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J Case Rep Images Pathol 2021;7(2):18–22.
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A case report of eccrine porocarcinoma with metastatic spread

Bradley Kaptur, Nicholas Peterman, Keith Cordner, Ikechukwu Uzoaru, Juan Jimenez

ABSTRACT

Introduction: Eccrine porocarcinoma (EPC) is a rare malignant neoplasm arising from the eccrine sweat glands. Metastatic presentation of EPC is a rare complication of the disorder.

Case Report: A 62-year-old man presented with a longstanding skin lesion on his right forearm. A punch biopsy specimen demonstrated ulcerative, highly infiltrative carcinoma extending throughout the dermis into the cutis. The tumor cells were positive for cytokeratin 5, cytokeratin 7, p63, and p40. Additionally, immunostains showed positive epithelial membrane antigen (EMA), focal positive S100, negative carcinoembryonic antigen (CEA), and negative Melan A. They showed areas of ductular differentiation, and lymphovascular invasion was found to be present. Positron emission tomography (PET) and magnetic resonance imaging (MRI) demonstrated distant metastases, with a prominent large necrotic mass in the left leg.

Conclusion: Eccrine porocarcinoma is often misidentified initially, and there is no clear consensus on the best treatment approach.

Keywords: Eccrine porocarcinoma, Eccrine poroma, Metastatic skin malignancy, Skin lesion

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Received: 25 August 2021
Accepted: 05 December 2021
Published: 23 December 2021

How to cite this article

Kaptur B, Peterman N, Cordner K, Uzoaru I, Jimenez J. A case report of eccrine porocarcinoma with metastatic spread. J Case Rep Images Pathol 2021;7(2):100057Z11BK2021.

Article ID: 100057Z11BK2021

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doi: 10.5348/100057Z11BK2021CR

INTRODUCTION

Eccrine porocarcinoma (EPC), or malignant eccrine poroma, is a rare skin malignancy that develops from the intraepidermal ductal portion of the eccrine sweat glands [1]. It has been estimated to account for between 0.005% and 0.01% of cutaneous tumors [1, 2]. Eccrine porocarcinoma was first described in 1963 by Pinkus and Mehregan and termed epidermotropic eccrine carcinoma [3].

Eccrine porocarcinoma usually arises in elderly patients (typically in the sixth and seventh decades of life), affects men and women equally, and typically arises the extremities [3, 4]. Often, elderly patients do not seek medical treatment immediately for the condition [4, 5]. Eccrine porocarcinoma has potential to be locally aggressive and spread to both regional lymph nodes and distant organs [6]. The typical presentation is as a nodule associated with warty epidermal changes with or without ulceration [7].

The treatment of EPC is constantly evolving [8]. The first step is to take a surgical resection [9]; however, beyond that there is no definitive conclusion whether radiotherapy or chemotherapy is preferred for sweat gland carcinomas [10]. A variety of chemotherapeutic approaches have been used with varying degrees of responsiveness; however, there is little evidence to support an optimal chemotherapy regimen [11, 12]. Metastatic

EPC has demonstrated resistance to many cytotoxic agents [13, 14]. Some combination chemotherapy regimens have yielded short remission times; though, they often produce severe adverse effects [15].

Eccrine porocarcinoma is rare and metastatic presentation is even rarer. The purpose of this case report is to discuss a clinically challenging presentation of EPC and the imaging abnormalities associated with metastatic lesions.

CASE REPORT

A 62-year-old man presented with a history of a longstanding skin lesion on his distal right forearm. The patient reported the lesion persisting for at least 20 years; however, it started enlarging and becoming inflamed in the six months preceding his office visit. The patient described the lesion as originally "spongy" until it began to change in size and become tender. He had initially presented to his primary care physician who suspected an infected sebaceous cyst and subsequently attempted incision and drainage. During the attempted incision and drainage, a mass was noted, and punch biopsy was performed (Figure 1).

The biopsy specimen demonstrated extensively ulcerated, highly infiltrative carcinoma extending throughout the dermis into the subcutis involving the deep margin (Figure 2A and B). The tumor cells were positive for cytokeratin 5, cytokeratin 7, p63, and p40 (Figure 2C). They showed areas of ductular differentiation. They were negative for cytokeratin 20 and TTF-1. In a subsequent analysis, the biopsy specimen was found to be negative for estrogen, progesterone, and HER2. The immunostains showed positive EMA, focal positive S100, negative CEA, and negative Melan A (Figure 2D and E). Lymphovascular invasion was found to be present (Figure 2F).

After the biopsy results were obtained, a PET scan was ordered that demonstrated additional lesions (Figure 3A). The lymph nodes in the neck, mesenteric, retroperitoneal, and inguinal lymph nodes appeared to be clear and were neither enlarged nor hypermetabolic. Within the chest, hypermetabolic nodularity along the left hemidiaphragm was noted. Additionally, a mass was noted in the upper anterior thigh with a maximum standardized uptake value (SUV) of 17 (Figure 3B and C)—as well as a satellite intramuscular lesion laterally with a maximum SUV of 9. There was also a hypermetabolic intramuscular lesion posterior to the left scapula (Figure 3D and E). These results were overall consistent with metastatic disease involving the lungs, left hemidiaphragm, and intramuscular lesions in the left back and left thigh. Additionally, a lytic bone lesion was noted on the proximal left femur.

The patient was initiated on a chemotherapy treatment plan consisting of gemcitabine and protein-bound paclitaxel.

An MRI was performed six months later. This demonstrated a large necrotic mass in the vastus

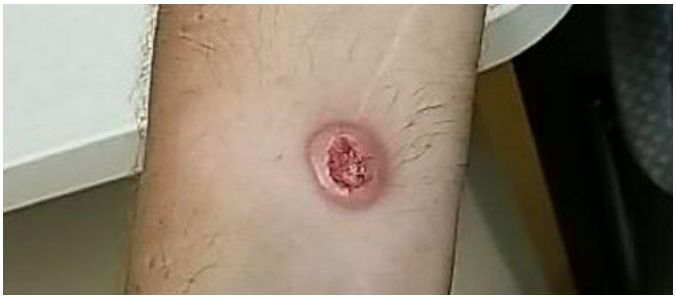

Figure 1: Photograph of patient's right distal volar arm with the presenting lesion subsequent to biopsy.

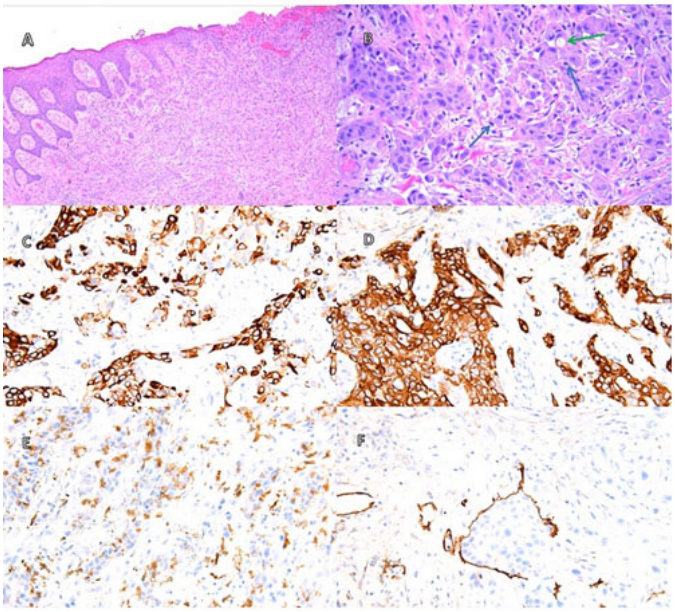

Figure 2: Photomicrographs of the biopsied arm tumor. (A) H&E (×40) showing ulcerative skin with diffuse cellular infiltrate. (B) H&E (×400) showing infiltrate of malignant cells arranged in sheets and nests. (Green arrow noting eccrine ductal differentiation. Blue arrows noting mitotic activity). (C) CK 5/6 (×200), (D) EMA (×200), (E) S100 (×200) with focally positive regions. (F) D2-40 (×200) showing lymphovascular invasion.

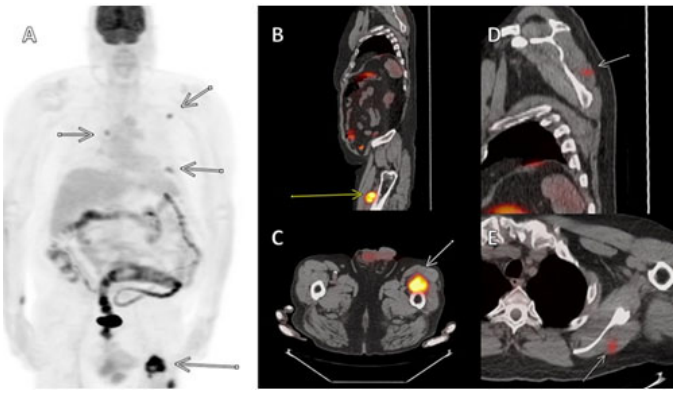

Figure 3: Positron emission tomography (PET) images demonstrating metastatic lesions on initial presentation. (A) PET scan showing at least four distinct metastatic lesions (arrows). (B and C) Sagittal and axial views of PET scan showing hypermetabolic mass in left upper anterior thigh (arrows). (D and E) Sagittal and axial views of PET scan showing hypermetabolic intramuscular lesion posterior to left scapula (arrows).

intermedius muscle with peripheral and thick septal enhancement on postcontrast sequence with a centrally necrotic component. A similar smaller necrotic mass was noted in the vastus lateralis muscle. On postcontrast axial sequence, multiple additional smaller enhancing lesions were seen in the vastus lateralis muscle. These multiple necrotic intramuscular lesions were suggestive of an increased degree of neoplastic involvement from the patient's scan six months prior. Additionally, diffuse intramuscular edema and hyperemia in the muscles of the anterior thigh were noted that could represent nonspecific myositis or changes of acute or subacute denervation.

A repeat PET scan was also performed that demonstrated the previously described lesions had a significantly decreased tracer uptake, which was suggestive of response to therapy. The lesion in the left thigh had enlarged, but it was characteristic of fluid density without metabolic activity. This scan was valuable in that it helped clarify the MRI findings. In isolation, the MRI results may be misleading as they suggest progression; however, that progression could now be more accurately characterized as necrosis—as the PET showed decreased metabolic activity. Together, these findings highlight the relative usefulness of PET in staging with imaging studies.

The original chemotherapeutic plan was ceased after seven months as the patient reported negative effects including nausea, anorexia, and weight loss. The patient was started on sunitinib as an alternative chemotherapeutic agent.

DISCUSSION

Eccrine porocarcinoma is a rare primary malignant adnexal tumor of the skin [3]. There are four primary adnexal structures of the skin from which adnexal tumors can arise: the hair follicle, sebaceous, eccrine, and apocrine glands. Eccrine porocarcinoma arises from the eccrine sweat glands, which open directly onto the surface of the skin and are widely distributed [3, 6].

Our patient presentation of an elderly adult having a skin lesion for many years that has recently begun to change is a typical presentation of EPC. In terms of clinical diagnosis, eccrine gland carcinomas have no distinctive clinical features—making diagnosis by gross appearance effectively impossible [6, 16]. As in the case of our patient, histology is typically necessary for accurate diagnosis [17].

The classic histological findings are acanthotic epithelial proliferation containing clear cell nests with radial extension of polygonal nuclei, eosinophilic cytoplasm, and ductal structures with intraepidermal atypia [1, 18]. The duct differentiation varies from intracytoplasmic lumina to mature ducts with eosinophilic cuticles [19]. The cells of excretory coil of eccrine sweat glands are typically positive for the expression of low molecular weight keratin, EMA, CEA, and S100 protein in the basal layer [20]. Ki-67 and p53 may be used to differentiate benign from malignant lesions [20]. Some eccrine carcinomas are positive to estrogen and progesterone receptors, which has clinical implications for potential hormonal therapy [20].

The differential diagnosis of EPC is vast and a range of pathologies from basal cell carcinoma to metastatic adenocarcinoma need to be considered [11]. The differential should include eccrine poroma, squamous cell carcinoma, malignant melanoma, and occult visceral neoplasms [2, 21]. The lesions of EPC are verrucous plaques or polypoid protrusions that mimic squamous cell carcinoma or Bowen's disease [22]. Both clonal seborrheic keratosis and Bowen's disease are negative for CEA, CK 7, and S100, which are often positive in EPC [23]. Melanoma in situ can be ruled out by negativity for Melan-A and HMB-45 [23]. The metastatic deposits from visceral and breast adenocarcinoma are microscopically indistinguishable from eccrine carcinoma and thus should be considered before diagnosing the lesions as metastatic presentation of sweat gland carcinoma [6].

Porocarcinomas present with two models of growth: horizontal and nodular [24]. There has been disagreement in the literature whether EPC can be described as aggressive with a high rate of recurrence and metastasis [25]. A large case series by Robson et al. (69 EPC cases) found metastasis and recurrence rates of less than 20% [19, 25], suggesting that metastasis and recurrence are exceptions—rather than the norm. In metastatic presentation, nodal involvement is the most frequent form, with cutaneous metastasis being more rare [7]. Pedunculated tumors appear to be less aggressive and less likely to recur than tumors that present with ulceration and multinodularity [26]. Other previously described prognostic factors include depth of invasion, number of mitoses, and growth patterns of neoplastic margin [25]. The mortality among patients with EPC with nodal metastasis can be as high as 67% [27].

There is consensus that the initial treatment for EPC is typically total surgical excision [28]. Local excision with broad margins has traditionally been considered the standard of care; however, Mohs micrographic surgery (MMS) has gained prominence [9, 28]. The routine dissection of regional lymph nodes is, however, controversial—though lymphadenectomy may be indicated for patients with nodes that are clinically enlarged [15, 29]. In chemotherapeutic treatment, the agents such as methotrexate, cisplatin, doxorubicin, bleomycin, isotretinoin, and interferon alpha have been used with partial or little response [30].

For our patient, the lymph nodes were clear on computed tomography (CT) scan. The presenting lesion on his arm was resected, and chemotherapy was undertaken for the metastatic lesions using combination therapy consisting of gemcitabine and protein-bound paclitaxel. While this therapy demonstrated initially promising results, the patient's reporting of unbearable side effects made the switch to sunitinib necessary.

CONCLUSION

Eccrine porocarcinoma is a rare skin malignancy that can be difficult to identify initially—with biopsy and immunohistology being key diagnostic tools. Eccrine porocarcinoma should be considered in the diagnosis of cutaneous malignant tumor.

REFERENCES

1. Aaribi I, Mohtaram A, Ben Ameur El Youbi M, et al. Successful management of metastatic eccrine porocarcinoma. Case Rep Oncol Med 2013;2013:282536.
2. Lloyd MS, El-Muttardi N, Robson A. Eccrine porocarcinoma: A case report and review of the literature. Can J Plast Surg 2003;11(3):153–6.
3. Khaja M, Ashraf U, Mehershahi S, Ayyadurai P, Malik S. Recurrent metastatic eccrine porocarcinoma: A case report and review of the literature. Am J Case Rep 2019;20:179–83.
4. Gonda K, Hatakeyama Y, Rokkaku Y. Eccrine porocarcinoma arising from the knee. Clin Case Rep 2018;6(2):350–2.
5. Jarocki C, Kozlow J, Harms PW, Schmidt BM. Eccrine porocarcinoma: Avoiding diagnostic delay. Foot Ankle Spec 2020;13(5):415–9.
6. Kshirsagar AY, Wader JV, Nagur B, et al. Case report: A rare case of eccrine carcinoma. Int J Surg Case Rep 2015;15:149–51.
7. Lehmann C, Rodriguez Ossa P, Vargas Manrique M, Acosta AE, Quintero Pérez Y. Eccrine porocarcinoma with zosteriform metastasis. Cureus 2020;12(2): e6873.
8. Denisova E, Westphal D, Surowy HM, et al. Whole-exome sequencing in eccrine porocarcinoma indicates promising therapeutic strategies. Cancer Gene Ther 2021.
9. Nazemi A, Higgins S, Swift R, In G, Miller K, Wysong A. Eccrine porocarcinoma: New insights and a systematic review of the literature. Dermatol Surg 2018;44(10):1247–61.
10. Shen J, Pan X, Lu Y, Pan D, Ma Y, Zhan R. A case of eccrine porocarcinoma characterized by a progressive increase in the level of Ki-67 index: Case report and review of literature. BMC Surg 2019;19(1):142.
11. Marone U, Caracò C, Anniciello AM, et al. Metastatic eccrine porocarcinoma: Report of a case and review of the literature. World J Surg Oncol 2011;9:32.
12. Fukui K, Fujioka M, Matsuo H, Noguchi M. Eccrine porocarcinoma on the lateral nose wall: A rare case report. Case Rep Dermatol 2019;11(2):215–9.
13. Westphal D, Garzarolli M, Sergon M, et al. High tumour mutational burden and EGFR/MAPK pathway activation are therapeutic targets in metastatic porocarcinoma. Br J Dermatol 2021;185(6):1186–99.
14. Comito F, Nigro MC, Sperandi F, Melotti B, Ardizzoni A. Immunotherapy-refractory, EGFR overexpressing metastatic porocarcinoma responding to cetuximab. Eur J Cancer 2021;146:84–6.
15. Fujimine-Sato A, Toyoshima M, Shigeta S, et al. Eccrine porocarcinoma of the vulva: A case report and review of the literature. J Med Case Rep 2016;10(1):319.
16. Das D, Datta C, Chatterjee U, De A. A case of eccrine porocarcinoma: A treacherous mimic and diagnostic challenge. Indian J Pathol Microbiol 2021;64(1):186–8.
17. Koh M, Telang G, Fonseca A, Ghanian S, Walker J. Clear cell differentiation in eccrine porocarcinoma as a high-risk feature: Epidemiologic and pathologic features of eccrine porocarcinoma in a single-center case series. Am J Dermatopathol 2021;43(9):647–52.
18. Salih AM, Kakamad FH, Baba HO, et al. Porocarcinoma; presentation and management, a meta-analysis of 453 cases. Ann Med Surg (Lond) 2017;20:74–9.
19. Robson A, Greene J, Ansari N, et al. Eccrine porocarcinoma (malignant eccrine poroma): A clinicopathologic study of 69 cases. Am J Surg Pathol 2001;25(6):710–20.
20. Idrissi Serhrouchni K, Harmouch T, Chbani L, et al. Eccrine carcinoma: A rare cutaneous neoplasm. Diagn Pathol 2013;8:15.
21. Llamas-Velasco M, Fernández-Flores A, Saeb-Lima M, Cassarino DS. Porocarcinoma with areas of mucinous differentiation suggesting multilineage differentiation. J Cutan Pathol 2021;48(1):90–4.
22. Kurisu Y, Tsuji M, Yasuda E, Shibayama Y. A case of eccrine porocarcinoma: Usefulness of immunostain for s-100 protein in the diagnoses of recurrent and metastatic dedifferentiated lesions. Ann Dermatol 2013;25(3):348–51.
23. Ramasenderan N, Shahir H, Omar SZ. A synchronous incidence of eccrine porocarcinoma of the forearm and facial squamous cell carcinoma: A case report. Int J Surg Case Rep 2018;42:116–20.
24. Urso C, Bondi R, Paglierani M, Salvadori A, Anichini C, Giannini A. Carcinomas of sweat glands: Report of 60 cases. Arch Pathol Lab Med 2001;125(4):498–505.
25. Kurashige Y, Minemura T, Nagatani T. Eccrine porocarcinoma: Clinical and pathological report of eight cases. Case Rep Dermatol 2013;5(3):259–66.
26. Salih AM, Kakamad FH, Essa RA, et al. Porocarcinoma: A systematic review of literature with a single case report. Int J Surg Case Rep 2017;30:13–6.
27. Thuruthil RR, Jayalakshmy PS, Sukumar V. A case of recurrent eccrine porocarcinoma with regional lymph nodal metastasis, arising on a Bowen's disease patch. Indian J Surg 2015;77(Suppl 1):182–4.
28. Rabi S, Syder N, Ragab O, In G, Hu JC. Rapidly progressive porocarcinoma of the ear and multidisciplinary approach to management. Dermatol Surg 2021;47(3):404–6.
29. Chow KL, Peters X, Mashbari H, Shokouh-Amiri M, Benjamin M, Warso M. A case report of malignant eccrine porocarcinoma involving the palm requiring surgical excision and free flap reconstruction. Am J Case Rep 2020;21:e925231.
30. Chang O, Elnawawi A, Rimpel B, Asarian A, Chaudhry N. Eccrine porocarcinoma of the lower extremity: A case report and review of literature. World J Surg Oncol 2011;9:94.

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Author Contributions

Bradley Kaptur – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Nicholas Peterman – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.