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Dermatofibrosarcoma protuberans post basal cell carcinoma excision: A case report

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ABSTRACT

INTRODUCTION: Dermatofibrosarcoma protuberans (DFSP) is a rare, malignant, soft tissue neoplasm of the dermis. Tumor recurrence is common following resection, and can be locally devastating if not identified in a timely manner.

We report a unique case of this rare tumor. This case poses the question of an association between basal cell carcinoma (BCC) and DFSP, and presents the possible need for increased awareness of DFSP for healthcare providers and patients with a history of non-melanoma skin cancers as well as surgical or burn scars.

PRESENTATION OF CASE: A 77-year-old male with a history of surgical excision of BCC presented with several palpable lesions in the superficial cutaneous tissue of the right anterior abdominal wall. Most of the lesions were consistent with lipoma; however, one lesion near the excision site of the BCC was more solid in consistency. The mass was removed with wide local excision encompassing all layers down to the abdominal fascia. Subsequent pathology findings included CD34 positive spindle cells in a whorled pattern consistent with DFSP. Resection margins were positive and a wide re-excision was performed with margins being negative.

DISCUSSION: DFSP comprises approximately 0.01% of all malignant tumors. There are no known precipitating factors of DFSP, but its presence in surgical and burn scars is not uncommon.

An association between DFSP and basal cell carcinoma has been suggested in the literature. Dermatofibroma and rarely DFSP may demonstrate basaloid proliferation of the overlying epidermis with characteristics of BCC. One case reporting coexistent DFSP and BCC located to the ear also suggested an association, but concluded that the finding was likely incidental due to sun exposure. In our case, the lesion's location is less routinely subjected to sun exposure and points more towards a possible association.

The mainstay of treatment for local DFSP is wide local excision. Negative margins with the removal of fascia and muscle tissue as necessary is essential and the most significant prognostic factor. Three-dimensional reconstructions of DFSP have shown villous finger like projections of primary tumors, which is believed to be responsible for local recurrence. Recurrence can be devastating, as several cases have demonstrated rapid growth of remaining cells with increased morbidity following further resection.

CONCLUSION: Based on this case and those found in the literature, we believe an association may exist between DFSP and BCC and further study of this association is needed. DFSP is a rare malignancy unknown to many healthcare providers, but in the presence of increased awareness and physician vigilance in surgical resection and follow up, the potential morbidity of DFSP may be prevented.

1. Introduction

We report a unique case of a rare tumor, dermatofibrosarcoma protuberans (DFSP) at the site of previous basal cell carcinoma (BCC) excision. The patient presented to clinic out of concern for several palpable lesions on the right anterior abdominal wall. All but one lesion was consistent with lipoma, which was removed by wide local excision. Pathology returned with immunohistochemistry positivity for CD34, which is consistent with DFSP. One other case in the literature suggests an association between the two neoplasms. This case presents the need for increased awareness of DFSP and its management as well as the possible association with non-melanoma skin cancers and surgical or burn scars.

DFSP is a rare, malignant, soft tissue neoplasm of the dermis. The clinical presentation of this tumor ranges from a single painless

Abbreviations: DFSP, Dermatofibrosarcoma protuberans; BC, CBasal cell carcinoma.

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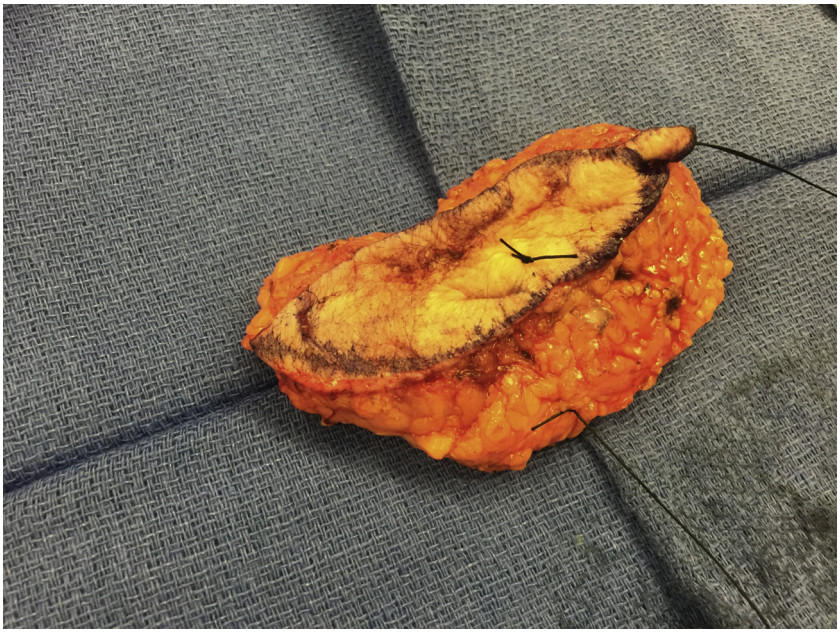

Fig. 1. Gross Specimen.

papule to an ulcerated nodule or plaque. Local recurrence of DFSP is common following primary resection, and can be locally devastating if not identified in a timely manner. We report a unique case of this tumor which suggests an association with BCC and presents the possible need for increased awareness of DFSP for healthcare providers and patients with a history of non-melanoma skin cancers, as well as surgical or burn scars.

2. Presentation of case

A 77-year-old male with a history of surgical excision of BCC seven years ago presented to the clinic out of concern for several palpable lesions in the superficial cutaneous tissue of the right anterior abdominal wall. Most of the lesions were consistent with lipoma; however, one lesion near the excision site of the BCC was more solid in consistency. The mass was removed with wide local excision encompassing all layers down to the abdominal fascia (Fig. 1). Subsequent pathology findings included immunohistochemistry positivity for CD34 (Fig. 2) and positive spindle cells in a whorled/storiform pattern consistent with DFSP (Fig. 3). Resection margins were positive and a wide re-excision was performed with margins being negative. The patient is currently being monitored as an outpatient without evidence of local recurrence.

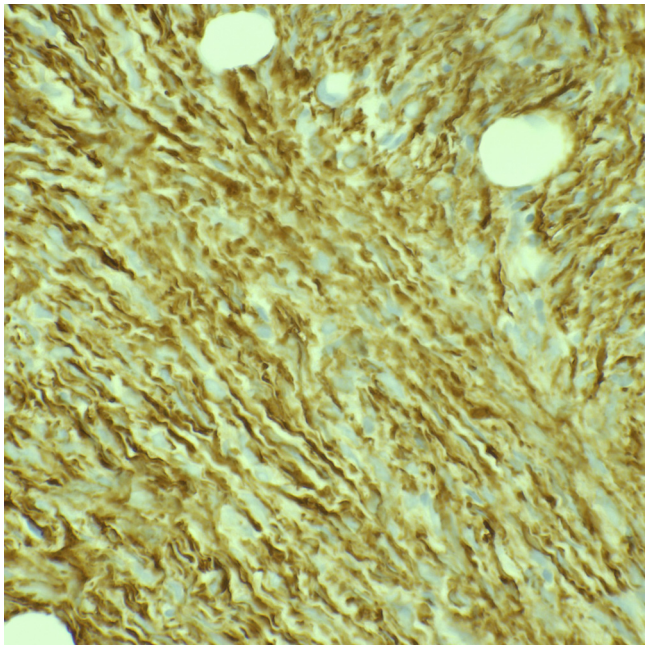

Fig. 2. Immunohistochemistry, CD34.

3. Discussion

DFSP comprises approximately 0.01% of all malignant tumors, and has an annual incidence of 4.2 per million population [1,2]. Epidemiological studies have shown no genetic predisposition, almost equal sexual distribution, and no racial predilection. This tumor is seen most commonly in adults aged 20–50, but may present in individuals of all ages [3].

DFSP is often mistaken for lipoma, morphea, hypertrophic scars, dermatofibroma, and insect bites. The tumors may present as a single painless papule, non-indurated patch on the skin, or as an area of cutaneous thickening with red or bluish discoloration at the periphery. The lesion may enlarge into a lumpy nodule, ulcerate, hemorrhage, or possibly evolve into an atrophic/sclerotic plaque.

It has been well demonstrated in the literature that the finding of DFSP in surgical and burn scars is a relatively common clinical presentation [4]. DFSP is routinely located to the trunk, with less common locations of the head and neck [5]. Metastasis is rare, with a rate of 4–6%, and if present, is most commonly to the lungs [6]. Due to the slow glowing nature of DFSP, diagnosis is often delayed up to several years.

Histologically, the tumor cells have partial features of fibroblastic, histiocytic, and neuroectodermal cells, which suggests undifferentiated mesenchymal cells as a possible origin. DFSP is considered to be of low-grade malignancy and identified by a pattern of monomorphous spindle cells with a storiform or whorled pattern [7]. Positive immunohistochemistry for CD34 expression is consistently seen in DFSP and is often useful in differentiating DFSP from benign dermatofibroma and other soft tissue neoplasms [8].