Pilomatrixoma of the Ear Helix

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Pilomatrixoma is a rare benign tumor originating in the hair follicles and usually appears in the head and neck region, trunk and extremities. Several cases of pilomatrixoma in the pre-auricular area and ear lobe have been reported. We describe a patient with pilomatrixoma of the ear helix; to the best of our knowledge this is the first report of this tumor presenting in this location.

A 72 year old man presented with a tender, rapidly growing mass in the helix of his right ear. This was not responsive to topical treatment and he was referred to our care. There was no bleeding from this site, nor was there a history of other skin tumors. His history was positive for diabetes and benign prostate hypertrophy. Physical examination of the right helix demonstrated a 1.5 cm nodular mass without the appearance of overlying skin adherence although the skin was slightly erythematous [Figure 1A]. The rest of the physical examination was normal. The lesion was removed under local anesthesia with primary closure and the postoperative course was uneventful [Figure 1B and C].

Pathological examination revealed a 2.8 x 1 x 0.6 cm skin fragment. On the surface there was a dome-shaped tan-brown lesion 0.5 cm in diameter. On the cut surface, in the lower dermis, a pinkish, sharply demarcated nodular mass 0.8 cm in diameter was present.

Microscopic examination [Figure 2A] demonstrated a tumor consisting of two epithelial cell types – basophilic cells and eosinophilic shadow cells consistent with pilomatrixoma. The basophilic cells were located mostly at the periphery of the cell islands. They had little cytoplasm, indistinct cell borders, hyperchromatic nuclei and rare mitoses [Figure 2B]. The eosinophilic shadow (mummified) cells were found toward the center of the cell masses. They had more cytoplasm, distinct cell borders, and lacked nuclear staining [Figure 2B]. In addition, small calcifications and moderate chronic inflammation with focal foreign-body giant cell reaction [Figure 2C] were seen. Inked surgical resection margins were free.

Comment

Pilomatrixoma, also known as pilomatricoma, trichomatricoma or calcifying epithelioma of Malherbe, was first described

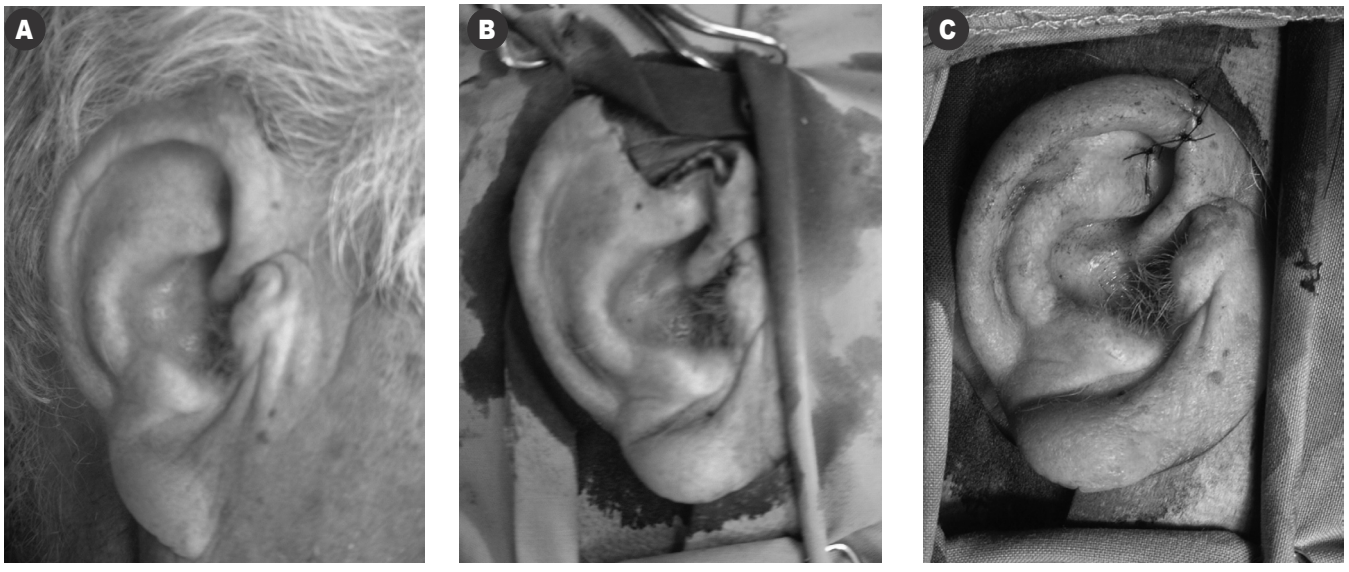

Figure 1. [A] Macroscopic findings of a 2.5 cm mass in the patient's helix. **[B]** Lesion resected. **[C]** Wound closure.

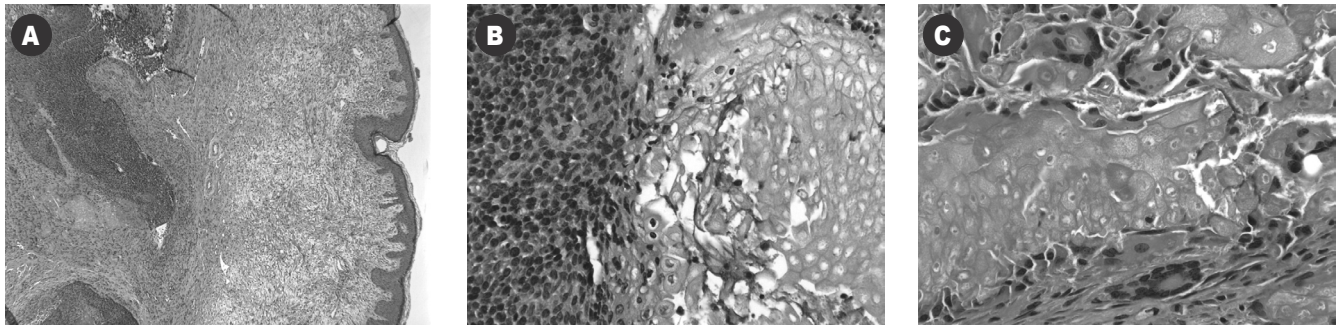

Figure 2. [A] Skin with intradermal tumoral mass (hematoxylin & eosin x 40). **[B]** Two epithelial cell types – basal cells (left) and shadow (mummified) cells (right) (H&E x 200). **[C]** Focal foreign-body giant cell reaction (H&E).

by Malherbe and Chenantais in 1980 [3]. This rare benign neoplasm is slightly more common in females and usually appears in children, so that 60% of cases are reported in the first two decades of life [1,2]. The etiology of pilomatrixoma is not completely understood, but there are suggestions that an activating mutation in the β-catenin gene mapped to chromosome 3 p22-21.3 plays a major role in this tumor genesis [2,4]. Although pilomatrixoma usually appears sporadically, some association was found between this tumor in the pediatric population and disorders such as myotonic dystrophy, Rubinstein-Taybi syndrome, Turner syndrome, Gardner syndrome, xeroderma pigmentosum and basal cell nevus syndrome [2].

This tumor is usually palpated as a firm, solitary dermal or subcutaneous nodule most commonly in the head and neck region or in the upper extremities [1,2]. Pilomatrixoma is usually a slow-growing tumor, but it may begin to grow rapidly as a result of inflammation or hemorrhage and, if left untreated, may also invade the epidermis [4]. A high index of suspicion is important since studies have shown that a correct preoperative diagnosis was made in only 28.9–43% of cases [3]. When the presentation is classic, diagnosis may be made by clinical findings. Fine-needle aspiration is not recommended for the diagnosis of this lesion, and in atypical cases diagnosis should be made by biopsy [5]. The treatment of choice for this condition is surgical excision, and recurrence is rare [1].

References

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