Pilomatrixoma

23

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Introduction

Pilomatrixoma (pilomatricoma) is a benign skin neoplasm arising from the matrix cells at the base of the hair. It was originally described as calcified epithelioma of sebaceous glands, but later the term pilomatrixoma was suggested to denote its origin. Some authors have reported a bimodal peak during the first and sixth decades of life, with a male:female ratio of 2:3. The most frequent location is the head and neck region (>50% of cases). Involvement of the face has been reported in frontal, temporal, cheek, periorbital and preauricular areas. It must be considered in the differential diagnosis of hard masses involving the facial region.

Clinical Scenario

A 46-year-old male with no significant past medical history presented with a left eyebrow lump of 1 month's duration. The lump was progressively enlarging and associated with mild discomfort and redness, but did not cause any discharge or visual symptoms. He denied any previous history of trauma.

On examination, there was a left eyebrow mass measuring 2 × 2 cm (Fig. 23.1). It was mobile, fixed to the skin but not to underlying bone. It was pointing superiorly, but had no evidence of preseptal cellulitis. The rest of the ophthalmic examination did not reveal any abnormalities (Table 23.1).

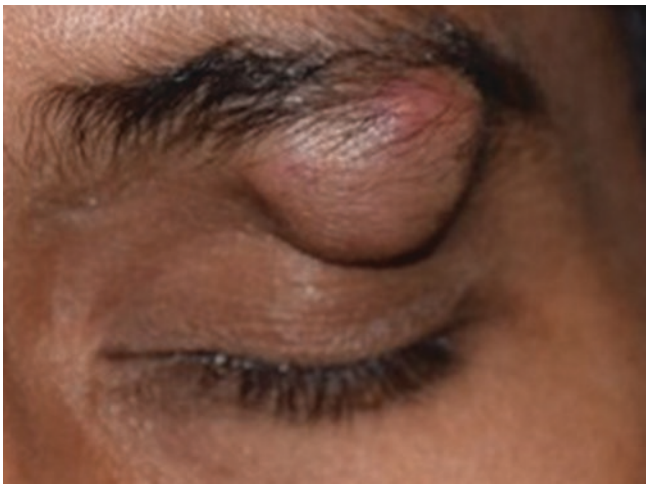

Fig. 23.1 Left sub-brow nodule, attached to overlying skin but mobile and not fixed to underlying bone

Table 23.1 CLOSE summary

*C*linical process: progressively enlarging lump
*L*ocation: sub-brow skin
*O*nset: subacute
*S*igns and symptoms: mass lesion
*E*pidemiology: adult Malay male

Differential Diagnosis

- Sebaceous cyst
- Dermal adnexal tumours

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- Sweat gland tumours: Syringoma, eccrine hidradenoma, eccrine hidrocystoma and apocrine hidrocystoma
- Hair follicle tumours: Trichoepithelioma, trichilemmoma and pilomatrixoma (pilomatricoma)

Intervention

The patient underwent excision biopsy of the cyst (Fig. 23.2).

Histopathology

There was a well circumscribed dermal nodule lined by squamous and basaloid cells with round to oval nuclei, occasional nucleoli and scant cytoplasm. Areas of the lining epithelium appeared to transit to masses of eosinophilic material containing ghost cells within the cyst. The ghost cells showed holes where nuclei once were and had more eosinophilic cytoplasm. The features are consistent with a pilomatricoma (Fig. 23.3).

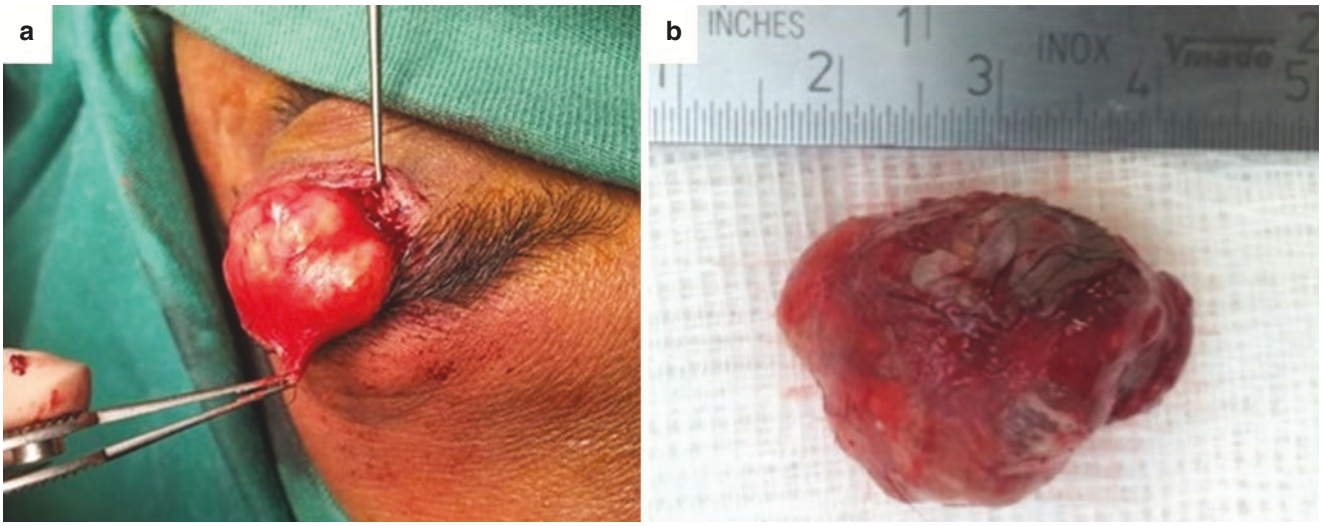

Fig. 23.2 (**a**) Excision biopsy of sub-brow lump performed through an elliptical skin excision, (**b**) well-encapsulated dermal tumour measuring approximately 2 cm in diameter

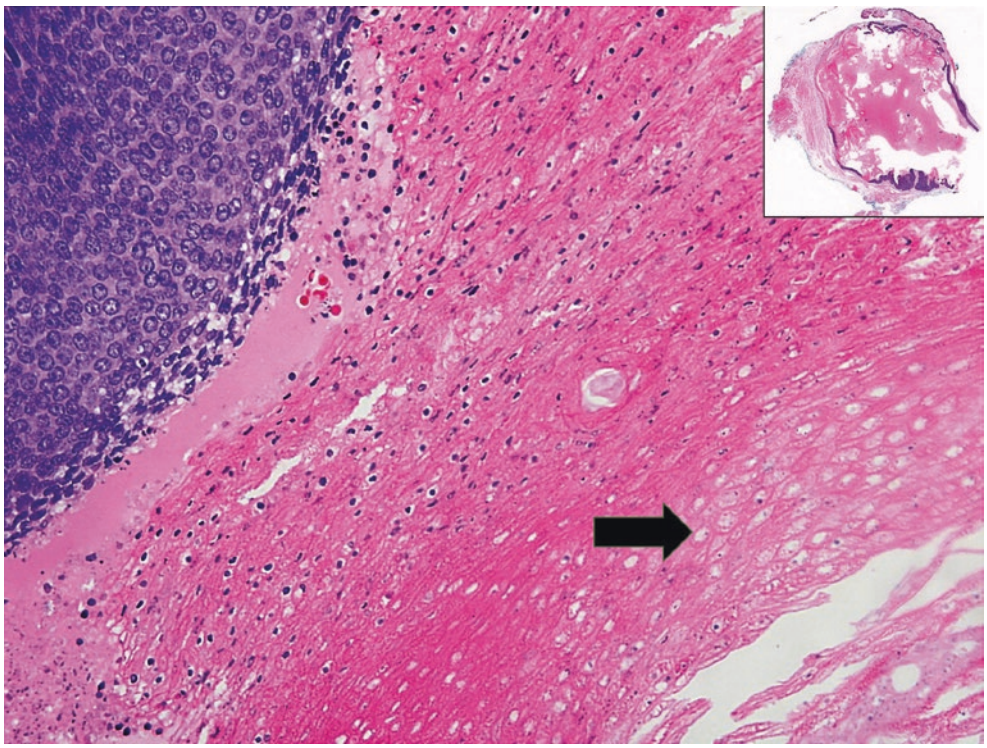

Fig. 23.3 Basaloid cells (top left corner) transitions into eosinophilic material containing "ghost cells". Some empty holes where nuclei used to be are indicated by the black arrow HE stain, 200× magnification. Inset shows a circumscribed nodule in the dermis. HE stain, 1× magnification

Discussion

The most common site for pilomatrixomas is in the head and face. In the periorbital region, pilomatrixoma usually involves the eyebrows, possibly because of high density of hair follicles. It usually presents as a solitary, slow-growing, superficial, mobile, hard mass ranging in size from 0.5 to 5 cm in diameter. It is often asymptomatic, and the overlying skin may exhibit a bluish-red discoloration or ulceration. Multiple occurrences and familial disease are associated with Gardner syndrome, sarcoidosis, and Turner syndrome.

The clinical diagnosis of a pilomatrixoma is usually difficult, and they have variably been diagnosed preoperatively as eyelid cysts (mainly sebaceous and dermoid cysts) and rarely as eyelid tumours such as papilloma, keratoacanthoma, sebaceous cell carcinoma and basal cell carcinoma. Histopathologic diagnosis is based on the presence of basophilic, as well as shadow cells, which may be associated with foreign-body giant cells, calcification, and ossification. The malignant counterpart (pilomatrix carcinoma) is extremely rare, and characterized by actively proliferating, hyperchromatic, pleomorphic basaloid cells, atypical mitosis, tumour necrosis, architectural asymmetry, and infiltrative growth pattern.

Treatment

Surgical excision is the treatment of choice. The mass is usually well encapsulated. If the margins are ill-defined or adherent to the surrounding tissues, malignancy can be suspected. Cytological sampling errors with fine-needle aspiration cytology (FNAC) may lead to misdiagnosis, especially if there are no ghost cells in the aspirate.

Learning Points

Pilomatrixoma is an uncommon, benign, cutaneous tumour with a characteristic histological appearance. Preoperative diagnosis is difficult, and thus it must be considered in the differential diagnosis of hard masses involving the facial region.

Further Reading

1. Forbis R Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch Dermatol. 1961;83:606–18.
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3. Levy J, Ilsar M, Deckel Y, Maly A, Anteby I, Pe'er J. Eyelid pilomatrixoma: a description of 16 cases and a review of the literature. Surv Ophthalmol. 2008;53:526–35.