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Case Report

Eccrine Porocarcinoma of the Young Adult: Case Report and Literature Review

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Abstract: Eccrine porcarcinoma is an extremely rare malignancy of the skin with few well documented cases reported in literature. It is a malignant cutaneous adnexal neoplasm originating from the intraepidermal ductal portion of eccrine sweat glands. This tumor predominantly affects elderly patients, with a high potential for locoregional metastasis, significant morbidity, and mortality. We present a rare case of porocarcinoma affecting a 22-year-old patient on the lower limb, without extension. The initial tumor was biopsied, excised and diagnosed as an eccrine porocarcinoma. Reconstruction of the heel was achieved with a medial plantar flap.

Keywords: Porocarcinoma, Young Adult, Surgery, Medial Plantar Flap, Guidelines.

INTRODUCTION

Porocarcinoma is a rare malignant tumor that grows in the intraepidermal portion of the duct of eccrine glands, known as acrosyringium. It was first described by Pinkus and Mehregan in 1963 as the malignant counterpart of poroma [1]. This rare tumor generally affects the elderly, with peak incidence in the seventh and eighth decades of life and is rarely described in young adults. We report a never described case of eccrine porocarcinoma of the foot in a 22 year old patient.

CASE PRESENTATION

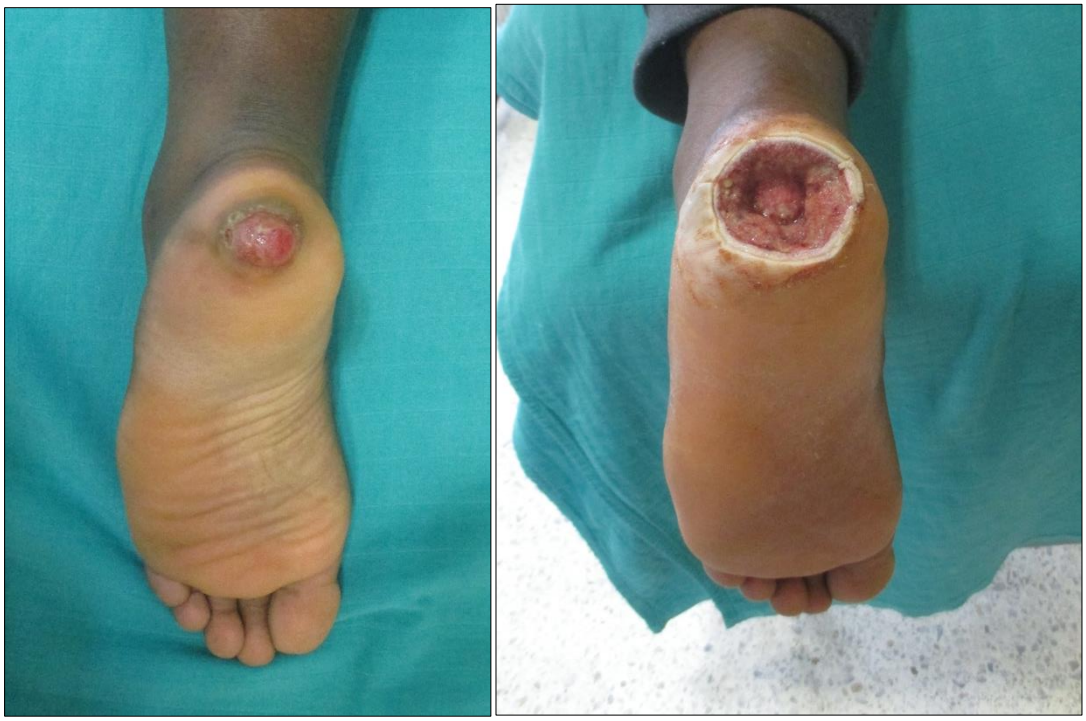
A 22-Year-old male, with no medical history, presented to our department with an eight months history of an enlarging mass over his left heel. The tumor, located on the bearing zone of the heel, made it painful to walk and difficult for the patient to wear shoes.

Clinical examination found an ulcerated reddish nodule of the left heel, with slight serous discharge, measuring 3 x 2,5 cm. There were no other lesions and no palpable regional lymphadenopathy.

A biopsy was conducted, concluding at first in favor of an anexial tumor whose benign or malignant nature could not be determined. A re-reading of the biopsy specimen was requested, concluding in favor of a porocarcinoma. Laboratory investigations were non-contributory.

A CT scan of the foot showed no extension to the bone. Locoregional extension of the tumor was assessed with inguinal ultrasound and computed tomographic thoraco-abdomino-pelvic scan. No other lesion was identified, neither pathologic inguinal lymph node.

Under local anesthesia the patient underwent wide excision of the tumor with 1cm lateral margin and deep margin including the periosteum.



A **B**

Fig. 1: A/ Initial tumor of the heel B / Excision of the tumor with 1cm margin

Following histopathological results confirming complete tumor resection with clear margins, reconstruction of the tissue defect was performed using a medial plantar flap under general anesthesia. Harvested from a non-weight-bearing area, the flap provided our young patient with a sensitive, durable, and aesthetically pleasing coverage of the heel, enabling both walking and normal footwear.

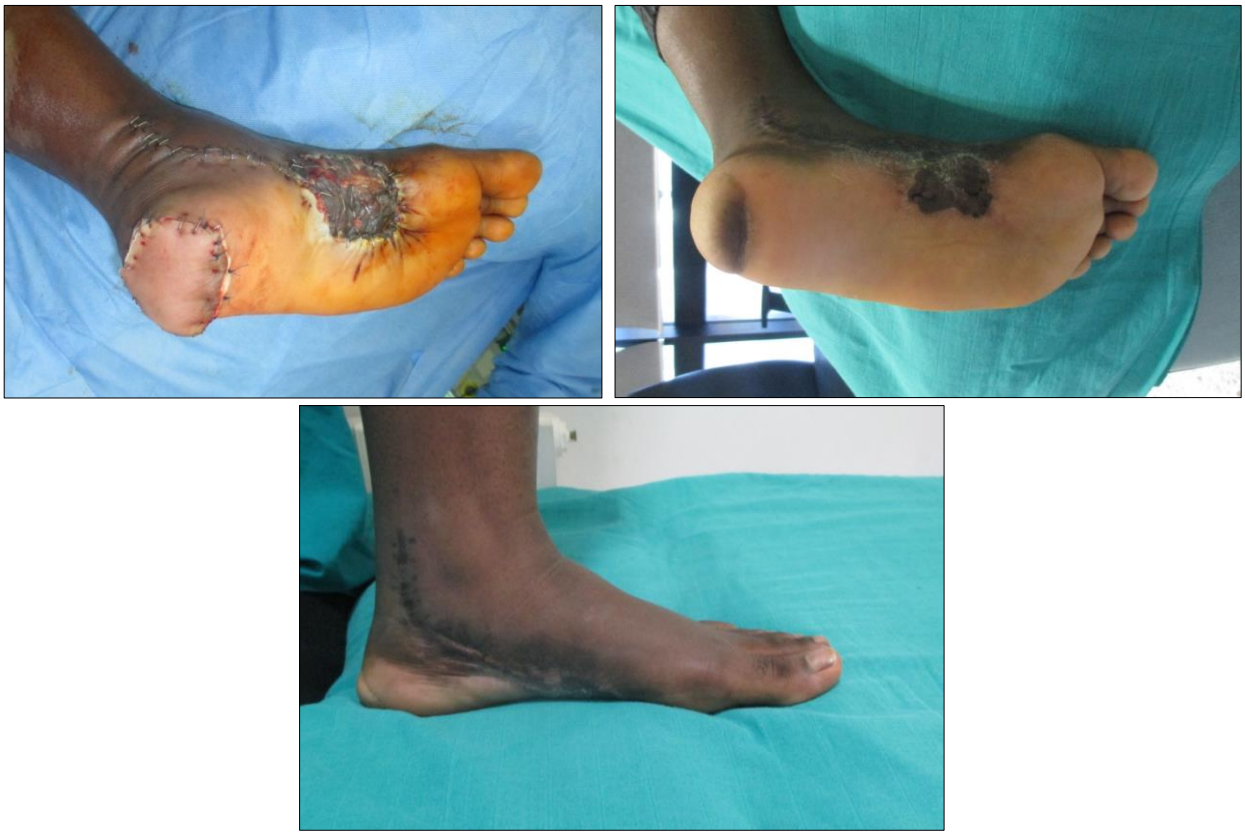


Fig. 2: Medial plantar flap, photograph on postoperative day 5 and after 18 months

There was no evidence of recurrence at 2 years of follow-up, and the patient continues to be followed up regularly.

DISCUSSION

Porocarcinoma is a rare malignant tumor of sweat gland origin, whose incidence and prevalence in the general population are not well established. However, it is known to be more common in the elderly, with peak incidence in the seventh and eighth decades of life, with a slight male predominance [2]. Cases reported in young adults are exceptional. To the best of our knowledge, there have been fewer than 10 cases seen in younger age group reported previously in literature. Only 3 cases have been described over the last 10 years: a case of pororcarcinoma of the scalp was reported in a 22-year-old woman in 2015 [3], and a 29-year-old woman in 2016 [4]. And a rare case of porocarcinoma of the abdominal wall was reported in a 21-year-old patient in 2016 [5].

Given its multiple clinical mimics, especially within the young age group, porocarcinoma is prone to misdiagnosis. It is often misdiagnosed as a squamous cell carcinoma. The rarity of malignant cutaneous adnexal tumors makes the diagnosis even more challenging for pathologist. The rate of misdiagnosis is around 37 to 50% of cases analyzed by pathologists in primary care clinics [6]. For our patient, after a first non-conclusive histopathologic diagnosis, we requested a rereading from a different anatomopathology laboratory. Then after wide resection of the tumor, the diagnosis was confirmed again by a third and different pathologist. Therefore, surgeons should proactively seek additional histopathological reviews of the pathology slides as needed to ensure an accurate diagnosis and provide the patient with the most effective and, whenever feasible, curative treatment strategy. Indeed, confirmation is essential for the prognosis. It is an aggressive tumor with a high recurrence rate and a tendency to metastasize to regional lymph nodes. Once metastasis has occurred, mortality rate increases to 75%-80% and thus survival is dependent on adequate diagnosis and timely wide resection of the lesion.

Porocarcinoma most commonly arises on the lower limbs (33%) as for our patient and on the head and neck (32%), but can can also be found in the trunk (14,7%), genitalia/buttocks (11,2%), and upper limbs (7,4%), including the nail bed. Porocarcinoma can arise spontaneously but is frequently associated with a preexisting benign eccrine poroma [7]. Several case reports and case series have described the development of porocarcinoma in association with other conditions such as Bowen's disease, squamous cell carcinoma, and hidradenitis suppurativa.

As of today, there is no standard guideline for porocarcinoma treatment. It primarily relies on surgery for localized forms and can be curative if the tumor is completely resected [2]. The recommended surgical margins for porocarcinoma vary significantly across studies. Some authors have recommended 2 cm margins, similar to those used for melanoma [7, 8]. More recently, Mohs micrographic surgery is emerging as an effective treatment method particularly for lesions on the limbs or face, requiring a more conservative resection. When the disease is inoperable or advanced, systemic treatment represents a challenge. Various therapeutic schemes have been evaluated but neither chemotherapy nor radiotherapy have shown real clinical benefits in metastatic stage of the disease.

Moreover, porocarcinoma has a poor prognosis, with a 35% recurrence rate even following surgery with clear margins [8]. This recurrence rate is among the highest for cutaneous neoplasms, exceeding those of dermatofibrosarcoma and melanoma.

CONCLUSION

Porocarcinoma is a rare, aggressive, and radio and chemoresistant skin cancer arising from eccrine duct called the acrosyringium. Although a rare entity, it should be considered in the differential diagnosis of cutaneous tumors among younger patients. Accurate histopathology diagnosis is often delayed, and optimal management is lacking. Surgery is the first and only therapeutic option that could significantly affect patient's outcome.

Given the lack of standardized protocols, and the rarity of this tumor, further studies, including case reports, would help improve the understanding of porocarcinoma.

Competing Interests: None.

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