

# Pilomatrix carcinoma: a rare cutaneous adnexal tumor

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## Abstract

Pilomatrix carcinoma is a rare tumor that is generally not diagnosed clinically. An 80-year-old man presented with a 5-month history of rapidly growing nodule of the submandibular area. Histological examination revealed a pilomatrix carcinoma, an aggressive malignancy with metastatic potential.

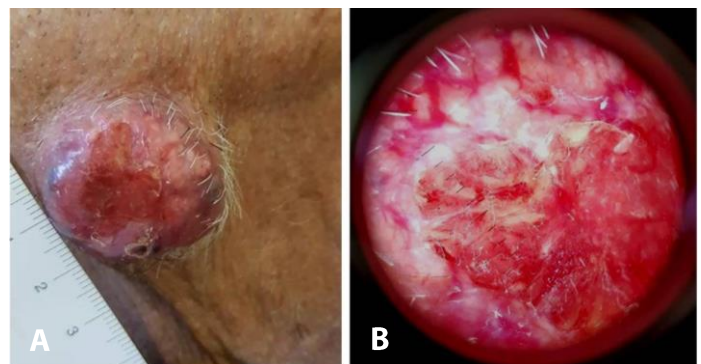
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## Introduction

Pilomatrix carcinoma is a rare cutaneous malignant tumor of hair matrix cell origin [1]. There are approximately 125 cases reported in English literature to date [2]. Owing to its rarity, it is not usually considered in differential diagnosis of skin tumors and it is misdiagnosed preoperatively. We present here a case of this very rare tumor.

## Case Synopsis

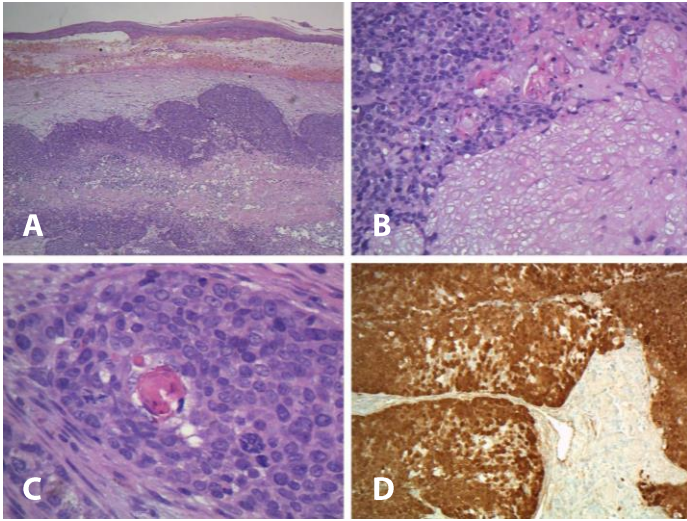
An 80-year-old man presented to the dermatology clinic with a 5-month history of a rapidly growing ulcerated nodule located on the left submandibular region. Physical examination revealed a firm, reddish-violaceous, ulcerated nodule of 3.5cm×3.0cm, which was not adherent to deep planes (**Figure 1A**). Dermoscopy revealed telangiectasias, white structureless areas, and yellowish hues on an erythematous background (**Figure 1B**).



**Figure 1. A)** Nodule on the left submandibular region. **B)** Dermoscopy of the lesion.

The nodule was excised with a wide margin and histopathology examination demonstrated a prevalent dermal proliferation with an infiltrative growth pattern of basaloid cells (**Figure 2A**). Foci of necrosis (**Figure 2A**), increased mitotic rates, atypical mitosis (**Figure 2C**), and enlarged anucleated cells with eosinophilic cytoplasm (**Figure 2B**) were also observed. On immunohistochemistry, the beta-catenin immunoreactivity was diffusely positive (**Figure 2D**) and the cells were negative for cytokeratin 20, cytokeratin 7, neuron specific enolase, synaptophysin, and chromogranin A. Histopathological findings led us to the diagnosis of pilomatrix carcinoma.

Pilomatrix carcinoma is a rare malignant neoplasm that originates from hair matrix cells. It was first described in 1980 [1] and approximately 125 cases have been reported since then [2]. This neoplasm most occurs in male patients from the fifth-to-eighth decades and it is commonly located on the head or neck, although it has been described on upper and lower extremities, trunk, and genital region [1,2]. Pilomatrix carcinoma can develop de novo or can



**Figure 2.** **A)** Histopathology shows derma-based proliferation of basaloid cells with central necrosis. H&E, 10x. **B)** Ghost-cells. H&E, 40x. **C)** Increased mitotic rates with atypical mitosis. H&E, 40x. **D)** Beta-catenin immunostain, 10x.

arise through a malignant transformation of a previous longstanding pilomatrixoma [1,2]; in our case the lesion was very recent and rapidly growing, thus supporting a de novo presentation.

Histologically, pilomatrix carcinoma is characterized by a dermal proliferation of basaloid cells with infiltrative growth pattern, and foci of central necrosis. Basaloid cells usually show atypia, pleomorphism, nuclear hyperchromatism, and raised mitotic index with numerous atypical mitoses.

## References

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Another abundant population is represented by enlarged anucleate epithelial cells with eosinophilic cytoplasm, named “ghost cells,” which are otherwise expressed by pilomatrixoma, craniopharyngioma, and odontogenic tumors [3], which must be considered in the differential diagnosis. Currently, pilomatrix carcinoma is believed to arise from a mutation in the WNT signaling pathway which is involved in cell adhesion, differentiation, and proliferation [2]. Similarly to pilomatrixoma, immunohistochemistry shows a hyperexpression of beta-catenin, a downstream effector in the WNT-pathway, owing to mutations in the *CTNNB1* gene [2].

The tumor is locally aggressive with a tendency to recur and metastasizes in about 10% of cases, mainly into regional lymph nodes and lungs; regular patient follow-up is advisable [4]. Owing to its rarity there are no well-defined standards for the surgical management and patient follow-up. In our case a wide margin excision was curative and no signs of local recurrence and metastasis have been identified after 5 years of six-monthly clinical and radiological follow-up.

## Potential conflicts of interest

The authors declare no conflicts of interest.