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CASE REPORT

Eccrine porocarcinoma of the scalp: diagnosis and importance of early surgical intervention

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Abstract

Eccrine porocarcinoma is a rare and potentially fatal malignant adnexal tumor that can arise de novo or develop from its benign precursor, poroma. Diagnosis is difficult due to its rarity and resemblance to many other (benign) skin tumors. We present the case of a 71-year-old woman presenting with a long-standing case of eccrine porocarcinoma on the scalp. After multiple incomplete excisions over the last 15 years, no metastases were found. Major reconstructive surgery was necessary after complete excision of the lesion. Complete removal of the eccrine porocarcinoma was achieved in 2 stages. The defect was closed with a large rotation flap of the scalp. No adjuvant radio- or chemotherapy was given. Twenty-one months later, the patient presented with a rapidly growing lymph node recurrence, which was treated by lymph node dissection followed by chemoradiation. This case demonstrates the consequences of suboptimal surgical treatments and follow-up of eccrine porocarcinoma and its benign precursor, poroma. Early recognition, proper pathological diagnosis, and adequate surgical treatment are highly recommended in order to obtain a good prognostic outcome.

Level of evidence: Level V, Risk/Prognostic study.

Keywords Poroma · Eccrine porocarcinoma · Histopathology · Immunohistochemistry · Mohs micrographic surgery · Plastic surgery

Introduction

Eccrine porocarcinoma (EP) is a rare malignant neoplasm arising from the intraepidermal ductal portion of the eccrine sweat glands [1]. Porocarcinomas represent only 0.005 to 0.01% of all malignant epithelial neoplasms [2, 3]. This malignant tumor can arise de novo or evolve from a pre-existing benign eccrine poroma [4], with malignant transformation reported in 18 to 50% [5, 6]. EP is usually characterized by a naturally slow growth pace (average of 4 years to clinical presentation) but has the potential to develop rapidly, over a few weeks to months, in certain cases [7]. Although excision is curative for benign eccrine poromas [8], 20% of porocarcinomas recur and about 20% metastasize [3, 9]. Aggressive treatment of benign poromas is therefore advised, in order to prevent malignant conversion [1]. Female predominance had been suggested; however, recent literature failed to show a significant difference in incidence between genders [1–3, 5, 10]. The mean age at diagnosis is approximately 65, with the vast majority of patients being 50 to 80 years old [1–3, 5]. The primary sites of porocarcinomas are the head and neck, as well as the lower extremities [1, 3, 5]. Primary tumor

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location was significantly correlated with the presence of lymph node and distant metastases, with genitalia and buttocks having the highest risk and the head/neck region having the lowest risk [3]. Previously reported case series suggest immune compromise, sun exposure, and radiation therapy as risk factors for the development of porocarcinoma; however, the precise etiology of this malignancy remains unknown [1, 8]. Various clinical appearances have been described: subcutaneous masses, ulcerated nodules, plaques, polypoid, or verrucous papules [1, 9]. Recurrence, spontaneous bleeding, ulceration, sudden itching, pain, or accelerated growth is alarming for possible malignant transformation of a pre-existing poroma [8]. Tissue biopsy followed by histological examination in combination with immunohistochemistry is necessary for (differential) diagnosis. The main morphological features are mature duct formation, intracytoplasmic lumina, comedonecrosis, a bowenoid pattern, and clear cell change [3, 5]. Epithelial membrane antigen (EMA), periodic acid-Shiff (PAS), cytokeratin (CK) AE1/AE3/7/19, CD117, and carcinoembryonic antigen (CEA) are the most important immunohistochemical markers, reaching up to 100% sensitivity, although with variable (and mostly poor) specificity [3, 11–13]. Histologically, the most important prognostic factors are lymphovascular invasion, a high number of mitoses (> 14 mitoses per 10 high power fields), increased tumor depth (≥ 7 mm), and the presence of infiltrative or pagetoid tumor borders [5, 14]. An overview of the features of EP is presented in Table 1. Despite wide local excision (WLE) with broad tumor margins (10–20 mm), being the most widely used treatment for primary tumors, modified Mohs micrographic surgery (MMS) is gaining popularity due to the superior results demonstrated in recent literature [1, 3, 9, 14]. Furthermore, surgical margins should be at least 3 mm with excision and should be extended to > 5 mm with MMS, if an infiltrative or pagetoid subtype is seen [14]. In the case of recurrence and/or metastasis, adjuvant chemo- and or radiotherapy are necessary [3, 9, 15]. We present the case of a female patient that had to undergo major reconstructive surgery due to inadequate surgical treatment and follow-up of the scalp tumor.

Case

A 71-year-old Caucasian woman presented with a 15-year history of (incomplete) excisions of a relapsing exophytic tumor of the scalp. The first excision took place in 2003; the excised specimen was described by the pathologist as a hidradenoma of the scalp. The patient was reassured of the benign aspect of this lesion. Due to local relapse of the lesion, a similar local excision was performed 2 years later. The last surgery occurred in 2011, with closure of the defect using a split-thickness skin graft. The patient did not show for follow-up. In 2016, she was referred by her general practitioner to the dermatology department of the Antwerp University Hospital (UZA) due to recidivating lesions and ulcerations in the grafted area of the scalp. The past medical history of the patient is unremarkable. There has been little to zero sun exposure of the skin in the past. Her family history of malignancy was positive only for her brother, who suffered from lung cancer. The patient denied any constitutional symptoms (fever, malaise, night sweats, fatigue, or weight loss). There were no complaints of pain, pruritus, or bleeding concerning the scalp lesions. Clinical examination showed a large crusting and ulcerating tumor located in the left parietal region of the scalp (Fig. 1a). Physical examination showed no occipital, pre-auricular, or cervical lymphadenopathy. A 4-mm punch biopsy of the lesion was sent for histopathological evaluation. Furthermore, older histological sections from previous excisions were requested and compared. Microscopy showed a tumoral lesion with comparable morphological characteristics as the adnexal tumor that was incompletely removed years ago. The pathologist now described an adnexal tumor consisting of large tumor nests (Fig. 2a). In some nests, we recognized duct formation. The cells within the nests were round, partly with an eosinophilic and partly with a clearer cytoplasm. The nuclei were slightly enlarged. There was a striking mitotic activity. In some cell nests, there was central comedo-type necrosis and the nests were surrounded by dense eosinophilic stroma. A significant intraepithelial component was described (Fig. 2b). It consisted of large tumor cells located in the squamous epithelium, especially at the level of the hair follicle infundibulum. Those intraepithelial cell nests also showed necrosis. Carcinoembryonic antigen (CEA), keratine-7, and epithelial membrane antigen (EMA) staining were positive in the duct formations, but not in the tumor cells (Fig. 2c). Cervical ultrasound showed some lymph nodes with limited cortical thickening, but no convincing adenopathy. CT scan of the brain showed an extracranial soft tissue mass with exophytic components, suspicious for recurrence. However, no destruction of the bone, suspicious lymph nodes, or intracranial lesions was seen. PET scan showed an intense heterogenous annular cutaneous hearth located in the left parietal region. No evidence for metastasis was found. The case was presented on the dermatological multidisciplinary oncological consultation. In this meeting, as wide as possible resection of the porocarcinoma by the Department of Plastic Surgery was decided to be the preferred therapy, if necessary to be followed by radio- or chemotherapy. The patient was informed that after the large excision, a reconstruction would be required to close the defect. Multiple valid anterior scalp reconstruction options exist, such as a galeal rotation flap or a more advanced island flap based on branches from the occipital or temporal artery [16]. During the operation, the lesion was macroscopically excised with dimensions of 13 × 8 × 0.6 cm and sent for pathology (Fig. 1b). The defect was closed using the galeal scalp flap and the donor site was closed with a standard split-thickness skin graft from the left upper thigh. Microscopically, surgical margins were positive at the medial excision edge. Therefore re-excision was performed 1 month later with

Table 1 Features of eccrine porocarcinoma

Origin [1]	De novo or from pre-existing poroma
Mean age [3]	± 65 years
Sex ratio (male:female) [3]	± 1:1
Clinical features [1, 3, 5, 8, 9]	
Morphology	Mass and nodule (71.2%)
	Ulcer (18.3%)
	Plaque (9.8%)
	Swelling (1.3%)
	Papule (0.6%)
Characteristics	Spontaneous bleeding
	Ulceration
	Recurrence of previously removed lesion
	Sudden itching
	Accelerated growth
Location	Head and neck (40–44%)
	Lower extremities (24–34%)
	Upper extremities (9–24%)
Color	Violaceous
	Erythematous
	Reddish
	Pink
	Skin color
Size	Usually < 2 cm
Histologic characteristics [3, 5]	Mature duct formation
	Intracytoplasmic lumina
	Comedonecrosis
	Bowenoid pattern
	Clear cell change
Immunohistochemical markers, sensitivity [3]	Epithelial membrane antigen (EMA), 100%
	Periodic acid-shiff 9 (PAS), 100%
	Cytokeratin (CK) AE1/AE3/7/19, 100%
	Proto-oncogene c-Kit (CD117), 100%
	Carcinoembryonic antigen (CEA), 95%

± approximately

dimensions of 9.5 × 1.5 × 0.5 cm, showing no residual microscopical tumor lesion. The patient did not receive any radio- or chemotherapy afterwards. Twenty-one months later, the patient presented with a rapidly growing cervical mass. There were no distant metastases or local recurrence (Fig. 3). Cervical lymph node dissection showed 49 lymph nodes of which five were invaded by tumor with extracapsular spread in one. Adjuvant chemoradiation (70 Gy in 33 fraction) with weekly carboplatin at an area under the curve of 1.5 and paclitaxel 70 mg/m^2.

Discussion

In hindsight, our case represents a rather "typical" presentation of EP that occurred on the scalp of an elderly woman. Nevertheless, due to its rarity and similarities with many other benign conditions (seborrheic keratosis, verucca vulgaris, pyogenic granuloma), recognizing, diagnosing, and treating a EP (or its benign precursor) adequately is not that self-evident [17]. The first important aspect we would like to address is reducing the chances of developing an EP. It has been advocated that most of EP arise from a long-standing eccrine poroma [18]. Therefore, early complete excision of the latter should be considered [2]. Although recurrence of poromas rarely occurs, follow-up and patient education about recurrence and/or deterioration of the lesion is required [8]. In contrast to its benign precursor, EP has a recurrence rate up to 20%, with metastasis seen even after complete excision (probably due to micro metastasis at the time of surgery), and therefore requires intensive follow-up [5, 8, 12].

The second aspect we want to address is the importance of adequate diagnosis. The clinical appearance is extremely variable

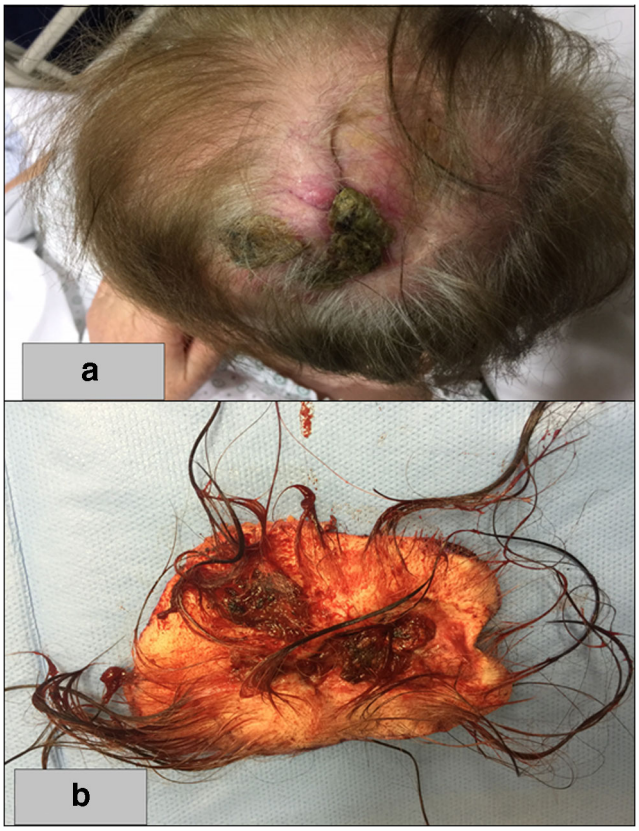

Fig. 1 **a** Top view of patient scalp, preoperative state: large crusting, ulcerating tumor of the scalp. **b** Resection specimen

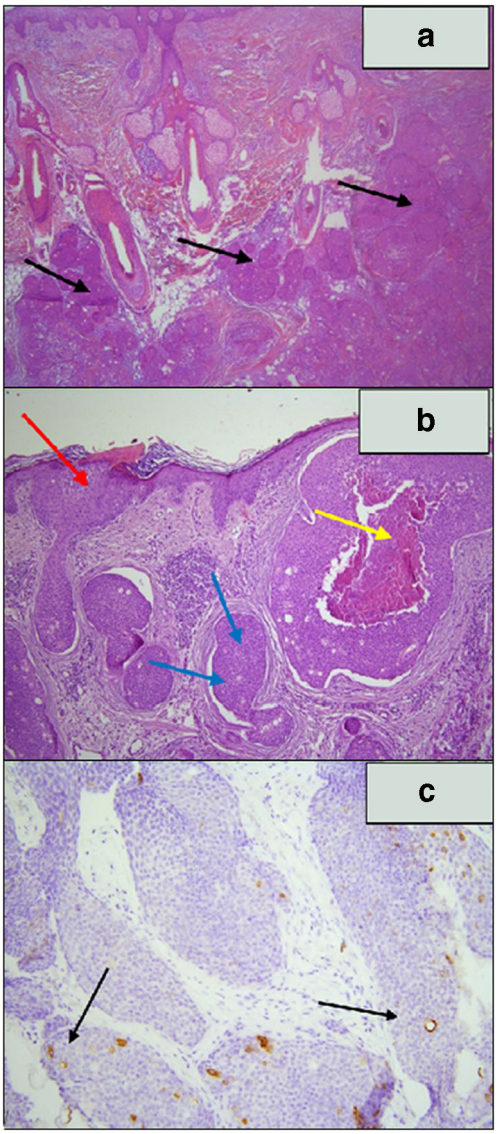

Fig. 2 **a**–**c** Microscopic examination. **a** H&E (Hematoxylin and eosin), × 40: skin specimen overview with epidermis, dermis, and subcutis. There is extensive invasion of the dermis and subcutis from large tumor nests (black arrows). **b** H&E, ×100x: the tumor cells are round with eosinophilic and focally pale cytoplasma. In some nests, there is central comedo-type necrosis (yellow arrow). There is ductal differentiation (blue arrow). There is also a in situ component (red arrow). **c** EMA (epithelial membrane antigen), DAB (3,3′-Diaminobenzidine), × 200: there is obvious duct formation. The duct-cuticle stains strong by immunhistochemistry with EMA (arrow)

and the contribution of dermoscopy is limited. At the slightest suspicion of EP, an excisional biopsy should be performed. Since histological features alone can lead to misidentification of the lesion (e.g., the close histologic resemblance to squamous cell carcinoma), the role of immunohistochemical staining is crucial [3, 19]. Distinguishing the appendageal nature of the tumor is done by staining for low molecular weight keratins such as CK7/19, EMA, and CEA. The latter in combination with the typical morphological features of EP should be sufficient to ensure diagnosis [3, 13]. When, eventually, the diagnosis of EP is made, MMS should be performed in order to guarantee a > 3 mm or > 5 mm (if an infiltrative or pagetoid subtype is observed) tumor-free resection margin. With recent literature showing far fewer cases of recurrence and/or metastasis after treatment with MSS in comparison with WLE, MSS should be the preferred initial therapy from here on out.

Benign poromas are most often located in the acral regions (palms and soles), where the highest concentration of eccrine sweat glands is found. It is therefore worth noticing that most eccrine porocarcinomas develop in the lower extremities or the head/neck area. Nazemi et al. recently investigated the importance of primary tumor location of EP, showing that the more "atypical" primary tumor locations (e.g., genitalia/trunk), had higher risk of metastasis. This combined with the histological high-risk features should be of great value in diagnostic (e.g., imaging for staging) and therapeutic decision-making. However, due to the lack of prospective studies and overall scarcity of data, only limited information is available about optimal treatment and follow-up of eccrine porocarcinomas [12]. Considering the relatively small number of cases, studied in case series and systematic reviews, the current conclusions should be regularly reassessed. The importance of basic knowledge of this disease for physicians must be emphasized. Considering EP in the differential diagnosis of not-so-obvious skin lesions,

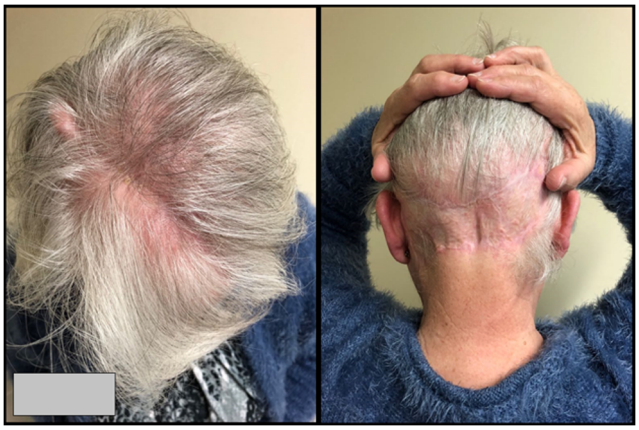

Fig. 3 Status 1 year post-operative

especially when recurrence, spontaneous bleeding, ulceration, or accelerated growth is noted, is vital.

Conflict of interest Maxime De Fré, Katrien Smets, Michal Ulicki, Veronique Verhoeven, Vasiliki Siozopoulou, Tine Strobbe, Specenier Pol, Olivier Aerts, Julien Lambert, Thierry Tondu, and Filip E. F. Thiessen declare that they have no conflict of interest.

Ethical approval All applicable international, national, and/or institutional guidelines were followed.

Informed consent Informed consent was obtained from all individual participants included in the study.

Patient consent Patient consent was obtained for publication of this manuscript and complementary images.

References

1. Salih AM, Kakamad FH, Baba HO et al (2017) Porocarcinoma; presentation and management, a meta-analysis of 453 cases. Ann Med Surg 20:74–79 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/28721214
2. Riera-Leal L, Guevara-Gutiérrez E, Barrientos-García JG, Madrigal-Kasem R, Briseño-Rodríguez G, Tlacuilo-Parra A (2015) Eccrine porocarcinoma: epidemiologic and histopathologic characteristics. Int J Dermatol 54:580–586
3. Nazemi A, Higgins S, Swift R, et al. (2018) Eccrine Porocarcinoma. Dermatologic Surg. [internet]. [cited 2018 Sep 18];1. Available from: http://www.ncbi.nlm.nih.gov/pubmed/29894433
4. Brown CW Jr, Dy LC (2008) Eccrine porocarcinoma. Dermatol Ther 21:433–438 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/19076620
5. Robson A, Greene J, Ansari N, Kim B, Seed PT, McKee PH, Calonje E (2001) Eccrine porocarcinoma (malignant eccrine poroma): a clinicopathologic study of 69 cases. Am J Surg Pathol 25:710–720 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/11395548
6. Shaw M, McKee PH, Lowe D et al (1982) Malignant eccrine poroma: a study of twenty-seven cases. Br J Dermatol 107:675–680
7. Lloyd MS, El-Muttardi N, Robson A (2003) Eccrine porocarcinoma: a case report and review of the literature. Can J Plast Surg 11:153–156 [cited 2019 Jun 3] Available from: http://www.ncbi.nlm.nih.gov/pubmed/24115860
8. Sawaya JL, Khachemoune A (2014) Poroma: a review of eccrine, apocrine, and malignant forms. Int J Dermatol 53:1053–1061 [cited 2018 May 19] Available from: http://doi.wiley.com/10.1111/ijd.12448
9. Song SS, Wu Lee W, Hamman MS et al (2015) Mohs micrographic surgery for eccrine porocarcinoma. Dermatologic Surg 41:301–306 [cited 2018 may 19] Available from: http://content.wkhealth.com/linkback/openurl?sid=WKPTLP:landingpage&an=00042728-201503000-00001
10. Salih AM, Kakamad FH, Essa RA et al (2017) Porocarcinoma: a systematic review of literature with a single case report. Int J Surg Case Rep 30:13–16 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/27898349
11. Ramasenderan N, Shahir H, Omar SZ (2018) A synchronous incidence of eccrine porocarcinoma of the forearm and facial squamous cell carcinoma: a case report. Int J Surg Case Rep 42:116–120 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/29245095
12. Horwich MD, Finch J, Ibrahimi O et al (2017) Eosinophilic variant of eccrine porocarcinoma of the scalp: case report and review of the literature. Int J women's dermatology 3:157–160 [cited 2018 May 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/28831427
13. Goto K, Takai T, Fukumoto T, Anan T, Kimura T, Ansai SI, Oshitani Y, Murata Y, Sakuma T, Hirose T (2016) CD117 (KIT) is a useful immunohistochemical marker for differentiating porocarcinoma from squamous cell carcinoma. J Cutan Pathol 43:219–226 [cited 2018 Oct 19] Available from: http://www.ncbi.nlm.nih.gov/pubmed/26449497
14. Belin E, Ezzedine K, Stanislas S et al (2011) Factors in the surgical management of primary eccrine porocarcinoma: prognostic histological factors can guide the surgical procedure. Br J Dermatol 165:985–989 [cited 2018 May 19] Available from: http://doi.wiley.com/10.1111/j.1365-2133.2011.10486.x
15. Kurashige Y, Minemura T, Nagatani T (2013) Eccrine porocarcinoma: clinical and pathological report of eight cases. Case Rep Dermatol 5:259–266 [cited 2018 Sep 22] Available from: http://www.ncbi.nlm.nih.gov/pubmed/24403888
16. Tenna S, Brunetti B, Aveta A, Poccia I, Persichetti P (2013) Scalp reconstruction with superficial temporal artery island flap: clinical experience on 30 consecutive cases. J Plast Reconstr Aesthetic Surg 66:660–666 [cited 2019 May 30] Available from: http://www.ncbi.nlm.nih.gov/pubmed/23415876
17. Gerber PA, Schulte K-W, Ruzicka T, Bruch-Gerharz D (2008) Eccrine porocarcinoma of the head: an important differential diagnosis in the elderly patient. Dermatology 216:229–233 [cited 2018 May 21] Available from: https://www.karger.com/Article/FullText/112931
18. Jeon H, Smart C (2015) An unusual case of porocarcinoma arising on the scalp of a 22-year-old woman. Am J Dermatopathol 37:237–239 [cited 2018 Sep 22] Available from: http://content.wkhealth.com/linkback/openurl?sid=WKPTLP:landingpage&an=00000372-201503000-00009
19. Boam T, Szczap A, Mendes da Costa T et al (2017) A case of massive porocarcinoma. Ann R Coll Surg Engl 99:e230–e232 [cited 2018 Sep 23] Available from: http://www.ncbi.nlm.nih.gov/pubmed/29022792