BRIEF ARTICLES

Pilomatrical Carcinoma in a 70-Year-Old Hispanic Female

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ABSTRACT

Pilomatrical carcinoma is a rare, locally aggressive malignancy of follicular matrix origin. It presents as a rapidly growing, flesh-colored, or blueish, exophytic nodule with tumor asymmetry and poor circumscription. Histological features of these tumors include dominant hyperchromatic basaloid cells with high mitotic rate and nuclear pleomorphism; along with anucleate matrical cells and central necrosis. Pilomatrical carcinomas demonstrate a high rate of recurrence and tendency to metastasize. The most described treatment is wide excision of the tumor with negative margins with or without adjuvant radiation therapy. We present the case of a 70-year-old female patient who presented to the office with a 3-year history of a growing, exophytic lesion on the right nasal ala. A biopsy of the lesion was obtained, and histologic examination was consistent with a diagnosis of pilomatrical carcinoma.

INTRODUCTION

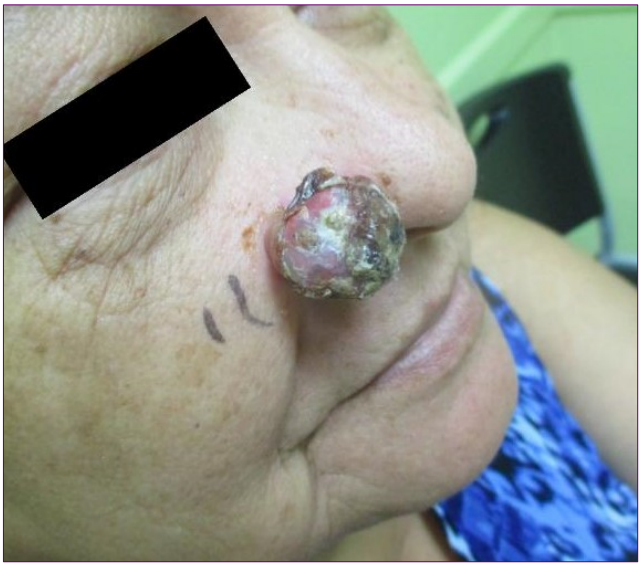
Pilomatrical carcinoma is a rare cutaneous malignancy of follicular matrix origin that was first described in the English literature by Lopransri and Mihm in 1980.[1] Since that first description over 130 cases have been reported.[2] Pilomatrical carcinoma is associated with mutations in the *CTNNB1* gene responsible for encoding β-catenin, a protein implicated in cell differentiation and proliferation.[3] It demonstrates locally aggressive behavior with high recurrence rates; however, lymphovascular invasion is rare.[2] Histological features include predominant hyperchromatic basaloid cells with high mitotic rate and nuclear pleomorphism. Anucleate matrical corneocytes or “ghost cells”, central necrosis and occasional dystrophic calcification can also be present. Lesions are most prevalent on the head and neck and predominate in a geriatric population and among male patients.[4] Treatment of the lesion is wide excision with negative margins. Radiation treatment can be considered in the case of recurrence.[5] We report a case of a pilomatrical carcinoma in a 70-year-old female patient.

CASE PRESENTATION

A 70-year-old Hispanic woman and former-smoker presented with an exophytic tumor on the right nasal ala. No other significant dermatologic history was reported. The patient reported a 3-year history of the lesion with rapid growth in the past 7 months. She denied pain and pruritus of the area. She admits to manipulating the lesions

with her fingers, as she reports “trying to pop” the lesion. On physical exam, a 1.1 cm pedunculated friable tumor with crust is noted on the right nasal ala (Figure 1). On initial visit, a shave biopsy was obtained, and histology demonstrated zones of basaloid cells that predominate over areas of necrosis (Figure 2). Nuclear pleomorphism is noted on higher power (Figure 3). Immunostaining was positive for CEA, CK7, CK-AE1/AE3, chromogranin, CK 20 and synaptophysin. After discussion of treatment options, the patient elected to have the lesion treated with wide excision.

Figure 1. Friable Nodule, Pilomatrical Carcinoma on the right nasal ala.



DISCUSSION

Pilomatrical carcinoma is a rare cutaneous malignancy of follicular matrix origin. Other names for this malignancy include malignant pilomatricoma, matrical carcinoma, and calcifying epitheliocarcinoma of Malherbe. Clinically, the lesion can be described as a rapidly progressing, flesh-colored to bluish, exophytic nodule. Nodules have been reported from 0.05 to 20 cm in diameter, with a mean of 3.8 cm. The malignancy is usually found on the head and neck, but it has also been reported on the torso, extremities, buttocks, inguinal region and axilla.[4] Seventy-six percent of reported pilomatrical carcinomas were found in males and 81% in Caucasians, with the mean age of affected patients being 52 years-old.[2] Pilomatrical carcinoma is locally aggressive and tends to metastasize. Common sites of metastasis are regional lymph nodes, lungs, bones, and the brain.[6]

Figure 2. Hyperchromatic Basaloid cells predominate the histology of Pilomatrical Carcinoma. (H&E; 1x)

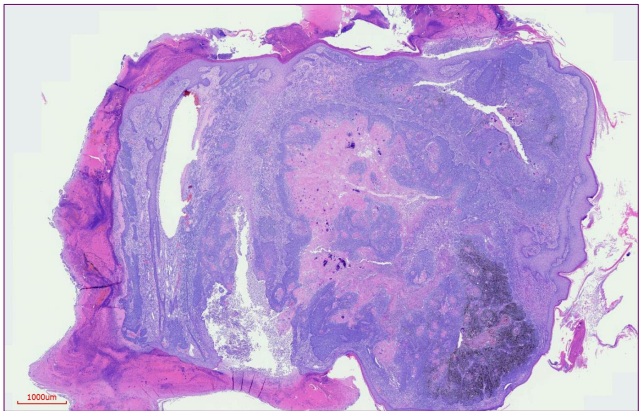
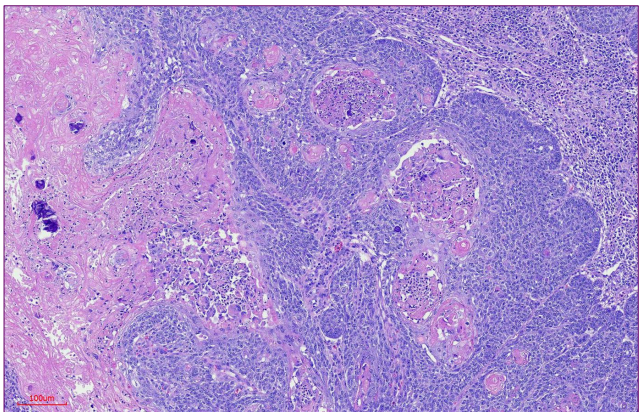


Figure 3. Hyperchromatic Basaloid cells in Pilomatrical Carcinoma. (H&E; 8.4x)



The pathogenesis of Pilomatrical carcinoma is unclear. Similarities exist between this neoplasm and the benign Pilomatricoma. Both are associated with mutations in the *CTNNB1* gene, suggesting a common pathogenesis.[3] In fact, a review by Herman

et al. found that 7% of reported cases of pilomatrical carcinoma were known to arise from a previously biopsied or surgically resected pilomatricoma. While similarities exist, a link between pilomatricomas and pilomatrical carcinoma has yet to be definitively established. Since pilomatrical carcinoma is usually found on sun-exposed areas, actinic-induced transformation has also been suggested to play a role in its pathogenesis. This link has not yet been established either, and sun exposure is not currently defined as a risk factor.[2]

Pilomatrical carcinomas can be differentiated from pilomatricomas through histological inspection. Both contain aggregates of anucleate matrical corneocytes, or ghost cells, and basaloid cells. In pilomatricomas the ghost cells dominate the histology. In a pilomatrical carcinoma, in comparison, the hyperchromatic basaloid cells with high mitotic rate and nuclear pleomorphism dominate. Central necrosis and occasional dystrophic calcification can also be seen. Macroscopically, pilomatrical carcinomas also demonstrate tumor asymmetry and poor circumscription.[4]

Pilomatrical carcinoma should also be distinguished from basal cell carcinoma (BCC) with matrical differentiation. The latter will show conventional histopathological traits of BCC with the additional presence of foci showing matrical cornification.[7]

Due to high rates of recurrence and possibility for lymphovascular invasion and metastasis, determining the correct treatment modality for pilomatrical carcinoma is important. The most described treatment is wide excision. Wide excision of the malignancy has demonstrated lower rates of recurrence. In the Herrmann et al. review, tumors removed with simple excision recurred at a rate of 83% while recurrence in wide excision was only 23%.[2] The data is less clear on whether wide excision is effective in preventing metastasis. A review by Melancon et al. showed reduced rates of metastasis in pilomatrical carcinomas treated with wide excision (10.3%) verses simple excision (20.5%), however, these differences (with p=.11) did not meet the criteria for statistical significance.[6] Wide excision of the tumor is effective in preventing recurrence, however reduction in metastasis has not yet been established.

Other treatment modalities for pilomatrical carcinoma mentioned in the literature are Mohs surgery, radiation therapy, both alone and as an adjuvant, and chemotherapy. Radiation has shown mixed results, while chemotherapy has not been proven effective.[6] Mohs surgery shows promise as a treatment modality due to the ease of identifying pilomatrical carcinoma with hematoxylin-eosin stain.[8] However, due to the limited reports in literature of treatment with Mohs surgery, further research is needed.

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References:

1. Refs Lopransri S, Mihm MC Jr. Pilomatrix carcinoma or calcifying epitheliocarcinoma of Malherbe: a case report and review of the literature. Cancer 1980 May;45(9):2368-73
2. Herrmann JL, Allan A, Trapp KM, et al. Pilomatrix carcinoma: 13 new cases and review of the literature with emphasis on predictors of

metastasis. J Am Acad Dermatol. 2014 Jul; 71(1):38-43.e2
3. Lazar AJF, Calonje E, Grayson W, et al. Pilomatrix carcinomas contain mutations in CTNNB1, the gene encoding β-catenin. Journal of Cutaneous Pathology. 2005 Feb;32(2):148-57
4. Hardisson D, Linaves MD, et al. Pilomatrix carcinoma: a clinicopathologic study of six cases and review of the literature. Am J Dermatopahol. 2001 Oct;23(5):394-401
5. Aherne NJ, Fitzpatrick DA, et al. Pilomatrix carcinoma presenting as an extra axial mass: clinicopathological features. 2008 Nov 29;3:47
6. Melancon JM, Tom WL, et al. Management of pilomatrix carcinoma: a case report of successful treatment with Mohs micrographic surgery and review of the literature. Dermatol Surg. 2011 Dec;37(12): 1798-805
7. Aloi F G, Molinero A, and Pippione M. Basal Cell Carcinoma with Matrical Differentiation. Matrical Carcinoma. American journal of dermatopathology. 1988 Dec;10(6): 509–513.
8. Xing L, Marzolf SA, et al. Facial pilomatrix carcinomas treated with Mohs micrographic surgery. JAAD Case Rep. 2018 Mar 3;4(3): 253-255