Case Report

Eccrine Porocarcinoma (Malignant Eccrine Poroma) with Bone Invasion and Lymph Node Metastases

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Abstract

Objective: Eccrine porocarcinoma is a rare malignant sweat gland tumor. This skin neoplasm represents only 0.005% of epithelial cutaneous neoplasms. We have diagnosed and treated a case of eccrine porocarcinoma in a cancer center institution, which is a regional reference for skin cancer treatment in Curitiba, Brazil. We report the clinicopathological data of this case. **Material and Methods**: We reviewed the medical records and notes, including surgical specimens of the skin tumor excised from a patient treated at our institution. All microscopic slides in our files from this case were reviewed. **Results:** A 58-year-old woman with no previous disease presented with a 9-month history of a progressively enlarging lesion on the occipital region. It was partially fixed to periosteum and associated with retracted adjacent tissues. A soft tissue biopsy revealed an eccrine porocarcinoma of the scalp. **Conclusions:** Because of the aggressive progression of this neoplasm, the best treatment option includes associated surgical excision and no adjuvant therapy.

Keywords: Carcinoma; Eccrine glands; Skin neoplasms

Introduction

Eccrine poroma is a benign tumor of the intra-epidermal component of the eccrine sweat duct.[1] The first malignant variant reported in the literature has been attributed to Pinkus and Mehregan (1963). They described a glycogen rich malignant tumor in an 82-year-old woman which showed multiple epidermotropic metastases and lymph node deposits, subsequently killing the patient.[2] Since this original case report of eccrine porocarcinoma, (EP) there have been numerous examples of EP described in the literature. Most of these have either been isolated cases or small series with frequently detailed aggressive tumors. This neoplasm represents only 0.005% of epithelial cutaneous neoplasms.[3] There are no more than fifty cases with complete follow-up related in international literature and this is one of the first cases with bone invasion and lymph node metastasis.

Report of a Case

A 58-year-old woman with no previously disease presented with a 9-month history of a progressively enlarging lesion on the occipital region. Physical examination revealed a 6.5cm x6.5cm irregular, ulcerative and infiltrating lesion of the midline occipital region. It was partially fixed to periosteum and associated with retracted adjacent tissues.

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On cervical exam we detected three enlarged lymph nodes at the posterior cervical drainage chain and another two enlarged lymph nodes at the right posterior cervical chain. The findings of laboratory tests demonstrated an elevated lactate desidrogenase and alkaline phosphatase. A chest x-ray film was otherwise normal. A soft tissue biopsy revealed an eccrine porocarcinoma of the scalp (Figure 1).

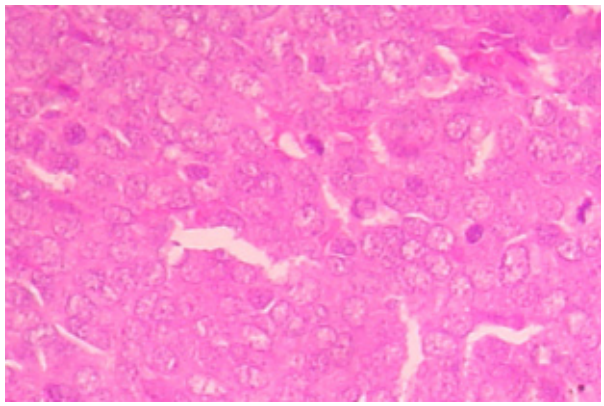

Figure 1 – Photomicrography of neoplasm composed of malignant cells with large, hyperchromatic, irregularly shaped nuclei. Mitotic activity is notable. Necrosis and ductal differentiation was not observed

Six months later, another local recurrence at adjacent bone was treated with palliative chemotherapy in a regimen that included paclitaxel, ifosfamide and cisplatin. The partial response gave the patient eight months of overall survival after the second surgery.

Discussion

Despite its rarity, eccrine porocarcinoma (EP) remains the most frequently encountered malignant eccrine sweat gland tumor, occurring predominantly in adults 50 to 80 years old.[2] Accurate data on the incidence of EP are difficult to obtain. Mehregan et al. and Wick et al. quoted EP incidences of 0.01% and 0.005%, respectively, of cutaneous tumors, with EP representing the most prevalent of malignant eccrine tumors.[4] Eccrine porocarcinoma has a propensity to appear on the lower limbs (44%), trunk (24%) or head and neck region (24%). Tumors vary greatly in size from less than 1 cm to 10 cm. A longstanding history is often encountered (up to 50 years) partly reflecting that some of these tumors originate from a preexisting BEP. Regional lymph node metastases are found in about 20% and distant metastases in about 10% of patients.[4-6]

Broadly, the tumor may appear as a nodule: verrucous, cauliflower-like infiltrative plaque or; as a polypoid growth that is frequently ulcerated and may bleed. Multinodularity, ulceration, and rapid growth may be associated with either local recurrence or metastasis. Microscopically, the diagnosis is rendered on either an invasive architectural pattern and/or significant cytological pleomorphism in a tumor showing eccrine differentiation.[1-2] This tumor is typically formed of cohesive basaloid epithelial cells. Others histologic patterns can contribute to the difficultly in making a pathologic diagnosis.[1] This morphologic appearance can include squamous cell, clear cell, spindle cell differentiation, mucous cell metaplasia, a Paget phenomenon and colonization by melanocytes.[6-7] The clear cell changes are frequently observed and are attributed by moderate amounts of glycogen, as detected by periodic acid of Schiff (PAS) and PAS after digestion with diastase.[7-8] A more aggressive clinical course may be indicated by a mitotic index of more than 14 per high power field, lymphvascular invasion and a tumor depth exceeding 7 mm. Although the gross size of tumor had no significant relationship to prognosis, tumor depth >7 mm is predictive of death or lymph node involvement

The patient underwent a radical excision of tumor with oncological margins and bilateral radical modified neck dissection. The prior diagnosis was confirmed and showed metastasis in 3 of 15 lymph nodes. Twelve months later, she developed a local recurrence and underwent another local excision (Figure 2).

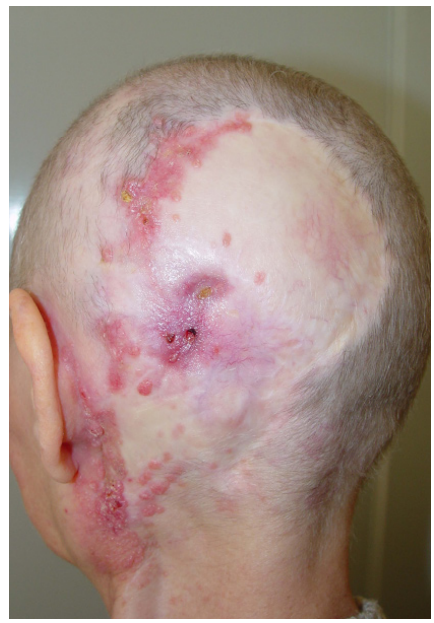

Figure 2 – Occipital recurrence of an ulcerative lesion. Note the area of previous skin graft used in reconstruction after first resection

(HR 2.5). Finally, an "infiltrative" tumor margin had a dramatic influence on local recurrence (HR 6.8). On a small biopsy, the accurate distinction among EP, SCC, and basal cell carcinoma may be impossible.[1-3] In our case, a good specimen and an expert Pathology Oncology department helped us to achieve the rare and definitive diagnosis.

Immunohistochemical techniques may be used to aid in the diagnosis of EP, because the cells that line the neoplastic ducts and clefts are positive for carcinoembrionic antigen and negative for S100 protein (in contrast, the myoepithelial cells of glandular portion are positive for S100 protein).[3-4] These findings confirm a primitive eccrine ductal differentiation.

Penile involvement by eccrine porocarcinoma is rare, as there are few cases related globally.[9-10]

The clinical differential diagnoses of these lesions include seborrheic keratosis, pyogenic granuloma, amelanotic melanoma, squamous cell carcinoma, basal cell carcinoma, verruca vulgaris, and metastatic adenocarcinoma.[1]

Cutaneous EP is rarely diagnosed preoperatively; thus, initial surgical management is not usually specifically planned. Although wide local excision of any cutaneous tumor is to be recommended, the results from historical series suggest EPs with an infiltrative pattern may benefit from further surgery if doubt exists regarding completeness of excision.[2-4,8] This case represents a particular situation in which initial resection may have not been as fully radical as needed; instead, surgical margins were clear on all slides. Similarly, once this diagnosis has been made, extra care should be taken by the surgical pathologist to evaluate resection margins. Because of this and the tendency to develop local recurrences, a wide excision of the primary tumor, with histologically clear margins, is indicated. A wide excision of the primary lesion is curative in 70% to 80% of cases.[3-5] The adjuvant treatment of this tumor has been incompletely characterized because his rarity. Bree et al. described a case in which topical 5-fluorouracil application and intra-arterial chemotherapy with docetaxel resulted in a histologically confirmed complete response of multiple regional skin metastases for more than 2 years. Despite intravenous administration of docetaxel, slow progression of systemic disease was observed.[11] Bazi et al. proposes a new protocol that includes isotretinoin and interferon alpha to metastatic disease with good results. Nevertheless, there are few studies about schemes or protocols of chemotherapy to EP without metastasis.[12]

Because EP has been a rare neoplasia and only a limited numbers of cases have been reported, there are many problems remaining to be resolved, including those concerning the pathophysiologic mechanism and treatment. Publication of additional case reports should improve our understanding of this uncommon neoplasia.

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