Keratoses Obturans: A Disease of the Tropics?

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Abstract Keratoses obturans appears to be an obscure and relatively uncommon entity, even in literature search of journals and reference texts, so much so that there is not even any prevalence or incidence statistics available. However, the condition did not appear to be as uncommon based on our clinical observations. We have managed to obtain 64 patients representing 67 ears with keratoses obturans in our study period of about 18 months with a pattern of occurrence during this period. Humid weather seemed to play a role in the frequency of its appearance during certain period in our observation. There also appears to be a correlation between the severity of symptoms (predominantly pain and hearing loss) and the presenting appearance of the condition, i.e., presence or absence of granulation tissue, as well as that the degree of difficulty in exenteration of the keratoses obturans (matrix and content) depending on the expansion of the bony canal. Our figures showed the majority of the patients are females and young individuals, the majority of them occur unilaterally. The condition also appear to stop short of involving the tympanic membrane with only the bony canal being expanded with the surrounding oedema creating an apparent “canal stenosis”.

Keywords External ear · Keratoses obturans · Pain

Introduction and Background

Although Toynbee was frequently credited for noting a pearly white shining mass occupying the posterior surface of the external canal which he called mollusum contagiosum [1] in 1850, keratoses obturans was in fact first properly described and named by Wreden of St. Petersburg in 1874, who differentiated the condition from that of impacted wax (which was then called ceruminosis obturans) [2]. Schofield further added another terminology, cholesteatoma of the external auditory canal in 1893, which he attributed to an insect [3] sting or bite.

Piepergedes and Behnke [4] clarified the two conditions defining the distinction between the two, which until then had been considered different presentations of the same disease; keratoses obturans was defined as an accumulation of keratin plugs within the ear canal which may result in some widening of the external auditory canal, and external auditory canal cholesteatoma as bone erosion resulting from squamous tissue at a specific site in the external auditory canal. Further distinction has been drawn between the two conditions on the grounds of differing clinical [4] and pathologic features [5, 6].

Both conditions are considered uncommon, with cholesteatoma of the external auditory canal quoted as 0.1–0.5% of patients with otologic disorder [7–9]. Several classifications have been proposed for external auditory canal cholesteatoma [10, 11] none was available for keratoses obturans. Further, no incidence or prevalence figures are available for keratoses obturans despite extensive search, further emphasising its rarity of occurrence.

Due to the paucity of information, we present findings from our institution where keratoses obturans appear to present more frequently than in articles already published.

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Materials and Methods

The period of data collection was from March 2012 to October 2013. As inclusion criteria we used the accepted clinical features of

1. Presentation of pain (may be acute and severe) and/or deafness (conductive) in the affected ear(s);
2. A plug of “organized” keratin, usually circumferential, surrounded by a layer of pearly-white matrix;
3. A widened bony canal;
4. The presence of granulation tissue within the external auditory canal usually at the bony-cartilaginous junction; and
5. Intact, mobile tympanic membrane.

Exclusion criteria were:

1. Presence of bony erosion;
2. Foul-smelling mucopurulent otorrhoea, a feature of periosteitic activity, involving the temporal bone in cholesteatoma.

Other demographic details noted include age, gender, seasons (month of occurrence).

Results

A total of sixty-four (64) patients, which represented sixty-seven (67) ears, who fulfilled the criteria for diagnosis were entered into the study; there were 2 patients who had recurrence and one (1) patient with bilateral disease at the time of presentation.

The age ranged from 9 to 81 years, with the mean of 27.3. Fifty-nine (59) were in the third and fourth decades, with 2 in the first, 1 in the second, 1 in the sixth and 1 in the ninth decade of life; the majority being in their early- to mid-twenties. Females outnumbered males with 61 females against only 3 male patients (ratio of 20:1). All the three males patients were in the early twenties. The 2 patients with recurrence and one patient with bilateral were females.

There were notably 2 peaks when the number of patients were significantly higher, i.e., in April–June and November–January, which generally corresponded with the changing of the monsoon seasons experienced in this country. This is the intermediate period of the weather change, i.e., extremely hot and dry weather followed by heavy downpours and vice versa. This corresponds to the peak which the condition presented, i.e., beginning from end-April, end-June, and end-October, end-January.

Pain was the predominant symptom in all the patients (severe in 9 (14%) of the patients, causing disturbed sleep

and interrupted daily routine). Hearing loss was the next common complaint with 51 (79.6%) patients having varying degree of hearing impairment. In 13 (20.4%) there was no perceivable hearing loss.

All the patients had exenteration (removal) of the keratosis obturans which involved careful peeling off of the tightly adherent matrix. On some occasions this may be achieved in a single session of patiently multi-staged aural toilet by vacuum suctioning. However, many required more than one sitting for the complete removal of the disease. In 2 cases involving children under 12-years of age, the procedure had to be done under general anaesthesia.

Widened canal due to the expansile nature of the disease, specifically the bony canal, was noted in every patient, except one. The area of involvement was exclusively in the posterior bony canal extending inferiorly. This finding was not found in the anterior wall of the bony canal. There is associated apparent narrowing at the isthmus at the osteo-cartilaginous junction; this is due to the oedema and swelling in the cartilaginous portion of the external auditory canal in relation to the widened bony canal.

There were two (2) degrees of expansion of the bony canal noted, ranging from (1) minimal (Fig. 1) or no expansion [noted in (1.5%) ears], and (2) presence of recognisable expansion (arrow—Fig. 2). Even in extensive disease, there was not a single case where the facial nerve was involved causing facial palsy. We have staged the disease process based on the symptoms and extent of expansion noted as this will provide an estimate as to the aggression displayed by the disease (see Table 1; Figs. 1, 2, 3).

Fig. 1 No discernible widening of the canal noted after clearing of the tightly adherent matrix (grade 1)

Fig. 2 Widening of the bony canal is seen in the inferior direction (after keratosis obturans is removed) as indicated (grade II)

Fig. 3 Presence of matrix and keratin (yet to be completely removed). Also indicating a “widened” bony canal (arrow **a**). Granulation tissue (**b**) seen at the osteo-cartilaginous junction (arrow) (grade III)

Table 1 Keratosis obturans (KO) (grades I–III)

Grade I	Mild pain $\pm$ ear block with the presence of accumulated keratin enveloped by a tightly adherent matrix; no discernible expansion of external canal (Fig. 1)
Grade II	Moderate to severe pain $\pm$ conductive deafness; presence of accumulated keratin enveloped by a tightly adherent matrix with mild expansion of the bony canal (arrow) in the presence of keratosis obturans (Fig. 2)
Grade III	Moderate to severe pain $\pm$ conductive deafness; presence of accumulated keratin enveloped by a tightly adherent matrix with expanded bony canal (arrow a) with granulation tissue (arrow b) at the osteo-cartilaginous junction (Fig. 3)
Grade IV	Presence of accumulated keratin enveloped by a tightly adherent matrix (grade III) with exposure of the mastoid air cells with/without facial nerve involvement

There was no grade IV noted in our series

Granulation was found in 46 (68.7%) ears (Fig. 3—arrow-b), in whom pain appeared to be more severe than those in whom granulation tissue was absent. The removal of keratosis obturans in these patients was more difficult when granulation tissue resulted in further “constricted” view in the canal. In 3 cases, the granulation tissue completely obstructed the canal and excision of the granulation by snaring was necessary to facilitate the exenteration of keratosis obturans. The histopathology examinations of the granulation tissues sent were reported as “acute inflammatory tissue” with no specific pathology identified. No fungus was reported in the specimens.

None of the patients had a perforated tympanic membrane. All these patients recorded conductive hearing loss of mild to moderate severity in Pure Tone Audiometry (10–30 dB air-bone gap).

Examination of the ear after clearance of the ear canal revealed an unaffected intact tympanic membrane. There was no complication post-clearance except for the widened bony canal. Subsequent follow-ups at 1 week, 3 weeks and three months post-clearance follow up did not present further changes or recurrence. None needed a meatoplasty to ease future care.

One common denominator among all the subjects was chronic use of cotton buds to clean the ears.

Discussion

It is difficult to differentiate between keratosis obturans and impacted wax at first presentation. Keratosis Obturans is a clinical entity which is often only diagnosed when attempts at removal of accumulated desquamated keratin from deep in the ear canal elicits excruciating pain and the visualisation of the silvery white matrix at its periphery. Stripping or peeling off of the matrix may result in bleeding may be due to neovascularization; the formation of new capillaries around the matrix is probably the result of the inflammation or irritation of the surrounding skin of the bony canal.

Granulation tissue as a result of inflammation of the surrounding tissue typically forms at the bony-cartilaginous junction of the apparently “stenosed” external canal. The surrounding oedema, together with the granulation tissue, further exaggerate the appearance of a tight and narrow external canal. This apparent stenosis makes the removal of the accumulated keratin and the surrounding matrix a challenge, a test of patience and perseverance to both

patient and doctor. Often more than one session may be required to completely and satisfactorily remove the entire content along with the matrix. The granulation tissue subsides spontaneously after keratosis obturans is completely exenterated.

While it is commonly seen in younger persons, in the 3rd and 4th decade, it may also occur less commonly in the young as well as in older population, as shown in our observation. The most common presenting complaint in our patients is pain, ranging from a constant throbbing ache to such excruciating pain that the sufferer could not even sleep the entire night. Hearing impairment is the next common complaint, but this is usually elicited by the attending doctor rather than volunteered by the patient; perhaps the pain so greatly overwhelmed the patient that the hearing deficit became a secondary concern.

The pain experienced by each of our patients is dependent on both the patient's tolerance of pain as well as the extent of the disease, i.e., the extent of bony expansion, and the activity of the inflammatory process. In almost every case in our observation, the predominant stages noted are grades II (19.4%) and III (68.7%).

Although complications such as tympanic membrane perforation have been reported [12] there was none observed in our series.

Keratosis obturans remains a rare and obscure condition in most Western countries, hence the paucity of information in journals and reference texts. The definition of the disease condition is just as vague and imprecise.

Many hypotheses have been propounded but none has satisfactorily been able to explain the contributing factors, nor the actual aetiology of the disease; these included risk factors associated with sinusitis and bronchiectasis [2], faulty migration of the squamous epithelial cells from the tympanic membrane and adjacent canal wall [4, 7–9], etc. Inflammation caused by virus, fungus and parasite have been proposed as the cause which led to initiation of the disease process. There is also much confusion between whether KO and external canal cholesteatoma are two different entities, or one entity but at opposite ends of the disease spectrum [4–6].

In our observation, none of the above suggested possibilities was seen. However, two patterns are noted in nearly everyone of the patients, i.e., regular use of cotton bud to “clean” the ear of wax and the frequency of these patients presenting coinciding with the transitional season in our tropical climate. While cotton bud use may contribute to disrupting the normal epithelial migration of the desquamated keratin, this phenomenon has not been observed in other patients. Frequency of use also did not feature as a contributing factor, again keratosis obturans did not occur in many who fervently and regularly use cotton buds (sometimes several times a day).

However, we noted an emerging pattern in our observation in which patients with keratosis obturans present more frequently in the transitional period between hot, dry weather and the rainy season of the monsoon (a period of moderate heat and humidity). The numbers dropped after the onset of the monsoon until the next cycle, which generally happens twice annually in the tropics. Although initial thoughts of fungal infection may be a cause of the disease, this was dispelled when no fungus was grown from the specimen of KO removed from a patient's ear.

Conclusion

Despite the relatively large number of patients who presented to our clinic who were subsequently found to have keratosis obturans, there is no typical presentation aside from it being common in young adults and pain as the main presenting complaint. Hearing deficit is of conductive type, but it is often only a secondary problem (the symptom is usually elicited by the doctor and only occasionally volunteered by the patient).

We have staged the disease based on the extent of bony expansion that is observed in each patient. Although this observation is subjective, we feel that this may be helpful in anticipating the degree of pain, especially when granulation tissue is encountered.

In closing, we would like to propose a working definition which may help throw suspicion on this condition: “Accumulation of desquamated keratin surrounded by a tightly adherent matrix in the expanded bony (inner two-thirds) of the external auditory canal with associated moderate to severe pain, with or without noticeable hearing deficit. This condition is more commonly seen in young adult females living in high humidity tropical climate, but is of unknown aetiology.”

We are of the opinion that keratosis obturans is strictly a disease of the external auditory canal and any localised erosion into the external bony canal and middle ear cleft must be considered as external auditory canal cholesteatoma.

Compliance with Ethical Standards

Conflict of interest None.

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