

# A Rare Breast Tumor: Dermatofibrosarcoma Protuberans

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## ABSTRACT

Dermatofibrosarcoma protuberans is a slow-growing, local aggressive fibrous tumor of the subcutaneous tissue, frequently seen in the proximal extremities and the trunk. Its occurrence in the breast is very rare. Herein, we present a female who presented with a breast mass, and aim to discuss pathological features and differential diagnosis of dermatofibrosarcoma protuberans. A 44-year-old female presented to our clinic with a mass on her breast. Physical examination revealed a 8x5.5 cm mass with multilobular nodules on the skin in the lower inner quadrant of her right breast. Her mammography revealed a hyperdense, 7.5x6.5 cm, well-demarcated, lobulated mass in the right breast, which caused nodules on the lower para-areolar portion of the breast skin. There was no axillary lymphadenopathy on both clinical and radiologic examinations. A core needle biopsy had been performed prior to her referral to our center, which revealed a 'spindle cell lesion'. The patient underwent simple mastectomy. On macroscopic examination; the skin over the lesion appeared ulcerated, and there was a well-defined solid mass, which was pale white-tan on the cut surface. Microscopic examination revealed monotonous spindle cell proliferation arranged in storiform pattern within the collagenous stroma with irregular extensions into deep adipose tissue. There were no necrosis or nuclear pleomorphism. The mitotic rate was 2-3/10 HPF. Immunohistochemically tumor cells showed diffuse CD34 positivity, and S100, EMA and SMA negativity. Based on histopathological and immunohistochemical findings, the lesion was diagnosed as dermatofibrosarcoma protuberans. Local recurrence is expected in 20-50% of these cases. Its treatment requires complete surgical excision with wide margins. Distant metastases, although rare, have been reported.

**Keywords:** Dermatofibrosarcoma, skin neoplasm, breast, diagnosis, differential

## Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare tumor of intermediate malignancy. Alteration in Thrombocyte Growth Factor  $\beta$  chain (TGF- $\beta$ ) is held responsible in its pathogenesis. It was first described by Darier and Ferrand in 1924 as a "progressive and recurrent dermatofibroma", and in 1925 it was named as DFSP by Hoffman (1, 2). Clinically it is usually seen as a small red-brown nodule localized in the dermis. As the lesion progresses, it forms swelling on the skin surface, infiltrates through subcutaneous adipose tissue, muscles and bones thus forming a multinodular, hard, fixed mass with ulcerated and hemorrhagic areas (3, 4). Recurrence has been reported in some cases despite wide local excision (3). Magnetic resonance imaging and computerized tomography are useful methods for detecting local recurrence (5). Distant metastasis of DFSP is rare. Its standard treatment is excision of the lesion with wide surgical margins. Chemotherapy and radiotherapy play a limited role. Radiotherapy is only recommended in cases with positive surgical margins (6).

## Case Presentation

A 44-year-old female patient of foreign nationality was referred to our clinic with a mass on her right breast. Her physical examination revealed a 8x5.5 cm mass showing multilobular nodules and ulceration on the skin surface in the lower inner quadrant of her right breast. Bilateral mammography revealed a hyperdense, 7.5x6.5 cm well-demarcated, lobulated mass in the right breast, which caused nodules on the lower para-areolar portion of the breast skin (Figure 1). On ultrasound, the same lesion was heterogenous and hypoechoic. Bilateral dynamic contrast-enhanced magnetic resonance imaging was performed for preoperative staging and to rule out any accompanying lesions, which showed a 7.5x6.5 cm mass in the right breast and was interpreted as malignant. There were no axillary lymphadenopathy on both clinical and radiologic examinations. A core needle biopsy had been performed prior to her referral to our center, which revealed a 'spindle cell lesion'. The patient underwent simple mastectomy. On macroscopic examination, the skin over the lesion appeared ulcerated and necrotic. There was a well-defined solid mass which was pale white-tan on the cut surface (Figure 1). Microscopic examination



**Figure 1.** Macroscopic and radiologic (bilateral mammography) images of the lesion

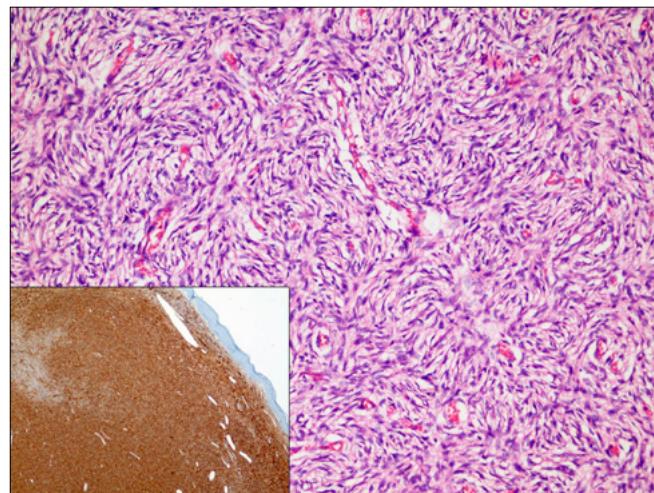
revealed monotonous spindle cell proliferation arranged in storiform pattern within the collagenous stroma (Figure 2). The lesion showed irregular extensions into deep adipose tissue (Figure 3). Necrosis and nuclear pleomorphism were not detected. The mitotic rate was 2-3/10 HPF. Immunohistochemically tumor cells showed diffuse positivity for CD34 (Figure 2-inset) and negativity for S100, EMA and SMA. Based on histopathological and immunohistochemical findings, the lesion was diagnosed as dermatofibrosarcoma protuberans. For this study, written informed consent was obtained from the patient.

## Discussion and Conclusion

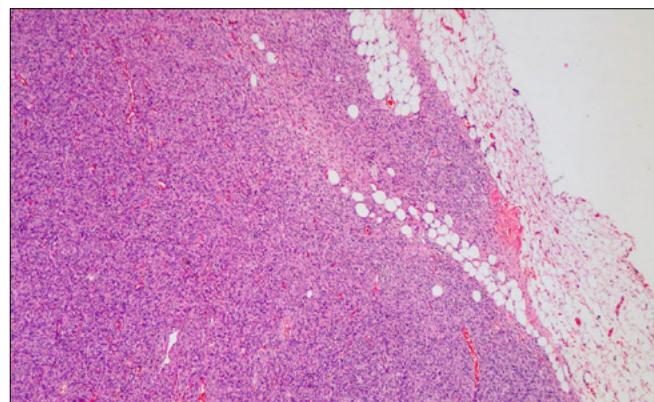
Dermatofibrosarcoma Protuberans is a local aggressive soft tissue sarcoma derived from the dermis. Local recurrence is expected in 20-50% of cases, usually within the first 3 years after excision (7-9). It can rarely metastasize by hematogenous and lymphatic route (3, 7). Brain, bone and heart metastases have been reported (8, 9). Distant metastases typically occur only after recurrences. Due to the significant role of surgical margins on local recurrence and metastasis, safe surgical margins are reported to be as wide as 4-5 cm (10, 11). In our case, simple mastectomy was performed and the distance from the tumor to the fascia was 4.5 cm. In one of the largest series in the literature, the 5-year and 10-year mortality rates were reported to be less than 2% and 3%, respectively (3). The patient is being followed-up for 9 months with no recurrence.

Microscopically, the tumor consists of uniform fibroblasts proliferation arranged in a storiform or chartwell pattern with mild nuclear pleomorphism and low mitotic activity (2-3 mitosis/10 HPF). Inflammatory cells, xanthoma cells and giant cells can rarely be detected in the tumor. The tumor can also have focal myxoid and fibrosarcomatous areas, and the local recurrence rate in such cases is reported to be high (8, 9). In our case, the tumor did not contain myxoid or fibrosarcomatous areas.

The most important entity in the differential diagnosis of DFSP is dermatofibroma. Prominent storiform pattern, increased mitotic activity and CD34 positivity in tumoral cells are not detected in dermatofibroma. Dermatofibroma is a tumor of the dermis, whereas DFSP frequently infiltrates the subcutaneous adipose tissue. Our case was distinguished from dermatofibroma with its prominent storiform pattern, increased mitotic activity (2-3/10 HPF) and diffuse positive immunohistochemical staining for CD34 (8, 9, 12). CD34, which is a myeloid progenitor cell antigen, is an important marker for the diagnosis of DFSP. It is found in endothelial cells; therefore, all vascular



**Figure 2.** Monotonous spindle cell proliferation arranged in storiform pattern within the collagenous stroma. Inset: Immunohistochemical positive staining for CD34 in tumor cells



**Figure 3.** Lesion showing extensions into the adipose tissue, low magnification view

lesions show positive staining for CD34. Its presence has also been described in many fibroblast like cells. For these reasons, CD34 is an important antigen in the differential diagnosis of DFSP (6, 8, 9, 12, 13).

Bednar tumor is known as the pigmented variant of DFSP, and it has similar morphologic properties with DFSP. The only difference is presence of melanin containing dendritic cells in the Bednar tumor. These dendritic cells show positive immunostaining for HMB-45 and S100 (8, 9, 12). Our case was immunohistochemically negative for S100.

Another important tumor in the differential diagnosis of DFSP is malignant fibrous histiocytoma (MFH). Marked nuclear pleomorphism, increased mitotic activity and necrosis are detected in MFH, and the tumor cells are negative for CD34 (5).

Our case was differentiated from leiomyoma and leiomyosarcoma by immunohistochemical positivity for CD34 and negativity for SMA, along with its morphological characteristics. These features combined with the storiform growth pattern and irregular borders of the tumor helped in distinguishing this tumor from myofibroblastoma. A spindle cell sarcoma was ruled out due to the absence of marked nuclear atypia, pleomorphism, high mitotic activity, necrosis and the presence of CD34 positivity (8, 9).

In conclusion, breast skin is a rare location for DFSP. Its definite diagnosis relies on histopathologic examination. Differential diagnosis includes various benign and malignant spindle cell lesions such as dermatofibroma, leiomyoma, myofibroblastoma, MFH and leiomyosarcoma. Diffuse positive immunohistochemical staining for CD34 is an important finding supporting the diagnosis of DFSP. Excision with wide margins is suggested due to high rates of local recurrence. Although very rare, distant metastasis can also be encountered. Clinical follow-up once in 6 months in the first 5 years, and once a year in the following 5 years is recommended in patients with DFSP (13).

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