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Clinical Record

Neglected keratosis obturans causing facial nerve palsy

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Abstract

Keratosis obturans is characterized by the accumulation of desquamated keratinous material in the bony portion of the external auditory canal. Classically, it is reported to present with severe otalgia, conductive hearing loss and global widening of the external auditory canal. Extensive erosion of the bony meatus, with exposure of the facial nerve, has been previously reported, but no case of facial nerve palsy has as yet been published. We report the first published case, to our knowledge, of a unilateral facial nerve palsy secondary to neglected keratosis obturans.

Key words: Ear Canal; Keratosis; Cholesteatoma; Facial Palsy; Cerumen

Introduction

Keratosis obturans (KO) is a condition in which a cholesteatoma-like mass fills the deep external auditory meatus (EAM). It was first described by Toynbee in a brief case report in the *London Medical Gazette* in 1850.[1] In 1874, Wreden presented 12 cases of a 'peculiar form of obstruction of the external auditory meatus consisting of a compact mass of cerumen'.[2] The characteristic clinical manifestations are severe otalgia and hearing loss due to accumulation of large plugs of desquamated keratin in the ear canal. There may be circumferential widening of the bony external auditory canal (EAC), but no evidence of osteonecrosis or bony sequestration, as is found in EAC cholesteatoma.[3]

We report an atypical, previously undescribed presentation of KO.

Case report

A 56-year-old gentleman with a background history of bronchiectasis and KO presented to the emergency department with a 48-hour history of a left-sided facial nerve palsy. This gentleman had attended the out-patient department regularly for microdebridement for the preceeding 26 years. However, he had failed to attend for the previous two years. On presentation, on this occasion, the patient also complained of severe pain and reduced hearing in his left ear. Otoscopy revealed impacted keratinous debris in the left ear. A computed tomography scan of the temporal bones (Figure 1) revealed a left-sided, rounded, 2 cm soft tissue mass causing erosion of adjacent temporal bone, with an apparently normal tympanic cavity, ossicles and scutum. The patient proceeded to examination of his left ear under anaesthetic. A waxy, keratinous plug was removed, revealing deeply eroded anterior, inferior and posterior canal walls. The tympanic membrane was mildly thickened and the vertical portion of the facial nerve was partially exposed. The cavity was lined with mildly erythematous squamous epithelium. There was no evidence of osteonecrosis or sequestrum formation. The canal was completely debrided, and facial nerve function fully recovered.

Discussion

Keratosis obturans and external auditory canal cholesteatoma (EACC) have been considered separate entities for the past 20 years, after being previously regarded as variations of the same disease for at least 87 years. While both disorders are distinct, they do have some overlapping characteristics which make definitive diagnosis difficult. In 1980, Piepergerdes *et al*. reviewed the literature and highlighted the differences between the two disorders; they did this by comparing the clinicopathological findings of both conditions.[4] Patients with KO presented with acute conductive hearing loss, severe pain, a widened ear canal, thickened tympanic membrane and, rarely, otorrhoea. Deafness and pain are usually secondary to accumulation of keratin in the ear canal. In contrast, EACC is characterized by otorrhoea and chronic, unilateral, dull pain secondary to invasion of squamous tissue into a localized area of periosteitis in the canal wall. Hearing and the tympanic membrane are usually normal in EACC. Piepergerdes *et al*. also reported that most cases of EACC occurred unilaterally and in the elderly population. Seventy-seven per cent of children and 20 per cent of adults with KO have associated sinusitis or bronchiectasis.[5] Bilateral KO is more common in children.[6]

Bunting stated that bony erosion of the canal wall can occur in both KO and EACC.[6] In an attempt to differentiate the disorders, Piepergerdes *et al*. described two different erosion patterns. Keratosis obturans is associated with a greatly widened ear canal due to circumferential erosion of bone, whilst, in EACC, the erosion is localized to the postero-inferior aspect of the bony canal.[4] Naiberg *et al*. stated that erosion of the underlying bone does not occur in KO.[7] However, Hawke and Shanker reported in 1986 a case of automastoidectomy caused by KO and suggested that bony erosion may be due to the pressure exerted by the silently accumulating mass of keratin in the EAC.[8] In

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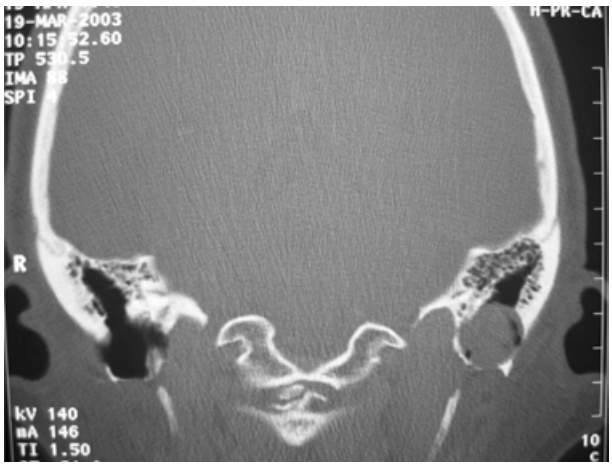

FIG. 1

A rounded, well defined mass is seen in the left external auditory canal. Note widening or ballooning of both external auditory canals.

the case presented, there was extensive bony erosion in the anterior and postero-inferior segments, with exposure of the vertical portion of the facial nerve. There was no bony necrosis or sequestrum of the underlying bone, and all areas were covered by mildly erythematous epithelium. In EACC, there is usually bony necrosis or sequestrum of the underlying bone,[9] and this is distinctly absent in KO.[7]

The aetiology of KO remains uncertain, although it has been related to eczema, seborrhoeic dermatitis and furunculosis[10,11] and even to bronchiectasis, leading to reflex sympathetic nervous system stimulation of ceruminal glands and subsequent formation of an epidermal plug.[5] Paparella and Shumrick suggested that the epidermal plug may also be due to excessive production of epithelial cells or faulty epithelial migration.[12] This was confirmed by Corbridge *et al*. in 1996.[13] Ink dot markers placed in the centre of the ear drum normally move laterally, whilst markers placed on the drum over the malleus handle move upwards to the pars flacida and then postero-superiorly. In KO, there may be abnormal migratory pathways, no migration or abnormally slow movement of epithelium, leading to accumulation of squamous debris. The pathogenesis of the abnormal migration may be associated with damage to the migratory basal epithelial cells following inflammation involving these cells, and a similar aetiology may be responsible for the inflammation of the lower respiratory tract and paranasal sinuses that often co-exists.[14]

As both KO and EACC arise in the EAC, there is an overlap of signs and symptoms. Examination of cases presented in the literature revealed that there is no consistent mode of presentation. However, a careful assessment of the reported cases revealed that osteonecrosis or bony sequestration is found in EACC and not in KO, and is the main finding which consistently differentiates the two disorders.[3]

This case illustrates dramatically the atypical and extensive erosion that can occur in a neglected case of KO. During a two-year absence from follow up, a facial nerve dehiscence was produced by erosion of the posterior bony canal wall and adjacent mastoid air cells, due to pressure exerted by the accumulating mass of keratin within the EAC. This case emphasizes the importance of regular aural toilet in patients with this condition.

- **Keratosis obturans and external auditory canal cholesteatoma are two distinct clinical entities which share similar characteristics**
- **Bony erosion or sequestrum formation are absent in keratosis obturans**
- **Neglected keratosis obturans may cause pressure necrosis of underlying bone, thereby causing a facial nerve palsy**

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