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Case Presentation

Pilomatrix carcinoma: a rare cause of facial tumor

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Abstract

Pilomatrix carcinoma is a rare malignant tumor that originates from hair matrix cells. It is not usually considered in a differential diagnosis owing to its low incidence. We present a case of this uncommon entity and review the literature.

Keywords: pilomatrix carcinoma, calcifying epitheliocarcinoma of Malherbe, pilomatrixoma.

Introduction

Pilomatrix carcinoma is a rare malignant tumor that originates from hair matrix cells. It is not usually considered in a differential diagnosis owing to its low incidence. We present a case of this uncommon entity and review the literature.

Case synopsis

An 87-year-old man with no relevant previous history was referred to our clinic. He presented with a 1-month history of a rapidly growing tumor located on the right preauricular area and the root of his right helix. There was no history of preceding trauma or previous lesion.

Physical examination revealed a firm, ulcerated tumor of 1.5x1.3 cm, which was not adherent to deep planes (Figure 1). No local or regional lymphadenopathy was detected.

A biopsy was performed, and the histological sections showed a neoplastic proliferation of epithelial character and an infiltrative growth pattern consisting of intermediate size basaloid cells with scant cytoplasm and hyperchromatic nuclei (Figure 2). Ghost cells were seen in the center of the nests; multinucleated giant cells, dystrophic calcifications, and melanophages were also observed. This epithelial proliferation showed positivity for β-catenin (Figure 3), p63, and CD10; a focal positivity for EMA and BerEP4 was also seen. HMB45, Melan A, and S100 stains were negative.

A full-body computed tomography was performed. Neither infiltration of underlying structures nor lymphadenopathy, nor metastases were detected.

A diagnosis of pilomatrix carcinoma was established.

Wide surgical excisión and closure with a transposition flap was perfomed. No evidence of recurrence has been noted at 12 months follow-up.

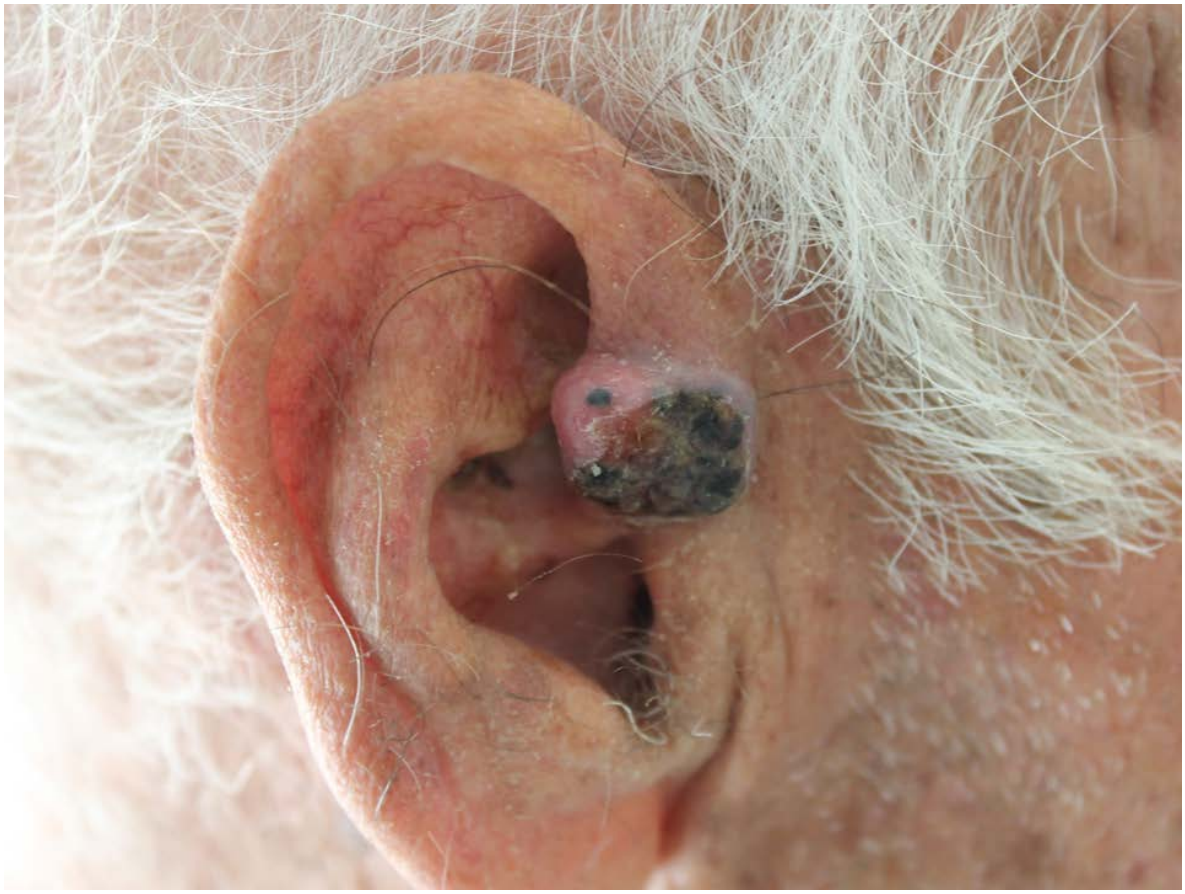

Figure 1. Ulcerated tumor located on the right preauricular area and the root of the right helix

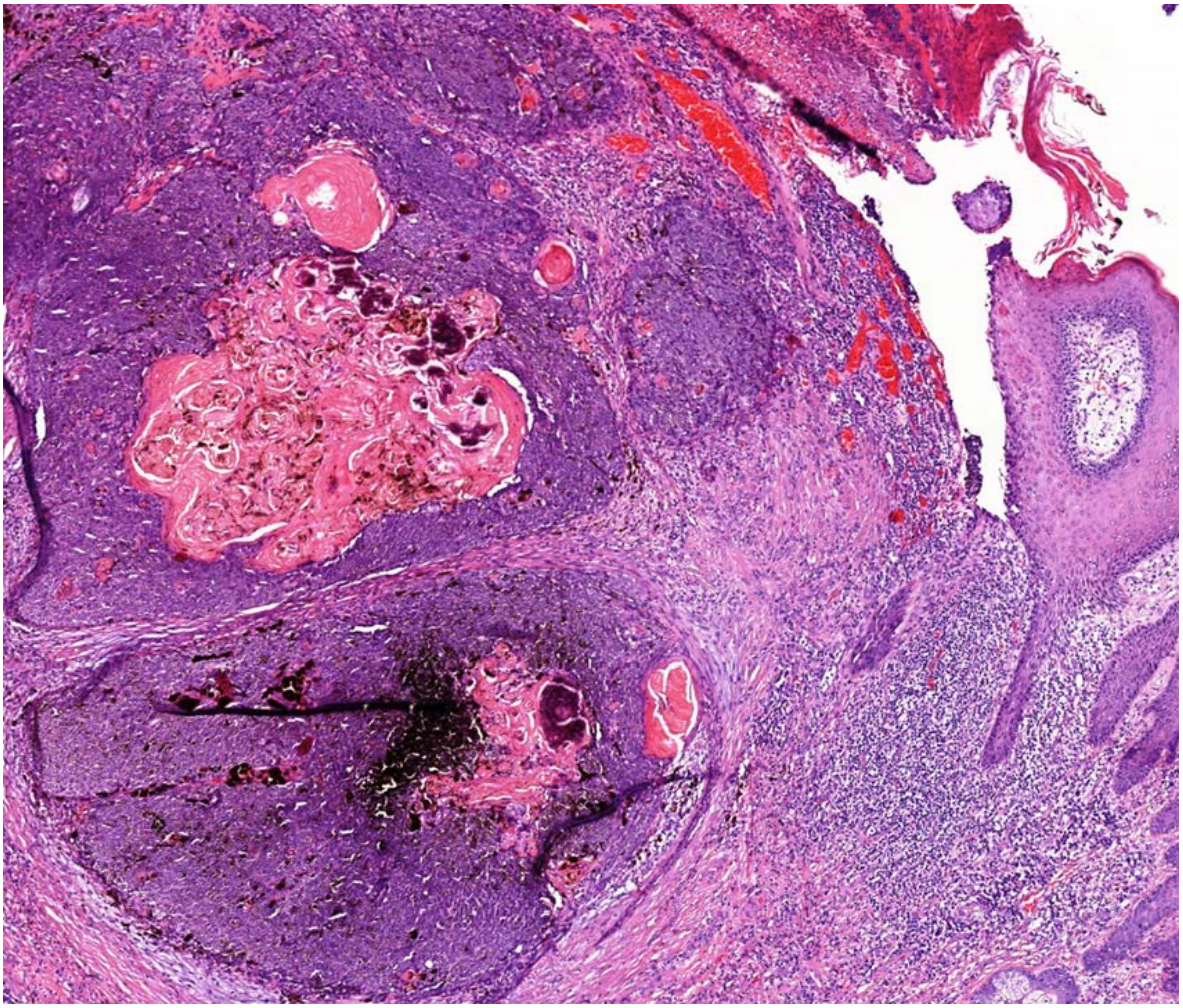

Figure 2. Neoplastic proliferation of epithelial character and infiltrative growth pattern consisting of intermediate size basaloid cells and "ghost cells" located in the center of the nests: Multinucleated giant cells, dystrophic calcifications and melanophages can also be observed.

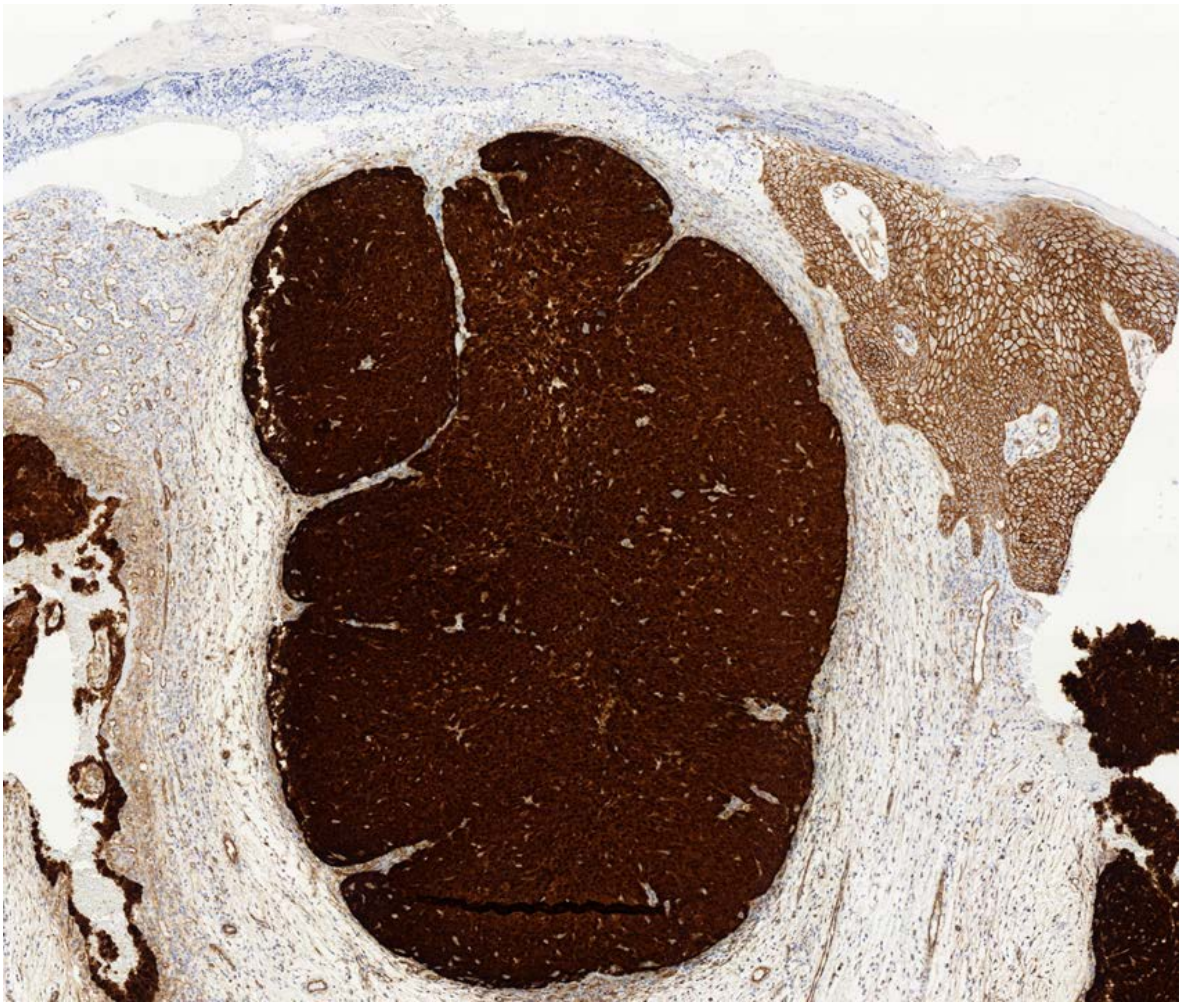

Figure 3. β-catenin positivity

Discussion

Pilomatrix carcinoma or calcifying epitheliocarcinoma of Malherbe is a malignant neoplasm that originates from hair matrix cells. It was first defined in 1980 and has been considered as the malignant variant of pilomatrixoma [1]. In contrast to pilomatricoma, which is more frequent in females during the first two decades of life, pilomatrix carcioma is most commonly found in men from the fifth to the seventh decades [2,3]. Pilomatrix carcinoma tends to appear on head or neck, although it has also been described on trunk, upper and lower extremities, and genital regions [2].

It is uncertain whether pilomatrix carcinoma develops de novo or if it is a malignant transformation of an existing pilomatrixoma. Some cases of pilomatrix carcinomas have been reported to develop from histologically confirmed pilomatrixomas [4]. It is also possible that both hypotheses are valid.

Similarly to pilomatrixoma, activating mutations in exón 3 of the CTNNB1 gene, which encodes β-catenin, are detected. The presence of a common mutation implies a common initial pathogenesis [5].

Histologically, pilomatrix carcinoma has eosinophilic ghost cells and basaloid matrical cells at the periphery. Melanophages, keratinization, and calcium deposits are frequently seen. Its distinction from pilomatrixoma can be complicated and there are currently no reliable immunohistochemical markers that allow their differentiation. Asymmetry, great size, poor circumscription, extensive areas of necrosis, ulceration, infiltrative growth pattern, marked anaplasia of the tumor cells, frequent atypical mitoses, prominent desmoplastic stromal reaction surrounding the tumor nests, and vascular, lymphatic, or perineural invasion help to their distinction [2,6]. Proliferating pilomatrixoma, matricoma, and basal cell carcinoma with matrical differentiation should also be considered in the differential diagnosis.

Wide surgical excision with confirmed negative margins remains the treatment of choice. However local recurrences are not uncommon (20.5%). If simple excision is performed recurrence rate can be up to 64.1% [2]. Therefore, some authors have used Mohs micrographic surgery to achieve control of tumor margins with promising results [2]. Radiotherapy has been proven useful in cases in which surgery is not feasible or as adjuvant therapy. In contrast, chemotherapy has been used without success.

The risk of metastases has been reported in approximately 16% of cases; regional lymph nodes, lung, bone, and central nervous system are the most frequently affected locations [2].

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