Primary Cutaneous Carcinosarcoma: A Case Report and Discussion of a Histological "Chimera"

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Abstract

Primary cutaneous carcinosarcoma is a rare and aggressive biphasic malignant neoplasm that exhibits both epithelial and mesenchymal components. This malignancy is more commonly described arising from organs such as the uterus, breast, bladder, and lung, and is rarely seen on the skin. The histopathogenesis of this neoplasm is unknown, but a prevailing divergence theory exists. It is imperative that this neoplasm be diagnosed and treated, as it can be fatal. Here we report a case of primary cutaneous carcinosarcoma presenting on the skin of an 86-year-old male.

Introduction

Primary cutaneous carcinosarcoma (PCC) is a rare neoplasm not commonly found on the skin. To our knowledge, fewer than 100 cases of PCC have been reported in world literature.[1] Carcinosarcoma is most often observed in organs other than the skin including the uterus, breast, urinary bladder, and lungs.[1,2] When it does occur on the skin, it is typically found on an elderly male in sun-exposed areas of the head, neck, and upper extremities.[3] Clinically, the lesion is often exophytic and ulcerated and develops rapidly.[4]

Case Report

An 86-year-old male presented with a lesion on his left cheek of 3 months' duration. On physical exam, there was a 1.0 cm x 1.2 cm, ill-defined, red, friable nodule on the patient's left cheek. The clinical impression was of basal-cell carcinoma.

After a shave biopsy, routine H&E stains of the lesion revealed a poorly differentiated, biphasic malignant neoplasm comprised of trabecular arrangement of pleomorphic cells with considerable cytoplasm juxtaposed with atypical cellular hyperchromatic malignant stroma (**Figure 1**). Immunohistochemical stains revealed the pleomorphic cells with considerable cytoplasm were positive for cytokeratin (**Figure 2**) and p63 (**Figure 3**), while the intervening atypical stromal cells were positive for vimentin (**Figure 4**) and CD10. Both cell populations were negative for neuroendocrine markers. Computed tomography of the neck and chest was negative for locoregional lymphadenopathy. This microscopic and radiographic analysis was consistent with primary cutaneous carcinosarcoma.

The patient was treated with Mohs micrographic surgery and remains disease-free at three months.

Discussion

Primary cutaneous carcinosarcoma is an aggressive tumor composed of carcinomatous and sarcomatoid cells. The epithelial component is most commonly a basal-cell carcinoma or squamous-cell carcinoma, but it can also be associated with adnexal-derived tumors including spiradenocarcinoma, porocarcinoma, proliferating trichilemmal cystic carcinoma, and metrical carcinoma. The mesenchymal component may be of osseous, cartilaginous or, more rarely, skeletal- or smooth-muscle lineage.[5]

Although the histopathogenesis of PCC is unknown, there are two common theories at present. The prevailing hypothesis, also known as the divergence or monoclonal hypothesis, states that a single stem cell undergoes divergent differentiation into separate epithelial and mesenchymal elements.[5] The less likely convergence hypothesis proposes that the tumor arises from two or more stem cells of epithelial and mesenchymal origin that independently converge.[1,2,5]

Immunohistochemical stains are important for the diagnosis of carcinosarcoma. Cytokeratin highlights the epithelial elements, while vimentin highlights the mesenchymal elements.[1] Two studies emphasize the role of p63, a homologue of the tumor suppressor gene p53, in confirming epithelial derivation of poorly differentiated or metaplastic carcinomas.[6,7] It is thought that p63 is involved in the prevention of terminal squamous stem-cell differentiation and can be the key to establishing an epithelial presence in a tumor.[6] Pure sarcomas and carcinomas are negative for p63, thus p63 staining is highly specific for diagnosing metaplastic carcinomas like PCC.[7] Our tumor demonstrated positive staining for

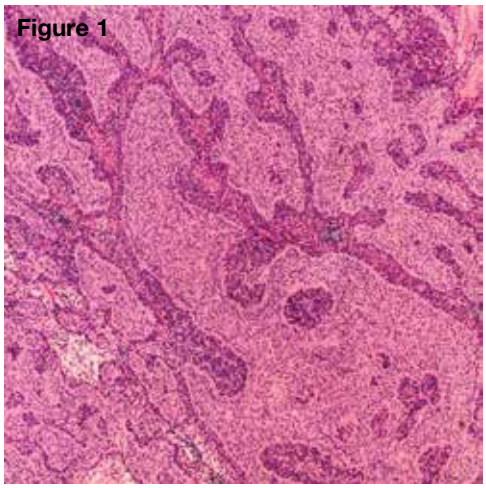
Figure 1

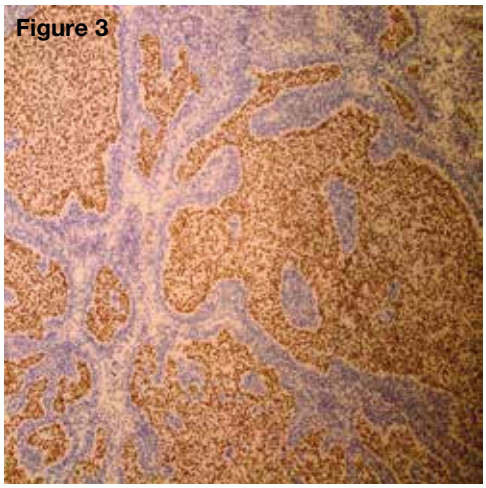
Figure 3

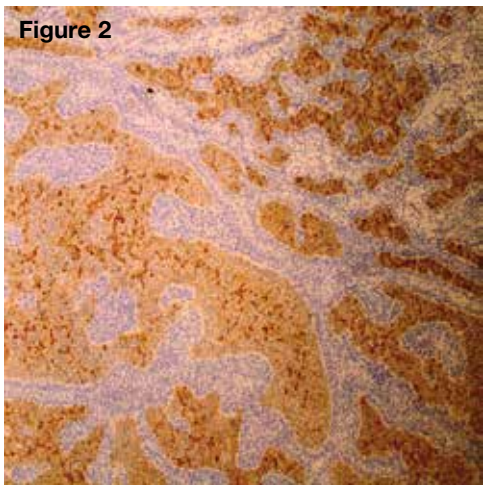
Figure 2

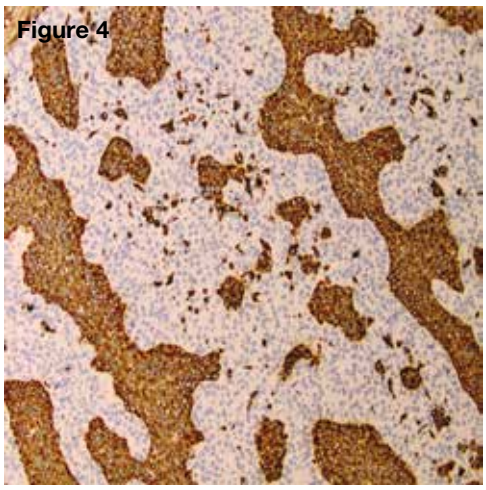
Figure 4

CD10 in the mesenchymal component, but the significance of this is unclear as this pattern is recognized in both basal-cell carcinomas (epithelial lineage) and atypical fibroxanthomas (mesenchymal lineage).[2]

Treatment of PCC is predominantly surgical with wide local excision or, as in the case of our patient, Mohs micrographic surgery. Adjuvant radiotherapy is not currently recommended.[4] Regular clinical follow-up is paramount.

Cutaneous carcinosarcomas typically have a better prognosis than carcinosarcomas arising in visceral organs, but nonetheless these tumors can be aggressive. Prognosis seems to be most closely linked to the origin of the epithelial component. One meta-analysis found that PCCs containing a basal- or squamous-cell carcinoma had a five-year survival rate of 70%.[8] Conversely, PCCs with an epithelial element of adnexal origin have a poorer prognosis, with a 25% five-year disease-free survival rate.[5] Other poor prognostic factors include age younger than 65, tumor size greater than 2 cm, a recent growth pattern, longer duration of existing skin tumor, and metastasis to lymph nodes.[3,5] Even after surgical excision, 7% to 19% of PCCs recur.[1,9] Diagnosis and treatment is necessary, with locoregional and distant metastases documented in 19% and 26% of cases, respectively.[1] PCC can also be fatal, with one report documenting PCC with cerebral metastases resulting in death.[10]

Conclusion

PCC is an admixed malignancy of epithelial and mesenchymal components. The diagnosis of this rare neoplasm is critical given its high rate of recurrence, metastases, and occasional mortality. These risks are especially notable when the lesion clinically resembles an unexceptional basal-cell carcinoma, as in our case. It is necessary to increase knowledge and awareness of this uncommon and aggressive histologic "chimera."

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