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An Unusual Case of Squamoid Eccrine Carcinoma of the Abdominal Wall with Inguinal Nodal Metastases: A Case Report and Review of the Literature

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Abstract

Introduction: Squamoid eccrine carcinoma is a very rare carcinoma with few reported cases in the literature. As a result, there is limited guidance on management and follow-up of these cases.
Case Presentation: We describe the case of a 39 year-old male with a large painful squamoid eccrine carcinoma of the right lower abdominal wall with inguinal nodal involvement. He underwent radical resection, superficial groin dissection, transposition of a sartorius muscle flap, and a pedicled anterolateral thigh perforator flap for reconstruction. The postoperative course was uneventful apart from a postoperative seroma which was treated with aspiration. He underwent adjuvant radiation following full recovery from his procedure.
Conclusions: Our case study represents a rare, large squamoid eccrine carcinoma of the abdominal wall treated with radical resection, superficial groin dissection, transposition of a sartorius muscle flap, and a pedicled anterolateral thigh perforator flap for reconstruction. Due to the limited number of reported cases, there are no guidelines for management of squamoid eccrine carcinomas. With this case report we aim to increase awareness of this rare carcinoma, and to summarize the existing literature on management of squamoid eccrine carcinoma.

Keywords: Eccrine, Porocarcinoma, Sweat Gland Carcinoma

1. Introduction

Primary squamoid carcinoma of the sweat glands is a very rare type of carcinoma, with only a few hundred cases recorded in the medical literature within the past several decades (1, 2). Squamoid eccrine carcinoma represents between 0.005 and 0.006 percent of all malignant epithelial neoplasms (1, 2). Squamoid eccrine carcinoma arises primarily from the intraepidermal portion of the sweat gland (3). There are various cell types found within a squamoid eccrine carcinoma, including squamous cells, clear cells, spindle cells, and melanocytes, making the diagnosis challenging (4). The squamoid eccrine carcinoma is histologically characterized by an asymmetrical solid tumor with nodular growth pattern and infiltrative borders. The neoplastic cells within are seen with cytonuclear atypia, hyperchromatic nuclei, and enlarged nucleoli (4). These carcinomas are often mistaken for squamous cell carcinomas, however squamous cell carcinomas lack a benign poroid component, which helps to distinguish between the two (4).

Only a few case reports and small series of squamoid eccrine carcinoma have been reported in the medical literature. Clinical presentation is highly variable and often asymptomatic leading to the common misdiagnosis as squamous cell carcinoma, basal cell carcinoma, or seborrheic keratosis (1, 3, 5). A review of previous case studies suggest that the majority of cases occur in the lower extremities, head and facial region (1, 5, 6). In rare cases,

regional lymph node metastases and distant metastases have been reported (4-7). In cases with nodal metastasis, the tumor is highly aggressive with mortality near 67%, and is often associated with pain near the primary tumor site (4, 5). In a study by Wick et al. (7), all ten of the cases presented with nodal metastasis and had a mortality rate of 50%.

2. Case Presentation

Here, we describe the case of a 39 year-old man with a right lower abdominal wall squamoid eccrine carcinoma with superficial inguinal lymph node metastasis. He underwent wide local excision, superficial groin dissection, transposition of a sartorius muscle flap, and closure with a pedicled flap for reconstruction followed by adjuvant radiation therapy. Presentation of this case study will provide further guidance for the management of this rare disease.

The patient is a 39 year-old male with no significant past medical history who noted a "pimple" along his right lower abdominal wall when he was a child. The lesion progressively increased in size and became painful approximately one year prior to presentation. Multiple physicians noted this mass in 2017, and this was evaluated with cross-sectional imaging. However, the patient received no follow up thereafter due to difficulty with access to consistent medical care. After visiting an urgent care center in September 2017, a biopsy was performed demonstrating a poorly-differentiated carcinoma with squamoid differentiation proliferating into the dermis and subcutaneous tissue raising concerns for a sweat gland carcinoma. Staging studies confirmed no distant metastasis, although there was extension into the right inguinal canal, with a questionable satellite lesion versus nodal involvement. Following consultation with medical and radiation oncology, and presentation of the case at our multidisciplinary tumor board, recommendations were made to undergo wide local excision of the squamoid eccrine carcinoma and superficial right groin dissection.

The patient was taken to the operating room and placed in the supine frog leg position. A large scaly and exophytic mass was discovered in the right lower abdomen with scattered areas of weeping pustules (Figure 1). The mass measured 9 $\times$ 5 centimeters with induration spanning 15 $\times$ 9 centimeters. Two centimeter margins were taken around the induration. The mass extended to the fascia of the rectus muscle, including the oblique musculature, the anterior portion of the anterior superior iliac spine laterally, as well as to the level of the inguinal ligament. There was no obvious invasion of the periosteum. A firm node adjacent to the mass, over the inguinal ligament, and centered within the inguinal triangle was also noted. This was removed separately along with the nodal packet and Cloquet's node (Figure 2). A sartorius muscle flap was raised to provide adequate coverage of the exposed femoral vessels.

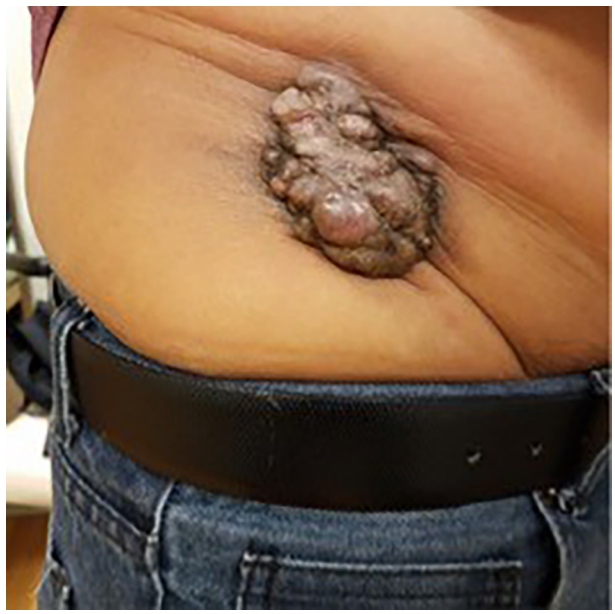

Figure 1. Tumor

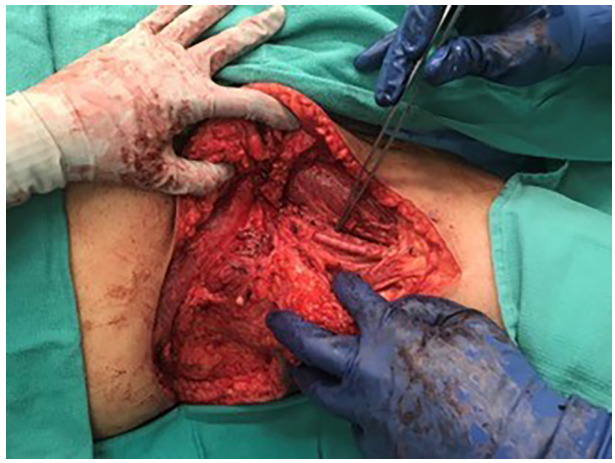

Figure 2. Defect

The size of the resulting defect of the lower abdomen precluded primary closure. The location of the defect was well suited for closure with a pedicled anterolateral thigh perforator flap. A single distal perforator was identified with an intramuscular course. Dissection of this perforator was performed, and the descending branch of the lat-

eral circumflex femoral vessel was identified and dissected proximally. The posterior incision was then made to complete the skin paddle and flap dissection. A subcutaneous tunnel connecting the abdominal defect to the donor site was created, and the flap was delivered through this tunnel into the defect. The posterior portion of the defect was closed primarily in layers, and the flap was inset to close the remainder of the defect (Figure 3). The donor site and the groin dissection site were closed in multiple layers over drains.

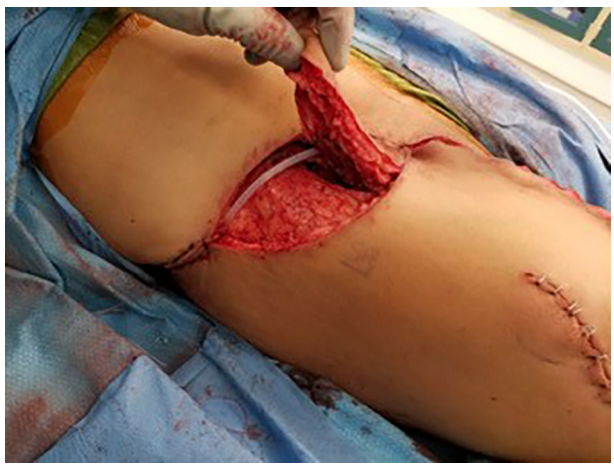

Figure 3. Flap

The immediate postoperative course was uneventful and the patient was discharged home on postoperative day six. In follow up he was noted to have a small seroma in the abdominal wall resection site after removal of his Jackson-Pratt drains. This was aspirated, and clear fluid was returned (Figure 4).

Pathology demonstrated a 6.8 centimeter poorly-differentiated carcinoma with squamous differentiation consistent with a squamoid eccrine carcinoma. Polyclonal CEA immunostain highlighted ductules within the tumor (Figure 5). Where the tumor abutted the epidermis, there were focal connections to the epidermis. However, an in-situ squamous cell carcinoma is not identified. The tumor was diffusely positive for HMW keratin (CK903) with focal positivity for L < W keratin (CAM5.2) and occasional single cells staining for cyclin D1. The tumor was negative for beta-catenin and weakly positive for EMA. The tumor involved the dermis and subcutaneous tissue, and directly invaded one lymph node. One additional lymph node was positive without extracapsular extension for a total of 2/7 lymph nodes. Due to tumor focally approximating the inferior margin and the aggressive nature of this carcinoma, adjuvant radiotherapy was recommended (Figure 5). The patient was treated with 30 fractions of intensity modulated radiation therapy, delivering 66 Gy to the site of positive margin, 60 Gy to the high risk region including the tumor bed and 54 Gy to the regional lymph nodes.

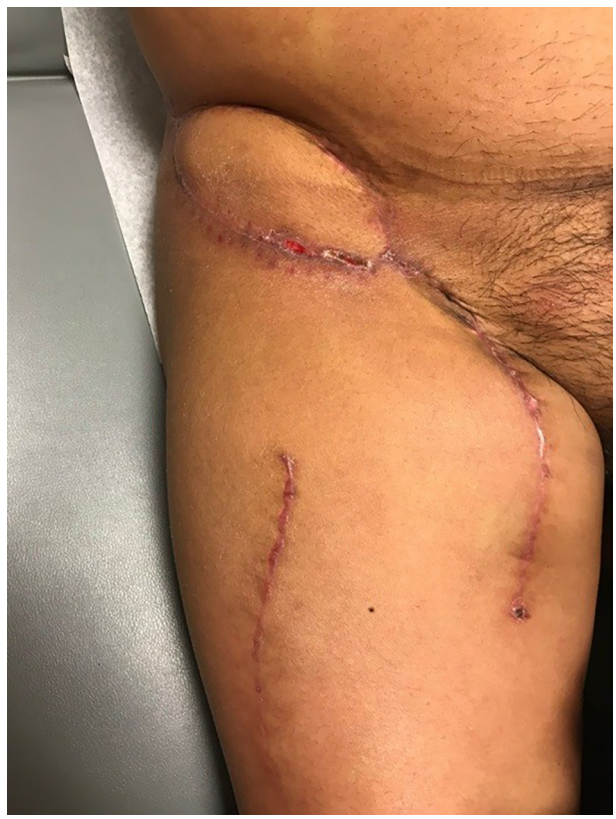

Figure 4. Post-operative follow up

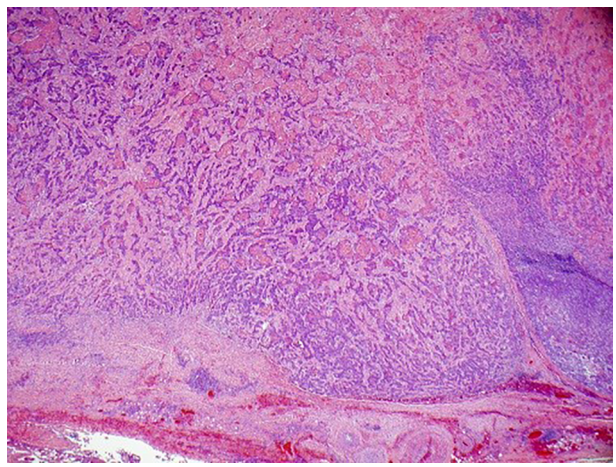

Figure 5. Path

Sections show a deeply invasive tumor extending from the upper dermis to the subcutaneous tissues composed of nests of squamoid cells. On higher power view (left insert), eccrine ductules can be found within some of the tumor nests. The eccrine ductules are also highlighted by a polyclonal CEA immunohistochemistry stain (right insert).

3. Discussion

Squamoid eccrine carcinoma remains an extremely rare type of cancer with only a few cases reported in the literature. The reported overall prevalence of squamoid eccrine carcinoma is less than 0.01% of all skin tumors (7, 8). The earliest documented case of squamoid eccrine carcinoma was described by Pinkus and Mehregan in 1963, as a cutaneous cancer that arose from the cells of the eccrine sweat glands (9). The majority of reported cases occur in females with an average age in the late sixties to early seventies (1, 4, 9-11). While the majority of cases are documented in the elderly, incidence has been reported in a wide age range from 12 to 99 years old (11-16). Currently there are fewer than 300 cases reported of squamoid eccrine carcinoma with most of the reported cases occurring in the head, neck and peripheral extremities (1, 5, 6, 17). Squamoid eccrine carcinoma of the abdominal wall is an extremely rare primary site of disease with few studies reporting it as the location of the primary tumor (8, 12). In a study by Martinez et al., only 16.8% of the cases were reported with the trunk as the primary anatomical tumor location (18). Another study by Blake et al. reviewed 126 cases of porocarcinomas with 14.6% of those patients having primary tumor site on the trunk (6).

Patients diagnosed with squamoid eccrine carcinoma have been shown to develop both regional lymph node involvement and distant metastases (1, 5). Regional lymph node involvement was reported in 3% to 32% of historic cases (8, 11-16). In a study by Yugueros et al., out of the 36 patients included, 13 presented with local recurrence, 4 with regional recurrence and 2 with distant metastases (2). These results were consistent with another review by Martinez et al., in which 12.2% of cases presented with metastatic disease and 7.4% of cases with lymph node metastases (18). Reviews by Mahomed et al. (19) and Kim et al. (20), showed distant metastases in approximately 12% of cases.

Management of squamoid eccrine carcinoma remains controversial due to the rarity of the diagnosis and a paucity of literature on the topic. All reported cases underwent surgical excision either by wide local excision or Moh's micrographic surgery, or in one case amputation (8). The reported curative rates after wide local excision alone range from 70 to 80% with a recurrence rate of approximately 20% in 60-months follow up (8, 21). There are no data to suggest what factors may put some patients at higher risk of recurrence and therefore whether adjuvant treatment may help to prevent recurrence in these higher-risk patients. In patients with other cutaneous malignancies such as squamous cell carcinoma, regional lymph node involvement, large size of the primary lesion, poorly differentiated histology, depth of invasion, perineural or vascular involvement are considered high-risk features. NCCN guidelines for squamous cell carcinoma of the skin suggest, in these cases, that adjuvant radiotherapy be considered (22). In the absence of data to prove that the same features confer higher risk in squamoid eccrine carcinoma, we believe that it is reasonable to extrapolate from the data on other cutaneous malignancies in making treatment decisions (22).

In patients who present with regional or distant metastases, use of chemotherapy and radiotherapy has been reported. Plunkett et al. were the first to describe using chemotherapy in the form of Docetaxel to treat metastatic eccrine porocarcinoma (23). A case report by de Bree et al. showed complete response of skin metastases for over two years when the patient was given an initial treatment of interferon (IFN)-alfa and Isotretinoin followed by topical 5-fluorouracil application and intra-arterial Docetaxel chemotherapy (24). Another case report by Khaled and Hassan described treatment of metastatic squamoid eccrine carcinoma using a regimen of Docetaxel, Cisplatin and infusion of 5-fluorouracil (25). There is unfortunately little evidence for the use of chemotherapy in other non-melanoma cutaneous malignancies as well. NCCN guidelines suggest that Cisplatin either alone or in combination with 5-flurouracil and Cetuximab can be considered in these cases (22). There is, however, growing evidence for use of immunotherapy in non-melanoma skin malignancies such as avelumab for Merkel Cell Carcinoma, hedgehog pathway inhibitors for invasive basal cell carcinoma and anti-PD-1 antibody for invasive squamous cell carcinoma (26-28).

There was a total of 10 cases reported that utilized radiation therapy. Wang et al. reviewed nine cases in which surgery and adjuvant radiation was performed for high-risk skin adnexal carcinoma and concluded this offered excellent locoregional control with minimal toxicity (29). One reported case of squamoid eccrine carcinoma of the vulva described the use of radiation therapy in the context of lung and sacral metastases (30). Adjuvant radiation therapy of the pelvis, bilateral inguinal area and vulva (50.4 Gy/28 Fr) with weekly Cisplatin (40 mg/m^2) for five cycles was completed in order to prevent local recurrence (30). Despite this, PET CT scans one month following com-

pletion of treatment showed metastases to the sacrum and multiple small nodules in the bilateral lungs (30). The patient then underwent three additional courses of chemotherapy using paclitaxel and carboplatin, in addition to palliative radiation therapy to the sacrum (30).

Our case demonstrates a very rare squamoid eccrine carcinoma of the right lower abdominal wall with inguinal nodal involvement who underwent radical resection of the mass, superficial groin dissection, transposition of a sartorius muscle flap, and a pedicled anterolateral thigh perforator flap for reconstruction. Post-operatively, he was treated with adjuvant radiation in hopes of decreasing the likelihood of locoregional recurrence. There is limited guidance on management of patients with squamoid eccrine carcinoma as only a few case series have been reported. Due to the rarity of this pathology, it is unlikely that large studies will be feasible. We believe that a multidisciplinary approach is crucial to making management decisions in these patients. In the absence of data to support management decisions, the approach at our center has been to extrapolate data and guidelines from other non-melanoma cutaneous malignancies.

Footnotes

Authors' Contribution: Brenna L. Hennessey , Logan S. Schwarzman and Dorotea Mutabdzic were major contributors in the writing and editing of the manuscript. Sameer A. Patel performed the anterolateral thigh perforator flap for reconstruction and was a contributor in the writing and editing of the manuscript. Anthony J. Olszanski managed the medical oncologic treatment of the patient and contributed as editor of the manuscript. Shelly B. Hayes and , Joshua E. Meyer managed the radiation oncologic treatment of the patient and contributed as editor of the manuscript. Hong Wu performed the histological examination of the specimens and contributed as writer and editor of the manuscript. Sanjay Reddy is the corresponding author, performed the radical resection of the carcinoma, and was a major contributor of the writing and editing of the manuscript. All authors read and approved the final manuscript.

Conflict of Interests: Anthony Olszanski discloses his participation on the advisory board at Bristol-Myers Squibb, Merck and Array BioPharma and his participation on the data safety monitoring board at Takeda. All other authors on this manuscript have no conflicts of interest disclosures to report.

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