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Two Cases of Scalp Eccrine Porocarcinoma with Review of Literature

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Abstract

Eccrine Porocarcinoma (EPC) is a rare malignant adnexal neoplasm arising from the eccrine sweat glands. Head and neck, lower extremities are the most common primary locations for the development of EPC. Scalp is a rare site of involvement, with less than 20 cases reported so far. This study aims to report two cases of scalp EPC, which showed different clinical courses that could be predicted by histopathologic features. In one case, a 62-year-old woman with EPC was treated by local excision with 30 mm margin. A mitotic count of 16/10 high-power field, invasion depth >7 mm, and infiltrative tumor margins were observed, which are suggested as poor prognostic factors. Pulmonary metastasis and local recurrence occurred rapidly after 4 months of disease-free interval. In the other case, local excision with 10 mm margin was performed in a 45-year-old woman with EPC. Poor prognostic histopathologic features were absent, and there was no evidence of the disease during the 2-year follow-up period.

Keywords: Eccrine porocarcinoma; Scalp; Review

Introduction

Eccrine Porocarcinoma (EPC) is a rare malignant tumor derived from the acrosyringium, the terminal intraepidermal segment of the eccrine sweat gland. Common primary locations of EPC are head and neck, lower extremities, trunk, and upper extremities [1]. Scalp is a rare site for the development of EPC, with less than 20 cases worldwide and 3 cases in Asian population reported so far [2]. Surgical excision is considered the treatment of choice for the primary localized disease. However, as this condition is rare, there is no consensus regarding the adequate surgical margin, indications for adjuvant treatment [3,4]. Here, we report two cases of scalp EPC with different clinical courses. Additionally, we reviewed the current literature regarding prognostic factors in relation to treatment strategies.

Case Reports

Case 1

Patient 1: A 45-year-old woman presented with a 2 cm nodule in the left parietal scalp (Figure 1). She had undergone an excision biopsy and the histopathology report suggested EPC. A wide local excision, deep into the temporalis muscle, with a 10 mm margin from the previous excision was performed. Histopathology examination demonstrated a 2.0 × 1.4 × 0.4 cm tumor with polygonal basaloid tumor cells displaying ductal differentiation, which was consistent with porocarcinoma (Figure 2). The tumor extended from the epidermis to the subcutaneous layer with a depth of 4 mm. Pushing growth pattern was observed at the tumor margin. The mitotic count was 1/10 High-Power Field (HPF). Lymphovascular invasion was not present. All surgical margins were negative. No evidence of local recurrence or distant metastasis was found during the 2-year follow-up period.

Case 2

Patient 2: A 62-year-old woman presented with a 3 cm sized painful occipital scalp mass. Biopsy result suggested a presence of malignant skin adnexal neoplasm. Cervical lymph node and distant metastasis were not identified in the computed tomography scan. A wide local excision with 30 mm margin was performed in full layer. Intraoperative findings showed no evidence of skull and surrounding tissue invasion. Histopathology examination revealed a 3.2 × 1.5 × 1.1 cm porocarcinoma infiltrating into the subcutaneous layer at a depth of 11 mm (Figure 3). An infiltrative growth pattern was observed at the tumor margin. The mitotic count was 16/10 HPF. Lymphovascular invasion was not present. Immunohistochemical staining showed that the tumor cells were positive for Cytokeratin (CK) 5/6; a few tumor cells were positive for p63, p40, and CK7. Although surgical margins were negative, the deep resection margin was very close to the tumor (0.1 mm). Adjuvant radiotherapy was prescribed for the close margin (30 Gy, 13 fractions). After 4 months of disease-free interval, local recurrence with multiple cervical lymph node and pulmonary metastasis were identified by radiologic and histologic evaluations. Palliative chemotherapy with a single dose of docetaxel (60 mg/m^2) was given; the patient was lost to follow-up.

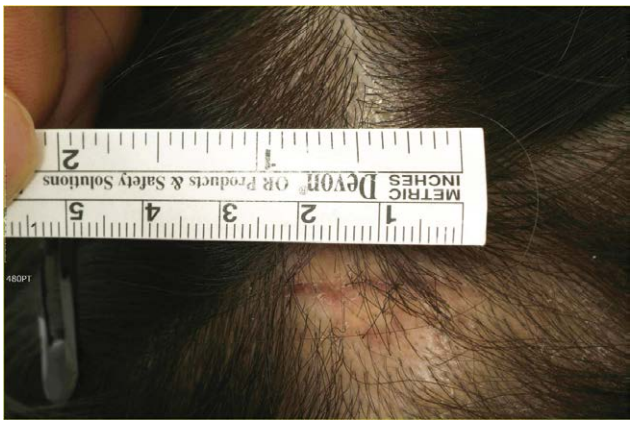

Figure 1: Clinical photograph of patient 1 (post-biopsy state), A 2 cm sized post-biopsy scar was observed at left parietal scalp.

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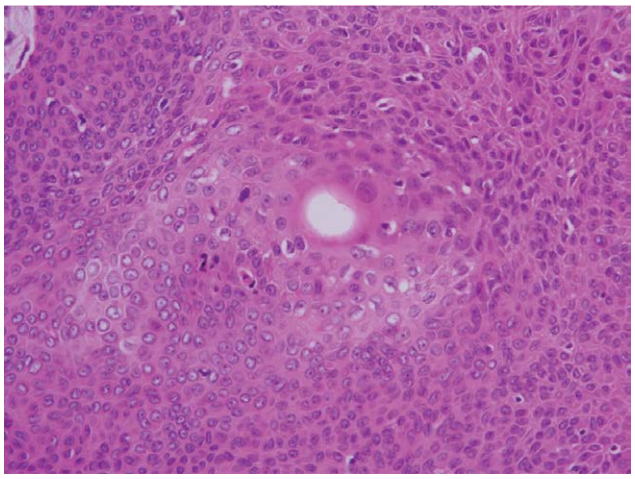

Figure 2: Microscopic findings of excised specimen in patient 1, Poroid cells with cellular atypia were observed, Small ductal structures were identified (H & E, 400x).

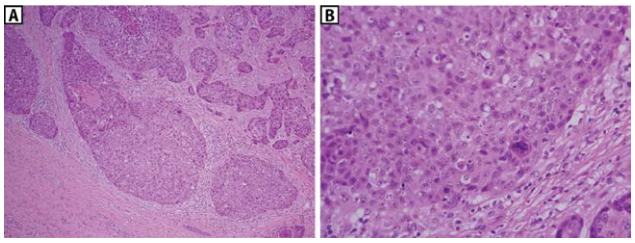

Figure 3: Microscopic findings of excised specimen in patient 2. (A) Nests of tumor cells with necrosis (H and E, × 100). (B) Marked cellular atypia with hyperchromatic nuclei, distinct nucleoli were observed, Small ductal structures were identified, Multiple mitotic figures were found (H & E, 400x).

Discussion

EPC is a rare malignant adnexal neoplasm, with an estimated incidence of 0.005-0.01% of all cutaneous tumor. As this condition is rare, research is limited and no prospective studies have been published, leading to poor understanding of this entity. The most common clinical presentation of EPC is a mass or nodule (71.2%); however, ulcerative (18.3%) and plaque-like (9.8%) morphology are also possible [1]. The initial clinical diagnosis is not accurate in most cases, with wide range of differential diagnoses, including squamous cell carcinoma, Bowen's disease, pyogenic granuloma, basal cell carcinoma, amelanotic melanoma, and metastatic adenocarcinoma [5]. Therefore, histopathological examination is essential to confirm the diagnosis. Pathology results also provide information to predict tumor behavior, which may provide guidance for the development of further treatment strategies.

EPC is reported as a relatively aggressive neoplasm displaying poor prognosis among non-melanoma skin cancer. Regional lymph node and distant metastasis are observed in 20% and 10% of the patients, respectively [6]. Recurrence after wide local excision is estimated in about 20% of the cases [7,8]. Lymph node and distant metastasis are detected on an average 11 and 9 months after operation, respectively [9]. Surgically excised localized primary EPC shows good survival outcomes in cases of no evidence of recurrence, while disease specific mortality rates are reported up to 67% if lymph node metastasis are present [3,6]. Therefore, it is important to evaluate the nodal status at initial presentation and during the follow-up period.

A few retrospective studies attempting to identify histopathologic prognostic factors have been reported (Table 1). Robson et al. [6] analyzed histopathology and clinical outcomes of 69 cases, in a study that has the largest sample size so far. In this study, mitotic count, tumor depth, and lymphovascular invasion was associated with death or lymph node involvement. The presence of infiltrative tumor margin, described as ill-defined lower limit with strands of atypical cells invading the dermis, was associated with local recurrence. Similarly, Skowron et al. [7] and Belin et al. [4] reaffirmed that infiltrative margin was associated with metastasis or recurrence. The role of the infiltrative margins is reproducible in majority of the studies, and thereby suggested as a reliable prognostic factor. However, treatment approaches and primary locations are not standardized in these studies, which make implementation on real clinical cases difficult [10].

We experienced two cases of invasive EPC on scalp, one with an indolent clinical course while the other presented an aggressive behavior. In the former case, known poor prognostic histopathologic features were absent, and there was no evidence of the disease following the local excision of 10 mm margin. On the contrary, the second case

Authors/ References	No. of Cases	Anatomic Site	Treatment	Poor Prognostic Factors
Robson [6]	69 (*in situ* 7)	Head and Neck (21%)	WLE (55)	Mitoses >14/HPF
		Trunk (24%)	Curettage (4)	Lymphovascular invasion
		Upper limb (11%)	Biopsy (6)	Depth >7 mm
		Lower limb (44%)	Amputation (1)	Infiltrative growth pattern
			Others (3)	
Shiohara [9]	12	Head and Neck (8%)	WLE, SM ≥ 2 cm (7)	Infiltrative growth pattern
		Trunk (8%)	WLE, SM <2 cm (4)	Lymphovascular invasion
		Upper limb (8%)	Biopsy (1)	--
		Lower limb (75%)	--	
Mahomed [10]	21	Head and Neck (19%)	WLE (17)	Mitoses >14/HPF
		Trunk (19%)	Biopsy (3)	--
		Upper limb (19%)	Amputation (1)	--
		Lower limb (28%)	--	--
		Vulva (5%)	--	--
Belin [4]	24	Head (33%)	WLE, SM ≥ 2 cm (6)	Infiltrative growth pattern
		Trunk (37%)	WLE, SM <2 cm (13)	--
		Upper limb (9%)	Mohs surgery (2)	--
		Lower limb (21%)	Others (3)	--
Skowron [7]	50 (*in situ* 6)	Head (38%)	WLE (49)	Depth >10 mm
		Trunk (10%)	Radiotherapy (1)	Infiltrative growth pattern
		Upper limb (18%)	--	--
		Lower limb (34%)	--	--
WLE: Wide Local Excision; SM: Surgical Margin				

Table 1: Summary of recent studies recruiting more than 10 cases which analysed poor histopathologic prognostic factors in eccrine porocarcinoma.

presented multiple poor histopathologic features. Infiltrative tumor margins, a mitotic count of 16/HPF, and a tumor depth >10 mm with very close deep margin were indicative of poor prognosis. Although wide surgical margin of 30 mm and adjuvant radiotherapy was applied, local recurrence and distant metastasis occurred rapidly within 4 months. Hence, our observations support the role of the previously reported prognostic factors.

Conclusion

In conclusion, we report two cases of EPC affecting the scalp, a rare site of presentation. These two cases showed different clinical courses, which could be predicted by histopathologic features described in the literature. Though there are no reliable guidelines for EPC treatment, we suggest that treatment strategies may be specified considering the histopathologic prognostic factors.

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