Eccrine Porocarcinoma: A Report of 2 Cases and Review of the Literature

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Practice Points

- The diagnosis of eccrine porocarcinoma is made by histologic examination, as the lesion has no consistent clinical presentation.
- The tumor has a high risk for metastasis.
- Timely biopsy of suspicious lesions and surgical treatment are important, as other treatment modalities have not been proven.

Eccrine porocarcinoma (EPC), or malignant eccrine poroma, is an uncommon malignant tumor presumably arising from the intraepidermal ductal portion of the sweat glands. The variable clinical appearance of EPC lesions can make diagnosis challenging for physicians and could delay appropriate treatment. We report 2 cases of EPC and review the salient features of this uncommon malignancy.

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Eccrine porocarcinoma (EPC) was first described by Pinkus and Mehregan[1] in 1963; they referred to it as epidermotropic eccrine carcinoma. Mishima and Morioka[2] subsequently introduced the term *eccrine porocarcinoma* in 1969. Eccrine porocarcinoma represents 0.005% to 0.01% of all malignant cutaneous tumors and carries a high risk for local recurrence, regional lymph node invasion, and distant metastasis.[3] Tumors can develop de novo, but EPC generally has been reported to develop within a preexisting benign poroma.[4]

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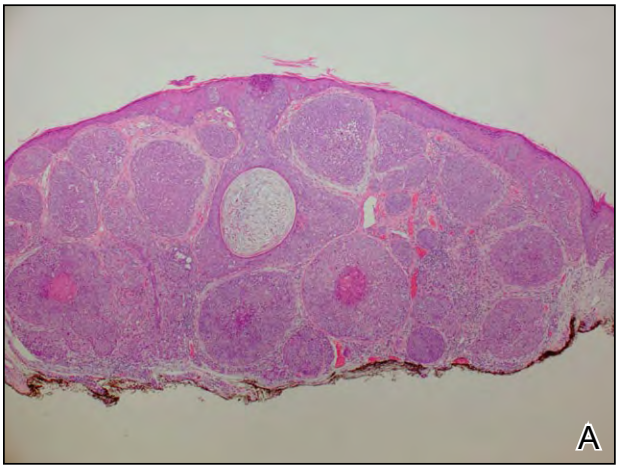
Case Reports

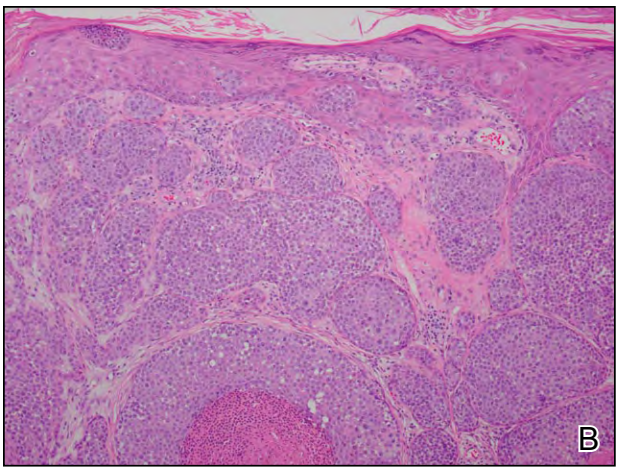
Patient 1—A 54-year-old woman with a history of hypertension presented with a slightly erythematous, crusty, 4×4-mm papule of uncertain duration on the left side of the upper neck adjacent to the proximal mandible. The patient did not report any symptoms, except for some mild irritation.

Based on our clinical examination, the differential diagnosis included irritated seborrheic keratosis, an irritated nevus, and a cutaneous malignancy. A shave biopsy revealed nests, lobules, and pagetoid spread of pleomorphic atypical poroid cells, many of them laden with glycogen in the cytoplasm (Figure 1). Histologic findings were consistent with EPC. Wide local excision with 5-mm normal skin margins showed no residual tumor. Workup for metastasis was negative. The patient was clear of clinical disease at 44-month follow-up.

Patient 2—A wheelchair-bound 72-year-old man with a history of diabetes mellitus and lymphedema of the lower extremities presented with a several year history of a crusty, erythematous, 12×9-mm nodule with slightly waxy borders on the vertex of the scalp (Figure 2). The lesion was asymptomatic aside from mild irritation that sometimes was induced when the patient combed his hair.

Based on our clinical examination, the differential diagnosis included an inflamed seborrheic keratosis or a cutaneous malignancy. A shave biopsy revealed cords and masses of pleomorphic atypical poroid cells with increased mitosis that extended from an eroded crusty epidermis into the dermis.





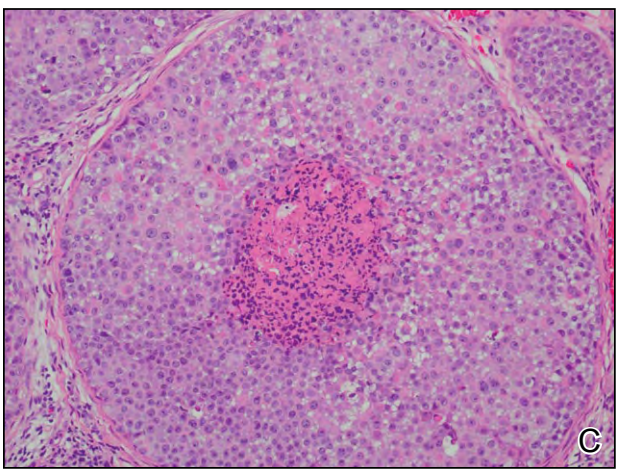


Figure 1. A shave biopsy revealed nests and lobules of poroid cells in the epidermis and dermis. Some lobules showed central necrosis (A)(H&E, original magnification ×4). Pagetoid spread of tumor cells also was noted (B) (H&E, original magnification ×10). Pleomorphic tumor cells with large hyperchromatic nuclei and prominent nucleoli were present. Many cells had clear cytoplasm. Two atypical mitotic figures were noted in the upper left corner of the field (C)(H&E, original magnification ×20).

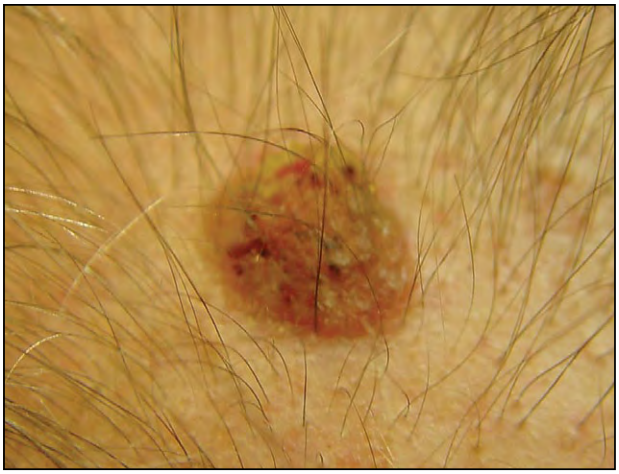


Figure 2. Crusty erythematous nodule with slightly waxy borders on the vertex of the scalp.

The histology supported a diagnosis of EPC. Initial workup, including a lymph node survey and chest radiographs, was negative for metastasis. Following discussion of treatment options, the patient was referred for Mohs micrographic surgery. The patient had no clinical disease at 22-month follow-up.

Comment

Eccrine porocarcinoma has been most frequently reported on the lower extremities (44%), followed by the trunk (24%) and head (18%).[4] Rare cases have been reported on the ear auricle,[5] eyelids,[6,7] fingers,[8] nail folds,[9-11] and genitalia.[12-14] Although the age at presentation ranges from 19 to 90 years, EPC is more common in the elderly population, with an average age of 68 years.[15] Eccrine porocarcinoma shows no predilection for either gender. Lesions can present as nodules or infiltrated plaques with or without verrucous, erosive, ulcerative, or polypoid characteristics. Multinodularity, ulceration, and rapid growth may be associated with local recurrence or metastasis.[4] The size of the lesion at the time of diagnosis can range from a few millimeters to 10 cm.[16] The differential diagnoses can include seborrheic keratosis, verruca vulgaris, pyogenic granuloma, basal cell carcinoma, squamous cell carcinoma, melanoma, and metastatic tumors.[4,17] The incidence of regional lymph node and distant metastases are 20% and 11%, respectively.[4,14] Approximately 20% of tumors recur after excision, and 5-year mortality rates may reach 67% in cases with regional lymph node involvement.[4,17,18]

Suzaki et al[19] studied the dermoscopic features of an EPC and suggested that the combination of an

atypical vascular pattern and milky red globules was specific to the diagnosis of EPC. Histologically, EPC can be recognized by cords and lobules of pleomorphic basaloid cells with large hyperchromatic nuclei and mitotic activity. In cases of EPC arising from benign poromas, areas of monomorphous poroid cells adjoining to or among areas of anaplastic cells may be observed. The presence of glycogen granules in the cytoplasm of tumor cells can be seen with the aid of periodic acid–Schiff stain. Ductal differentiation and intracytoplasmic lumen formation can be highlighted with epithelial membrane antigen and carcinoembryonic antigen.[2,20,21] Histologic findings of lymphovascular invasion, more than 14 mitoses per high-power field, and tumor depth of more than 7 mm are predictive of an aggressive clinical course.[4] A mutation in the tumor suppressor gene *TP53* could be involved in EPC carcinogenesis; however, p53 positivity cannot be reliably used as a diagnostic criterion for malignancy because it can be expressed in both benign poromas and malignant EPCs.[22] In one study, flow cytometry of DNA ploidy in benign poromas showed diploid cell population, while malignant poromas revealed aneuploid cell population in 40% (2/5) of cases.[23] Thus, aneuploidy could be utilized as an indicator of malignancy.

The mainstay of treatment of EPC is complete excision, which has resulted in cure rates of 70% to 80%[4,24]; however, the margins of resection have not been standardized. Although good results have been reported with 3- to 5-mm margins of resection,[25] other reports recommend applying clear margins of at least 10 mm.[26] Mohs micrographic surgery has been shown to be an effective treatment option for EPC.[27,28] The role of sentinel lymph node biopsy has not been defined, though it could be useful in cases with signs of histologic aggressiveness or intralymphatic permeation.[13,20] Neither chemotherapy nor radiotherapy has been proven to be clinically beneficial in treating metastatic EPC.

Conclusion

Eccrine porocarcinoma is a challenging tumor to clinically identify. Because chemotherapy and radiotherapy have not been proven in treating metastatic EPC, the best chance for cure is early diagnosis and surgical intervention. We hope to raise physician awareness of this rare tumor and highlight the need for further case reports and studies to improve diagnosis and treatment.

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