Surgical and Experimental Pathology

CASE REPORT Open Access

Occurrence of metastatic eccrine porocarcinoma in an immunosuppressed patient

Mitul B. Modi[1*], Ata S. Moshiri[2], Toru Shoji[3], Martin C. Mihm Jr[4], Xiaowei Xu[5] and David E. Elder[5]

Abstract

Background: Eccrine porocarcinoma is a rare malignancy accounting for 0.005 to 0.01% of all cutaneous tumors. However, its etiology is not well established. Herein we are reporting the occurrence of metastatic porocarcinoma of the back in a patient with a history of multiple cutaneous malignancies and chronic immunosuppression.

Case presentation: A 79-year-old male with a history of long-term immunosuppressive therapy for left lung transplantation and multiple cutaneous malignancies including melanoma, presented with an enlargening plaque on the right upper back. Biopsy demonstrated an infiltrative epithelial tumor with aggressive histology and ductal formation. Immunohistochemical stains for p63, CK-5/6, and CEA were diffusely positive, while CK-7 and CK-20 were negative. A diagnosis of eccrine porocarcinoma was made. He underwent complete wide excision with negative margins of the right upper back in January 2018. In November 2018, he was found to have a right posterior shoulder mass, positive right axillary lymph nodes, superficial right upper back soft tissue masses and lymphangitic carcinomatosis involving the left lung. He was transitioned to comfort care and subsequently passed from metastatic disease in December 2018.

Conclusion: The occurrence of eccrine porocarcinoma in a patient on long term immunosuppressive therapy for lung transplantation suggests a possible role for chronic immunosuppression in the induction of a subset of eccrine porocarcinomas, as in other cutaneous malignancies.

Keywords: Eccrine porocarcinoma, Infiltrating margin, Squamous cell carcinoma, Immunosuppression, Cutaneous lesions

Introduction

Eccrine porocarcinoma (EPC), was first described in 1963 by Pinkus and Mehregan. EPC, also known as "malignant eccrine poroma," accounts for 0.005 to 0.01% of all cutaneous malignancies, although the exact prevalence is unknown.(Salih et al., 2017a) It has been proposed that the tumor developed from a pre-existing eccrine poroma and factors such as exposure to chemical agents, chronic sun exposure, and immunosuppression could be the predisposing factors for de novo EPC.(Salih et al., 2017a) Despite these conjectures, its etiology is not well established.(Salih et al., 2017b) EPC develops from the intraepidermal ductal portion of the sweat gland.(Riera-Leal et al., 2015). The peak incidence occurs around 67 years of age and the majority of the patients are usually older, but it can affect children or young adults.(Salih et al., 2017b; Riera-Leal et al., 2015) A slight predominance in women has been observed.(Salih et al., 2017b; Riera-Leal et al., 2015). The head and neck are the most common location followed by lower extremity.(Salih et al., 2017b; Riera-Leal et al., 2015). The majority of published literature are single case reports, but few case series exist; some of them reporting only histopathologic findings and few describing clinical aspects only.(Riera-Leal et al., 2015)

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Herein we are reporting a case of 79-year-old male patient with a history of iatrogenic immunosuppression who died of metastatic eccrine porocarcinoma, with a detailed review of cases and studies from the literature to date.

Case presentation

A 78 year old male on a routine follow-up visit for melanoma of his lower back in December 2017, was found to have a large, asymptomatic, scaly plaque on the right upper back. His medical history was significant for left lung transplantation for chronic obstructive pulmonary disease. Biopsy of the lesion showed an epidermal based neoplasm with anastomosing cords of tumor cells extensively infiltrating into the dermis and involving the deep margins (Figs. 1 and 2). Immunohistochemical stains for p63, cytokeratin (CK)-5/6 and carcinoembryonic antigen (CEA) (Fig. 3) were diffusely positive, whereas CK-7 and CK-20 were negative. A diagnosis of eccrine porocarcinoma was made. Excision with appropriate negative margins, sentinel lymph node biopsy and workup for possible metastasis were recommended.

The patient underwent complete wide excision of the right upper back lesion in January 2018. Preoperatively he underwent computed tomography (CT) imaging of chest, abdomen and pelvis for staging. Chest CT was unremarkable concerning metastatic disease, but CT abdomen and pelvis showed indeterminate hepatic lesions as well as a prominent right inguinal lymph node. Magnetic Resonance Imaging (MRI) abdomen showed benign hepatic lesions as well as a cystic nodule in the right groin. He was deemed to have no evidence of distant metastasis at that time.

In November 2018, the patient was admitted to an outside medical institution for a two-month history of an enlarging right shoulder mass that began to ooze foul-smelling drainage. On review of systems, he also complained of worsening dyspnea on exertion, cough productive of dark brown/red sputum, and 10 lbs. weight loss. Given concerns for superficial skin infection, he was managed with intravenous (IV) antibiotics and transferred to our institution for further treatment. CT imaging of his neck showed a heterogeneous necrotic mass along the right upper back and shoulder (Fig. 4). Repeat chest CT was significant for likely malignant right axillary lymph nodes and superficial right upper back soft tissue masses, small left pleural effusion and lymphangitic carcinomatosis involving the left lung predominantly. The biopsy of the shoulder mass showed recurrent eccrine porocarcinoma (Figs. 5 and 6).

The patient was treated with radiation to the right shoulder and axillary masses for 12 days in December 2018. The treatment was initially intended to continue, but the decision was made to shorten his course considering his worsening prognosis and inability to pursue systemic chemotherapy. He was eventually moved to comfort care and expired shortly thereafter.

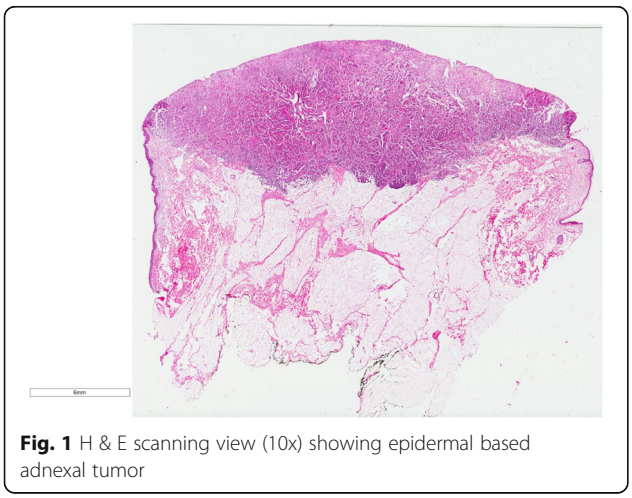

Fig. 1 H & E scanning view (10x) showing epidermal based adnexal tumor

Discussion

Eccrine porocarcinoma (EPC) is an exceedingly rare cutaneous adnexal tumor with high potential for morbidity and mortality.(Nazemi et al., 2018) Although any body site can be affected, EPC typically presents on the head and neck.(Salih et al., 2017b; Riera-Leal et al., 2015; Robson et al., 2001) At the time of presentation, tumor size generally varies from < 1 cm to 10 cm.(Robson et al., 2001) EPC cases have been reported to slowly arise from a preexisting benign eccrine poroma, however rapid development of de novo lesions is also well documented.(Robson et al., 2001)

Malignant tumor cells with nuclear atypia, hyperchromatic nuclei, foci of necrosis and frequent mitoses are characteristic features of EPC on histopathological examination.(Choi et al., 2011) Occasionally, cords and nests of polygonal tumor cells penetrating to adjacent dermis or extending into subcutaneous tissue have also been identified.(Choi et al., 2011) Epithelial cell abnormalities are not sufficient to make a diagnose of EPC according to Abenoza and Ackerman, as this may be apparent in benign eccrine poromas. The diagnosis of EPC is based on an invasive architectural pattern and the formation of cohesive basaloid epithelial cells.(Robson et al., 2001) Roaf et al. asserted EPC as seeming "benign" lesions on histological examination with locally aggressive behavior and further metastases.(Robson et al., 2001) Additionally, Shaw et al. contend two different classifications of EPC: an "infiltrative" type defined based on the nature of advancing tumor margin

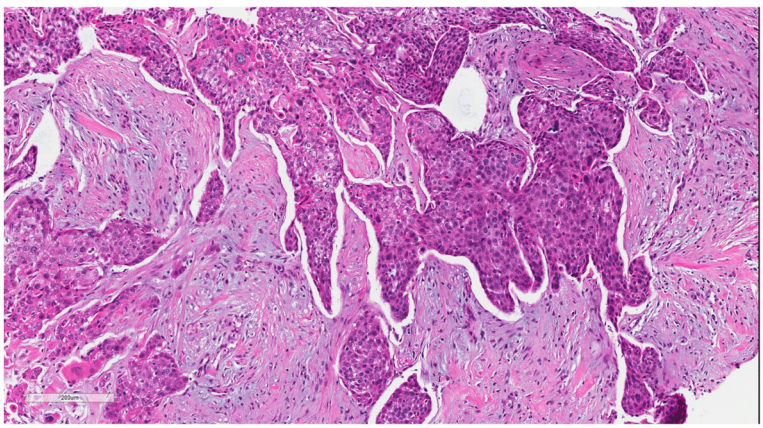

Fig. 2 H & E on high magnification (40x) showing tumor cells with anastomosing cords

irrespective of the degree of cytologic atypia; and the "cytologic" type with malignant cellular features and necrosis.(Robson et al., 2001) Because of synchronous epidermoid differentiation, diagnosis of EPC can be further supported by immunohistochemical staining for carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), and p53. (Robson et al., 2001; Parmar et al., 2016; Akalin et al., 2001) Positive staining for CEA and/or EMA usually accentuates ductal differentiation. (Robson et al., 2001; Parmar et al., 2016) Our case also showed infiltrative margins and positive CEA staining.

EPCs that are greater than 7 mm in thickness generally have more than 14 mitoses per high power field, or have lymphovascular invasion, and tend to carry a poor prognosis.(Robson et al., 2001; Parmar et al., 2016; Mahomed et al., 2008) However, generally tumor thickness is considered to be the main prognostic factor for EPC.(Robson et al., 2001; Parmar et al., 2016) Sentinel node biopsy has been proven to be advantageous tool for revealing metastasis to regional lymph nodes.(Robson et al., 2001; Parmar et al., 2016)

Mahomed et al reported five cases of porocarcinomas in patients with histories of immunocompromised conditions: two out of five cases were the acquired immune deficiency syndrome (AIDS) and the other three were recipients of renal transplants.(Salih et al., 2017b; Mahomed et al., 2008) Belin et al also investigated two patients (one for chronic lymphoid leukemia and the other for renal transplantation) with porocarcinoma who received immunosuppressing medications.(Salih et al., 2017b; Belin et al., 2011) Other authors also reported several of their patients who had similar histories of immunosuppressing treatment, and few others were

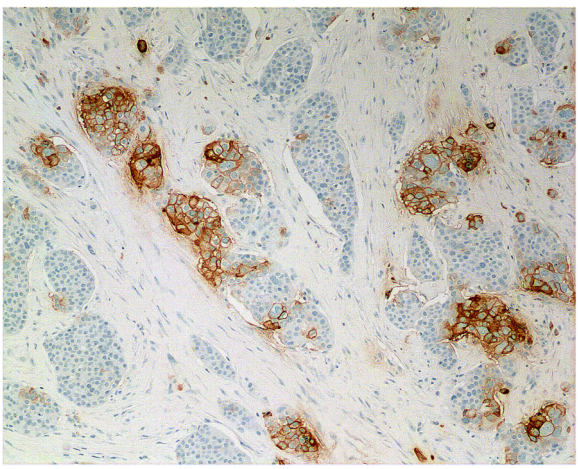

Fig. 3 Clinical image showing necrotic and fungating shoulder mass

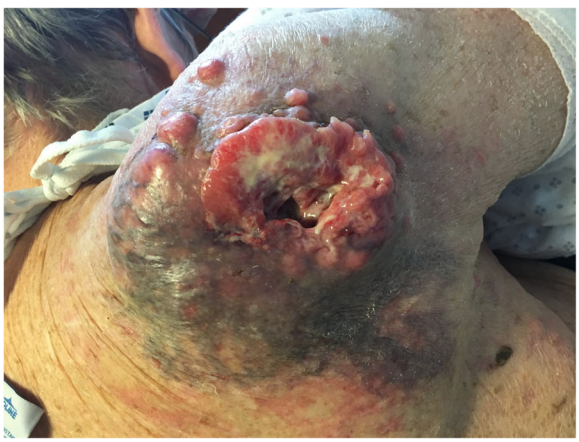

Fig. 4 Carcinoembryonic antigen (CEA) immunostain has a membranous pattern around a subset of tumor cells and highlights irregular ductal structures

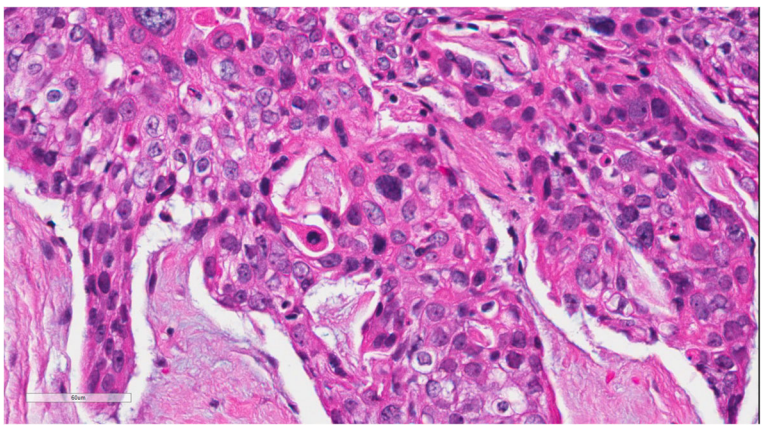

Fig. 5 H & E on high magnification (40x) showing malignant epithelial tumor cells forming ductal structures

immunocompromised due to underlying medical illnesses.(Salih et al., 2017b; Mahomed et al., 2008; Belin et al., 2011; Kim et al., 2005) Our case further supports the role of immunosuppression in the development of EPC given our patients' history of lung transplantation ten years before the diagnosis of EPC.

The presence of an infiltrating margin or marked cellular atypia should remind the pathologist to recommend reexcision and consideration of regional lymph node evaluation if there is doubt regarding the completeness of tumor removal. Surgery remains the principle management strategy for localized disease, with adjuvant chemoradiotherapy considered, especially if there is recurrence or metastasis.(Salih et al., 2017a; Salih et al., 2017b; Plunkett et al., 2001; Ramirez et al., 2012; Aaribi et al., 2013)

Summary

We are reporting an occurrence of metastatic eccrine porocarcinoma with an infiltrating margin on the back of an immunosuppressed patient. The presence of an infiltrating margin should alert the pathologist to recommend wider excision margins and regional lymph node dissection if there is uncertainty regarding completeness of excision. Radiotherapy and chemotherapy are necessary when presented with metastasis and recurrence. Due to the limited number of cases reported in the literature, it is very important to have a basic knowledge of this disease process as a differential diagnosis when dealing with such aggressive cutaneous lesions. Our patient's history of immunosuppression further supports a possible role for chronic immunosuppression in the carcinogenesis and metastasis of eccrine porocarcinomas. Further research may be useful

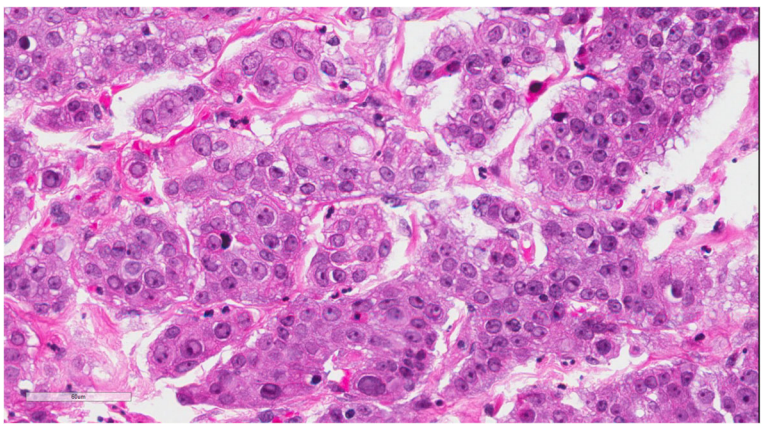

Fig. 6 H & E on high magnification (40x) showing pleomorphic tumor cells with enlarged, hyperchromatic nuclei and prominent nucleoli

in exploring the possibility of an infectious etiology, as has been found to drive several malignancies in the immunocompromised patient populations.

Abbreviations

EPC: Eccrine porocarcinoma; CK: Cytokeratin; CEA: Carcinoembryonic Antigen; CT: Computed tomography; MRI: Magnetic Resonance Imaging; IV: Intravenous; EMA: Epithelial Membrane Antigen

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Not Applicable.

Authors' contributions

MBM, AM, XX and DE edited and finalized the manuscript. DE performed the histological examination of the final resection. The authors read and approved the final manuscript. TS and MMihm provided the diagnosis on the initial biopsy with supporting immunohistochemical stains.

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Competing interests

The authors declare that they have no competing interests.

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