Carcinosarcoma: A Primary Cutaneous Tumor With Biphasic Differentiation

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Practice Points

- Carcinosarcoma is a rare biphasic tumor composed of 2 distinct malignant components that shows epithelial and mesenchymal differentiation.
- The tumor can be aggressive, with up to 19% of tumors recurring and metastases seen in 19% to 26%.
- The treatment of choice is surgical excision; no additional benefit is seen with adjunctive radiation.

We present a case of a primary cutaneous carcinosarcoma (PCS). Histopathologically, PCS is defined as a biphasic tumor composed of malignant epithelial (carcinoma) and mesenchymal (sarcoma) elements. The diagnosis of PCS can be challenging, not only because of its rarity but also because superficial biopsies can result in sampling errors. Accurate diagnosis is essential, as PCS carries a higher recurrence and metastatic rate than epithelial carcinomas, thus requiring wider excision or Mohs micrographic surgery as well as closer clinical follow-up.

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Case Report

A 63-year-old man presented with nonpainful scaly plaques on the scalp of several months' duration that intermittently bled. His medical history was remarkable for stage III neuroendocrine carcinoma on the scalp with 2 positive lymph nodes, which had been treated 2 years prior via wide local excision followed by adjunctive radiation. There was no evidence of recurrence at that site, and a review of systems was negative.

Physical examination revealed a 1.5×1.5-cm, eroded, erythematous plaque on the right vertex of the scalp (Figure 1). A shave biopsy of the lesion revealed an eroded acanthotic epidermis with elongated rete ridges composed of crowded atypical keratinocytes. The dermis contained a proliferation of atypical spindle cells arranged as irregular fascicles admixed with islands of pleomorphic keratinocytes containing keratinaceous material

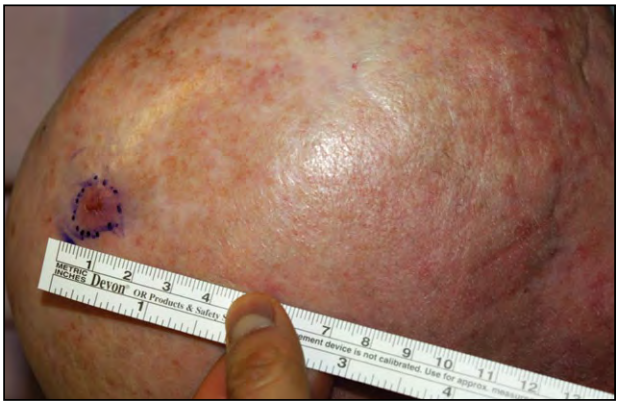

Figure 1. A 1.5×1.5-cm, eroded, erythematous plaque on the right vertex of the scalp.

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(Figure 2). Immunohistochemical stains confirmed 2 discrete cell populations. The spindle cells labeled with CD10, smooth muscle antibody, and vimentin (Figure 3), and were negative with high-molecular-weight cytokeratin, p63, epithelial membrane antigen, S-100, CD34, CD68, and CD99. The squamous islands only labeled with high-molecular-weight cytokeratin and epithelial membrane antigen (Figure 4). These findings supported a diagnosis of primary cutaneous carcinosarcoma (PCS) composed of an atypical fibroxanthoma admixed with a squamous cell carcinoma (SCC). Surgical excision was performed, and 33 months after primary excision, the patient remains free of recurrence.

Comment

Carcinosarcomas are rare tumors that most commonly arise in visceral organs. Although the true prevalence of PCS is unknown due to a lack of consensus in nomenclature and failure to report some cases, there are at least 94 cases. These cases have been reported under various names including metaplastic carcinoma, trichloblastic sarcoma, malignant mixed tumor, biphasic sarcomatoid carcinoma, sarcomatoid basal cell carcinoma, and sarcomatous carcinoma, with PCS most commonly used.[1-9] Primary cutaneous carcinosarcoma consists of an intimate admixture of malignant epithelial (carcinoma) and mesenchymal (sarcoma) elements in various proportions with no transition between the 2 components.[2] The epithelial component usually is a basal cell carcinoma or SCC and/or an adnexal carcinoma (eg, spiradenocarcinoma, pilomatrical carcinoma, porocarcinoma, trichilemmal carcinoma).[1,2] The mesenchymal component typically is an atypical fibroxanthoma, osteosarcoma, or chondrosarcoma, but leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, synovial sarcoma, and angiosarcoma also have been reported.[1,2,4,5] Immunohistochemical stains clearly demonstrate the biphasic nature of the tumor, with cytokeratins labeling the epithelial component and vimentin labeling the mesenchymal component.

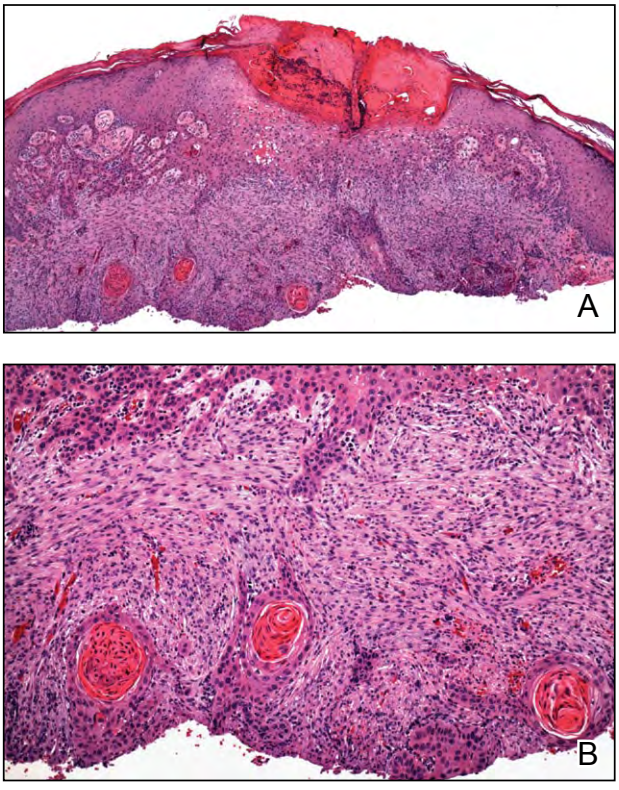

Figure 2. A shave biopsy of the lesion revealed a biphasic tumor composed of an atypical epithelial proliferation admixed with an atypical spindle cell proliferation (A) (H&E, original magnification ×40). A higher-power view showed the intimate admixture of a squamous cell carcinoma with an atypical fibroxanthoma (B)(H&E, original magnification ×100).

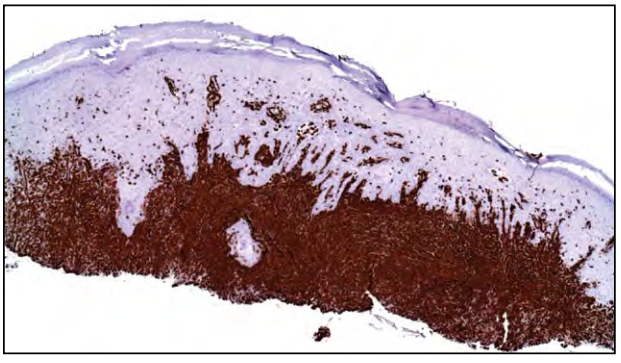

Figure 3. The spindle cell component diffusely labeled with vimentin (H&E, original magnification ×40).

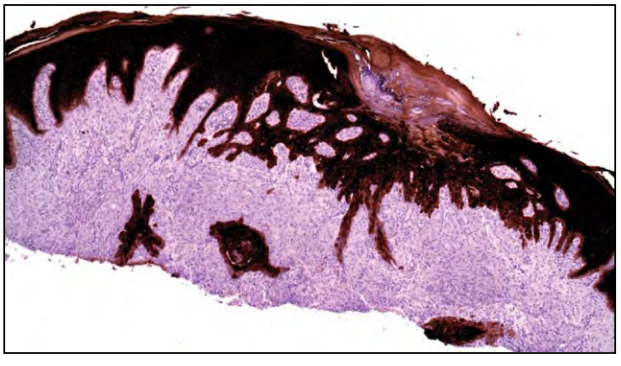

Figure 4. The epithelial component diffusely labeled with high-molecular-weight cytokeratin (H&E, original magnification ×40).

The etiopathogenesis of PCS is unknown, but 3 main hypotheses exist. The composition theory suggests the mesenchymal component is a pseudosarcomatous reaction to the epithelial carcinoma; however, because PCS can metastasize, this theory is unlikely.[4,5] The multiclonal hypothesis proposes that 2 or more stem cells of epithelial and mesenchymal origin independently converge. Currently, the favored hypothesis is that PCS has a monoclonal origin and undifferentiated pluripotent stem cells diverge into epithelial and mesenchymal components either synchronously or metachronously; this hypothesis is supported by studies showing identical p53 and K-*ras* mutations in both components.[2,5]

The diagnosis of PCS can be challenging, not only because of its rarity but also because superficial biopsies can result in sampling errors. Recognition of PCS is critical, as approximately 19% of tumors recur, and 19% to 26% metastasize to the lymph nodes or viscera, respectively.[1,2] Tran et al[2] proposed that the clinical presentation and prognosis of PCS may be influenced by the nature of the epithelial component. This hypothesis was supported by a subsequent study that showed similar findings.[1] Tumors containing a basal cell carcinoma or SCC typically arise as slow-growing lesions in elderly men with actinically damaged skin with a 5-year survival rate of 70%. Conversely, adnexal PCS has a 5-year survival rate of 25% and usually presents in middle-aged women as rapidly growing nodules within long-standing benign tumors.[2]

Treatment typically includes surgical excision or Mohs micrographic surgery with close clinical follow-up. Adjunctive radiotherapy has shown no therapeutic benefit.[3]

Conclusion

Primary cutaneous carcinosarcomas are uncommon cutaneous tumors of uncertain etiology. Recognition of the 2 distinct components of this tumor is essential to differentiate it from more common epithelial carcinomas because the prognoses are substantially different.

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