



A patient with porocarcinoma of the lower extremity and lung metastasis: a rare case report

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Abstract

Background. Porocarcinoma is a rare cancer of the developing sweat glands. It often occurs in older adults and frequently affects the head, neck, and extremities. We report a rare case of metastatic porocarcinoma with intriguing approach of the diagnosis and management due to limited resource setting.

Case Report. A 60-year-old man with a history of type 2 diabetes mellitus presented with nodules on the left foot with no palpable lymph node. A chest radiograph revealed multiple coin lesions and histopathological findings were suggestive for porocarcinoma. We assessed the case as a metastatic porocarcinoma in a patient with uncontrolled type 2 diabetes mellitus. Surgery was performed in the initial phase of treatment, consisting of below-knee amputation. The patient refused the planned chemotherapy in the referral hospital and then underwent other modalities of palliative care. He passed away eleven months after the initial diagnosis.

Conclusions. Metastatic porocarcinoma is a rare oncological case with a challenging approach of the diagnosis and management. High awareness of clinical clues in rare cancer is needed for early diagnosis and prompt treatment, especially in limited resource settings.

Keywords: porocarcinoma of lower extremity; lung metastasis; metastatic porocarcinoma; elderly patient

Introduction

Porocarcinoma is an extremely rare malignancy of the sweat glands that has a high rate of extracutaneous spread [1,2]. Its etiologies are not well-understood [1,3]. Clinical diagnosis is a challenge in porocarcinoma and histopathological examination is mandatory. Surgery is the mainstay treatment of porocarcinoma, but the treatment modalities of porocarcinoma with metastasis are limited.

To our knowledge, this is the first case report of porocarcinoma in Indonesia. This case report demonstrates the urgent need for meticulous clinical approach for its diagnosis and

management, specifically in hospitals with limited resources in developing countries, such as Indonesia.

Case report

We present a case of a 60-year-old man with a history of uncontrolled type 2 diabetes mellitus (T2DM) who was initially diagnosed with diabetic foot. He is a farmer. He was referred from a primary healthcare clinic with nodules merely on the left foot that had begun to develop 3 months before admission. No history of trauma or insect bites was recorded. No fever or weight loss were reported. There was no neurologic deficit. The ankle-brachial index was normal

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(1.1/1.0). The Eastern Cooperative Oncology Group (ECOG) performance status was 2 and the pain visual analog scale (VAS) was 3–4. The examination of the lower extremity revealed hyperemic exophytic nodules displaying an erosive surface on the left foot (Figure 1). No lymph nodes (clavicular, axillary, or inguinal) were palpable. The complete blood count demonstrated normal range (hemoglobin: 12.1 g%, hematocrit: 34%, WBC: 6,000/mm³, platelet: 251,000/mm³). Hyperglycemia at admission was documented (random blood glucose: 322 mg/dL). Blood chemistry showed normal range for hepatic enzymes (ALT: 33.3 mg/dL, AST: 32.9 mg/dL), kidney function test (urea: 26.7 mg/dL, creatinine: 0.96 mg/dL, eGFR CKD-EPI: 85.5mL/min/1.73m²), and coagulation panel test (PT: 14.3second, aPTT 42.1second).



Figure 1a.



Figure 1b.

Figure 1. Physical findings. **1a.** Plantar view; **1b.** Medial view.

The imaging tests were performed. A chest radiograph showed variable sizes of rounded nodules, scattered throughout both lungs (Figure 2). A radiograph of the left foot revealed a soft tissue mass with a normal bone structure (Figure 3). The abdominal ultrasound was normal and negative for metastatic lesions.



Figure 2. Chest radiograph showing multiple coin lesions in both lungs.



Figure 3. Foot radiograph showing soft tissue mass in left foot.

The complete excisional biopsy was obtained from the nodule in the left foot and fixation was carried out by using formalin solution. Embedding was performed in a paraffin embedding medium in order to carry out the sectioning process. The hematoxylin and eosin-stained sections were then examined. The histopathologic findings revealed a hyperplastic epidermis with some ulcerations (Figure 4a). Cell clusters were present under the epidermis with an irregular and invasive pattern, basaloid pattern (Figure 4b), and squamous-like differentiation (Figure 4c). Ductal differentiation varied from small intracytoplasmic lumens to mature, well-differentiated ducts lined with a thin-eosinophilic layer (Figure 4d). Numerous mitotic activities (>14/HPF) and cytological pleomorphisms were observed (Figure 4e). This pathology report was suggestive for porocarcinoma.

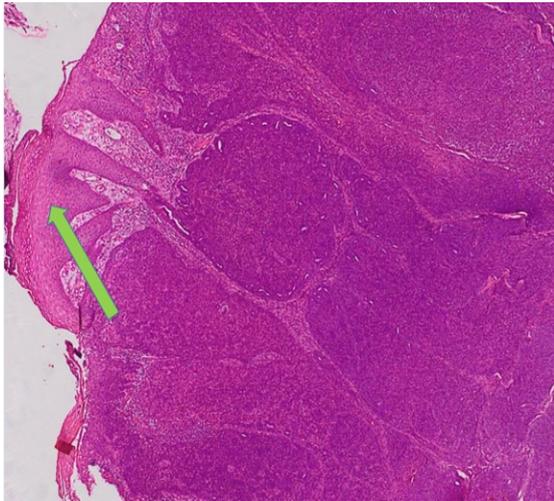


Figure 4a.

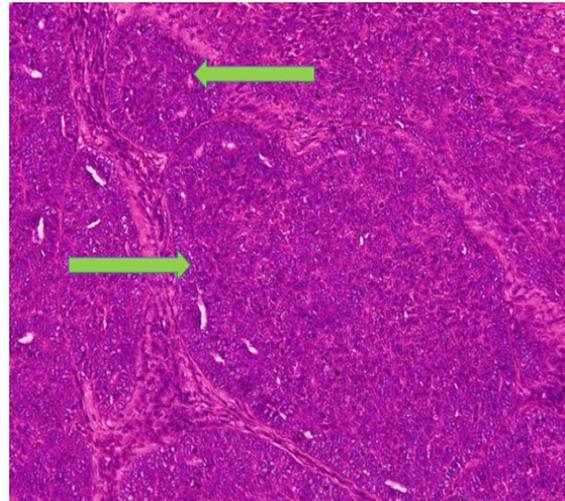


Figure 4b.

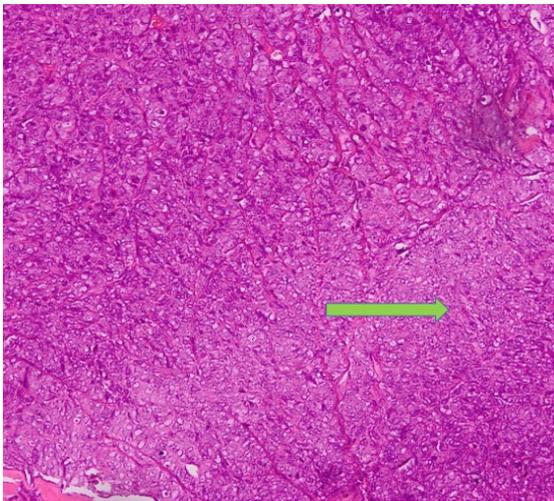


Figure 4c.

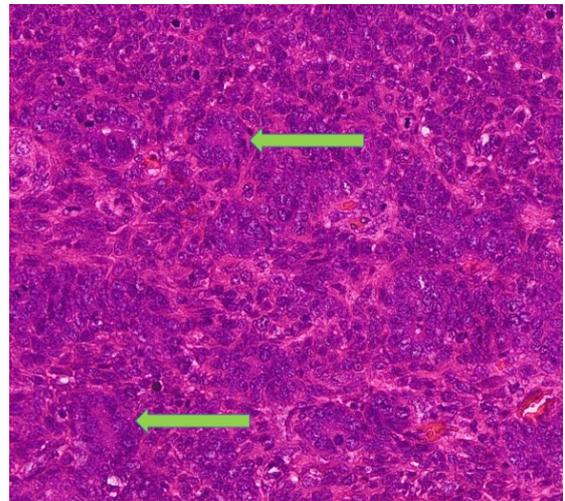


Figure 4d.

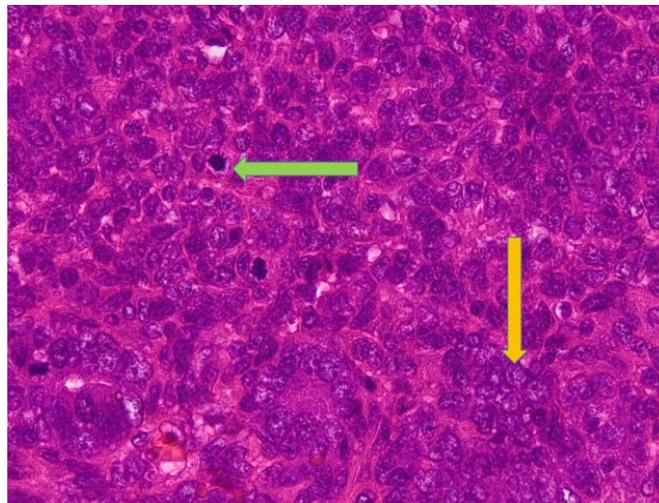


Figure 4e.

Figure 4. Histopathological findings. **4a.** Hyperplastic epidermis (light green arrow) with ulcerations (4×4); **4b.** Basaloid pattern (light green arrow) (10×4); **4c.** squamous-like differentiation (light green arrow) (10×4); **4d.** ductal differentiation (light green arrow) (10×4); **4e.** high mitotic activities (light green arrow) and pleomorphisms (orange arrow) (40×4).

The patient was diagnosed as having porocarcinoma with lung metastasis and the management planning encompassed surgery followed by systemic chemotherapy. The patient underwent below-knee transtibial amputation (Figure 5). Because the patient refused systemic chemotherapy, other modalities of palliative care were planned for further management of the case. However, the patient was lost to follow-up; approximately 11 months after his last hospitalization, the patient was found not breathing at home, taken to the emergency room, and pronounced dead.



Figure 5. After below-knee amputation.

Discussion

Porocarcinoma, also known as eccrine porocarcinoma, is a malignant tumor derived from the intraepidermal portion of the eccrine sweat duct. This type of rare malignant sweat gland tumor accounts for 0.01% of all cutaneous epithelial neoplasms. It occurs most often in older adults, with a mean age of 67 years old. It affects both sexes relatively equally. This case report presents an older male patient with porocarcinoma of the left lower extremity [2,4–7].

The macroscopic appearance of porocarcinoma is of a solitary tumor or nodule, typically with epidermal changes, ulceration, or both [5,7]. Other clinical forms may present as exophytic tumors. Most cases grow slowly or remain stable for several years without symptoms, which may cause delayed diagnosis. The primary sites of the lesions are the head and neck, followed by the lower extremities; the risk factors of porocarcinoma are not well understood [1]. Azadeh et al. reported that more than one-fifth of patients with porocarcinoma had metastatic disease at the time of diagnosis [2]. These cases included regional lymph node metastasis, distant metastasis and locoregional cutaneous metastasis.² Furthermore, Abdulwahid et al. demonstrated that the frequent distant metastasis sites were the lungs, liver and brain [1]. In this case, the physical findings revealed hyperemic exophytic nodules with eroded surfaces on the left foot, along with lung metastasis at the time of diagnosis, with no lymph node involvement.

Some considerable possible risk factors in this case were sunlight exposure due to his occupation and T2DM. Although it is still controversial, some reports notified that the sunlight exposure may play role as a predisposing factor for porocarcinoma [8,9]. Further, Hui-Wen Tseng et al. demonstrated that T2DM is associated with increased incidence rate (incidence rate ratio= 1.44, $p= 0.02$) and risk of developing overall skin cancer (adjusted hazard ratio= 1.46, $p= 0.01$) [10]. However, a direct association between T2DM and porocarcinoma has yet to be discovered.

Porocarcinoma has three patterns of histopathologic features. The first pattern involves pleomorphic cells with nuclear hyperchromasia and important mitotic activity surrounded by a ductal lumen [11]. The invasive porocarcinoma reveals extensive growth of cords or bands of polygonal tumorous cells with pleomorphic nuclei proliferating asymmetrically inside the dermis, with many mitotic figures and necrotic areas with ductal structures (ductal differentiation). Although it is rare, the porocarcinoma may invade lymphatic vessels and reinvade the epidermis, resulting in cutaneous metastasis [11]. The second pattern is the comedonecrosis pattern, consisting of tumor cell islets that contain central areas of dead cells surrounded by an inflammatory reaction. Squamous differentiation is rarely observed. The third pattern is the Bowenoid pattern, which is formed by pleomorphic cells with prominent nuclei and some multinucleated monster cells [11]. Robson et al. emphasized the role of histopathologic parameters in determining prognosis. Mortality was predicted independently by high mitotic activity (HR 17.0, 95% CI 2.71–107), lymphovascular invasion by tumors (HR 4.41; 95% CI 1.13–17.2), and a depth of >7 mm (HR 5.49, 95% CI 1.0–30.3) [12]. The histopathologic features in this case were a hyperplastic epidermis, ulceration, irregular and invasive patterns, basaloid and squamous-like differentiation, ductal differentiation, pleomorphisms, and high mitotic activity, suggesting a pattern of invasive porocarcinoma. This case had a poor prognosis because of high mitotic activity, an independent risk factor for mortality.

The diagnosis of this case was porocarcinoma with lung involvement as a distant site of metastasis. Surgery is the main modality for the management of porocarcinoma, although the management of porocarcinoma with distant metastasis has not been standardized [2]. Some therapeutic options have been reported with variable outcomes. Godilot et al. have stated that the overall survival of these patients is not well characterized, but it is probably poor [13]. The mortality rate is up to 80% in the presence of metastasis [14]. Salih et al. reported a patient with metastatic porocarcinoma who deteriorated and died after 2 months of chemotherapy and radiation therapy [4]. Imane et al. reported a patient with metastatic porocarcinoma who showed a complete response in the lung and regression of the breast mass with three cycles of cisplatin and

5-fluorouracil, but the authors did not state any information about progression-free survival (PFS) [15]. Godillot et al. reported a case of metastatic porocarcinoma in which the patient received paclitaxel and cetuximab with 6 months of PFS [13]. Azadeh et al. reported that among patients treated with chemotherapy, some have lymph node metastasis after an average of 13 months, and some experienced distant metastasis after an average of 7.5 months [2]. Rao et al. reported a patient with metastatic porocarcinoma without systemic chemotherapy who had one year of PFS [16]. Metastatic porocarcinoma seems to be chemoresistant [13,16]. Our patient underwent below-knee transtibial amputation and palliative care. Chemotherapy was not performed in this case because of the patient's refusal.

Conclusions

Porocarcinoma is a rare disease entity whose diagnosis and management are challenging, especially in developing countries. It should be suspected in patients with solitary or multiple tumors or nodules surrounded by epidermal changes, ulceration, or both, and in those with exophytic tumors. Histopathology findings are the gold standard in establishing the diagnosis and are associated with prognosis. Surgery is the primary treatment but a standardized management strategy for metastatic porocarcinoma has not yet been developed.

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