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Primary Apocrine Carcinoma of the Skin – a Case Report

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Sweat gland skin carcinomas are extremely rare. The differentiation between apocrine and eccrine neoplasms is very difficult, since composite adnexal tumors may often be encountered. We present a case of 72-year-old male with a slow-growing painless erythematous-livid nodule on the frontal scalp. According to the medical record, the lesion evolved on the site of a pre-existing cylindroma, excised 2 years before. Diagnostic punch biopsy showed solid apocrine carcinoma. Wide excision with en-bloc lymph node dissection was recommended. A dose of alertness, together with a thorough review of the clinical, histology and immuno-histochemical features of this very rare skin adnexal tumor is, herein, given.

Key words: sweat gland tumors, apocrine carcinoma, histogenesis

Introduction

Cutaneous apocrine gland carcinoma is a rare form of sweat gland neoplasm with distinct cytological appearance. No more than 100 cases have been described worldwide. Most common localization is axilla, followed by scalp, nipple, trunk, anogenital region, wrist, fingertip, foot, toe, lip, ear, chest and eyelid [14]. Usually, the cells show abundant eosinophilic cytoplasm and eccentric, basally located nuclei. Luminal cells decapitation secretion is the outstanding histological feature. Most tumors present periodic acid Schiff (PAS) positivity, and iron pigment. The luminal cells are reactive to low molecular weight keratin and androgenic receptor and do not express oestrogen receptors [6]. Those characteristics facilitate the proper assessment of suspicious lesions.

Case report

A 72-year-old man presented with a slow-growing soft painless erythematous-livid nodule with a diameter of 3 cm, located on the frontal scalp. The tumor mass was fixed to the skin and slightly ulcerative on the lower aspect. No palpable regional lymph nodes were identified. Other physical examination did not show any abnormalities. According

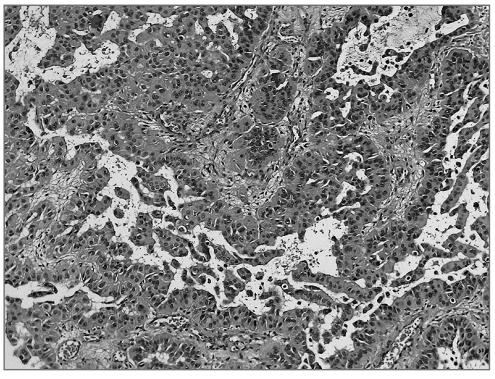

Fig. 1. Subcutaneous infiltrative tumor represented by un-uniform nests of tubular structures, covered by atypical cells with eosinophilic cytoplasm and central decapitation secretion (HE, × 100)

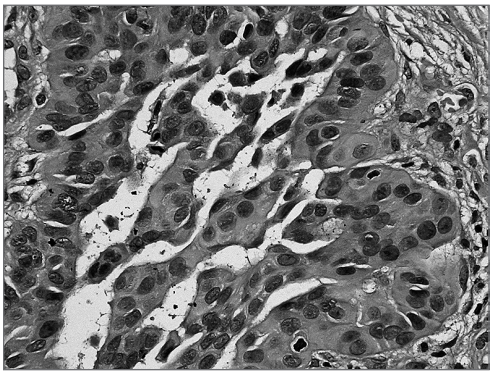

Fig. 2. Hyperchromic nuclei and frequent mitotic figures (HE, × 400)

to the medical record, this lesion evolved on the site of a pre-existing tumor, histologically verified as cylindroma, excised 2 years before. Unfortunately, no skin specimen was available for revision. Diagnostic punch biopsy was performed. Histological analysis showed invasive tumor, located on the border of the reticular dermis and subcutis, forming various-shaped nests of tubular structures, some of which were related to the follicular infundibulum (**Fig. 1**). The cells had eosinophilic cytoplasms and showed central apocrine secretion. Hyperchromic nuclei with frequent mitotic figures were also evident (**Fig. 2**). The immunohistochemical analysis showed positivity for cytokeratin 7 (70%) and cytokeratin 20 (30%). No expression of oestrogen receptor was found. The diagnosis of primary skin apocrine gland carcinoma was concluded. Wide excision with en-bloc lymph node dissection was recommended.

Discussion

A malignant counterpart of most benign sweat gland lesions have been described. Although very rare, those carcinomas deserve special attention because of their high recurrence rate and metastatic potential [1]. Features, suggestive for neoplastic degeneration, include architectural asymmetry, infiltrative borders, irregular arrangement of the nests, nuclear atypia, and increased mitotic activity. The presented case showed clear-cut cytological atypia and architectural dis-cohesiveness [11]. The striking feature, which gives the ground for concluding an apocrine origin, was the typical decapitation secretion. This finding was extremely interesting and did not correspond to the identification of pre-existing cylindroma.

Tumors of pure apocrine origin appear to be much less common than those of eccrine differentiation [4]. The very few cases described to date showed male predominance and old age of onset (average of 57.9 years). The tumors cannot be differentiated by virtue of localization. Moreover, many sweat gland tumors traditionally assumed to have an eccrine origin are now recognized to have apocrine analogues [10]. These examples encompass hidrocystoma, poroma, cylindroma, spiradenoma and chondroid syringoma.

Cylindromas usually affect adult females. Most common localization is head and neck, particularly scalp and face. They can be sporadic and present as a solitary lesion,

or represent multiple coalescing mass defined as turban tumor [12]. Histologically, cylindroma is easily recognized by non-encapsulated variable-sized islands and nodules of epithelial cells with peripheral hyalinised material, arranged in a "jigsaw puzzle". Some tumors may resemble spiradenoma, suggesting a continuous morphological spectrum of a single entity, or a transitional from one lesion to the other.

No apocrine secretion has been described in cylindromas and their malignant counterparts. Therefore, we may speculate that our case may have been an unrecognized apocrine gland carcinoma a priori. On the other hand, according to the histogenetic theory, multiple loss mutations of chromosome 16 q 12-13, essentially important for development of various adnexal skin tumors, may lead to stem cell degeneration towards apocrine gland neoplasm [3, 4, 7]. Apart from this hypothesis, a possible association of apocrine carcinoma and benign sweat gland tumors may also arise [5, 8]. Such co-morbidity seems to be not so rare, having in mind 5 cases of Japanese patients, Ogata et. al. [9] reported to have axillar apocrine carcinoma in association with apocrine hyperplasia and apocrine adenoma, and two cases of perianal apocrine tubulo-papillary hidradenoma and ductal carcinoma [8].

Standard treatment options for cutaneous apocrine carcinoma include wide excision with 2-3 cm margins [13]. Sentinel lymph node dissection prove to be beneficial, as almost 90% of cases return with recurrence in less than 5 years. Adjuvant radiotherapy is not recommended with the exception of unoperable cases.

The prognosis depends on tumor size, degree of histological differentiation, vascular invasion. Various literature sources pointed out a 10-year-recurrence rate up to 50% and fatal outcome between 20-40% [2, 8, 9, 13].

Conclusions

Apocrine carcinoma of the skin is a rare adnexal neoplasm, presenting as tender slow-growing dermal or subcutaneous nodule, located on sites, rich in apocrine glands. Histogenesis remains controversial. Sometimes stereotypical presentation may hardly be differentiated by other types of sweat gland tumors and primary breast or gastrointestinal adenocarcinomas. Due to its aggressive course and high recurrent rate, cutaneous apocrine carcinoma represents a diagnostic challenge and extreme therapeutic provocation.

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