

Eccrine Porocarcinoma Affecting An Unusual Site: Frontoparietal Scalp

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Abstract

Observation: Eccrine porocarcinoma is a rare malignant sweat gland neoplasm which may occur de novo or arise in a preexisting eccrine poroma. We describe a case with eccrine porocarcinoma of the scalp arising from eccrine poroma in a 74 year-old man. He was treated with wide local excision without any recurrence after 1 year follow-up.

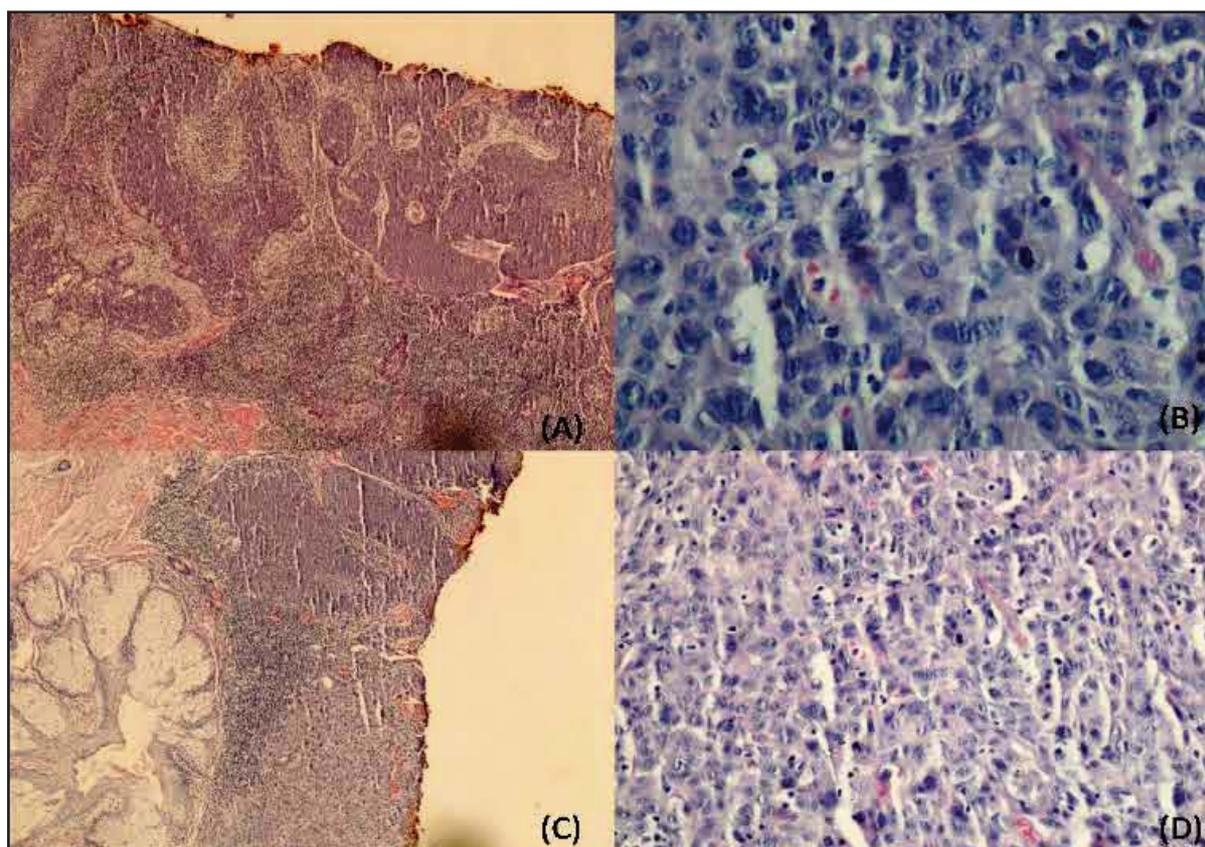
Introduction

Eccrine porocarcinoma [EPC] is a very rare cutaneous neoplasm which originates from the cells of acrosyringium of the eccrine sweat gland. It was first described by *Pinkus* and *Mehregan* in 1963 with the term of 'epidermotropic eccrine carcinoma' [1]. The exact pathogenesis of the disorder remains unclear though radiation and immunosuppressive agents were suggested to be the main triggers of malignant transformation of eccrine poroma [2, 3]. EPC which is the malignant counterpart of eccrine poroma may present in mid-life, but typically it affects elderly patients [4]. It may present with variable clinical forms including erythematous or violaceous, ulcerated, bleeding or asymptomatic, cauliflower-like papules or nodules. Most of the EPC lesions occur on the lower extremities followed by trunk, head, upper extremities, and neck in decreasing frequency. Scalp is the primary affected site rarely [5, 6]. Scalp

located EPC lesions mostly seen in occipital region however there are also reports of EPC affecting frontoparietal site in the literature [7, 8].



Figure 1. Verrucous nodulo-ulcerative lesion on the right frontoparietal scalp



Figures 2A ,B, C, and D. .Histopathological examination reveals interconnected epithelial downgrowths comprised predominantly of monomorphic basaloid cells and multiple foci of attachment to the epidermis of these epithelial bands (A), note the marked pleomorphic tumor cells and conspicuous mitoses in some areas(B,D), porocarcinoma arising in preexistent eccrine poroma (C). Original magnifications A;C: x40, B;D:x100, H&E

Case Report

74 year-old otherwise healthy male, presented to us with a gradually swelling of smaller lesion with a-year duration, over right frontoparietal region. His family history was unremarkable. He indicated that the preexisting lesion was present for about 10 years. The lesion was non-tender painless erythematous nodule with vegetative oozing surface. It was located on actinic damaged alopecic skin of scalp (**Figure 1**). His general and systemic examination was normal. Complete blood count, biochemical parameters and skull x-ray were normal. Erosion of calvarium or intracranial extension was not detected. Basal cell carcinoma, squamous cell carcinoma, verruca vulgaris were included in differential diagnosis. Wide local excision with primary closure was done. The specimen showed a mass without skin coverage measuring 2.1x1.7x0.5 cm. The tumoral nests composed of uniformly small basaloid cuboidal cells arising from epidermis and including tubular structures and cystic spaces in some areas were detected on microscopic examination (**Figure 2A B and C**). Some sections presented cytologic atypia as well

as infiltrative growth pattern (**Figures 2B and D**). Eccrine porocarcinoma within a eccrine poroma was observed (**Figure 2C**). The histopathological examination provided the diagnosis of eccrine porocarcinoma. Clinical examination and imaging analyses did not reveal any evidence of lymph node involvement or distant metastasis. No recurrence was observed during a 1-year follow up after surgery.

Discussion

EPC is a rare tumor to diagnose primarily based on clinical presentation especially in the case of atypical location. Therefore the role of the histopathological examination along with immunohistochemistry in determining the diagnosis is key. Scalp EPC is still one of the uncommon presentations of the tumor [9, 10, 11, 12]. Apart from the propensity for local recurrence and metastases, EPC those affecting scalp have potential risks

such as involvement of periosteum, calvarium and eventually compression brain lobes [13]. Even though it is commonly seen in occipital region, there are also a few reports of fronto-parietal location as the case presented here [7, 8].

Malignant transformation of eccrine poroma to porocarcinoma is well-known process however exact mechanism of this transformation has not been documented yet. Disorders causing immune deficiency including HIV, organ transplantation and diabetes are associated with eccrine porocarcinoma [14]. Trauma or radiation induced eccrine poromas are reported in literature [15, 16]. It has been suggested that radiation and trauma might contribute the malignant transformation to eccrine porocarcinoma [2, 3]. Clinical progression and histopathological findings revealed that our patient had eccrine porocarcinoma arising in preexisting eccrine poroma. Our case was healthy person without any immune deficiency, history of trauma or malignancy. The possible role of UV in the aetiology of EPC has been discussed in the literature [17]. On the other hand existence of EPC within area of Bowen disease has been reported [18]. In the light of the case presented here, we may speculate that UV induced damage has facilitated the progression of eccrine poroma. However, future studies are needed to unveil the exact pathogenic link between UV damage and EPC. In conclusion, because of aggressive behaviour and lack of distinct clinical appearance of this rare tumor we discussed this case to emphasize inclusion of the tumor in the list of differential diagnosis of exophytic tumors of scalp, in particular elderly patients with actinic damage.

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