Longstanding red, lobular nodule on an elderly male's thigh

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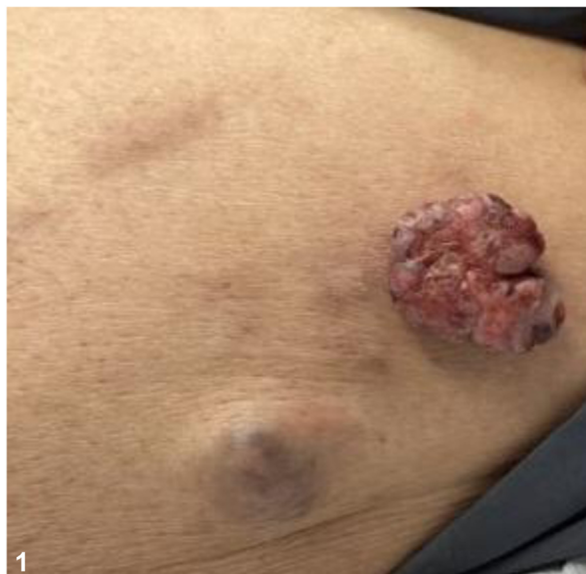

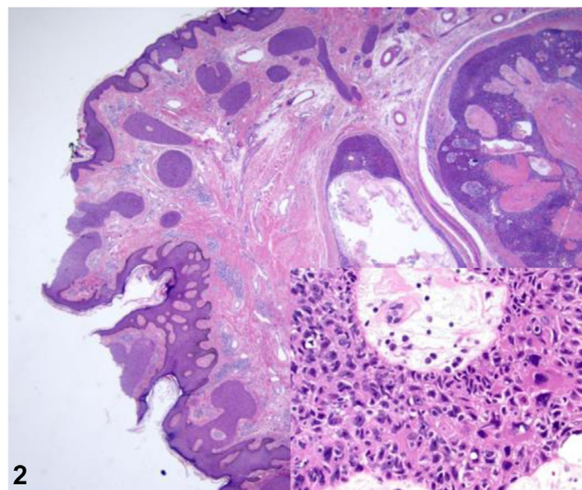

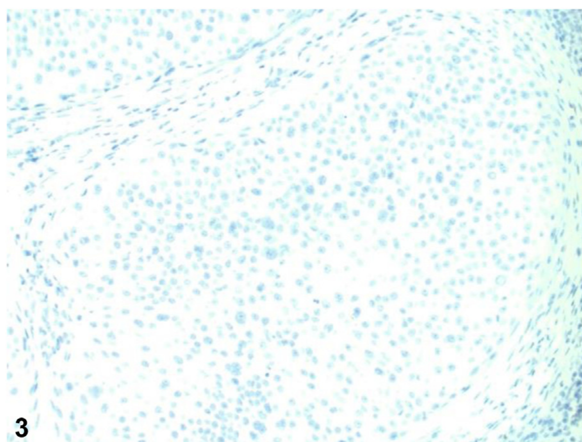

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CASE DESCRIPTION

A 69-year-old male presented with 2, adjacent lesions on the left thigh (Fig 1). The initial lesion was first noted 9 years prior and was reported to be slow-growing with associated bleeding. Months later, the patient noticed an adjacent asymptomatic lesion. Clinical examination demonstrated a 2.7 × 3.2 centimeter exophytic nodule and a distal flesh-colored, firm, subcutaneous nodule. He denied systemic symptoms. A biopsy of the exophytic nodule showed acrosyringeal cells, ducts, an invasive growth pattern, atypia, mitoses, and necrosis (Fig 2). Immunohistochemical staining for p53 demonstrated focal null phenotype (Fig 3).

Question 1: What is the most likely diagnosis?

A. Juxtaepidermal poroma

B. Eccrine porocarcinoma (EPC)

C. Malignant mixed tumor

D. Hidradenocarcinoma

E. Eccrine angiomatous hamartoma

Answers:

A. Juxtaepidermal poroma — Incorrect. Poromas are commonly found on the palms or soles as a solitary papule or nodule with a vascular appearance. A poroma is a benign sweat gland tumor and does not have cytologic atypia or mitoses. When a poroma occurs in the papillary dermis with continuity into the epidermis it is termed juxtaepidermal poroma.

B. EPC — Correct. EPC is an uncommon sweat gland malignancy and usually occurs in the elderly.[1] The location of the lesions is helpful to this diagnosis as the lower extremity is the single most common site for porocarcinomas. Histology resembles classic poroma but has infiltrative growth, cytologic atypia, mitoses, and necrosis.

C. Malignant mixed tumor — Incorrect. Malignant mixed tumor (malignant chondroid syringoma) is a rare, aggressive tumor that usually occurs on the distal extremities or foot. However, on histology there are malignant epithelial cells and myxoid or chondroid stroma.

D. Hidradenocarcinoma — Incorrect. Hidradenocarcinoma is an aggressive tumor that most commonly occurs on the head or neck. Histology shows sweat ducts within the tumor with atypia and mitoses and prominent dermal sclerosis with keloidal collagen.

E. Eccrine angiomatous hamartoma — Incorrect. Eccrine angiomatous hamartomas most commonly present in children as a tender, dusky nodule. Histology shows mature sweat glands surrounded by vessels.

Question 2: What immunohistochemical staining can help confirm the cell origin of this tumor?

A. CD31

B. Desmin

C. SOX10

D. Carcinoembryonic antigen

E. CK7

Answers:

A. CD31 — Incorrect. This tumor originates from eccrine ductal differentiation. CD31 is an endothelial cell marker and helps confirm a tumor of vascular origin.

B. Desmin — Incorrect. Desmin is a mesenchymal marker for skeletal and most smooth muscle cells.

C. SOX10 — Incorrect. SOX10 is a nuclear marker of melanocytes and Schwann cells and stains positively in melanoma.

D. Carcinoembryonic antigen — Correct. EPC is a tumor of eccrine ductal differentiation. Immunohistochemical staining with carcinoembryonic antigen positively stains the neoplastic eccrine cells lining the ducts and clefts. Additional helpful positive stains include epithelial membrane antigen, keratin, p53, and p63.[1]

E. CK7 — Incorrect. CK7 stains glandular epithelium and helps determine the origin of metastatic carcinoma when used in conjunction with CK20. It is also positive in Paget's disease and extramammary Paget's disease. It is not specific to eccrine cells.

Question 3: What is the initial treatment of choice?

A. Wide local excision of the lesion

B. Pembrolizumab

C. Chemotherapy

D. Acitretin

E. Radiation

Answers:

A. Wide local excision of the lesion – Correct. Given the rarity of this tumor there are no work-up or treatment guidelines, however wide local excision is currently considered the mainstay of treatment for EPC.[1,2] Use of Mohs micrographic surgery for treatment of EPC is increasing and its use can be considered for tumors with high-risk features.[3] Radiologic staging should be considered to evaluate for lymph node involvement or metastasis. EPC holds a high risk of recurrence and mortality, and multistep management is frequently required for successful treatment.[2]

B. Pembrolizumab – Incorrect. Immunotherapy with pembrolizumab is not currently considered a primary treatment for EPC. There are reports of successful pembrolizumab use for metastatic porocarcinoma; however further research is necessary to elucidate its safety and effectiveness for EPC.[4]

C. Chemotherapy – Incorrect. Chemotherapy has been used for EPC management as an adjunct therapy, for metastasic disease, or recurrence of disease. However, no standardized regimen exists and evidence of its effectiveness is lacking.[1]

D. Acitretin – Incorrect. Acitretin is still being studied as a treatment for certain nonmelanoma skin cancers and is not currently used to treat EPCs.

E. Radiation – Incorrect. Although radiation has been used concomitantly with wide local excision, radiation alone is not considered first-line treatment of EPCs. High metastasis rates have been seen in patients treated with radiotherapy for EPC.[2]

Abbreviation used:

EPC: eccrine porocarcinoma

Key words

adnexal neoplasm; hidradenocarcinoma; malignant mixed tumor; porocarcinoma; poroma

Conflicts of interest

None disclosed.

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