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Case Report

Huge Dermatofibrosarcoma Protuberans Mimicking a Breast Malignant Tumor with Abscess

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Abstract.

Dermatofibrosarcoma protuberans (DFSP) is an uncommon skin cancer that most commonly occurs on the trunk and extremities. DFSP of the breast has rarely been reported, and then is almost always of small size. We report a case of rapid-growing DFSP of the breast with abscess formation mimicking breast cancer, and also make a review of related literature.

Keywords **:** dermatofibrosarcoma protuberans, breast, fibrosarcomatous transformation

病例報告

巨大隆突性皮膚纖維肉瘤臨床上擬似乳房惡性腫瘤合併膿瘍

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中文摘要

隆突性皮膚纖維肉瘤是一種罕見的皮膚癌，一般常見是發生在軀幹和四肢。而發生在乳房的案例更是少見報導，而且幾乎是以體積小的腫瘤呈現。我們報告一名快速增長的隆突性皮膚纖維肉瘤而在型態上類似巨型乳癌合併膿瘍的個案，並和一系列的相關文獻作一比較。

關鍵字: 隆突性皮膚纖維肉瘤、乳房、纖維肉瘤轉化

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous soft tissue sarcoma, usually appearing on the torso, arms, legs, head, and neck. The size is commonly limited to less than three centimeters [1,2]. DFSP of the breast has rarely been reported. We report one case of a huge breast DFSP of more than 20 centimeters in size that presented as a malignant breast tumor and will review the literature related to DFSP of the breast.

CASE REPORT

A 45-year-old premenopausal woman presented with no significant prior medical history. She reported the existence of a non-tender mass lesion in her right breast for two years. She tried to ignore it, but the mass continued to increase in size over time. About three months ago, she began to feel intermittent right breast pain. She asked for medical assistance due to fever, diffuse painful sensations in the right breast, and pus discharge from the skin ulcer. Physical examination revealed a huge tender mass about 20 centimeters (cm) in size with a skin ulcer on admission. Laboratory studies showed leukocytosis (white blood cell 12100/uL), high serum C-reactive protein (> 250 mg/dl), and normal tumor marker (carcinoembryonic antigen, 1.67 ng/ml). Initial treatment included oxacillin 8 g/day intravenous drug infusion and debridement of the abscess from the breast ulcer. The pathology only showed necrotizing inflammation and the culture showed the *Staphylococcus* species. The breast sonography revealed a huge heterogenic echoic mass of more than 20 cm (Figure 1). A core-needle biopsy was performed, but only an abscess was discovered. She still had a low-grade fever and persistent mastalgia after medical treatment.

Then the suspicion of a huge tumor of unknown nature exhibiting an uncontrolled inflammatory process, a modified radical mastectomy with split thickness skin graft was performed. Intraoperatively, we discovered a 23-cm soft tissue mass occupying the entire breast and a residual abscess within the tumor near the previous debridement site. Final pathology revealed dermatofibrosarcoma protuberans with fibrosarcomatous transformation. Large areas of tumor necrosis could also be discerned. The lymph nodes and section margins were free of tumor involvement. Postoperative cancer systemic work-up revealed no definite evidence of metastases in the abdominal sonography and whole body bone scan. This patient received systemic chemotherapy with 3 cycles of Epirubicin 80 mg and Endoxan 1200 mg. She made an uneventful recovery, and was free of disease one year postoperatively.

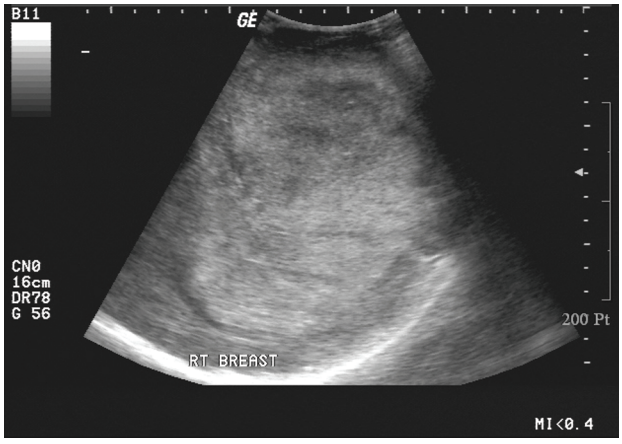

Figure 1. The breast sonography revealed a huge heterogenic echoic mass occupying the whole right breast

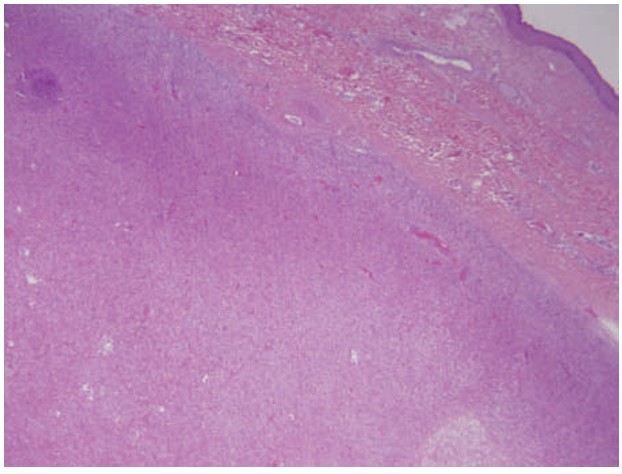

Figure 2. H&E, dermatofibrosarcoma protuberans, 40X: The figure shows a well-defined dermal tumor revealing a pushing border

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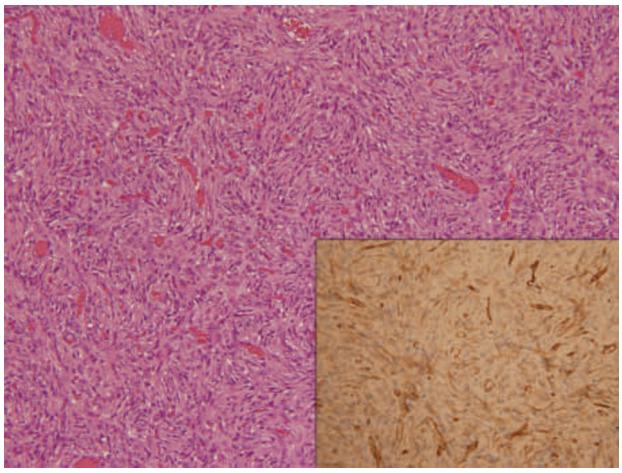

Figure 3. H&E and CD34 (inset), dermatofibrosarcoma protuberans, 200X: This tumor is mainly composed of hyperchromatic and monotonous spindle cells arranged in compact storiform patterns supported by a rich vasculature. Immunohistochemically, these tumor cells are diffusely and strongly positive for CD34 (inset)

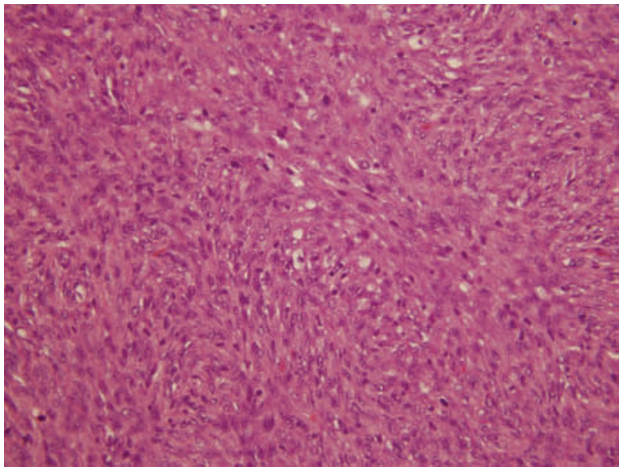

Figure 4. H&E, dermatofibrosarcoma protuberans, 400X: About 30% of the tumor demonstrates enlarged nuclei and a herringbone pattern with elevated mitotic activity indicating fibrosarcomatous transformation

PATHOLOGIC FINDINGS

Histologically, the tumor extended from the dermis to the deeper fibroadipose tissue (Figure 2). The tumor was mainly composed of hyperchromatic and monotonous spindle cells arranged in compact storiform patterns supported by a rich vasculature. Large areas of tumor necrosis could also be discerned. Around 20% of the tumor cells demonstrated enlarged nuclei and a herringbone pattern with elevated mitotic activity indicating fibrosarcomatous transformation (Figure 3).

From the immunohistochemical study (Figure 3), we learned that these tumor cells were positive for CD34. The Ki-67 labeling index was markedly elevated (at 20%) at the fibrosarcomatous transformation area (Figure 4). The final pathology was then dermatofibrosarcoma protuberans with fibrosarcomatous transformation, without nodal metastasis.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) was first described by Darier in 1924 [3]. It is an uncommon cutaneous sarcoma that constitutes less than 1% of all sarcomas [1]. It originates in the dermis and has an aggressive invasive tendency [4]. DFSP presents as a slow-growing tumor and its symptoms are long-lasting [5,6]. Grossly, it usually presents as painless, irregular nodules or plaques varying in color from flesh to reddish brown and occasionally mimics hemangioma [1,5].

DFSP most commonly occurs on the skin of the trunk in approximately 42% of cases, while in 18% of cases it involves the lower extremities and hips, and in 15% affect the head and neck [5]. Tumors presenting as breast tumors are rare. A literature search for DFSP of the breast revealed several cases, the clinical details of which are listed in Table 1. In most of these cases, the tumor size is usually limited to three centimeters, and they presented as a tumor slowly growing across several months up to more than ten years.

Complete surgical resection is the mainstay of therapy for DFSP. Unfortunately, even when the tumor has been resected with good safe margins, there still has been a high recurrence rate of up to 60% after

Table 1. Patient characteristics of 13 patients with dermatofibrosarcoma protuberans of the breast

Patient	Author	Age	Sex	Duration	Clinical information	Size	Location		Sonography	Mammography
1	Kamiya [9]	75	♀	3 years	Developed on the skin with chronic radiation dermatitis	1.2×1.2 cm	Left breast	skin	NA	NA
2	Fukushima [15]	19	♀	5 years	A redness and swelling tumor in her left breast	5.6 cm	Right breast	Skin-based	A hypoechoic area with a slight irregular internal echo	An oval, homogenous shadow with smooth margins
3	Karcnik [16]	57	♀	5 years	A palpable, firm skin mass with gradually enlarging borders	NA	Left breast	Intramammary	A superficial, solid, well-defined mass	A well-circumscribed nodule in the outer aspect of the breast
4	Sandberg [17]	57	♀	6 years	Two tumors at her left breast and left jaw	1×2 cm	Left breast	Skin-based	NA	NA
5	Cavusoglu [18]	26	♀	6 years	Keyhole-pattern surgery with preservation of cosmetic appearance	1×2 cm	Right breast	Skin	NA	NA
6	Swan [19]	39	♀	20 years	Dermatofibroma excision history, stretched scar transformation	5×2.5 cm	Left breast	Scar (Skin)	NA	NA
7	Rama-krishnan [20]	23	♀	1 month	A small, non-changing, non-tender mass	1.0 cm	Right breast	Intramammary	An irregular nodule, suggestive of a complex cyst or a solid nodule	NA
8	Kim [21]	74	♀	20 years	An acute, painful, growing mass	5×4.5 cm	Left breast	Intramammary	A well-defined ovoid mass with lobulated contours, focally indistinct margins, and mixed internal echogenicity	NA
9	Su-Ju Lee [22]	48	♀	NA	A recurrent intramammary tumor following two previous supramammary chest wall tumors excised at ages 20 and 31	2.3 cm	NA	Intramammary	NA	NA
10		30	♀	NA	NA	2.5 cm	Right breast	Skin-based	An oval, parallel, circumscribed mass with gentle lobulation	NA
11		25	♀	NA	NA	2.1 cm	Right breast	Subcutaneous	A heterogeneous predominantly hyperechoic mass in subcutaneous tissue	An oval intramammary mass with smooth anterior margin with microlobulation and mild spiculation
12		29	♀	NA	NA	3.0 cm	Left breast	Skin based	An oval, heterogeneously echogenic mass with microlobulation extending from dermis into breast tissue	Oval-shaped, smoothly marginated mass with a focal microlobulation mass
13	Present	45	♀	2 years	Rapid growth with abscess formation	22x18 cm	Right breast	Intramammary	An heterogenic echoic huge mass over whole of breast	No examination

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surgical resections, with substantial morbidity being reported in the past [5]. In our recent review, many authors still mentioned the same opinion that completive surgical resection with wide, pathologically negative margins of at least 2-3 cm is the optimal treatment option for primary or recurrent DFSP [7]. Distal metastasis is very rare. Metastatic disease is usually preceded by multiple recurrences. The lung is the most predominant site of metastasis via hematogenous spread. Regional lymph nodes are rarely involved [7].

Some principal reports suggest postoperative radiation to reduce the risk of local recurrence [4,8]. However, there was also a report of radiation therapy causing chronic radiation dermatitis as an inducer of DFSP [9,10]. DFSP is associated with t(17,22) (22; q13) translocation, which generates a Col1/PDGF (PDGF: platelet-derived growth factor) fusion gene and constitutively activates the PDGF receptor (PDGFR) [11]. Recent reports mention that imatinib, which inhibits the PDGF receptor, and can be used as the target therapy and neoadjuvant therapy [12-14].

CONCLUSIONS

Dermatofibrosarcoma protuberans is a very rare malignant soft tissue sarcoma that rare occurs in the breast. Most DFSP grows slowly, but it can occasionally enlarge rapidly due to the inflammatory process. Complete surgical resection with safe margins of at least 2-3 cm is currently an optimal treatment, but there is a very high recurrence rate. The radiation therapy for the postoperative treatment and imatinib for the target or neoadjuvant therapy need to be considered in patients with positive surgical margins and unresectable or metastatic tumors.

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