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## A synchronous incidence of eccrine porocarcinoma of the forearm and facial squamous cell carcinoma: A case report

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## ABSTRACT

**INTRODUCTION:** Cutaneous appendageal tumor can differentiate towards or arise from either pilosebaceous apparatus or the eccrine sweat glands. Appendageal tumors are relatively rare, their clinical appearance is non-specific, and the vast majority are not diagnosed until after excision. Eccrine porocarcinoma (EP), also known as malignant eccrine poroma is a rare adnexal tumor arising from the intraepithelial ductal parts of the sweat gland.

**CASE PRESENTATION:** We presented a 65-year-old, Asian, female with medical co-morbidities, who came with both a facial squamous cell carcinoma and a long-standing lesion over her left forearm. Histopathological finding of the left forearm demonstrated eccrine porocarcinoma.

**CONCLUSION:** Mohs micrographic surgery is the mainstay treatment of cutaneous carcinoma. It is important to rule out associated syndromes in patient who present with multiple cutaneous appendageal tumors.

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## 1. Introduction

The majority of appendageal tumors differentiate towards or arise from either pilosebaceous apparatus (hair follicle, sebaceous gland and in some body sites the apocrine gland) or the eccrine sweat gland. While the great majority of appendage derived tumors are benign, occasionally they can be cancerous or associated with various important syndromes. The pilosebaceous unit are concentrated in the head and neck area with the pilar element predominant on the scalp and the sebaceous element on the face, chest and upper back. The apocrine sweat glands are mainly found in the axilla, breast and perineal area. The eccrine sweat glands are found on all body sites. Eccrine and apocrine neoplasms present a bewildering array of morphologies which often defy precise classification [1]. Eccrine porocarcinoma (EP), also known as malignant eccrine poroma is a rare adnexal tumor arising from the intraepithelial ductal parts of the sweat gland. These tumors account for 0.005% to 0.01% of all epidermal skin neoplasms [2]. It was first described by Pinkus and Mehregan [3] in 1963, as ‘epidermotropic eccrine carcinoma’, later in 1969, Mishima and Morioka [4] introduced the term ‘eccrine porocarcinoma’. Since that time, Salih AM et al., has published a meta-analysis of 453 porocarcinoma cases reported worldwide [5]. EP is typically a disease of the elderly, with the mean age at presentation of 60–80 years, although rare

cases have been reported in children [6,7]. It is thought to arise *de novo*, but reports of an adjacent benign component on histology suggest that it can also be associated with a pre-existing benign poroma [7]. The presentation is extremely variable, and initial clinical impression is seldom accurate [7]. In line with SCARE criteria [8], we report an unusual case of both squamous cell carcinoma and eccrine porocarcinoma found on a patient.

## 2. Patient information

A 65-years old, Asian, female, with a medical history of diabetes mellitus, hypertension and dyslipidemia was referred to the Plastic Surgery Clinic from a district hospital for melanoma. The patient’s complaint began with a papular lesion over the left forearm 5 years previously, which was circular, gradually increasing in size. She then developed a nodule over her right cheek about 1 year ago. Both were slow growing lesions with no history of ulceration. She denied any history of trauma or discharge previously with no history of skin malignancy in her family. None of her siblings have been screened for any chromosomal studies. There were no constitutional symptoms from this patient. Her main complaint was itchiness and discomfort from both the lesions. She was keen for removal of the lesions. Patient has no known drug or food allergy history.

Patient presented to the clinic on 5th of March 2017.

Excision of both lesions (right cheek and left forearm) were performed under local anaesthesia on 15th March 2017. We were unable to excise the lesion over the left forearm due to exten-

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**Fig. 1.** Excision of left forearm lesion under general anaesthesia. Dissection done to identify the vascular bundle.

sive vascular supply; however, a punch biopsy sample of the left forearm was sent.

Histopathological results were validated on 29th March 2017. Right cheek sample; squamous cell carcinoma, well differentiated. Left forearm biopsy; consistent with eccrine poroma.

Excision of the left forearm lesion was performed by the plastic surgeon under general anaesthesia on 10th April 2017. She was kept for observation at the ward for 2 days post operatively before being discharged home.

### 2.1. Clinical finding

Generally, the patient was well, not cachexic looking. No lymphadenopathy was noted (cervical and axillary lymph nodes). Pedunculated lesion over the right cheek measuring about 1.0 × 0.5 centimeters, non-ulcerative. A hard-pedunculated mass over the anteromedial aspect of left mid-forearm measuring 6 × 5 centimeters, erythematous, cauliflower-like growth with a stalk measuring approximately 1 centimeter, and visible feeding vessels.

### 2.2. Therapeutic intervention

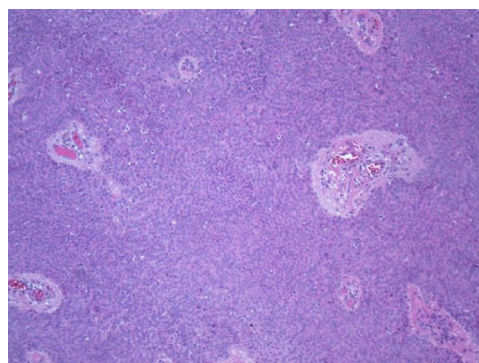
Under local anaesthesia, a transverse elliptical incision was done over the right cheek with clear margin of 5 millimeters. A punch biopsy was taken initially from the left forearm, a combination of both, the skin lesion tissue with normal tissue were sent for investigation. A transverse elliptical incision over the left forearm lesion with a 1-centimeter margin was done under general anaesthesia and the lesion was removed. Wound was closed primarily (Fig. 1).

### 2.3. Histopathological finding

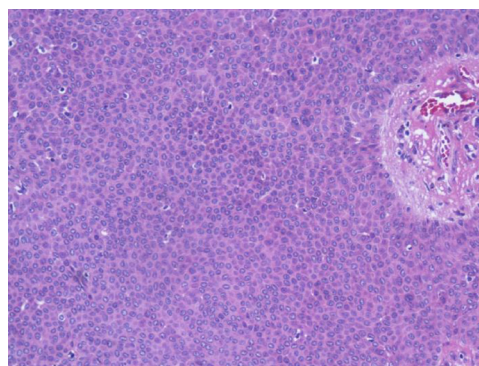
#### a) Right cheek

A hyperpigmented polypoidal tissue measuring 4 × 5 × 6 millimeters. A polypoidal skin lesion with malignant dermal infiltration derived from keratinocytes in epidermal layer. The malignant cells exhibit mild to moderate nuclear pleomorphism with vesicular nuclei, prominent nucleoli and abundant eosinophilic cytoplasm. Keratin pearls and intercellular bridges are seen in areas. Mitotic figures are increased. The surrounding stroma shows moderate lymphoplasmacytic infiltrates. The deep margin is free of tumor. Interpretation: squamous cell carcinoma, well differentiated (Figs. 8 and 9).

#### • Left forearm:



**Fig. 2.** Eccrine poroma at 10x magnification.



**Fig. 3.** Eccrine poroma at 20x magnification.

A polypoidal skin lesion composed of proliferation of small keratinocytes attached to the epidermis and extends into the dermis as broad columns. The tumor cells have monomorphic ovoid nuclei, inconspicuous nucleoli and distinct cytoplasmic margins. Occasional ductal lumina and cystic spaces within lobules of the tumor cells are also observed. No increase in mitosis, marked nuclear atypia or necrosis is seen. The surrounding dermis show focal dilated blood vessels. No evidence of malignancy seen. (Figs. 2 and 3).

Interpretation: Eccrine poroma.

A large pedunculated papillomatous skin lesion measuring 50 × 45 × 35 millimeters. The papillomatous lesion is pinkish in color with multiple hyperpigmented areas. The section shows proliferation of tumor cells with inconspicuous intercellular bridges extending from the lower epidermis into the dermis in broad expanded columns with pushing border. The tumor cells exhibit two types of atypical cells which is eosinophilic cells with polyhedral round to oval hyperchromatic nuclei, distinct nucleoli and variable amount of eosinophilic cytoplasm. The other types of cells are clear cells type which appear enlarged with round to oval nuclei, inconspicuous nucleoli and have abundant clear cytoplasm. Some of the cells contain pigments with focal squamous differentiation and ductal-like structures. In many areas, obvious nuclear atypia with frequent mitoses and focal necrosis are evident. The surrounding stroma shows proliferation of reactive vessels and mild chronic inflammation. The tumor cells are immunoreactive to CK 7 and non-reactive to S100. Focal reactivity towards SMA is seen in areas. The tumor is completely excised, 10 mm away from margin (Fig. 4–7).

Interpretation: Porocarcinoma arising from eccrine poroma.

### 2.4. Follow-up and outcome

She was seen at the plastic outpatient clinic 2 weeks later. The scars were well healed. Literature review showed that porocar-

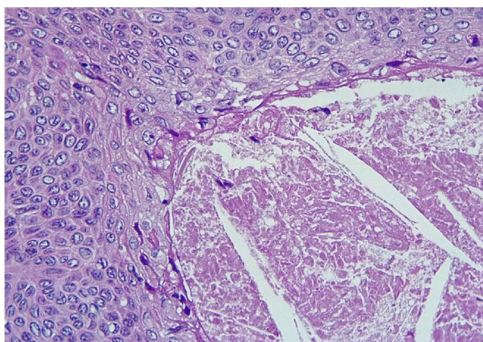


Fig. 4. Eccrine porocarcinoma with necrosis area seen at 20x magnification.

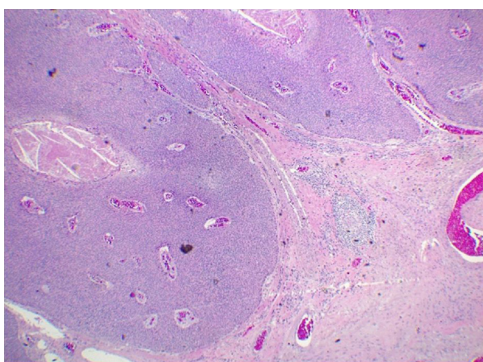


Fig. 5. Eccrine porocarcinoma with mitosis and prominent nucleoli seen at 40x magnification.

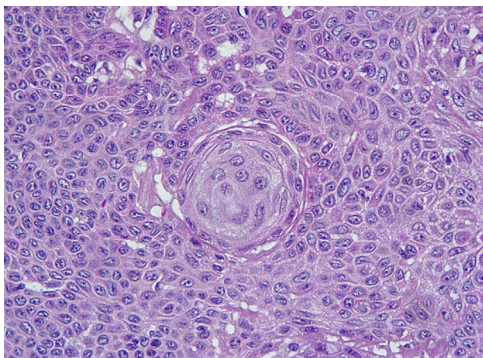


Fig. 6. Squamous differentiation seen in porocarcinoma at 400x magnification.

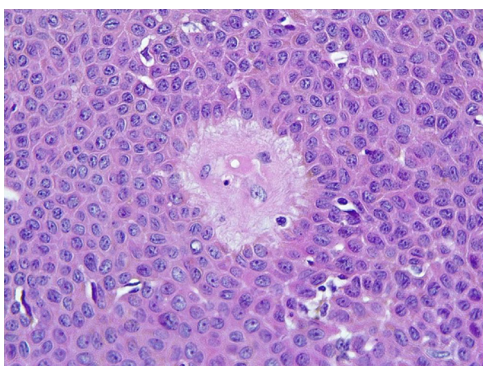


Fig. 7. Ductal like structure in porocarcinoma at 400x magnification.

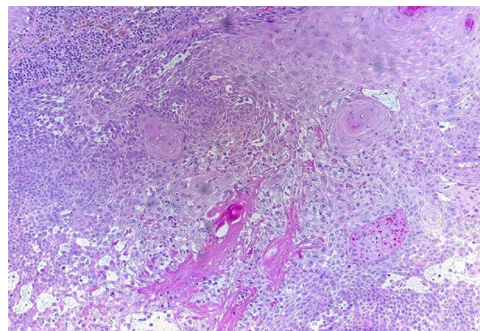


Fig. 8. Dyskeratotic cells and keratin pearls seen in squamous cell carcinoma of the right cheek at 100x magnification.

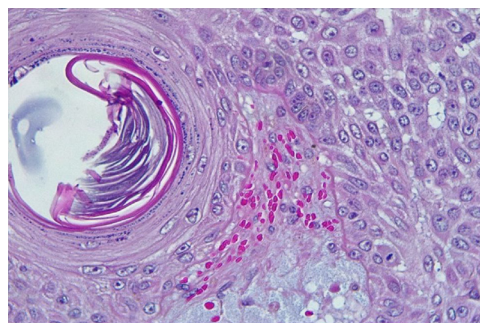


Fig. 9. Dysplastic keratinocytes with mitosis in squamous cell carcinoma at 400x magnification.

cinoma has a high recurrence rate and therefore, we proceeded with a computed tomography (CT) scan of thorax, abdomen and pelvis. There were no nodal involvement and distant metastasis. It has been 6 months post operatively (presented first on 5th March 2017), and the patient is still being seen at the clinic with no new lesions observed. Patient is happy with the post-operative scar outcome.

### 3. Discussion

The presence of eccrine type ducts may be a prerequisite for the reliable diagnosis of EP; however, using recognizable ducts as a major criterion for diagnosis implies that some poorly differentiated tumors may well be overlooked. Several associated cell types are found with EP, such as squamous cells, spindle cells, clear cells, mucin-producing cells and melanocytes [9], making all these as possible differential diagnoses. To be able to distinguish an appendageal tumor, the role of immunohistochemical is crucial. Eccrine sweat glands are widely distributed almost everywhere in the skin. The cells in the excretory coil express positivity for low molecular weight keratin (LMWK) such as CK 7 and CK 19, epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA), as well as S100 protein in the basal layer only. The myoepithelial cell layer may be highlighted by smooth muscle actin (SMA), p63 and calponin [10].

Porocarcinomas usually arise in pre-existing benign poroma, and are histologically characterised by asymmetrical, solid, nodular growth pattern, with infiltrative or pushing borders. The neoplastic cells in porocarcinoma may have basaloid features, resembling those of poroma, but show varying degrees of cytonuclear atypia, hyperchromatic nuclei, and prominent nucleoli. Large polyhedral cells with abundant clear cytoplasm can be present. Foci of squamous and spindle cell differentiation are common. Evidence of eccrine differentiation in the form of CEA-positive ductal formation is present in most of the cases. Brisk mitotic activity, necrosis and

desmoplastic stromal reaction can be prominent. Continuity with a benign pre-existing poroma and prominent intraepidermal component, occasionally showing severe cellular pleomorphism and resembling Paget's disease, can be present [10].

The main differential diagnosis of porocarcinoma is squamous cell carcinoma, which lacks intracytoplasmic lumina and ductal formation. The presence of an adjacent benign poroid component is a helpful feature in establishing the diagnosis. In clonal Bowen's disease, the tumor cells are more atypical and there is more severe architectural abnormality. Clonal seborrhoeic keratosis and Bowen's disease are negative for CEA, CK 7 and S100, which are often positive in porocarcinoma. Negativity for melan-A and HMB45 helps rule out melanoma in situ. In Paget's disease, the cells are large, with clear PAS-positive cytoplasm [11].

In the past decade the molecular basis of many inherited syndromes has been unraveled. Skin appendageal tumors present as papules (bumps/lumps) on the skin that are difficult to distinguish clinically from one another. They can be solitary or multiple. They are typically multiple when they are associated with an inherited syndrome. In all of these genodermatoses, the cutaneous tumors represent the pathognomonic finding of the disease [17]. In a review by Lee DA et al., it emphasizes that every clinical sign enables the detection of a hereditary predisposition to visceral cancer which will be of great value for general cancer prevention, and therefore knowledge of these recent molecular advances in inherited skin appendage tumor syndromes is essential [12]. We were unable to conduct genetic screening due to lack of technical knowledge and limited resources available. However, in that review there were no direct syndrome link between eccrine porocarcinoma and squamous cell carcinoma reported.

Ideal management of EP remains controversial as there is no consensus on treatment [13]. Most patients undergo surgical excision, often with wide margins. Despite negative-margin primary excision, many patients with EP [14,15–17] will go on to develop regional nodal involvement (20%) or distant metastases (10%) [14,15–17]. Mortality among patients with EP with nodal metastases is high at 67% [18].

To predict more aggressive behavior, two case series examined the prognostic implications of histologic factors for recurrence of EP. In their study of 69 patient cases, Robson et al. [7] found an infiltrative, as opposed to a pushing, histologic subtype to be strongly predictive of local recurrence after controlling for both tumor depth and mitoses [7]. They also found that tumor depth >7mm, >14 mitoses per high-power field, and lymphovascular invasion were predictive of death resulting from EP. In a second study of 24 patient cases, in which 35% presented with recurrences, Belin et al. [13] likewise divided EPs into infiltrative, pushing, and pagetoid based on the Robson et al. report. Although none of the seven pushing EPs recurred, four of 10 infiltrative and two of two pagetoid EPs were associated with local, regional, and distant recurrences. In our case, we did not see vascular invasion, neither radiologically nor surgically, however histopathological examination showed increased mitotic activity with a tumor depth of 35 mm. We classified our case as a high risk histologic type.

Our findings are limited by the number of cases and the follow-up period, which was around 8 months post-operative. In addition, follow-up visits were not always with a plastic surgeon and therefore careful inspection for local recurrence may not have been performed during every visit; however, this would not be expected to affect overall survival.

#### 4. Conclusion

Eccrine porocarcinoma is a rare aggressive form of skin cancer with unknown etiology and little guidance available in the litera-

ture on exact protocols for treatment and follow up. It should be on the differential diagnosis of any suspicious skin lesion seen by the plastic surgeon. Histologic assessment is indicated in suspicious lesions.

#### Conflict of interest

There is no conflict of interest.

#### Funding

None.

#### Ethical approval

Ethical approval has been requested from the ethical committee from my institution's Clinical Research Centre, Hospital Sultanah Nur Zahirah. In view of this being a case report, with no interventions/comparisons done, the committee agreed upon that we may proceed by obtaining an informed consent from the patient.

#### Consent

The consent was taken from the patient herself. Her family that was present during her clinic appointment also agreed for us to write a paper on her condition.

#### Author contribution

Nandinii ramasenderan was the main author writing this case report. The pathology input was provided by Siti Zarqah Omar. The surgeon who performed this surgery was Hasliza Shahir, assisted by Nandinii Ramasenderan.

#### Guarantor

Nandinii Ramasenderan.

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