Case report: Eccrine porocarcinoma of the scalp in an immunosuppressed patient

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Abstract

Eccrine porocarcinoma (EPC) is an extremely rare malignancy of the eccrine sweat glands. They arise from the intraepidermal portion of the eccrine glands and are locally aggressive, with a high propensity to metastasize. Compromised immunity may be a contributing risk factor for developing EPC. We report a case of EPC presenting on the scalp in a patient on immunosuppressive therapy for bilateral lung transplants.

Introduction

Eccrine porocarcinoma (EPC) was first described in 1963 by Pinkus and Mehregan. They used the term "epidermotropic eccrine carcinoma" to describe this tumor due to its origin from the intraepidermal portion of the eccrine sweat glands. EPC is a very rare type of skin cancer, representing 0.005% to 0.01% of all skin tumors.[1-5] EPC tumors occur most commonly in older age groups, with peak incidence at age 67, and have a slight female predominance.[3,4,6] They have various clinical appearances, presenting as an ulcerated nodule, plaque, polypoid or verrucous papule.[4] The most common location is the lower extremities, but EPC can occur on the upper extremities, trunk and abdomen. EPC arising in the setting of immune compromise is rare. Here, we report a case of EPC presenting on the scalp in a patient on immunosuppressive therapy for bilateral lung transplants.

Case Report

A 49-year-old Caucasian male presented with a rapidly enlarging tumor on his left posterior scalp for five weeks. The lesion was mildly tender to palpation but was otherwise asymptomatic. No regional lymphadenopathy was palpable. Past medical history was significant for cystic fibrosis resulting in bilateral lung transplant. Medications included mycophenolate mofetil 1.5 g twice daily, prednisolone 5 mg daily, and tacrolimus 1 mg twice daily. Family history was noncontributory.

Clinical exam revealed a 2.7 cm, exophytic, pink, keratotic nodule on the left posterior scalp (**Figure 1**). Upon initial examination, squamous cell carcinoma was suspected, and excision with Mohs micrographic surgery (MMS) was planned for the following week. The patient was prophylaxed with cephalexin 500 mg twice daily due to his immunosuppression. The tumor was first debulked with a flexiblade, and a specimen was sent for permanent sections. Deep and peripheral margins were examined during MMS, and squamous cell carcinoma was present in the first two stages. The tumor was cleared after removal of the third stage. The resulting defect was 4 cm, and the patient was sent to plastic surgery for closure immediately following MMS.

Histopathological evaluation of the permanent sections revealed additional findings beyond what was seen during MMS. Low-power microscopy revealed a proliferation of atypical epithelium emanating from the intraepidermal portion of the eccrine sweat glands, extending into the dermis (**Figure 2**). Areas of epithelioid cells with intercellular bridges and keratinization were present. However, cells with ductal differentiation were present (**Figure 3**) and were highlighted with both carcinoembryonic antigen (CEA) (**Figure 4**) and cytokeratin 7 (CK7) (**Figure 5**). No areas of lymphovascular or perineural invasion were identified. Depth of involvement was at least 6.5 mm, Clarks level IV. A diagnosis of eccrine porocarcinoma with squamous differentiation was made.

Subsequently, the patient was sent to medical oncology for evaluation and recommendations for further workup. The consultant endorsed only close clinical surveillance. At the time of publication, this patient has been recurrence-free for two months.

Discussion

Pinkus and Mehregan first described the tumor as epidermotropic eccrine carcinoma in 1963, and it was later termed eccrine porocarcinoma (EPC) by Mishima and Morioka in 1969.[2]

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Figure 1

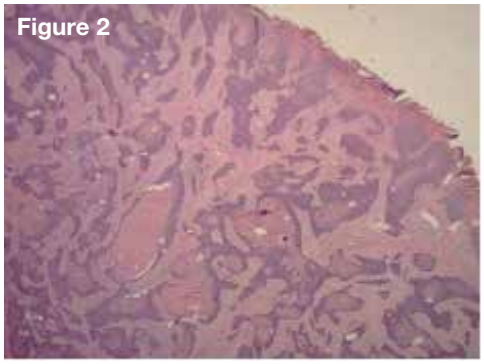
Figure 2

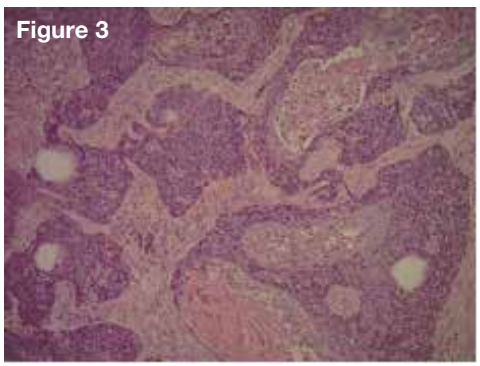
Figure 3

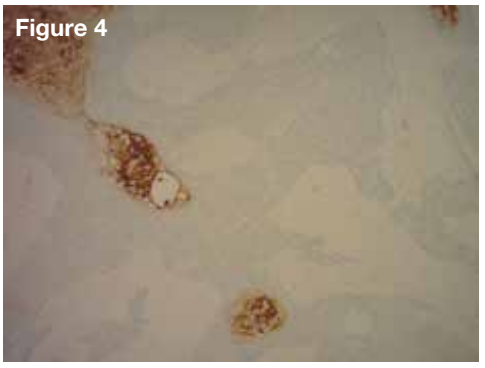
Figure 4

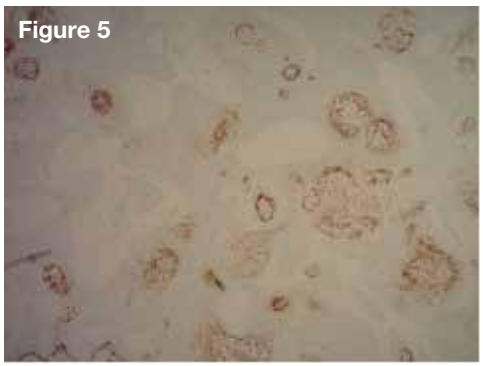
Figure 5

the eccrine glands and are locally aggressive, with a high propensity to metastasize. EPCs most commonly occur in adult life, with various reports of peak incidence ranging from 67 to 74 years.[2,3] A majority of authors report no difference in incidence between genders; however, Riera-Leal et al. recently described a slight female bias (64%).[6] The most frequent location of presentation is the lower extremities (50%), followed by the trunk (24%), head (18%), and upper extremity (8%).[2-4] Our case involved an atypical presentation on the scalp in an immunosuppressed patient.

Clinically, eccrine porocarcinomas can have a variety of appearances, such as ulcerated nodule, verrucous plaque, or polypoid papule. Due to their protean clinical presentations, these tumors are often misdiagnosed preoperatively and can be mistaken for pyogenic granuloma, basal cell carcinoma, seborrheic keratosis, amelanotic melanoma, and, as in our case, squamous cell carcinoma.[4]

Histologically, EPCs are located in the epidermis and dermis, as they arise from the intraepidermal portion of the eccrine sweat glands. The tumor is characterized by epithelioid cells with ductal lumen associated with pleomorphic, hyperchromatic nuclei and mitotic figures. When confined to the epidermis, they are termed "in situ." They can also invade the dermis with a pushing or infiltrative pattern. In our case, there was involvement of atypical cells extending into the dermis. There was no evidence of cutaneous or systemic metastases (no palpable lymphadenopathy); however, due to the aggressive nature of the lesion (and the patient's immunosuppressed state), our patient was referred to medical oncology.

EPCs are locally aggressive with a high likelihood of metastasis. Twenty percent of cases have reported local and lymph-node metastases (even though margins were free on pathological exam), while 10% of cases have reported further metastases to viscera and bone.[2,4] With metastases, prognosis is poor, with reports of mortality rates ranging from 67% to 80%. Prognosis has been associated with clinical characteristics such as multinodularity, ulceration, and rapid growth.[2,4] Histologic features also correlate with prognosis: A mitotic index of more than 14 mitotic cells/hpf, lymphovascular invasion, and a tumor depth greater than 7 mm indicate a worse prognosis.[2,4] High-risk features in our patient's tumor included ulceration, rapid growth, and a possible depth of greater than 7 mm.

In addition, compromised immunity may be a contributing risk factor for developing EPC. Six previous cases of EPC in immunosuppressed patients have been described: three patients with renal transplants, two patients with human immunodeficiency virus (HIV), and one patient with chronic lymphocytic leukemia (CLL).[4,12] Our patient, on treatment after bilateral lung transplant with cystic fibrosis, is the seventh reported case of EPC with immunosuppression.

Due to the rarity of this lesion, as well its aggressive nature, an optimal treatment plan has not been defined. A variety of treatment options are used for EPCs and include wide local excision (WLE), chemotherapy, radiation therapy and Mohs micrographic surgery (MMS). While most cases have been treated with wide excision, Song et al. proposed that MMS has proven success and should be considered.[4] MMS allows for examination of 100% of peripheral margins vs. traditional bread-loaf sections with WLE.

Conclusion

Eccrine porocarcinoma (EPC) is a rare skin malignancy presenting with a variety of clinical appearances, making identification difficult. In our case, an exophytic, pink, keratotic nodule was located on the scalp and initially thought to be squamous cell carcinoma. EPC recognition is important due to its aggressive growth, propensity for recurrence and spread, and resultant poor prognosis. No concise guidelines have been established for the treatment of EPC due to the paucity of reports in the literature. We report the 22nd case of EPC treated with MMS. It is a unique case of eccrine porocarcinoma due to its uncommon location on the scalp, as well as its presentation in an immunosuppressed patient. To the best of our knowledge, there have been fewer than 10 cases of porocarcinoma involving the scalp reported in the literature.[1,7,8] Our patient is also only the seventh reported case of EPC with compromised immunity, and, to our knowledge, the first case in a patient with lung transplantation.

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