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Case Presentation

Malignant combined squamomelanocytic tumor: a clinical case

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Abstract

A combined squamomelanocytic tumor is an exceedingly rare occurrence; little is known about its pathogenesis. A definitive diagnosis can only be made via histological examination. We describe herein an 83 year-old man who was discovered to have this combined tumor and recommend the appropriate management for such a lesion.

Introduction

A combined squamomelanocytic tumor can be defined by the presence of both squamous cell carcinoma (SCC) and malignant melanoma (MM) within a single lesion. Both populations are distinct and intermingled within one another. It is unknown exactly how these tumors come to form, but one theory suggests the SCC interacts with the MM cells, influencing its development [1]. Considering the nature of both of these cell types individually, the initial treatment recommendation is wide local excision.

Report

An 83 year-old man presented with a fairly well demarcated dark brown, and focally ulcerating tumor measuring 0.7 cm in diameter in the temporal region (Figure 1). The lesion was firm and nodular. The initial differential diagnosis included MM, pigmented basal cell carcinoma (BCC), and pigmented SCC. The surgeon performed a wide excision of the tumor with a 2 cm margin and sent the specimen to pathology.

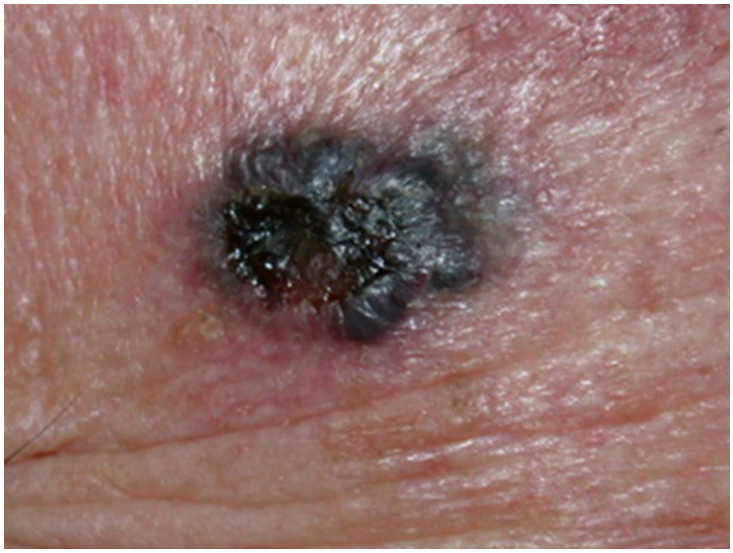
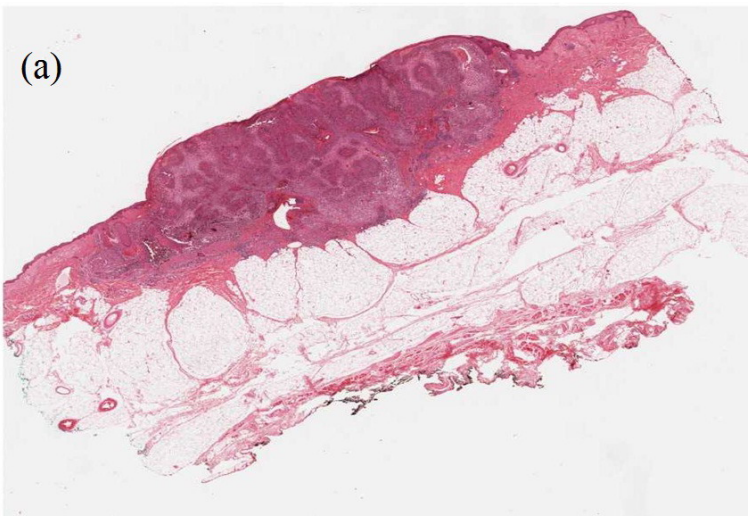


Figure 1. Dark brown focally ulcerated nodule.

On histological examination, there was a nodule with borders extending deep into the reticular dermis. (Figure 2a, H&E stain). At high power, the tumor originated from the surface epithelium and there were two populations of atypical epithelioid cells. (Figure 2b, H&E stain). Mostly these epithelioid cells showed squamous features with moderate to abundant pink cytoplasm and focal keratin pearls (Figure 2c), but there was a distinctly different population of atypical epithelioid cells containing granular melanin pigment (Figure 2d). In areas these pigmented cells were closely intermingled with squamous cells (Figure 2e).

Immunohistochemical study showed strongly positive staining of a majority of cells for HMW Cytokeratin (Figure 2f) confirming squamous differentiation. The pigmented cells were negative for HMW Cytokeratin (Figure 2g, arrows). Stains for MELAN-A (Figure 2h), S100 and HMB45 were positive in the pigmented cells and negative in squamous cells. Stains for P63 showed diffuse nuclear staining in squamous cells with scattered pigmented cells negative (Figure 2i, arrows for negative staining of pigmented cells). Electron microscopic study demonstrated melanosomes and keratin tonofilaments in some cells.



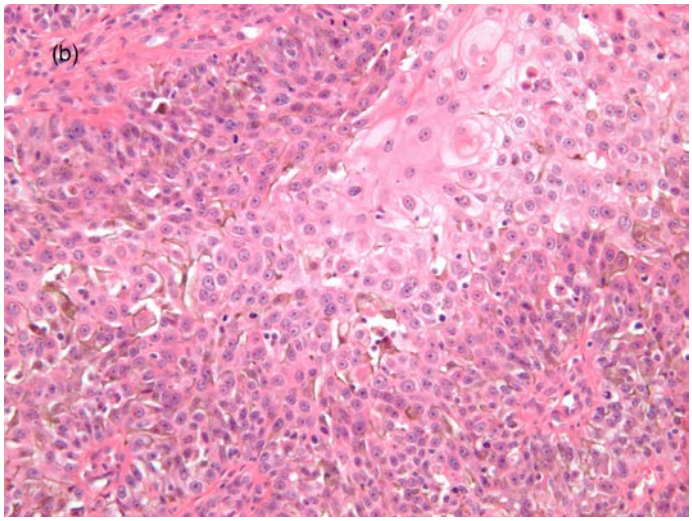
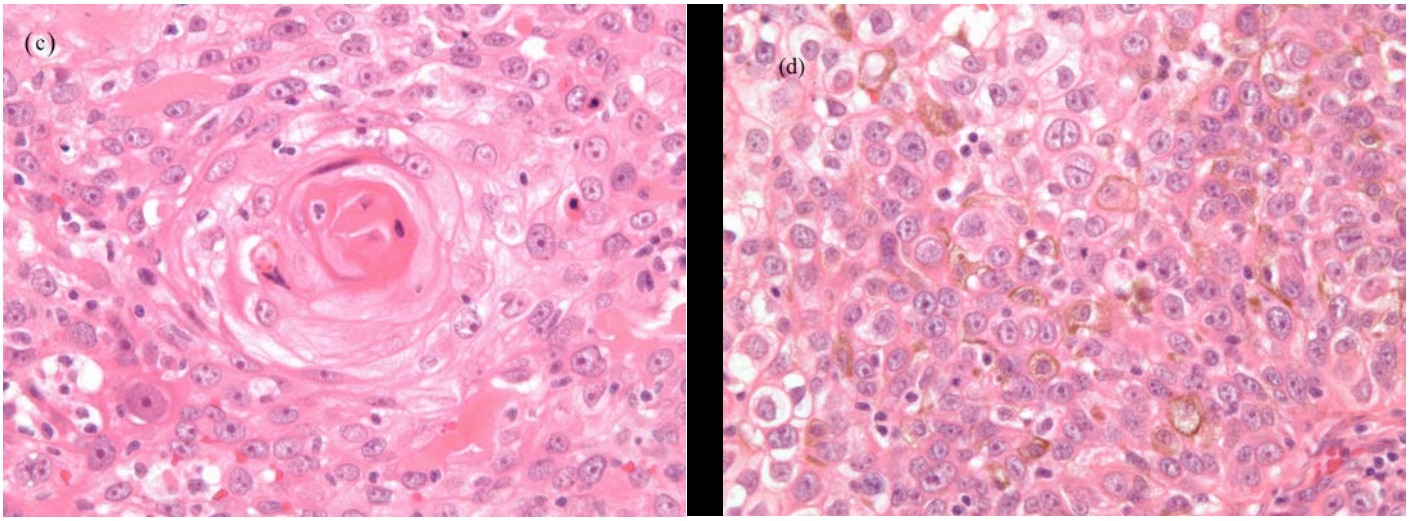


Figure 2. (a) Histology of dermal nodule showing pushing borders, H&E stain (magnification x 40).

Figure 2. (b) Low power of squamomelanocytic tumor showing 2 distinct populations of cells (magnification x 200)



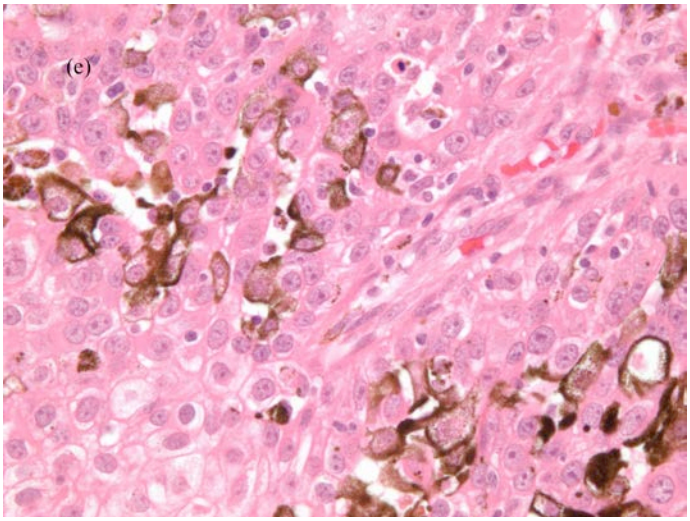


Figure 2. (c,d,e) Atypical squamous cells showing keratin pearl (c) and atypical melanocytes containing melanin pigment (d, e) on higher magnification. (H&E stain, magnification x 400).

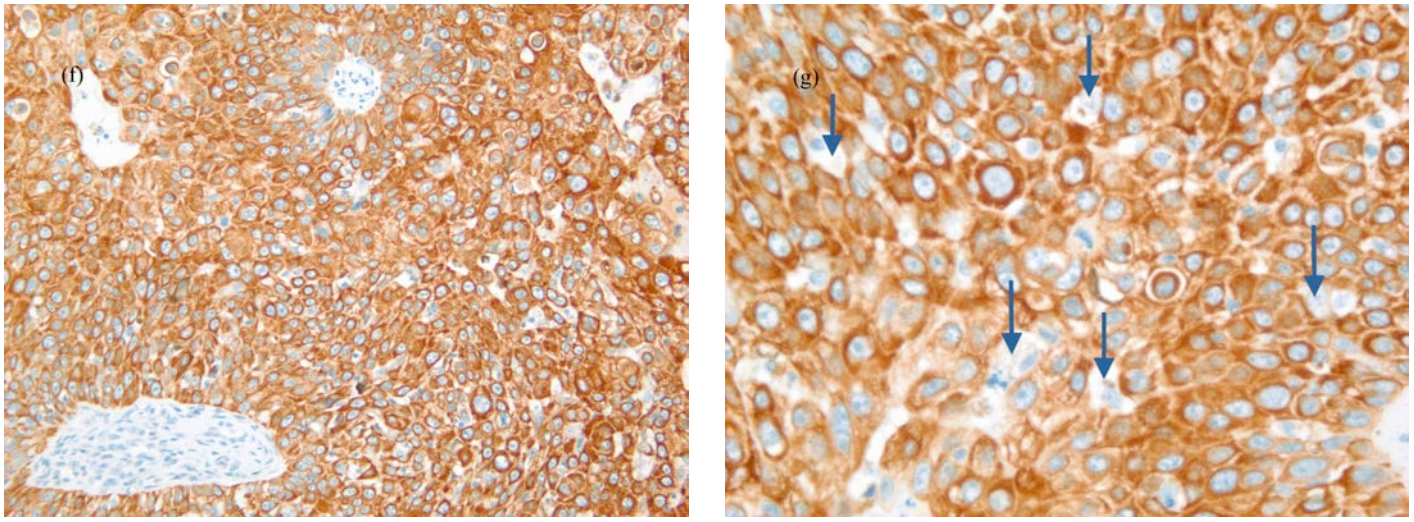


Figure 2. (f, g) Immunohistochemistry shows HMW cytokeratin staining in atypical squamous cells; arrows show negative staining in atypical melanocytes (f: magnification x 200 and g: magnification x 400).

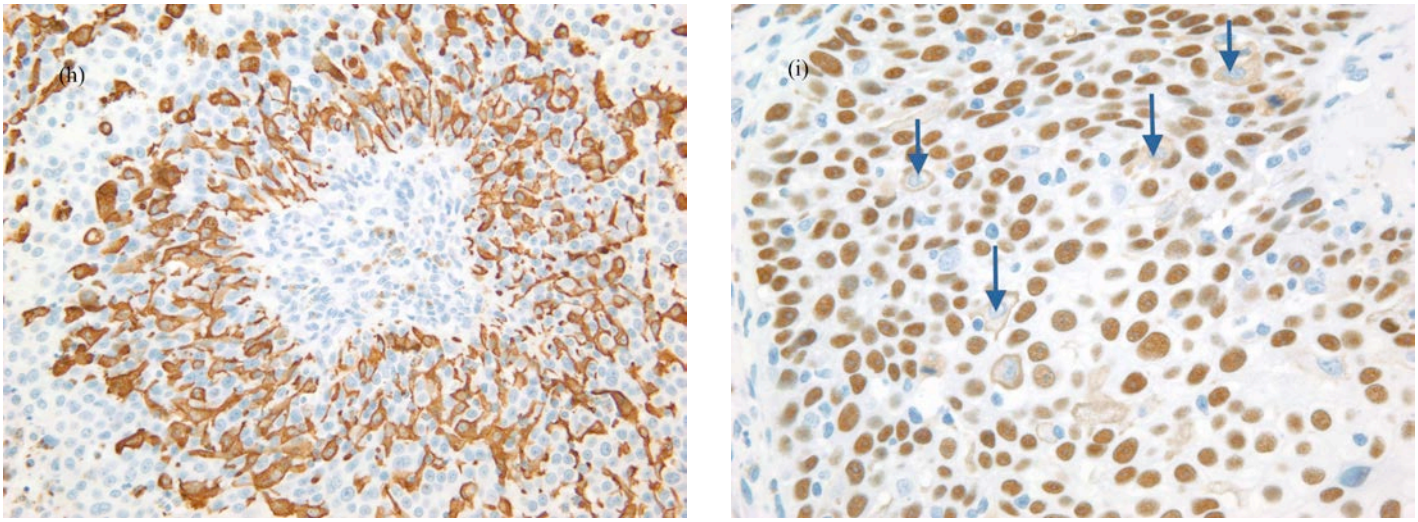


Figure 2. (h) Immunohistochemistry shows Melan-A staining in atypical melanocytes (magnification x 200).

Figure 2. (i) Immunohistochemistry shows P63 nuclear staining in atypical squamous cells; arrows show negative staining in atypical melanocytes (magnification x 400).

Discussion

The clinical differential diagnosis for a malignant pigmented skin tumor usually includes malignant melanoma (MM), pigmented basal cell carcinoma (BCC), and pigmented squamous cell carcinoma (SCC). The diagnosis of a malignant combined squamomelanocytic tumor is an exceedingly rare occurrence. It involves the presence of two distinct yet intertwined populations of malignant squamous epithelial and melanocytic cells merged together. The histogenesis is speculative. One theory is that there is malignant proliferation of two distinct phenotypes with the primary tumor (squamous cell carcinoma) initiating a second neoplasm by paracrine effect (interaction theory) [1]. Our case shows two populations of cells. One shows squamous features and another shows epithelioid cells with melanin pigment, consistent with melanoma cells. Immunohistochemistry using appropriate markers for squamous and melanocytic cells is essential to evaluate both populations and to confirm the diagnosis. Histologically combined squamomelanocytic tumors should be distinguished from other combination tumors such as squamous cell carcinoma with benign melanocytic colonization and collision tumor. We do not think that this is a pigmented squamous cell carcinoma with benign melanocytic colonization because the pigmented cells appear neoplastic and they express melanocytic markers by immunostudy. In addition, there are non pigmented squamous cells that do not express melanocytic markers.

The most recent reviews report only 13 previous cases of malignant squamomelanocytic tumors [1,2]. These tumors have occurred mostly in middle-aged to older adults spanning an age range of 32-98. The majority of tumors arise in the head and neck region and there appears to be a predilection in male patients (approximately 2:1). Almost all reported cases occur in Caucasians as well

[3]. Most cases to date have been pigmented squamomelanocytic tumors. However, there are also several cases involving a non-pigmented form [1,4,5,6]. The depth of these tumors has generally been around 2 mm, but one case has reached up to 9 mm [1,5].

Despite the malignant nature of this neoplasm, there have not been any reported cases of metastasis in follow-up. This may be more representative of the limited number of cases and the lack of sufficient follow-up time for most of them. Alternatively it is feasible that the neoplastic melanocytes in this type of combined tumor are sparse and less aggressive in behavior compared to traditional melanoma cells and that a combined squamomelanocytic tumor likely would behave like a squamous cell carcinoma rather than a melanoma. Nevertheless, in this case report, our patient had a 3-year follow-up without any local recurrence or metastasis.

Malignant squamomelanocytic tumor is a unique biphasic neoplasm with uncertain biological behavior. The preferred treatment of choice for malignant squamomelanocytic tumors is wide surgical excision. Follow-up monitoring for recurrence and metastasis is an important precautionary measure at this point.

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