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Commentary

The Heteromorphic Gob-Keratosis Obturans

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Preface

Keratosis obturans is an exceptional, occlusive disease of the external auditory meatus. Characteristically, anomalous accumulation of a wedge of desquamated keratin within the ear canal is delineated. Keratosis obturans is accompanied by accumulation of lamellar keratinaceous debris which distends and obstructs the external auditory canal with consequent occlusion and expansion of bony segment of the external auditory canal.

Keratosis obturans as a clinical entity can be adequately diagnosed following attempted extrication of accumulated, deep-seated, desquamated keratin confined to the external auditory canal. Keratosis obturans was initially scripted by Tonynbee in 1850 and aforesaid terminology was adopted by Wreden in 1874 [1].

Keratosis obturans is appropriately segregated from impacted cerumen within the external auditory canal and frequently requires a demarcation from cholesteatoma of the external acoustic canal.

As the condition can be misinterpreted and inadequately treated, preliminary disease discernment and appropriate management is necessitated in order to circumvent complications and relieve pertinent clinical symptoms.

Disease characteristics

Of obscure, aetiology, keratosis obturans exemplifies accumulation of desquamated, lamellar keratin within the bony part of external auditory canal [2,3].

Generally, keratosis obturans arises in young individuals below < 40 years. An estimated 50% instances are bilateral wherein bilateral lesions are commonly discerned within children. Around 80% of paediatric subjects and 20% of adults with keratosis obturans of external auditory canal are associated with bronchiectasis or chronic paranasal sinus disease [2,3].

Of obscure pathogenesis. keratosis obturans is associated with diseases such as eczema, seborrheic dermatitis or furunculosis. Besides, keratosis obturans may concur with sinusitis or bronchiectasis, diseases which facilitate reflex sympathetic stimulation of ceruminous glands with consequent configuration of an epidermal plug [2,3].

Excessive production or defective migration of epithelial cells is posited to engender keratosis obturans. Although circumferential dilatation of bony external auditory canal may occur, osteonecrosis or bony sequestration is absent [2,3]. Keratosis obturans is subdivided into:

- Inflammatory variant which arises due to an acute infection such as infection due to viruses. Migration of epithelial cells can be transiently altered on account of the inflammatory process. Besides, keratin extraction may alleviate associated inflammation. Inflammatory subtype can be managed with singular extermination of the keratinous plug.
- Silent variant is engendered by anomalous aggregation of keratin associated with perpetual disease progression despite initial extrication of the keratinous plug [2,3].

Clinical elucidation

Typically, young individuals incriminated by keratosis obturans represent with acute conductive deafness and severe otalgia arising due to accumulation of keratin within the ear. Additionally, the external auditory canal is distended and tympanic membrane is thickened [4,5].

Characteristically, keratosis obturans demonstrates clinical features such as moderate to severe otalgia and decimated hearing due to accumulated spigot of desquamated epidermis within the external auditory canal [4,5].

Sensation of ear blockage and ipsilateral hearing loss can be manifested. Keratosis obturans manifests with extreme, circumferential distension of the external auditory canal accompanied by an intact tympanic annulus which exhibits an appearance of "suspended in the air". Otorrhea is exceptional [4,5].

Cogent clinical symptoms associated with keratosis obturans are acute, severe pain in the ear and conductive deafness [4,5].

Histological elucidation

Keratosis obturans characteristically displays densely impacted keratinous debris deep within the external auditory meatus. Extricated spigot demonstrates acellular, lamellar flakes of keratin and keratinous material [6,7].

Upon microscopic examination, densely accumulated keratinized squamous epithelial cells configure a lamellar pattern. Superimposed stratified squamous epithelium is intact and demonstrates diffuse acanthosis. Epidermal layer of the external auditory canal enunciates hyperkeratosis along with subjacent chronic inflammatory infiltrate composed of lymphocytes, histiocytes and plasma cells. Foci of osteonecrosis are absent [6,7].

Subjacent stratified squamous epithelium is hyperplastic. Subepithelial soft tissue demonstrates an exudation of chronic inflammatory cells. External auditory canal exhibits remodelling and expansion of bony segment of external auditory canal. Erosion or necrosis of subjacent bone is absent. Accumulated, deep-seated keratinous debris within the external auditory canal may engender inflammation of stratified squamous epithelium and remodelling of the bone. Adjacent petrous bone may display severe erosion [6,7].

Differential diagnosis

Keratosis obturans requires a segregation from conditions such as:

- External auditory canal debris which is composed of cerumen admixed with a chronic inflammatory exudate which can partially occupy the ear canal and demonstrate foci of impacted air [8,9].
- External auditory canal cholesteatoma is associated with bony erosion. The lesion is layered by keratinized, non-dysplastic stratified squamous epithelium intermixed with abundant granulation tissue and keratinous debris. Cholesteatoma is infiltrated by a chronic inflammatory infiltrate comprised of lymphocytes and histiocytes. Additionally, epithelioid cell granulomas, foreign body giant cell reaction, cholesterol clefts and foci of hemosiderin pigment deposition may be observed [8,9].
- External auditory canal carcinoma exemplifies as an irregular tumefaction which may or may not be associated with bony erosion. Commonly, tumefaction is composed of infiltrating nests of squamous epithelial cells commingled with keratin pearls. Foci of individual cell keratinization and intercellular bridges may be discerned. Nuclear atypia is variable, mitotic activity with atypical mitosis is frequent. Tumour cell invasion may be superficial and appears associated with irregular budding of basal epithelium or irregular, inferiorly displaced tongues of malignant squamous epithelial cells [8,9].
- Otitis externa exhibits inflammation of circumscribing adipose tissue. Superimposed stratified squamous epithelium is necrotic or ulcerated and demonstrates pseudo-epitheliomatous hyperplasia. Significant, admixed acute and chronic inflammatory cell exudate confined to the subcutaneous tissue may be associated with necrotizing vasculitis. Necrotic foci of bone and cartilage appear commingled with an intense inflammatory cell exudate infiltrating viable bone. Non viable foci of bone or cartilage appear as a variable component [8,9].

Investigative assay

Upon computerized tomography (CT), a well defined soft tissue mass appears confined within the bony external auditory canal. The canal appears diffusely enlarged and appears devoid of bony

erosion. The tympanic canal may be normal or mildly thickened [8,9].

Computerized tomography demonstrates a soft tissue component of lamellar, keratinous spigot frequently obstructing bilateral external auditory canal. Besides, ballooning of osseous segment of external auditory canal is delineated [8,9].

Upon otoscopy, impacted ear wax appears superimposed upon an intact, unremarkable tympanic membrane. Oropharynx and nasal endoscopic examination appears unremarkable, cervical lymph nodes are non palpable and accompanying cranial neuropathies are absent.

Microscopic examination of the ear under general anaesthesia [10,11] demonstrates impacted cerumen in conjunction with a thick, keratinous spigot within the external auditory canal which engenders dilatation of external auditory canal. Bone remodelling may ensue and bony pockets may be configured. External auditory canal demonstrates circumferential dilatation associated with a normal tympanic annulus [10,11].

Therapeutic options

Keratosis obturans can be adequately treated with lavage of the external auditory canal. Packing of incriminated ear along with topical administration of antibiotic ear drops may be employed [10,11].

Frequent extraction of keratin plugs from the external auditory canal is an optimal treatment strategy [10,11].

Regular and continuous aural lavage is necessitated. Additionally, lesions of keratosis obturans lacking associated inflammation of external auditory canal mandate periodic, lifelong aural lavage on account of localized metabolic discrepancies which incriminate normal migratory mechanisms of squamous epithelial cells [10,11].

Topical medication and frequent aural lavage is adopted and beneficial for eradicating the keratinous spigot. The procedure may be performed under general anaesthesia in non-cooperative individuals [10,11]. Refractory lesions can be treated with split- skin graft and canaloplasty.

However, therapeutic surgical intervention is required exceptionally.

Eradication of desquamated keratinous plug may engender intense pain and haemorrhage along the stripped keratinous matrix on account of neovascularization. Configuration of nascent capillaries surrounding the peripheral keratinous matrix ensues due to inflammation or irritation of circumscribing cutaneous layer of bony segment of external auditory canal. Gradually and with appropriate therapy hearing is normalized and pain disappears [10,11].

Reoccurrence of keratosis obturans is frequent [10,11].

Although a benign condition**,** keratosis obturans can engender severe complications. Complications of untreated keratosis obturans appear as sensorineural deafness, dehiscence of tegmen tympani and facial nerve palsy [10,11].

Figure 1: Keratosis obturans depicting fragments of lamellar keratin obstructing the external auditory canal [12].

Figure 2: Keratosis obturans exhibiting accumulated keratinous debris within the external auditory canal along with a disrupted tympanic membrane [13].

Figure 3: Keratosis obturans enunciating lamellar keratin flakes admixed with an inflammatory exudate of neutrophils, lymphocytes and macrophages [14].

Figure 4: Keratosis obturans displaying aggregates of lamellar keratin with a superimposed stratified squamous epithelial layer with acanthosis an hyperkeratosis [15].

Figure 5: Keratosis obturans exhibiting a keratinous plug obstructing the external auditory canal along with compression of the facial nerve [16].

Figure 6: Keratosis obturans exemplifying fragmented, whitish, keratinous accumulate impeding the external auditory canal [17].

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13. Image 2 Courtesy: Science photo library.

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