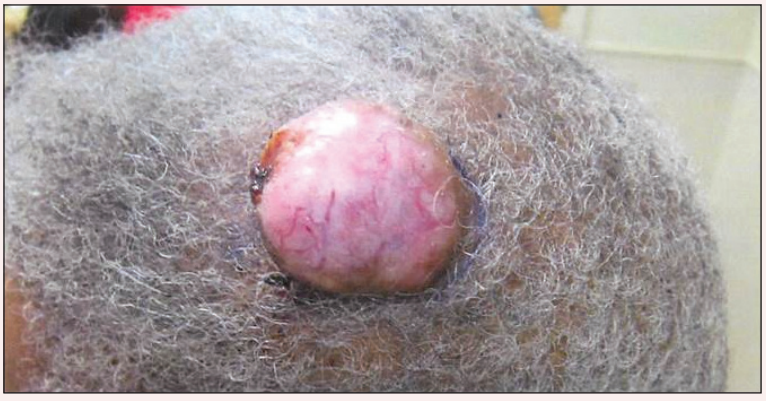
Exophytic Scalp Tumor

Gabrielle Cervoni, MD; William E. Steffes, MD; Katherina B. Kobraei, MD; Ann A. Church, MD



An 81-year-old man presented with a 3.5×3.0-cm pink exophytic tumor with an eroded surface and prominent vascularity on the left side of the parietal scalp. The patient reported that the tumor had been present for more than 30 years but recently had grown larger in size. He denied pain or pruritus in association with the lesion and did not report any systemic symptoms. He had received no prior treatments for the tumor.

What's the diagnosis?

a. keratoacanthoma
b. malignant eccrine spiradenoma
c. primary cutaneous carcinosarcoma
d. syringoid sweat duct carcinoma
e. trichoepithelioma

Dr. Cervoni is from the Department of Surgery, Beth Israel Deaconess Medical Center, Boston, Massachusetts. Drs. Steffes, Kobraei, and Church are from the Department of Dermatology, University of Florida, Gainesville.
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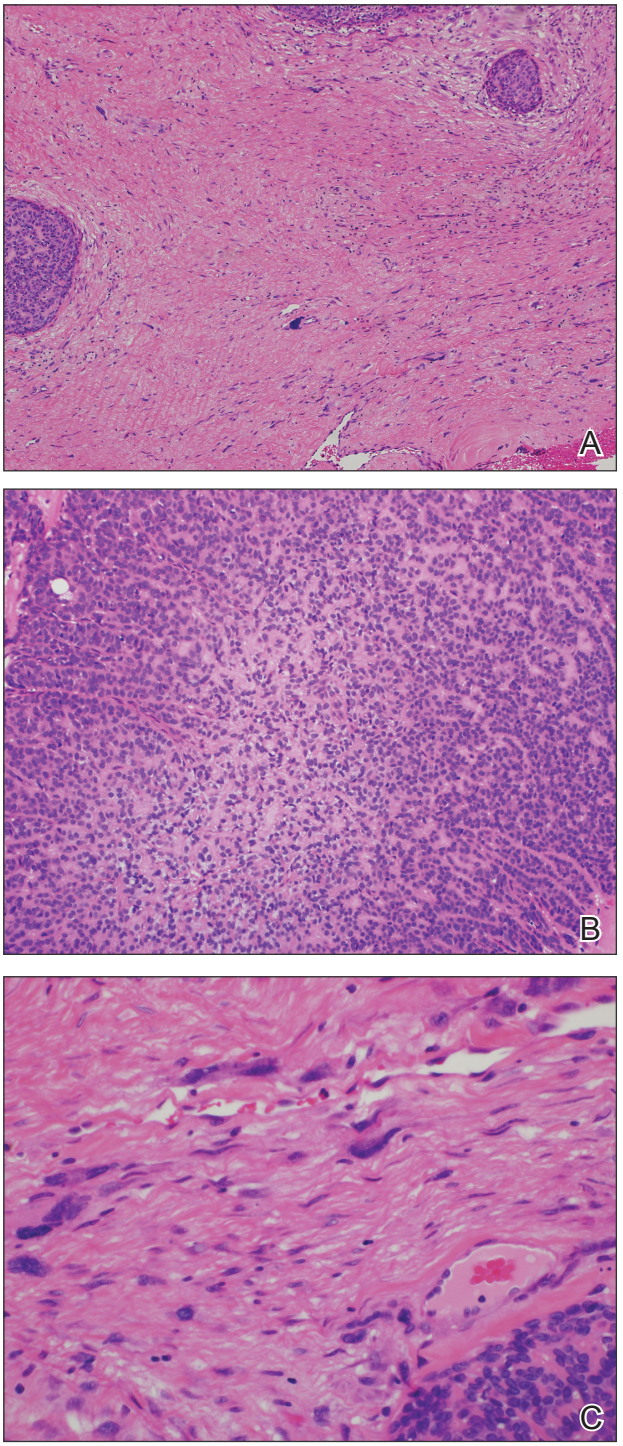
Correspondence: Gabrielle Cervoni, MD, 330 Brookline Ave, Boston, MA 02215 (gcervoni@bidmc.harvard.edu).

The Diagnosis: Primary Cutaneous Carcinosarcoma

A generous shave biopsy and debulking performed on the initial visit revealed an infiltrating tumor consisting of malignant epithelial and stromal components (Figure). The basaloid and squamoid epithelial cells were keratin positive. The stromal cells demonstrated positivity for CD10 but were keratin negative. The epithelial portion of the tumor was composed mostly of basaloid islands of cells with nuclear pleomorphism, scattered mitoses, and focal sebaceous differentiation. The mesenchymal portion of the tumor displayed florid pleomorphism and polymorphism, with many large atypical cells and proliferation. A diagnosis of primary cutaneous carcinosarcoma (PCC) was rendered. Head and neck computed tomography showed tumor penetration of less than 1 cm into scalp soft tissues with no involvement of the underlying bone. There was some evidence of swelling of the supragaleal soft tissues without indication of perineural spread. An 11-mm hyperlucent lower cervical lymph node on the left side that likely represented an incidental finding was noted. Surgical excision with margin evaluation was recommended, but the patient declined. He instead received radiation therapy to the left side of the posterior scalp with a total dose of 30 Gy at 6 Gy per fraction and 1 fraction daily. The patient was found to have a well-healed scar with no evidence of recurrence at 4-week follow-up and again at 5 months after radiation therapy.

Primary cutaneous carcinosarcoma is a rare biphasic neoplasm of unknown etiology that is characterized by the presence of both malignant epithelial and mesenchymal components.[1] Carcinosarcomas have been reported in both the male and female reproductive tracts, urinary tract, gastrointestinal tract, lungs, breasts, larynx, thymus, and thyroid but is uncommon as a primary neoplasm of the skin.[2] Epidermal PCC occurs with greater frequency in males than in females and typically presents in the eighth or ninth decades of life.[3] These tumors tend to arise in sun-exposed regions, most commonly on the face and scalp.[2]

Morphologically, PCCs typically are exophytic growths that often feature surface ulceration and may or may not bleed upon palpation.[4] Primary cutaneous carcinosarcomas may present as long-standing lesions that have undergone rapid transformation in the weeks preceding presentation.[4] It is not uncommon for PCC lesions to carry the clinical diagnosis



A generous shave biopsy and debulking performed on the initial visit revealed an infiltrating tumor consisting of malignant epithelial and stromal components (A–C)(H&E; original magnifications ×10, ×20, and ×40, respectively).

of squamous cell carcinoma, which suggests notable morphologic overlap between these entities. Histopathologically, PCC shows a basal cell carcinoma and/or a squamous cell carcinoma epithelial component intimately admixed with a sarcomatous component.[5] The mesenchymal component of PCC typically resembles a superficial malignant fibrous histiocytoma characterized by pleomorphic nuclei and cytoplasm, necrosis, and an increased number of mitotic figures.[2] Immunohistochemistry can be beneficial in the diagnosis of PCC. A combination of p63 and AE1/AE3 stains can be used to confirm cells of epithelial origin. Staining with vimentin, CD10, or caldesmon can help to delineate the mesenchymal component of PCC.

Epidermal PCC most commonly affects elderly individuals with a history of extensive sun exposure. It has been suggested that p53 mutations due to UV damage are key in tumor formation for both epithelial and mesenchymal elements.[5] Literature supports a monoclonal origin for the epithelial and mesenchymal components of this tumor; however, there is insufficient evidence.[6] Surgical excision is the primary treatment modality for epidermal PCC, but adjuvant or substitutive radiotherapy has been used in some cases.[4] The prognosis of PCC is notably better than its visceral counterpart due to early diagnosis and treatment of easily visible lesions. Epidermal PCC has a 70% 5-year disease-free survival rate, while adnexal PCC tends to occur in younger patients and has a 25% 5-year disease-free survival rate.[3] Due to the rarity of reported cases and limited follow-up, the long-term prognosis for PCC remains unclear.

We report an unusual case of PCC on the scalp that was successfully treated with radiation therapy alone. This modality should be considered in patients with large tumors who refuse surgery or are not good surgical candidates.

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