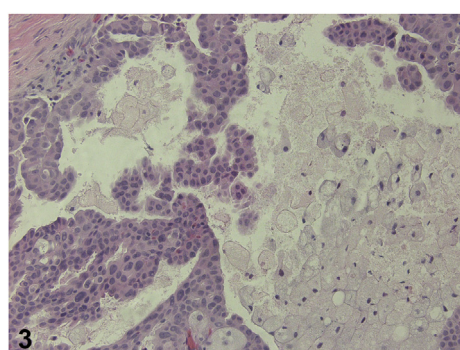
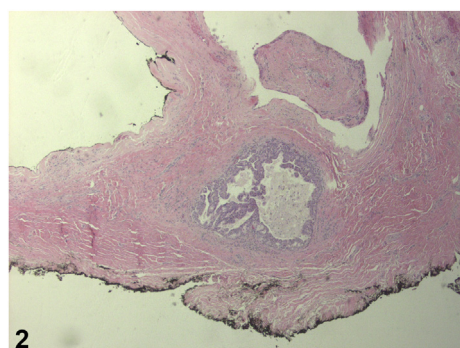
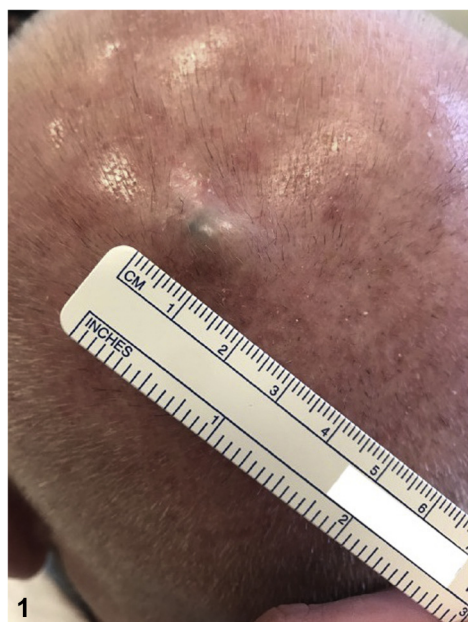


A slowly growing blue nodule on the scalp



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A 66-year-old man presented with a 1.5-cm subcutaneous blue nodule on the vertex of his scalp gradually increasing in size over several years (Fig 1). Excisional biopsy found dilated duct-like structures forming intraluminal papillation of variable complexity with loss of myoepithelial layers. Lumina were filled with amorphous eosinophilic material and histiocytes. The lining cells were atypical with prominent nucleoli (Figs 2 and 3). By immunohistochemistry, the cells were positive for AE1/AE3, GATA-3, p63, cytokeratin 7, EMA, and CEA. Estrogen receptor, progesterone receptor, CD117, CK5/6, and TTF-1 immunostains were negative.

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Question 1: Which of the following is the most likely diagnosis?

- A. Low-grade mucoepidermoid carcinoma
- B. Primary cutaneous apocrine carcinoma
- C. Adenoid cystic carcinoma
- D. Trichilemmal cyst
- E. Primary cutaneous mucinous carcinoma

Answers:

A. Low-grade mucoepidermoid carcinoma – Incorrect. Low-grade mucoepidermoid carcinoma is composed of mucinous and intermediate cells with bland nuclei forming glandular spaces.

B. Primary cutaneous apocrine carcinoma – Correct. The clinical presentation of a slowly enlarging nodule with the histopathology described is consistent with a diagnosis of primary cutaneous apocrine carcinoma.

C. Adenoid cystic carcinoma – Incorrect. Histologically, adenoid cystic carcinoma reveals a deep dermal tumor, often with subcutaneous extension, and is composed of nests of basaloid cells with cribriform and tubular pattern with mucin in the cysts and between the cells. It often displays perineural invasion, and, although it is positive for cytokeratins, it is diffusely positive for CD117 immunostain.

D. Trichilemmal cyst – Incorrect. Although these lesions often occur on the scalp, histology most often shows cystic structures lined by small layers of cuboidal epithelial cells with trichilemmal differentiation lacking a granular layer.¹ Prominent nucleoli and atypical cytologic features would not be seen without malignant transformation.

E. Primary cutaneous mucinous carcinoma – Incorrect. Histologically, these tumors are composed of an infiltrating duct pattern in a background of mucinous pools. Although cytokeratin 7 is positive in mucinous carcinoma, estrogen receptor and progesterone receptor immunostains are also positive in these tumors.

Question 2: What type of neoplasm morphologically resembles primary cutaneous apocrine carcinoma when viewed under the microscope and should be considered before making the diagnosis?

- A. Adenoid basal cell carcinoma
- B. Metastatic papillary lung adenocarcinoma

C. Metastatic papillary urothelial carcinoma of the bladder

D. Metastatic ductal mammary carcinoma

E. Sebaceous carcinoma

Answers:

A. Adenoid basal cell carcinoma – Incorrect. Adenoid basal cell carcinoma consists of infiltrating round nests of basal keratinocytes and scant mucin in the stroma.

B. Metastatic papillary lung adenocarcinoma – Incorrect. Papillary lung adenocarcinoma is composed of infiltrative glandular structures containing fibrovascular cores. The cells are positive for lung markers including TTF-1 immunostain.

C. Metastatic papillary urothelial carcinoma of the bladder – Incorrect. Papillary urothelial carcinoma of the bladder appears exophytic in nature with papillae containing fibrovascular cores.

D. Metastatic ductal mammary carcinoma – Correct. A distinction between primary cutaneous apocrine carcinoma and metastatic ductal mammary carcinoma is nearly impossible based on morphology alone, and both immunohistochemical staining and breast imaging are necessary to make a diagnosis. Positivity for the immunohistochemical markers p63, Ck5/6, GATA-3, and D2-40 has been reported as a sensitive marker for primary lesions of the skin, but these stains do not rule out metastatic disease. In any case of suspected primary cutaneous apocrine carcinoma, imaging such as breast ultrasound scan, mammography, or magnetic resonance imaging is essential to establish that a primary cancer from the breast has not metastasized to the skin.²

E. Sebaceous carcinoma – Incorrect. Sebaceous carcinoma is composed of infiltrative basaloid cells with scant cytoplasm, mitotic figures, and intervening foamy cells.

Question 3: Follow up computed tomography scan of the chest, abdomen, and pelvis found no evidence of metastatic disease. Breast ultrasound scan and colonoscopy show no signs of a visceral cancer, and a diagnosis of primary cutaneous apocrine carcinoma was made. Which of the following is the most appropriate next step in management?

A. Wide local excision with sentinel lymph node biopsy

B. Excision with 4 mm margins

- C. Denosumab therapy
- D. Chemotherapy and radiation
- E. Follow up examination in 3-6 months

Answers:

- A.** Wide local excision with sentinel lymph node biopsy – Correct. Many of these neoplasms are slow growing and indolent, but rapid progression and metastasis can occur. Wide local excision with or without lymph node biopsy is the standard of care.³
- B.** Excision with 4-mm margins – Incorrect. These margins are suitable for small, well-defined tumors like basal cell carcinoma with lower metastatic potential.
- C.** Denosumab therapy – Incorrect. Although RANK-L is expressed in cancers of apocrine origin and denosumab has been reported as successful treatment in advanced primary cutaneous apocrine carcinoma, wide local excision with lymph node biopsy is the next best step in treatment.⁴

D. Chemotherapy and radiation – Incorrect. Chemotherapy and radiation may be indicated if there is evidence of metastatic disease, but not before the primary lesion is adequately treated with wide local excision.

E. Follow-up examination in 3 to 6 months – Incorrect. Wide local excision with clear margins is indicated.

REFERENCES

1. Al Aboud DM, Patel BC. Pilar Cyst. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; 2020.
2. Mentrikoski MJ, Wick MR. Immunohistochemical distinction of primary sweat gland carcinoma and metastatic breast carcinoma: can it always be accomplished reliably? *Am J Clin Pathol*. 2015;143(3):430-436.
3. Loh SH, Oh YJ, Lew BL, Sim WY. Primary Cutaneous Apocrine Carcinoma. *Ann Dermatol*. 2016;28(5):669-670.
4. Furudate S, Fujimura T, Tsukada A, et al. Successful treatment of advanced primary cutaneous apocrine carcinoma on the scrotum with systemic chemotherapy and radiotherapy followed by denosumab. *Case Rep Oncol*. 2017;10(1):52-56.