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Authors
Dell'Antonia, Massimo
Ferreli, Caterina
Pilloni, Luca
et al.

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Pilomatrix carcinoma: a rare cutaneous adnexal tumor

Massimo Dell'Antonia[1] MD; Caterina Ferreli[1] MD; Luca Pilloni[2] MD; Laura Atzori[1] MD

Affiliations: [1]Dermatology Clinic, Department of Medical Sciences and Public Health, University of Cagliari, Cagliari, Italy, [2]Pathology Service, Department of Medical Sciences and Public Health, University of Cagliari, Cagliari, Italy

Corresponding Author: Dell'Antonia Massimo, Dermatology Clinic, Department of Medical Sciences and Public Health, University of Cagliari, Via Ospedale 54, 09124, Cagliari, Italy. Tel: 39-3462134028. Email: massimodellantonia@gmail.com

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.

Keywords: adnexal, carcinoma, pilomatricoma, pilomatrix, tumor

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**).

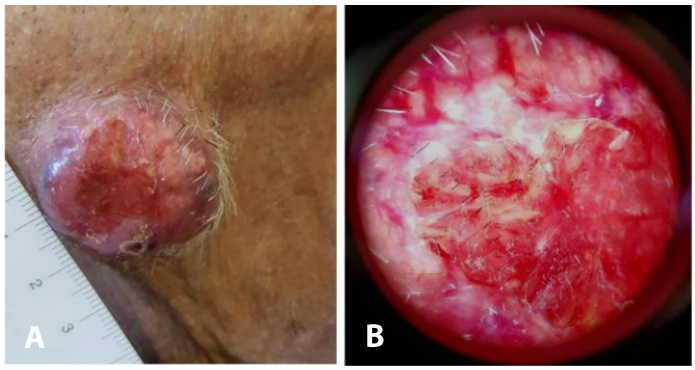

Figure 1. ***A)*** *Nodule on the left submandibular region.* ***B)*** *Dermoscopy of the lesion.*

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

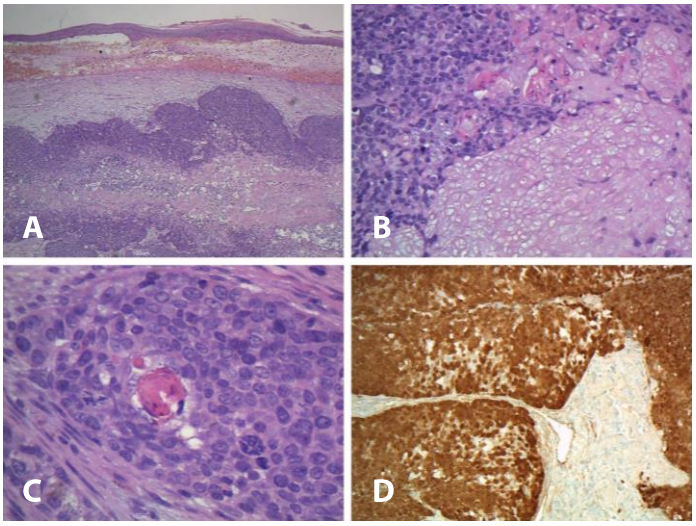

Figure 2. ***A)*** *Histopathology shows derma-based proliferation of basaloid cells with central necrosis. H&E, 10×.* ***B)*** *Ghost-cells. H&E, 40×.* ***C)*** *Increased mitotic rates with atypical mitosis. H&E, 40×.* ***D)*** *Beta-catenin immunostain, 10×.*

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. 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.

References

1. Alcántara-González J, Sánchez-Largo ME, Caminoa A, Erana I, Calzado-Villarreal L. Pilomatrix carcinoma: a rare cause of facial tumor. *Dermatol Online J.* 2014;20(7). [PMID: 25046461].
2. Jones C, Twoon M, Ho W, Portelli M, Robertson BF, Anderson W. Pilomatrix carcinoma: 12-year experience and review of the literature. *J Cutan Pathol.* 2018;45:33-38. [PMID: 28914451].
3. Rajesh E, Jimson S, Masthan KM, Balachander N. Ghost cell lesions. *J Pharm Bioallied Sci.* 2015;7(Suppl 1):S142–144. [PMID: 26015694].
4. Herrmann JL, Allan A, Trapp KM, Morgan MB. Pilomatrix carcinoma: 13 new cases and review of the literature with emphasis on predictors of metastasis. *J Am Acad Dermatol.* 2014;71:38-43. [PMID: 24739254].