Sarcomatoid squamous cell carcinoma: a long-standing case

R.S. Singh, P.B. Bhattacharjee, G.A. Youngberg, and M.A. Al-Abbadi

SUMMARY

Sarcomatoid, or spindle cell squamous cell carcinoma (SCSC), is relatively uncommon, but may be encountered. It poses a challenge in differential diagnosis that includes other spindle cell neoplasms. We present a case where the lesion existed for 20 years and raised the potential of chronicity as a factor in inducing spindle cell morphology. Detailed immunohistochemical features are demonstrated, and discussion of the differential diagnosis is offered. The patient was an 89-year-old African-American female with an exophytic and polypoid mass of the right upper arm measuring 5.5 × 5.5 × 3.0 cm. The mass had been present for the last 20 years, and was gradually and very slowly increasing in size. After refusing surgery several times, she finally agreed to have an excision. The tumor proved to be SCSC.

KEY WORDS

carcinoma, squamous-cell, sarcomatoid

Introduction

The differential diagnosis for cutaneous malignant spindle cell lesions poses special problems in dermatopathology. These typically include spindle cell squamous cell carcinoma (SCSC), "fibrohistiocytic tumors" such as atypical fibroxanthoma (AFX), superficial malignant fibrous histiocytoma (SMFH), and sarcomatoid malignant melanoma (SCMM) (1). These lesions manifest a great deal of clinical and histological overlap and pose the interesting challenge of separating them using conventional histopathology (2). These lesions may assume the form of crusted nodules or plaques, sometimes as large as 10 cm, with or without ulceration, and heaped-up white to gray margins of indurated tissue (3). These lesions mainly affect elderly individuals, and occur predominantly on sun-exposed areas. They may or may not show attachment between dermal tumor cells and the overlying epidermis, and usually display a varying proportion of spindle and pleomorphic cell morphology. Immunohistochemistry, despite certain limitations, plays a valuable role in the differentiation of problematic cases.

Case report

An 89-year-old African-American female patient checked into our hospital with a fungating and polypoid mass of the right upper arm. The mass displayed heaped up, elevated edges from the skin and apparent central ulceration measuring 5.5 x 5.5 x 3.0 cm. The process had been present for the last 20 years, and was gradually and

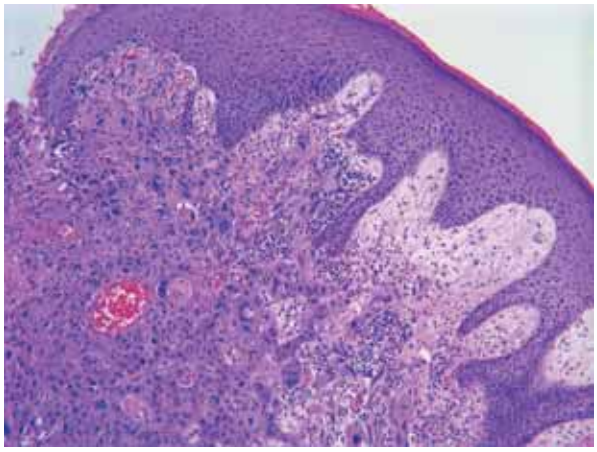

Figure 1. Section exhibits a high grade, pleomorphic spindle cell neoplasm with bizarre nuclei (H&E 200 ×).

very slowly increasing in size. The patient had been with her surgeon for more than 20 years, and no recent history of accelerated growth was mentioned. After refusing surgery several times over the years, she finally agreed to the excision, mainly because of its large size and unpleasant cosmetic effect. There was no history of burn or trauma. The histopathology revealed a high grade, pleomorphic spindle cell neoplasm with bizarre nuclei, and frequent abnormal mitotic figures within the dermis. The overlying epidermis was acanthotic and showed no connection to the dermal proliferation. Figure 1. As demonstrated in Figure 3, immunohistochemical stains were positive for Cytokeratin 7(CK 7), as well as Actin and AE 1/3; at the same time, CAM 5.2, cytokeratin 20 (CK 20), desmin, S-100 protein, HMB-45, vimentin and myogenin all tested negative. Based on histopathology morphology and immunohistochemistry features, the diagnosis of SCSC was made.

Discussion

It is not uncommon for poorly differentiated squamous cell carcinomas of the skin to assume an anaplastic fusiform morphology. Figure 2. The spindle cells have a large vesicular nucleus and scanty eosinophilic cytoplasm, often with indistinct cell borders. There is variable pleomorphism, usually with many mitoses. The presence of squamous differentiation, dyskeratotic cells, and continuity with the epidermis may assist in the diagnosis. However, these characteristics are not always detected even after thorough sampling. Similarly, the existence of overlying squamous cell carcinoma in situ or actinic keratosis has been reported only in a minority of cases (4). These cases are difficult to differentiate histologically from other spindle cell lesions, especially AFX. Whether chronicity and long duration play a role in transforming squamous cell carcinoma into spindle cell and sarcomatoid variants is not certain (5). One of the unusual findings in our case was the negative expression of vimentin by the tumor cells. The neoplastic cells of AFX are highly atypical and are variably fusiform or pleomorphic with few bizarre multinucleated cells (6). The predominant cells in AFX are plump, spindle-shaped, and occur in poorly arranged fascicles. The cells have a prominent nucleus which is often vesicular. Mitotic activity is usually brisk. Typically AFX cells shade off at the periphery to blend with the surrounding dermal fibroblasts; this feature when present can help make this diagnosis. Nevertheless, examples demonstrating deep infiltration of subcuticular adipose tissue are not uncommon.

Another important differential diagnosis is that of sarcomatoid melanomas. These are composed of amelanotic fusiform cells with no evidence of any atypical melanocytic proliferation in the overlying epidermis. Most demonstrate a disorganized tangle of atypical spindle cells in

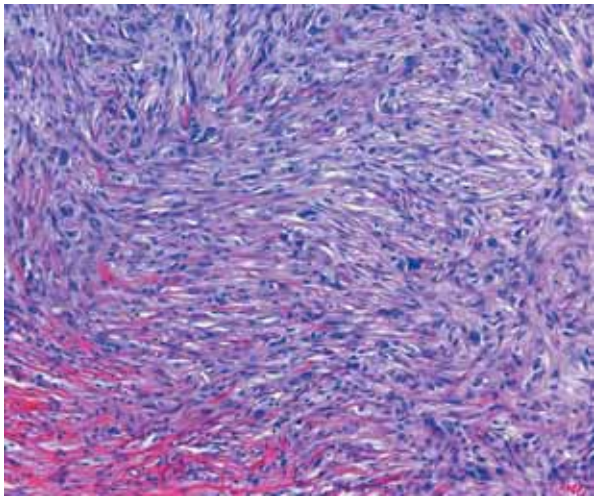

Figure 2. Pleomorphic spindleform cells with atypical mitoses are seen (H&E 200 ×).

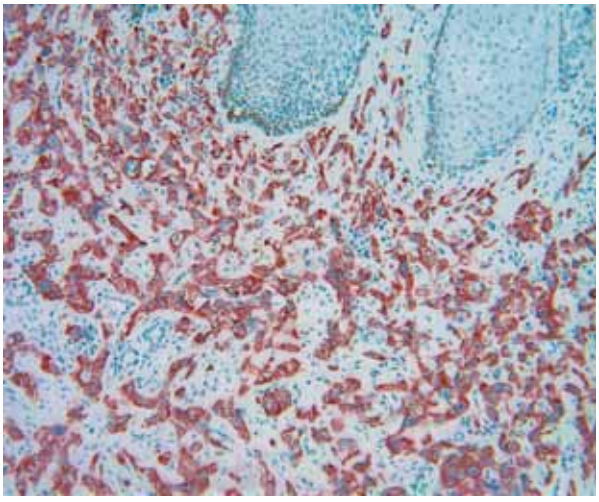

Figure 3: Immunohistochemical stain was positive for cytokeratin 7.

the dermis and subcutis, but some may assume a distinctly fascicular pattern. Nuclear pleomorphism, hyperchromatism, and nuclear enlargement are prominent; in rare cases the spindle cells may be frankly anaplastic mimicking high-grade sarcoma (7). Intranuclear inclusions and large eosinophilic nucleoli are helpful features when identified.

Due to the aforementioned histopathological overlap, immunohistochemical (IMH) evaluation using a panel of antibodies is essential. A battery of IMH stains are often used, including keratin, vimentin, EMA, S-100, HMB-45 and Factor X111a. SCSC is uniformly reactive for keratin, EMA, or both. However, each of these determinants may assume a focal distribution with positive tumor cells (8). Vimentin has been frequently reported to be expressed in SCSC, limiting its usefulness in the differential diagnosis (9). Sarcomatoid malignant melanomas are universally reactive for Vimentin and S-100 protein, but lack the expression of epithelial markers (10). AFX is usually characterized by its lack of epithelial and melanocytic markers, as well as its positivity for Vimentin and Factor X111a. Although some S-100 positive cells may be seen, these are usually Langerhans cells or nerve twigs (10).

Electron microscopic studies, though rarely needed, are potentially useful because SCSC retain a synthesis of cytoplasmic tonofibrils and intercellular desmoplastic attachments. SCMM seem to lose the capacity to form premelanosomes in most instances, and the neoplastic cells resemble fibroblasts or Schwann cells. Cells of AFX show abundant rough endoplasmic reticulum, small vesicles, and cytoplasmic filaments with small indentations in the nuclei. Other rare tumors that enter into the differential diagnosis of spindle cell lesions of the skin include dermatofibrosarcoma protuberans, leiomyosarcoma cutis, peripheral nerve sheath tumors, spindle cell angiosarcomas, carcinosarcoma and metastatic lesions. Because of its prognostic significance, it is important to recognize and separate the sarcomatoid malignant melanomas, as they have a more adverse biologic behavior in contrast to the others.

Although uncertain, this long-standing case of SCSC raises the possibility that chronicity and chronic irritation may be involved in inducing sarcomatoid morphology.

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AUTHORS' ADDRESSES

RS Singh, MD, Department of Pathology, Wayne State University, Detroit, MI
PB Bhattacharjee, MD, same address
George A. Youngberg, MD, Department of Pathology, East Tennessee State University, Johnson City, TN
Mousa A. Al-Abbadi, MD, Chief, Path & Lab Med Service, James H. Quillen Veterans Affairs Medical Center, Dept 113, Professor of Pathology, East Tennessee State University, P.O. Box 4000, Mountain Home, TN 37684, corresponding author, E-mail: mousa.al-abbadi@va.gov