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Pilomatrixoma of the eyelid

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Abstract

Background: Pilomatrixoma is a benign tumor of the hair follicle that can transform into a malignant lesion, the pilomatrix carcinoma. We present an unusual case of an eyelid pilomatrixoma.

Main observation: A 37-year-old white male presented with rapidly growing, pedunculated lesion located at the superior right eyelid of five months duration. The lesion was excised under local anaesthesia. The sample was fixed in 10% formalin and histopathological as well as immunohistochemical analyses were performed. Results of both examinations were consistent with a benign pilomatrixoma.

Conclusions: Even though the lesion had malignant clinical appearance, histopathology confirmed the diagnosis of a benign pilomatrixoma, supporting the decision not to make a more extensive surgery.

Introduction

Pilomatrixoma (pilomatricoma), a benign tumor of the hair follicle, was first described by Malherb in 1880 as "calcifying epithelioma" and was thought to be derived from the sebaceous gland.[1] Later on, in 1949 Lever and Griesemer suggested the hair matrix cells to be the origin of the tumor[2], and finally in 1961 Forbis and Helwig proposed the currently accepted name "pilomatrixoma".[3]

Clinically, the lesion appears as a slowly enlarging, irregularly shaped, rock hard, nodular, non tender mass freely movable over the subcutaneous tissue, but not from the overlying skin. The skin usually has reddish to blue discoloration due to dilated blood vessels and chalky white nodules may be seen through the skin. There is typically no history of inflammation or trauma.[4,5,6]

Pilomatrixoma needs to be differentiated from the malignant form of this tumour, pilomatrix carcinoma, that occurs more often in middle age or older individuals, more commonly in men than in women. Microscopically it is characterized by exuberant proliferation of basaloid cell masses arranged haphazardly throughout the tumor, varying degrees of cytological atypia, frequent mitoses, and areas of necrosis. The keratinisation with shadow cell formation is less extensive than in benign pilomatrixoma. The infiltrative growth pattern which extends into deeper soft tissues may display vascular and perineural invasion.[7]

In this report, we describe an unusual appearance of an eyelid pilomatrixoma.

Case Report

A 37-year-old white male presented with a rapidly growing lesion located at the superior right eyelid of five months duration. The history of the patient revealed no pain, nor previous inflammation or trauma. Clinically there was a pedunculated, not painful, rock hard, nodular mass, not movable over the subcutaneous tissue or the overlying skin. It had normal skin color, but chalky white nodules could be seen through the healthy appearing skin (Fig.1). The lesion was circumscribed by a fibrotic, continuous, capsule with uneven thickness. The lesion was excised under local anaesthesia. The tarsus, presenting

with an unusual yellowish appearance, was cauterised, but left in place, to avoid an extensive surgery.

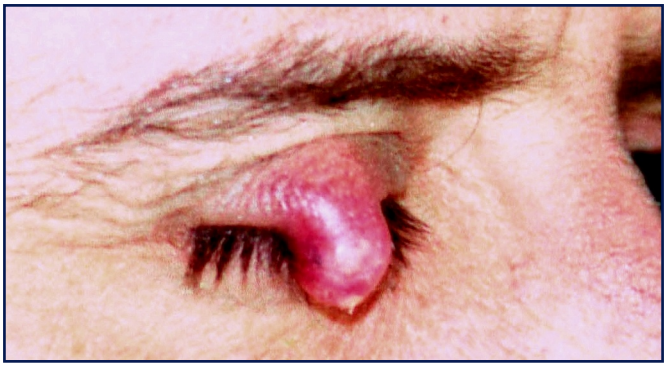

Figure 1
Pilomatrixoma of the eyelid.

The sample was fixed in 10% formalin in phosphate buffer and after dehydration and diaphanization embedded in histowax resin.

The internal structure was characterized by tissue islets and girders fold on themselves, the whole wrapped by a delicate loose fibrillar stroma; these consist of laid layers of basophilic cells located in the peripheral part of the lesion (epithelium basaloid), and in the central area of nucleus free shadow cell with diaphanous cytoplasm. Hemorrhagic sites, small calcium deposits and necrotic areas were also present (Fig.2). Inflammatory limfo-histiocitary infiltrates were prominent in the capsule, in the interepithelial spaces and in the free central areas.

Micron sections were prepared for immunohistochemical analysis (peroxidase-antiperoxidase technique of Sternberger). These sections were investigated with the use of mouse monoclonal antibodies (Mab) against human p53 protein, p27Kip1, Ki67-MIB (DAKO) to detect molecular expression of proliferative processes. Staining for p53 protein was negative, P27Kip1 was absent, Ki67-MIB was microfocal.

Discussion

To the best of our knowledge only one case of pedunculated benign pilomatrixoma has been described. The clinical appearance of this patients was quite unusual and the adherence to the subcutaneous tissue might have suggested a malignant lesion. Pilomatrixoma clinically appears usually as a slowly enlarging, irregularly shaped, rock hard, nodular, non tender mass freely movable over the subcutaneous tissue, but not from the overlying skin, the skin can have reddish to blue discoloration due to dilated blood vessels and chalky white nodules could be seen through the skin and no history of inflammation or trauma.[8]

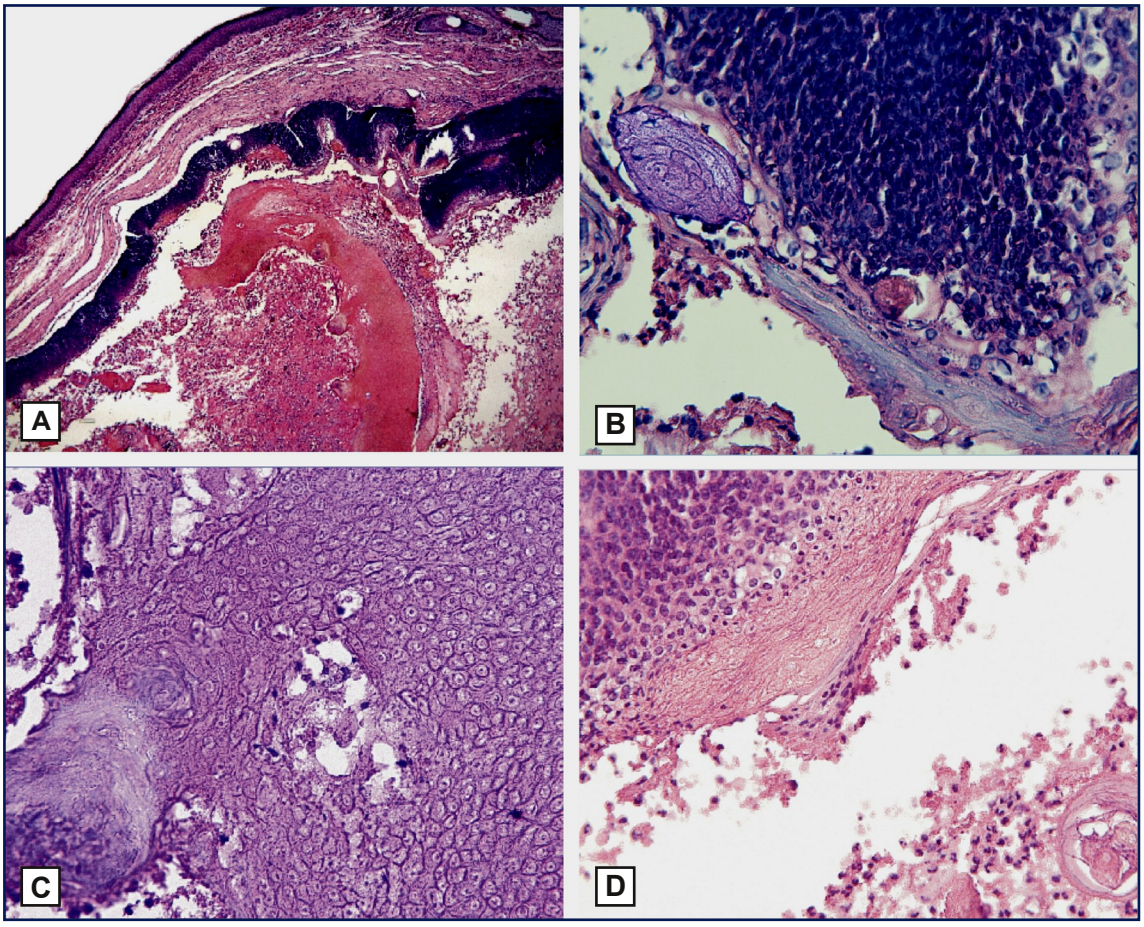

Figure 2
*Pilomatrixoma histopathology: **A)** cystic neoformation with a connectivous capsule. Internal surfaces are covered by a large band of epithelial cells (magnification 40X), **B)** epithelial layers, very close to the capsule, are made of very small cells similar to basal cells (magnification 400X) **C)** inner epithelial layers are made by plane cells with eosinophil nucleus and picnotic cytoplasm (magnification 400X) **D)** The central area of cystic cavity is full of cellular debris and not colourable elements without nucleous, some with a tendency to aggregate (magnification 100X).*

The lids can be affected by a wide variety of different benign and malignant lesions. Benign lesions are only three times more frequent than the malignant tumors.[9] Among lesions that need to be differentiate from a pilomatrixoma, there are: epidermal inclusion cyst, sebaceous adenoma, dermoid cyst, epidermoid cyst and vascular tumors.

In some cases, however, they need to be differentiated from other malignant lesions such as basal cell carcinoma, sebaceous gland carcinoma, Kaposi sarcoma. These tumors may have similar clinical appearance, making histopatological and immunohistochemical analyses mandatory.

Our immunohistochemical findings excluded the presence of malignancy. The epithelium was negative for P53 protein accumulation, that occurs in tumors and may be a neoplastic prognostic marker. The p27 (Kip 1) protein is a cyclin-dependent kinase (cdk) inhibitor of transition from G1 to S phase. The clinical significance of p27 (Kip 1) is not obvious and is currently investigated, as the loss of the negative cell-cycle regulator may contribute to oncogenesis. In spite of being a benign neoplasm, pilomatrixoma had a low p27 expression. This may be a reflection of the proliferative potential of the hair matrix. The Ki67- MIB is a protein nuclear expressed during all active phases of the cell cycle. The reaction serves as an indicator of the cellular growth fraction. The expression of Ki67-MIB was microfocal.

The benign nature of the lesion is further supported by the fact that after 1 year no recurrence of the tumor was present, and validate our decision not to make a more extensive surgery.

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