

Case Report

Eccrine Porocarcinoma: A Challenging Diagnostic and Therapeutic Tumoral Entity

Ha Mo Linh Le^a Laurence Faugeras^a Véronique De Moor^b
Caroline Fervaille^c Thierry Vander Borght^d Fanny Collette^a
Lionel D'Hondt^a

^aDepartment of Oncology, CHU UCL Namur – Site Godinne, Yvoir, Belgium; ^bDepartment of General Surgery, CHU UCL Namur – Site Godinne, Yvoir, Belgium; ^cDepartment of Pathology, CHU UCL Namur – Site Godinne, Yvoir, Belgium; ^dDepartment of Nuclear Medicine, CHU UCL Namur – Site Godinne, Yvoir, Belgium

Keywords

Eccrine porocarcinoma · Sweat gland tumors · Management

Abstract

Eccrine porocarcinoma is a rare malignant cutaneous tumor with high rates of extracutaneous spread, and its diagnosis and management can be quite challenging. This is a case of an 82-year-old woman presenting with an asymptomatic and chronic pubic skin lesion for whom the work-up required many investigations and procedures to confirm the diagnosis of metastatic eccrine porocarcinoma. Indeed, the patient underwent a wide local excision of the skin lesion, imaging with an FDG-PET scan, a colonoscopy, and two inguinal node dissections. As illustrated in this case, surgery should always be considered to achieve disease remission. Other treatments such as chemotherapy and radiotherapy have also been reported in the literature without clear standard guidelines.

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Introduction

Eccrine porocarcinoma (EPC) is a rare malignant cutaneous tumor with high rates of extracutaneous spread. It has first been described by Pinkus and Mehregan in 1963 [1]. EPC accounts for 0.005–0.01% of all malignant cutaneous tumors and arises from eccrine sweat



Fig. 1. A 2-cm ulcerated and erythematous skin lesion on the pubis.

gland tumors [2]. Due to its uncommon onset, EPC can be misdiagnosed as other skin neoplasms, and optimal management is lacking.

Here we report a case of EPC with lymphovascular invasion managed in our institution. Multi-step treatment was required to obtain a complete remission.

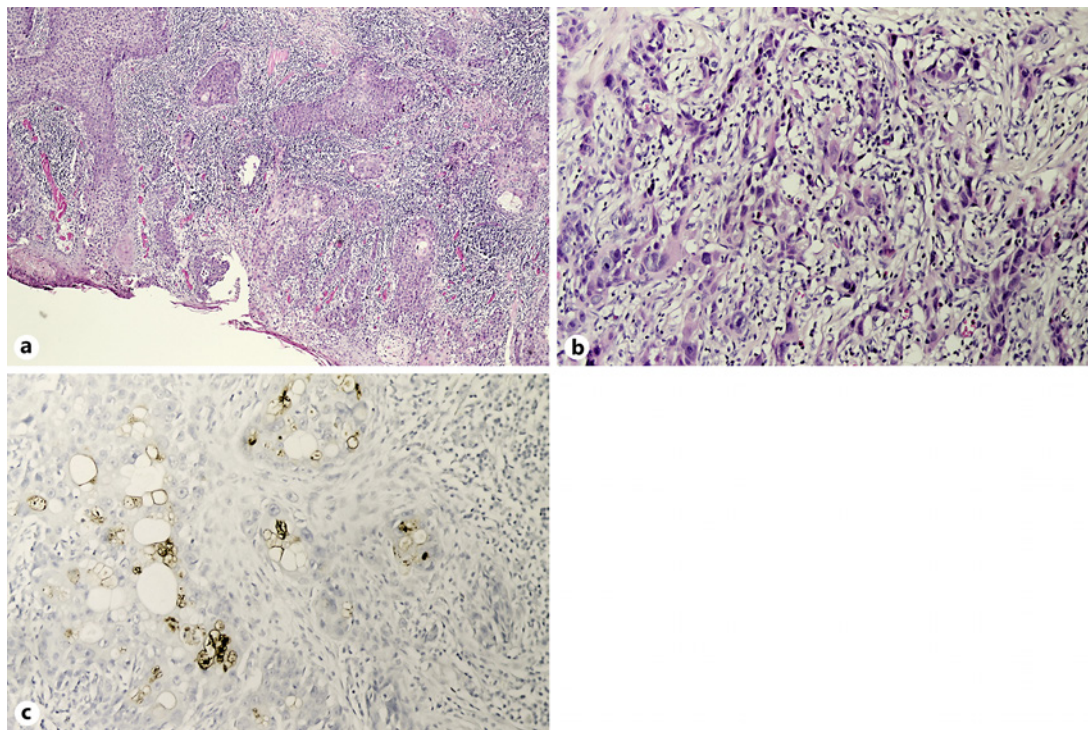
Case Report

An 82-year-old Caucasian woman with no major past medical history was referred to our center after undergoing biopsies of longstanding pubic skin lesions. The patient was otherwise feeling well and had no symptoms. Physical examination revealed a 2-cm ulcerated lesion on her pubis (shown in Fig. 1). There were no palpable inguinal lymph nodes, and examination of the genital parts was normal.

The biopsy results from another medical center came back with a diagnosis of poorly differentiated squamous cell carcinoma on all the samples. The excision was incomplete, and therefore this patient was addressed for further management.

The patient's case was discussed at our multidisciplinary meeting, and wide local excision was decided. The histological report of the reexcision revealed a lesion with a depth of 4 mm in the dermis reaching the limit of the hypodermis, with tumoral cells infiltrating the dermis and harboring nuclear atypia and high mitotic activity. The lesion was ulcerative with a fibrinous leukocytic exudate. Vascular emboli were reported, and clear margins of 1 cm were achieved (shown in Fig. 2). Immunohistochemistry showed positive staining for carcinoembryonic antigen and cytokeratin (CK) 7 on the luminal ductal borders. p40 and CK5 and 6 were also positive on the whole cell population, and epithelial membrane antigen was heterogeneously positive. Those characteristics were consistent with the diagnosis of EPC. TNM classification was pT2.

Given the presence of vascular emboli, a staging CT scan of the thorax, abdomen, and pelvis was performed, which showed an image compatible with pelvic carcinomatosis. An additional FDG-PET scan was then requested, which displayed a hypermetabolic left inguinal lymph node and focal uptake in the right colon (shown in Fig. 3). Further investigations were



conducted, which included a total colonoscopy and an inguinal left node biopsy. The ultrasound-guided node biopsy revealed distant infiltration of the porocarcinoma, whereas the endoscopy showed a large polyp which could not be endoscopically resected. Therefore, right colectomy combined with left inguinal and left iliac node dissection was performed, but the pathological findings showed no metastatic involvement. An additional ultrasound-guided resection was carried out to remove the targeted left hypermetabolic inguinal node, whose histology eventually revealed EPC and confirmed the disease's metastatic setting. The patient has been undergoing close follow-up for the last 4 months without any sign of relapse.

Discussion

EPC is an uncommon malignant adnexal cutaneous tumor deriving from terminal cells of the intraepithelial part of the eccrine duct called the acrosyringium. It occurs mainly in elderly patients, most frequently between the sixth and the eighth decade. It equally affects males and females and does not have any propensity for a specific ethnicity. There is currently no evidence suggestive of a hereditary factor. The etiology of EPC remains unknown, but risk factors such as radiation exposure and immunosuppression are correlated with EPC [3]. It arises de novo or develops from a benign preexistent lesion (poroma, nevus sebaceus, or actinic lesions). It is commonly found on the lower limbs and the head, but other sites have frequently been reported, such as the upper limbs, the trunk, and the abdomen. Head and neck locations seem to be associated with a lower incidence of lymph node involvement, whereas primary lesions on the genitalia and buttocks have a higher propensity for distant metastases [4].

Clinically, EPC appears as an asymptomatic erythematous or violaceous nodule or mass, although it can sometimes be ulcerative and painful [5]. It is often misdiagnosed as other skin lesions, particularly as squamous cell carcinoma. Other differential diagnoses include

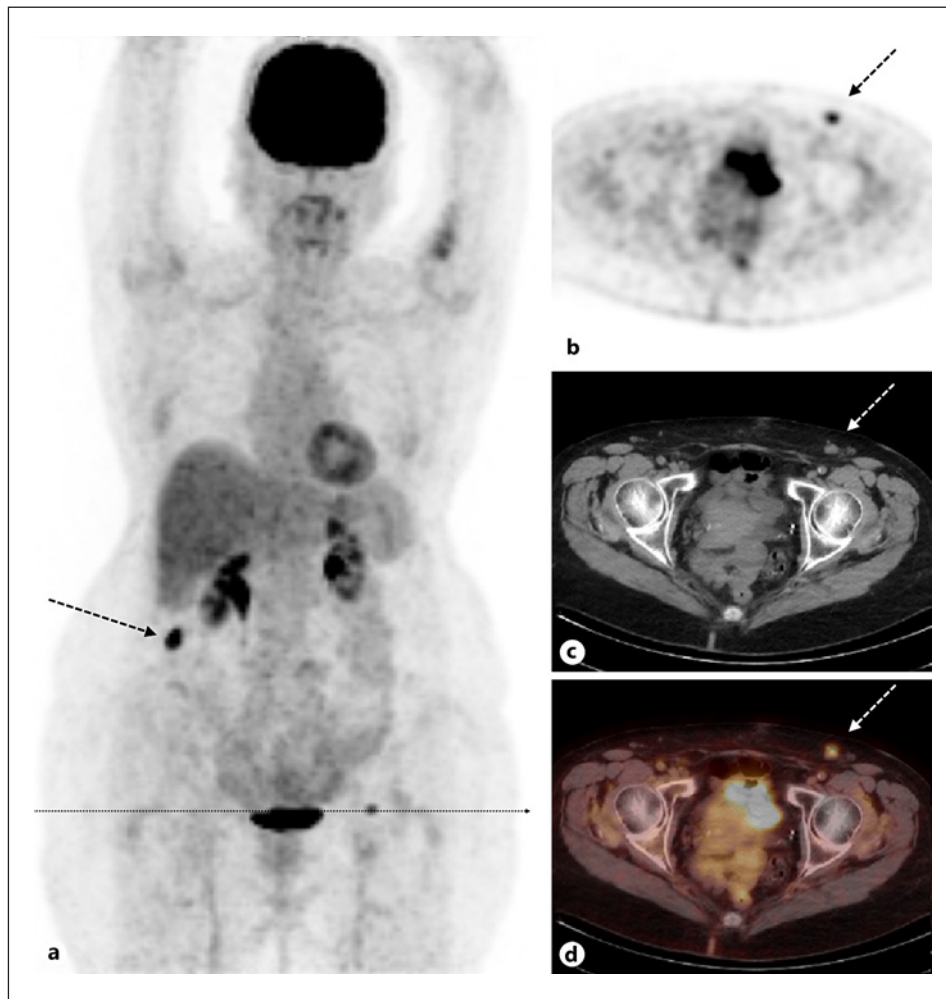


Fig. 3. In the left panel, the maximum-intensity projection image (**a**) shows foci of intense FDG uptake in the right colon (arrow) and in a left inguinal node through the level of which PET (**b**), noncontrast CT (**c**) and fused PET-CT (**d**) axial images are displayed. In the right panel, the left inguinal node is also highlighted by arrows on the three axial images.

pyogenic granuloma, amelanotic melanoma, seborrheic keratosis, Bowen’s disease, fibroma, verruca vulgaris, or metastatic adenocarcinoma.

Given its rare occurrence, there is currently no guideline for its work-up and management. Most available data are based on case reports and small case series. Robson et al. [3] presented a review of the disease with the largest series of 69 patients, describing the clinicopathologic features of EPC. Histology is important to confirm the diagnosis of EPC. This tumor shows various histopathological features, but some common characteristics are found, including nuclear atypia, an infiltrative growth pattern, increased mitotic activity, and necrosis. The architecture presents ductal differentiation and nests of basaloid cells [3, 6]. EPC is classified into three histological variant groups according to the infiltrative pattern of the margins. There is the “infiltrative” variant, which is characterized by malignant clusters infiltrating the dermis and often the hypodermis, the “pushing” form with a clear lower dermal limit, and the “pagetoid” lesion with an intraepidermal spreading of neoplastic cells [3, 4]. A high mitotic index, lymphovascular invasion, and a tumor depth greater than 7 mm are histopathological features that

have been associated with poor prognosis [3]. Immunohistochemistry is also a helpful tool, though there is no single marker for eccrine cells. These often show positive staining for keratin, epithelial membrane antigen, carcinoembryonic antigen, CK, p53, and p63 [3]. CD117 may be a useful marker in helping to distinguish EPC from squamous cell carcinoma [4]. The prognosis of EPC is not well established, due to its rarity and its varied clinical course. Five-year mortality rates reach 67% in case of positive regional lymph nodes [7].

Staging with radiology is useful for the evaluation of local and distant metastases, especially for tumors with high-risk features and poorly differentiated porocarcinoma. Varied modalities of diagnostic imaging are used, including whole-body CT scanning and PET-CT, but local imaging for lymph node assessment based on ultrasound, CT, or MRI has also been reported [4]. Sentinel lymph node biopsy remains controversial, but its use might be supported for high-risk EPC [8].

Treatment should always be considered, as it is an aggressive disease with a high potential for morbidity and mortality. Lymph node involvement is found in 20% of EPCs, and 10% have distant metastases at diagnosis [9]. The local recurrence rate is 20% in case of lymph node involvement [3]. Many treatment options have been reported, with no clear standard. Wide surgical excision is the mainstay of treatment, resulting in cure rates of 70–80% when the margins are clear. Some studies also reported good outcomes with Mohs micrographic surgery, and this technique could also be considered [4]. There is currently no evidence supporting adjuvant treatment, even in patients with poor prognostic factors [10]. Node dissection is recommended in the event of a positive sentinel node biopsy and clinical regional lymphadenopathy. Metastases occur mainly as cutaneous lesions in the primary area, but they can also develop in the lung, retroperitoneum, bone, liver, breast, bladder, peritoneum, or ovary [11]. Various chemotherapy regimens and radiation therapies have been used for the metastatic setting, but there is no standardized regimen. Treatment options are based on limited case series, and clinical responses have been achieved mostly with several combinations of chemotherapy [5, 8]. Piedbois et al. [12] achieved a durable complete remission using polychemotherapy with doxorubicin, mitomycin C, vincristine, and 5-fluorouracil (5-FU), alternating with cisplatin and bleomycin. In another case report, complete remission was seen in a patient with EPC of the leg after hyperthermic limb perfusion with 5-FU and melphalan [13]. A case of cutaneous metastatic EPC treated with topical 5-FU and intra-arterial docetaxel has also been reported [11]. Plunkett et al. [10] showed some benefit using a single cytotoxic agent with docetaxel. More recently, responses have been reported with the use of epidermal growth factor receptor targeted therapy and pembrolizumab [14, 15].

Conclusions

EPC is a rare malignant skin disease for which tissue biopsy is essential for a diagnosis. This clinical case illustrates the fact that EPC is a challenging diagnosis, and that clinical features as well as histological characteristics are key to differentiate this disease from other skin tumors. Early diagnosis and treatment are essential to decrease mortality, as EPC has a high rate of recurrence and metastasis. As in our case, multistep treatment is sometimes required to obtain complete remission.

Statement of Ethics

The authors certify that written informed consent was obtained from the patient to publish this case and the images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

H.M.L. Le reviewed the literature and wrote the Abstract, Introduction, Case Report, and Discussion sections. L. D'Hondt supervised the work. T. Vander Borgh and C. Fervaille contributed to the figures' legends. L. D'Hondt, L. Faugeras, V. De Moor, T. Vander Borgh, C. Fervaille, and F. Collette revised the paper and gave their final approval.

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