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Pilomatricoma of the external auditory canal

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A B S T R A C T

Pilomatricoma was first described in 1880 by Malherbe as a calcifying epithelioma, but this was later changed to a benign tumour arising from the hair follicle matrix cells. It typically presents as a firm, painless well-circumscribed mass in children. They are most commonly found in the head and neck, particularly the peri-auricular area. They are slow growing and asymptomatic, generally appearing as a solitary mass but multiple lesions can be seen.

While there is a paucity of case reports on pilomatricoma of the auricle, there are even fewer reported cases of external auditory canal pilomatricomas. We report the case of a 15-year-old male who initially presented with an enlarging external auditory canal lesion, which caused conductive hearing loss. An excision biopsy was performed under general anaesthetic. Histology returned as consistent with pilomatricoma.

1. Introduction

Pilomatricoma is a benign adnexal neoplasm composed of neoplastic cells with a derivation from the hair follicle cortex matrix keratinocytes [1]. Pilomatricoma is the most common non-melanocytic cutaneous neoplasm identified in childhood [2]. The classical presentation is of a slow growing, firm, painless mass [3]. As there are no characteristic macroscopic features, the clinical diagnosis may be difficult. Thus, surgical excision is required to provide a definitive diagnosis.

Within the head and neck region, the most common location is the pre-auricular area, however from a detailed literature review, there is a paucity of reported cases of external auditory canal pilomatricomas described [4,5]. This case report outlines key diagnostic points pertaining to the identification of a common tumour in an unusual site.

2. Case report

We present the case of a 15-year-old male referred to our outpatients department with a three-month history of a slow growing nodule/lesion/tumour in the right auricle. The patient reported progressive right side hearing loss. Pure tone audiometry demonstrated a right sided conductive hearing loss (Fig. 1). There was no prior medical, surgical or family history of note.

On examination, there was a firm, immobile, non-tender lesion extending from the medial aspect of the tragus into the external auditory canal (Fig. 2). The lesion was completely occluding his external auditory canal and the tympanic membrane was not visualised. Debris was seen at the external aspect of the lesion. An inflammatory process was suspected and a Betamethasone (0.1% w/v)/Fusidic acid 2% dressing was applied, with the aim of size reduction with topical corticosteroids.

The patient was reviewed six weeks post initial presentation. There was no alleviation of symptoms and the size of the lesion was unchanged.

At this time, a differential diagnosis included an epidermoid cyst, trichilemmal cyst, pyogenic granuloma (lobular capillary haemangioma), or a teratoma.

He proceeded to theatre and the neoplasm was excised at the base, with the bony margins curetted to ensure complete excision (Fig. 3). The wound was closed with non-absorbable sutures and the external auditory was again packed with a Betamethasone (0.1% w/v)/Fusidic acid 2% dressing. He had an uncomplicated post-operative course and was discharged on the same day.

Fig. 2 illustrates a right tragal lesion protruding within the ear canal. Macroscopic examination revealed an 8mm well circumscribed pale lesion. Microscopic examination of the lesion identified a dermal based well circumscribed non-encapsulated multinodular neoplasm with no epidermal connection. The neoplasm is composed of anucleate cells with abundant eosinophilic cytoplasm predominantly. These cells are characteristic ghost/shadow cells (Fig. 4). There are occasional foci illustrating a monomorphic basaloid population of cells at the rim, with transition to the ghost cells. Foci of calcification are also present. The histopathological findings are in keeping with a diagnosis of a

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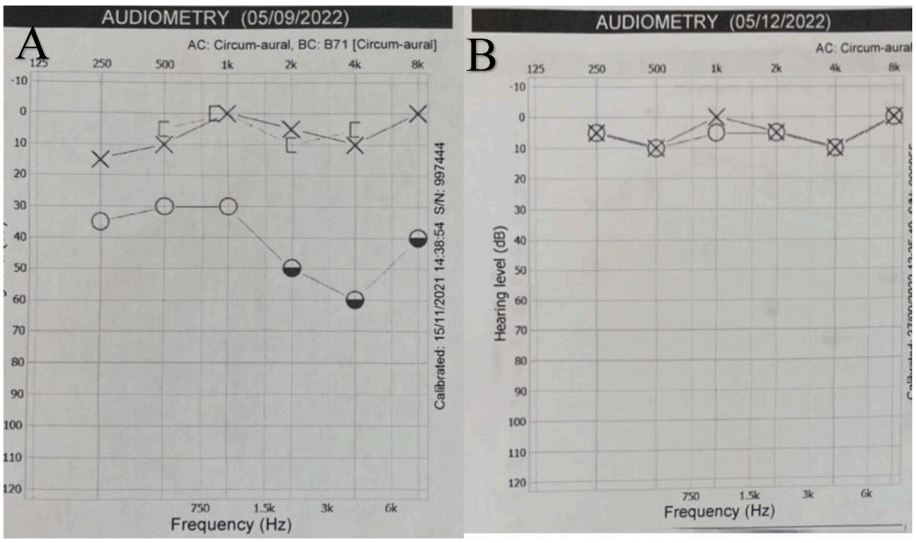

Fig. 1. Figure A: pre-operative pure tone audiometry demonstrating right conductive hearing loss Figure B: post-operative pure tone audiometry demonstrating normal hearing.

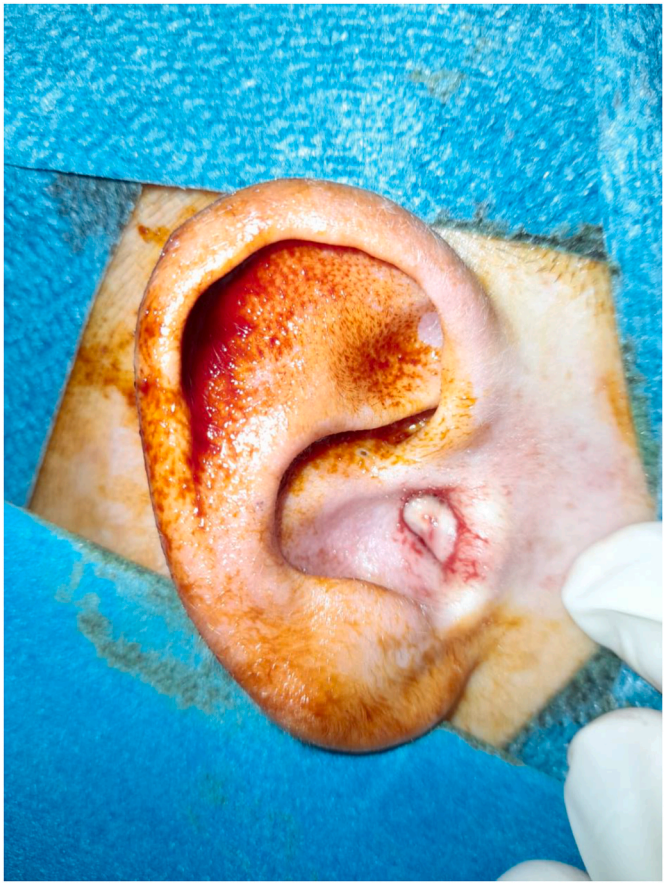

Fig. 2. Right auricle lesion completely occluding the external auditory canal.

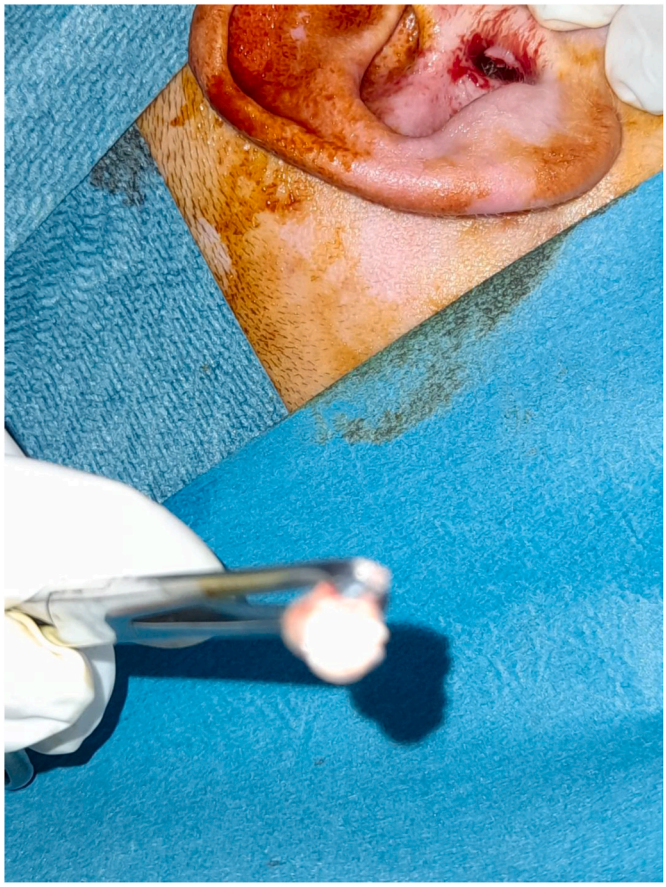

Fig. 3. Post excision of lesion.

Pilomatricoma.

The patient returned one week post-operatively and his sutures and dressing were removed, with a subjective improvement of his hearing.

3. Discussion

Pilomatricoma has been reported to account for almost 1% of all benign cutaneous lesions, with the pre-auricular area the most common site [4]. They tend to occur in the paediatric population with incidence decreasing in adolescence. There is female predominance, with a female to male ratio of 1.65:1 [4].

Clinical diagnoses of pilomatricomas can be challenging, with multiple other pathologies to be considered. While they are frequently seen by dermatologists, otolaryngologists often don't consider

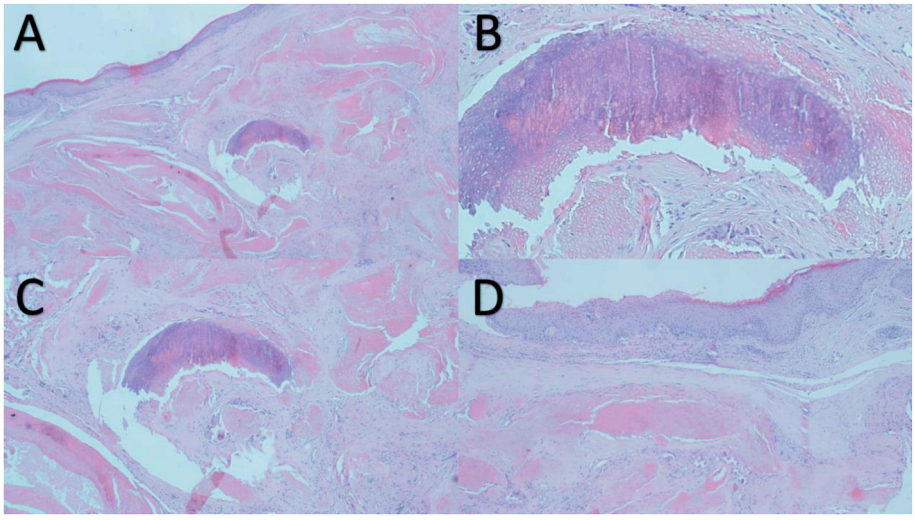

Fig. 4. Figure A: H & E 5 X magnification. This image illustrates the overlying squamous epithelium of the auditory canal with the underlying dermal based neoplasm. Figure D: H & E 10 X magnification. This image illustrates the overlying squamous epithelium of the auditory canal with the underlying dermal based neoplasm. Figure C: H & E 20 X magnification and Figure D: H & E 40 X magnification illustrate dermal based neoplasm composed predominantly of ghost/shadow cells.

pilomatricomas in their differential diagnosis. They generally present as firm, mobile, non-tender, slow growing lesions. Clinical signs can be used to elicit a diagnosis, in particular the "tent sign" (a multifaceted appearance is evident when the skin over the lesion is stretched) and the "teeter-totter" (skin bulging when the opposite end is palpated). A 2016 systematic review on the management of suspected pilomatricoma designed a management algorithm that recommended that lesions with a typical history and features should undergo surgical excision without pre-operative imaging [4]. If there is any doubt, an ultrasound can be performed, but this would not have been possible in this case due to its location. Pre-operative tissue sampling has not been shown to be beneficial.

Pilomatricomas are most commonly encountered benign lesions in the head and neck region, presenting as asymptomatically [4]. There are only four previously reported cases of external auditory canal pilomatricomas, however unlike our case, the canal was not completely occluded in the previously documented cases. Interestingly the extent of occlusion in our case was significant enough to cause conductive hearing loss. As such, excisional biopsy in our case was mandated for both symptomatic improvement and histopathological diagnosis.

There have been associations between pilomatricomas and genetic conditions, most notably Gardner syndrome, myotonic dystrophy and Turner's syndrome. Currently there are no recommendations for genetic screening for patients with pilomatricomas.

To the authors knowledge there have been no cases of spontaneous regression, with surgical excision the only treatment option. Post operative recurrence is rare, but recurrence can occur if incompletely excised. More aggressive subtypes with perineural and vascular invasion have been described and there have been cases of pilomatrix carcinoma which can occur in incompletely excised pilomatricomas [6–8]. Activating mutations of the Wnt signalling pathway, in the form of beta-catenin mutations in pilomatricomas of younger patients have been thought to be responsible for the malignant transformation over time to pilomatrix carcinoma [8,9]. Pilomatrix carcinoma has rarely been reported and histological criteria for diagnosis are not well defined. They present similarly, as a firm, non-tender lesion in the head and neck region. In contrast, they are locally invasive and have a tendency for local recurrence [8,9]. They are more common in adult males, and have not been reported in children or adolescents [10]. Surgical excision with wide margins is the mainstay of management, but in cases of recurrent or metastatic disease adjuvant chemotherapy or radiotherapy may be required.

This case report highlights the rare finding of an occlusive external auditory canal pilomatricoma, and re-enforces the importance of a broad differential when faced with lesions of the external auditory canal. Excisional biopsy provides both diagnoses and treatment for these lesions.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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