Primary Cutaneous Apocrine Carcinoma of the Scalp: A Case Presentation and Discussion

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Abstract

Primary cutaneous apocrine carcinoma (PCAC) is a rare, malignant adnexal neoplasm that tends to appear in the fifth decade of life with no predilection for sex or race. The diagnosis is complicated due to the vague clinical characteristics and the immunohistochemical similarities between PCAC and mammary apocrine carcinoma. Diagnosis relies on histologic examination, and treatment includes surgical excision with wide, clear margins. We report a case of PCAC in a cosmetically vulnerable location treated with Mohs micrographic surgery.

Introduction

Primary cutaneous apocrine carcinoma (PCAC) is an extremely rare, malignant adnexal tumor that presents a diagnostic challenge, as it shares many features with ductal mammary carcinoma with cutaneous metastasis.1 PCAC typically arises in areas of high apocrine-gland density, such as the axilla or anogenital region.2 There are reports, however, of PCAC arising on the eyelid, scalp, ear, chest, and extremities.3 We report a case of primary cutaneous apocrine carcinoma arising on the parietal scalp of a 73-year-old male treated with Mohs micrographic surgery.

Case Report

A 73-year-old Caucasian male with a past medical history including multiple basal cell carcinomas presented to an outpatient dermatology clinic for a routine annual full-body skin exam. An 8 mm by 5 mm, erythematous, asymptomatic but atypically-appearing nodule was noted on the patient’s right parietal scalp. The patient was unaware of the lesion, so the duration was unknown. A deep shave biopsy was performed, and the differential diagnosis included basal cell carcinoma, Merkel cell carcinoma, amelanotic melanoma, and epidermal inclusion cyst. The pre-biopsy photo was of poor quality and therefore was not included in this case report.

Histologic sections demonstrated compact stratum corneum cornorn with a slightly atrophic epidermis with flattening of the rete ridge pattern. Extending from the superficial dermis and throughout the reticular dermis were basoloid islands of varying sizes as well as duct formation with apocrine decapitation secretions. The basoloid islands showed slight atypia and scattered mitosis. Based on histology, a diagnosis of primary cutaneous adnexal carcinoma with apocrine differentiation (apocrine carcinoma) was favored. Because the apocrine carcinoma did not connect to the epidermis, the histopathologic differential included metastatic apocrine carcinoma; however, that diagnosis was not favored based on patient history and a lack of correlative clinical findings. Due to the location of the lesion and the high rate of recurrence, the patient was subsequently treated with Mohs micrographic surgery. Two stages were required for complete removal, and the primary defect was closed with a single advancement flap.

Histopathological variation of structural disorder, cellular atypia, and mitotic activity complicate histologic grading for PCAC. Furthermore, architectural patterns vary among different tumors, within tumors, and between primary tumors and recurrences. Despite this variability, three consistent criteria among cutaneous apocrine carcinomas reliably differentiate PCAC from other adenexal carcinomas: (1) decapitation secretion, (2) PAS-positive diastase-resistant material in the cell or lumina, and (3) positive immunostaining for gross cystic disease fluid protein.7

The standard of care for this rare malignant tumor has not been established; however, most cases are managed with wide local excision with 1 cm to 2 cm surgical margins. Mohs micrographic surgery is an option for cosmetically sensitive areas.1,4,6 Regional lymph node dissection or sentinel lymph node biopsy is recommended for palpable lymph nodes due to the high rate of lymph node metastasis at the time of diagnosis and the high rate of recurrence.2,5 Efficacies of radiotherapy and hormonal therapy as primary treatment methods are unknown, but they may be used as adjunctive treatment modalities. Although PCAC responds poorly to conventional chemotherapy, there are cases that demonstrate achievement of remission with adjunctive chemotherapy following wide surgical resection.1

Cutaneous apocrine carcinoma is usually associated with local recurrence and regional lymph node metastasis, yet this tumor is infrequently fatal. Disease-specific five-year survival has been reported at 75% to 88%.6 The five-year local recurrence rate is as low as 28%.3

Conclusion

Cutaneous apocrine carcinomas can be difficult to diagnose due to the vague clinical characteristics and rarity of the disorder. Immunohistochemical analysis is similar to that found in mammary apocrine carcinoma. Thus, the diagnosis of primary cutaneous apocrine carcinoma relies on histologic evaluation. In this case, microscopic findings were consistent with primary cutaneous apocrine carcinoma. We hope this review increases awareness of this neoplasm as a potential differential diagnosis for cutaneous neoplasms and provides insight into therapeutic modalities for successful remission.
References


