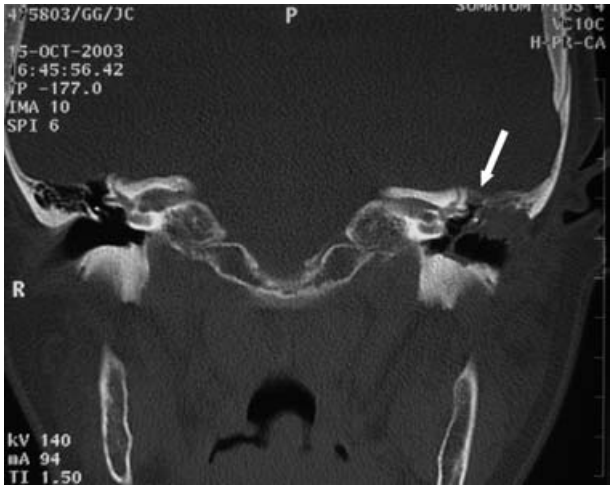

FIG. 5
Coronal computed tomography scan of patient two. The area of dehiscence of the middle fossa plate is arrowed.

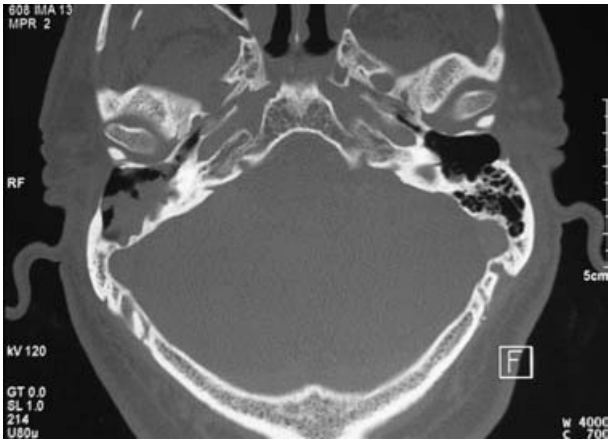

FIG. 6
Axial computed tomography scan of patient three. Extensive bony erosion is seen on the right. The facial nerve canal is not seen on this side and the mastoid cavity is filled with material of soft tissue density. On the left, there is a more limited bony defect. The canal of the descending facial nerve is partly eroded.

spontaneously over a period of four months, leaving a very mild residual weakness. He had also suffered a right facial palsy 30 years previously, which had resolved spontaneously and completely. Both of the previous episodes had been considered by the physicians treating him at the time to be idiopathic.

On further questioning, the patient admitted to a long history of right-sided ear problems, having had a polyp removed from that ear and having been aware of a gradual loss of hearing. Over the past 18 months, he had suffered a few episodes of slight imbalance. He also complained of a dull ache, which had been present for several weeks and which he localized to the right mastoid region. Of note in his past medical history was long-standing bronchiectasis.

On examination, the patient had a complete, right-sided, lower motor neuron facial palsy. The right ear was filled by extensive keratinous debris which was painful to aural toilet. On the left side there was also keratinous debris, with the facial nerve visible in the eroded posterior ear canal wall. Clinical tests of vestibular function were normal. An audiogram revealed a bilateral 30 dB air–bone gap superimposed on a high tone sensorineural loss consistent with early presbycusis.

A CT scan of the temporal bones (Figure 6) showed gross erosion of the right ear canal and mastoid, with the cavity filled by material of soft tissue density. The canal of the vertical segment of the facial nerve was not visible. The left ear canal also appeared expanded, with erosion of the facial nerve canal in the vertical segment.

The patient was scheduled for urgent surgery. At operation, the presence of a very extensive auto-mastoidectomy cavity was confirmed. The cavity was filled by layered squamous epithelial debris. The facial nerve had been completely skeletonized over a length of 1.5 cm and was stretched anterolaterally over a mass of epithelial debris (Figure 7). The temporomandibular joint was partially eroded and almost the entire tympanic bone was missing. The jugular bulb was widely dehiscent. The tympanic membrane was intact, thickened and medialized, with the fibrous annulus clearly visible (Figure 8).

After decompression of the facial nerve, removal and bevelling of the remaining cortical bone was undertaken to formalize the mastoid cavity. A generous meatoplasty was performed to provide access for cleaning of the cavity.

Discussion

In 1980, Piepergerdes and Behnke were the first to make a distinction between keratosis obturans and external auditory canal cholesteatoma (EACC).[3]

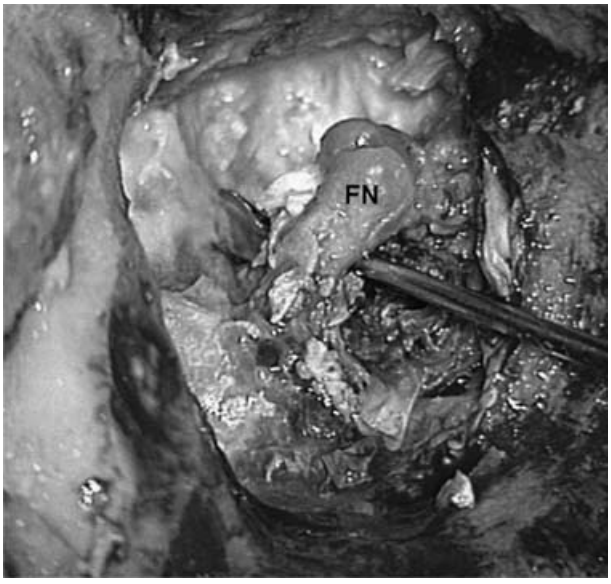

FIG. 7
Operative view of the right ear of patient three. The epithelial and keratinous debris has been largely removed. Some remains, deep to the facial nerve (FN), which is bowed forwards. A Fisch dissector has been passed medial to the nerve.

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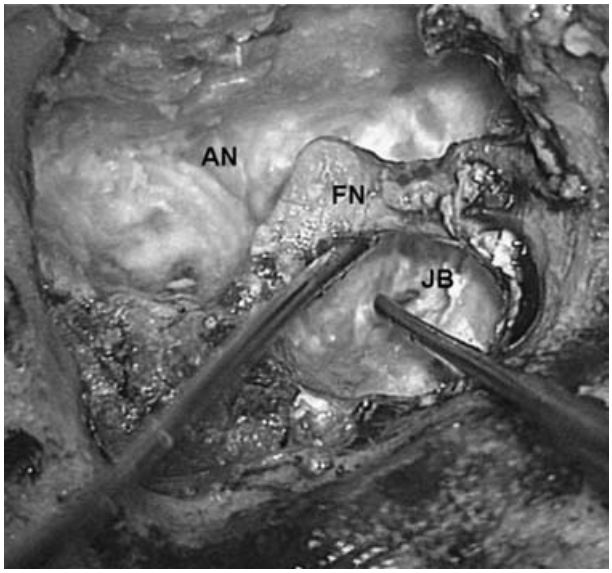

FIG. 8
The final appearance of the mastoid cavity of patient three. The skeletonized facial nerve (FN), exposed annulus (AN) and dehiscent jugular bulb (JB) are all clearly seen.

They described keratosis obturans as a condition presenting in a younger age group, with hearing loss and acute pain, due to the accumulation in the ear canal of large plugs of desquamated keratin. The ear canal may in severe cases be widened or 'ballooned',[6] and the tympanic membrane is intact but usually thickened. The fibrous annulus may be especially prominent. More extensive bone erosion has also been described in keratosis obturans. Hawke and Shanker[7] and Persaud *et al.*[8] both reported cases which had resulted in automastoidectomy cavities.

External auditory canal cholesteatoma was described by Piepergerdes and Behnke as presenting in older patients with otorrhoea and dull otalgia and with a localized area of cholesteatoma formation and bone erosion, usually in the posterior or inferior canal wall just lateral to the annulus. Bony erosion has more recently also been reported both in the anterior[9] and superior[10] canal walls and circumferentially.[11]

Keratosis obturans is associated with bronchiectasis and/or sinusitis in approximately 80 per cent of childhood cases and 20 per cent of adults,[12] whilst no such association has been demonstrated for EACC. Keratosis obturans may affect both ears in 44 per cent of cases.[12] There have been a number of case series including bilateral EACC,[4,9,10,13–15] with the estimated incidence of bilateral involvement ranging from between 20 and 50 per cent.

The pathogenesis of keratosis obturans is not clearly understood. Corbridge *et al.*[16] showed an association with delayed or aberrant epithelial migration. Several authors have described hyperaemia of the ear canal. It is unclear whether this in some way causes excessive desquamation and the build-up of debris or whether it is a consequence of it. In milder cases, just the tympanic membrane may be affected.[17] However, if the ear canal skin is affected, it is in a generalized rather than local fashion. The mechanism of bony erosion in keratosis obturans is unclear. Proteolytic enzymes may be involved,[11] or the process may be one of bony remodelling caused by the pressure of the expanding keratin plug. The bowing of the facial nerve evident in case three demonstrates that the plug can indeed exert significant pressure.

External auditory canal cholesteatoma may be primary or secondary to surgery, trauma, or stenosis or obstruction of the ear canal.[16] Primary EACC is believed to result from epithelial erosion into a localized area of periostitis and bone necrosis.[4,5] Overexpression of vascular endothelial growth factor may play a role in this process,[17] but although there may be extensive bone erosion and mastoid involvement, the bony defect in the ear canal wall itself is generally small.[18,19]

Distinguishing between keratosis obturans and EACC can be difficult, and the two conditions may appear to overlap. Classically, keratosis obturans is found in younger patients, is bilateral, causes more acute, severe pain and results in a conductive hearing loss which is not present in EACC. Otorrhoea is said to be rare in keratosis obturans but is common in EACC, and bone erosion is circumferential in keratosis obturans and usually focal in EACC. Keratosis obturans is associated with bronchiectasis, whilst EACC has no such association with any systemic disease. However, many patients present atypically and some may have features suggestive of both conditions. Persaud *et al.*[20] carefully examined the reported cases and concluded that clinical presentation alone is insufficient to enable a clear distinction between the two diseases. They felt that the most reliable indicators of EACC were the presence of osteonecrosis or bony sequestration and the focal loss of epithelium covering the external ear canal. Neither of these features is thought to be found in keratosis obturans.

- **This paper reports a series of patients with keratosis obturans**
- **The cases illustrate several previously unreported complications of this disease, including dehiscences of the tegmen, jugular bulb and temporomandibular joint, as well as facial palsy**

The authors believe that all three of our cases represent keratosis obturans rather than cholesteatoma[21] and in EACC. The presence of otorrhoea is in case 2 is atypical, although has been reported in up to 20% of cases.[21] Of particular note is that case 1 was largely silent and case 3 presented only when a major complication developed.

Both the degree and pattern of bone erosion seen in these three cases is highly unusual. Horizontal semicircular canal fistula is a well recognized complication of cholesteatoma associated with chronic suppurative otitis media, and it has been reported once in a case of EACC secondary to fracture of the

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temporal bone.[22] However, it has not been previously described as a complication of keratosis obturans.

Heilbrun *et al.*[11] described one case of EACC with a dehiscent tegmen tympani, although the extent of the dehiscence was not reported. Martin *et al.*[19] reported a case of EACC in which the sigmoid sinus and posterior fossa dura were exposed. However, the case of patient two constitutes the first report of significant erosion of the floor of the middle cranial fossa in this disease, and we feel that the size of the defect would have placed the patient at significant risk of intracranial complications had it not been explored and repaired.

Involvement of the facial nerve has been reported in both keratosis obturans and EACC. Persaud *et al.*[8] described a case of keratosis obturans in which the facial nerve canal was significantly eroded and the presenting symptom was a metallic taste. There have also been a number of reports of erosion of the facial nerve canal in EACC,[11,13,18,19,23] but only two cases of facial palsy have been described.[15,24] Facial palsy has not previously been described in keratosis obturans, nor has circumferential skeletonization of the facial nerve.

Case three demonstrates two other patterns of bone erosion that have not previously been associated with keratosis obturans, namely, erosion of the temporomandibular joint and dehiscence of the jugular bulb. Garin *et al.*[9] reported two cases of EACC, one with each of these complications.

Most cases of keratosis obturans may be treated conservatively with topical medications and regular removal of the keratin plug. This procedure may require general anaesthesia. Paparella and Goycoolea[25] described three cases of uncomplicated but refractory keratosis obturans treated by split skin grafting and canalplasty. External auditory canal cholesteatoma may similarly be managed conservatively, although only in very limited cases.[26] If there is significant bone destruction, surgical intervention is indicated, with removal of the cholesteatoma and debridement of necrotic bone. In very extensive cases, canal wall down mastoidectomy may be required.

All three cases described here required surgical intervention because of the onset or threat of major complications. In all three, the disease process itself had created very well contoured automastoidectomy cavities. These were formalized by extensive removal and bevelling of bone laterally and by creation of a generous meatoplasty. The authors believe this latter step to be particularly important, as the epithelium lining the cavity is likely to remain non-migratory and good access is required for regular cleaning.

Conclusion

Keratosis obturans is generally a relatively benign condition but can result in serious complications. The cases presented here illustrate several previously unreported complications, namely, labyrinthine fistula, tegmen dehiscence, facial palsy, and dehiscence of the temporomandibular joint and jugular bulb. The authors believe that the third case in this series is the most extensive example of keratosis obturans yet reported.

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Mr N C Saunders takes responsibility for the integrity of the content of the paper.
Competing interests: None declared