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## Case Report

# A Case of Dermatofibrosarcoma Protuberans in the Skin over the Breast

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**Dermatofibrosarcoma protuberans (DFSP) is a slow-growing but locally aggressive, fibrous tumor that has a high rate of local recurrence after surgical resection. This tumor occurs most commonly in the trunk and proximal extremities. In this report we present a case of dermatofibrosarcoma protuberans in the skin over the breast which was re-excised after pathological diagnosis, considering cosmetic aspects. Only three other cases of dermatofibrosarcoma protuberans of the anterior chest wall have been encountered in our hospital, and all of these cases were male. The presence of this tumor in the skin over the breast appears to be rare in females. Careful complete resection is recommended for this type of tumor.**

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**Key words:** Dermatofibrosarcoma protuberans, Cosmetic surgery, Breast tumor

Dermatofibrosarcoma protuberans (DFSP), currently categorized as a fibrohistiocytic neoplasm of intermediate malignancy, is a rare, slow-growing, but locally aggressive tumor that originates in the dermis. This tumor has a characteristic natural history and macroscopic features.

## Case Report

A 39-year-old premenopausal woman had a tumor in the left breast that had grown slowly over 10 years. She noticed the tumor growing and consulted our hospital. Her family history and past history were unremarkable. Upon gross inspection, the tumor was red plaque with an irregularly shaped lustrous central nodule measuring 2.8×2.4 cm (Fig 1). The plaque was located in the lower-inner region of her left breast. The physical examination showed no lymph node swelling in the axillary or supraclavicular regions.

Ultrasonography (US) revealed a well-defined, hypoechoic tumor without an acoustic shadow (Fig 2). However, the tumor was mainly located in the extraglandular tissue beneath the skin. Mammography showed a well-defined flat nodular mass without calcifications in the dense breast (Fig 3), which was diagnosed as mastopathy or a benign tumor. At this point, the tumor was clinically suspected to be inflammatory or locally-advanced carcinoma of the breast. None of the laboratory findings were abnormal. Excisional biopsy of the tumor was performed.

Macroscopically, the tumor was completely resected. The tumor was nodular, and was located between just beneath the epidermis and the subcutis above the mammary gland. On histopathological examination, the tumor was composed of a uniform population of fibroblasts, which revealed a distinct storiform pattern (Fig 4). In some peripheral regions, slender spindle tumor cells infiltrated adipose tissues. There was little nuclear pleomorphism and only moderate mitotic activity (3 per 10 high-power fields).

The tumor was diagnosed as dermatofibrosarcoma protuberans of the breast. Since the deep

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Abbreviations:
DFSP, Dermatofibrosarcoma protuberans

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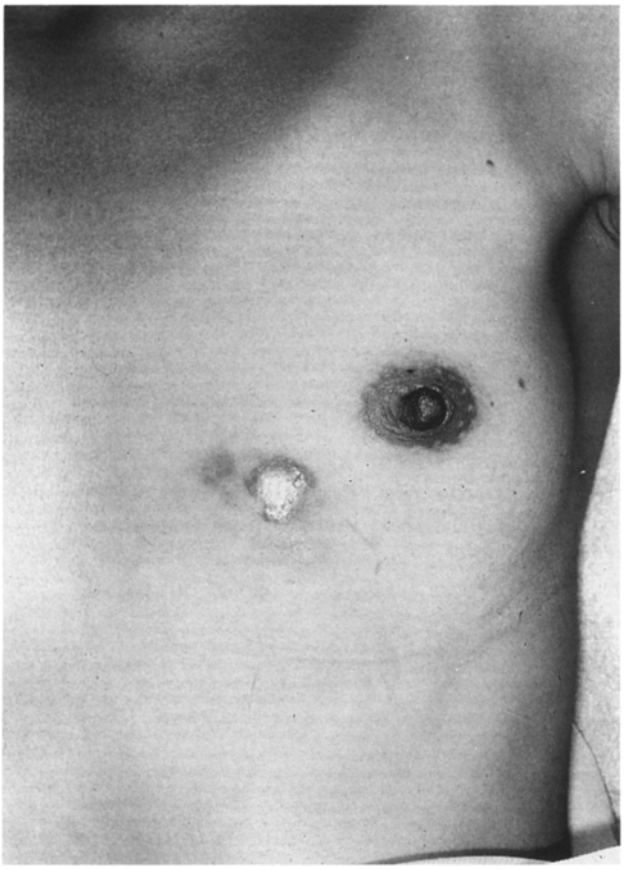

Fig 1. Preoperative appearance of the present case. The left breast tumor was 2.8×2.4 cm in diameter with irregular-shaped red plaques. The tumor was mobile and not tender.

surgical margin showed tumor cells microscopically, an additional wide resection was performed with a 2.5-cm surgical margin around the first incision. The major pectoral muscle just under the tumor was also resected. The margin of the re-excised tumor and the resected muscle was histologically negative for tumor cells. The patient remains disease-free 9 months after surgery.

## Discussion

Dermatofibrosarcoma protuberans is currently categorized as a fibrohistiocytic neoplasm of intermediate malignancy that has a high recurrence rate after surgical excision (11% to 73%)[1-4] and a low metastasis rate (4.7%)[5].

DFSP was first described by Darier and Ferrand in 1924[6] and was referred to as a progressive and recurrent dermatofibroma. It was first referred to as DFSP by Hoffmann in 1925[7]. Males (58.4%) appear to be affected more often than females (41.5%). It begins in early adulthood, most commonly between the second and fifth decade[8]. This tumor tends to occur in the trunk and proximal extremities[9]. It usually grows slowly as an indurated plaque which becomes red or bluish as it enlarges. This is a locally invasive tumor, and local recurrence invariably follows

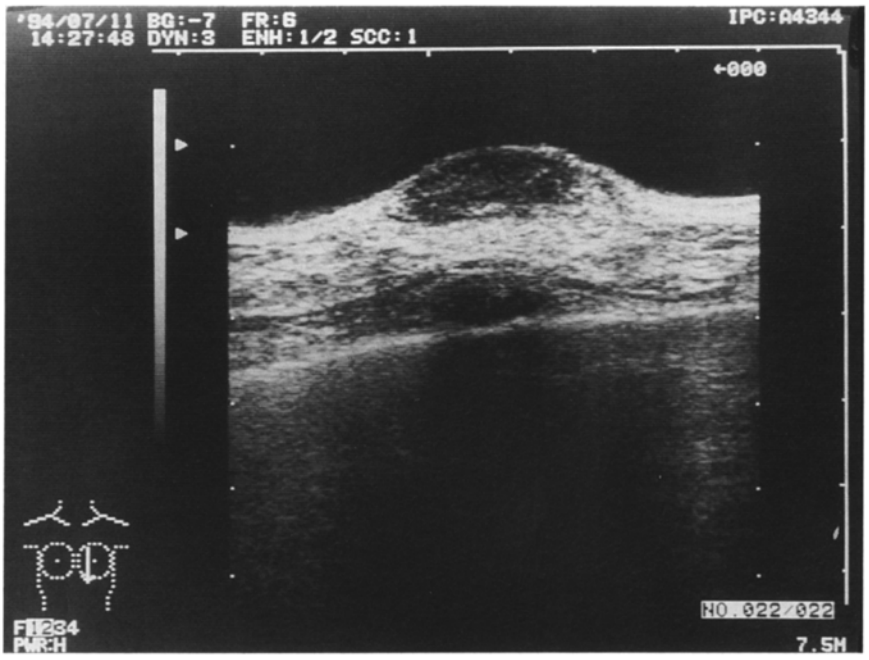


Fig 2. Ultrasonography showed a well-delimited, hypoechoic tumor shadow. There was no evidence of intraductal spread or any other tumors.

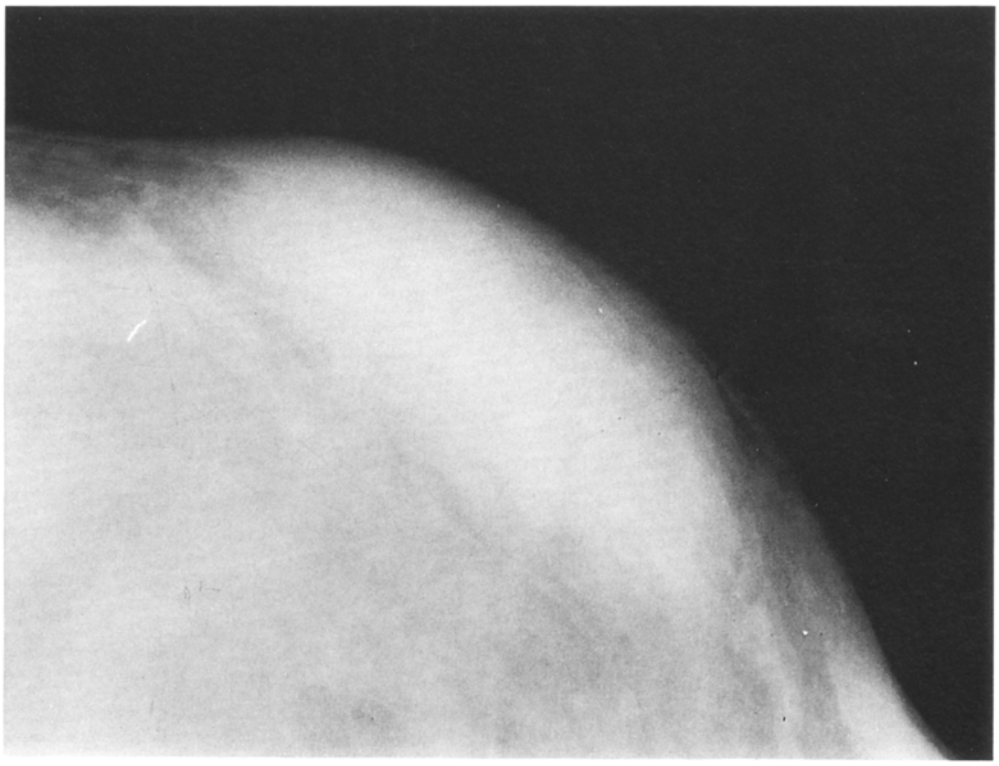

Fig 3. Mammography detected a well-defined flat nodular mass without calcifications.

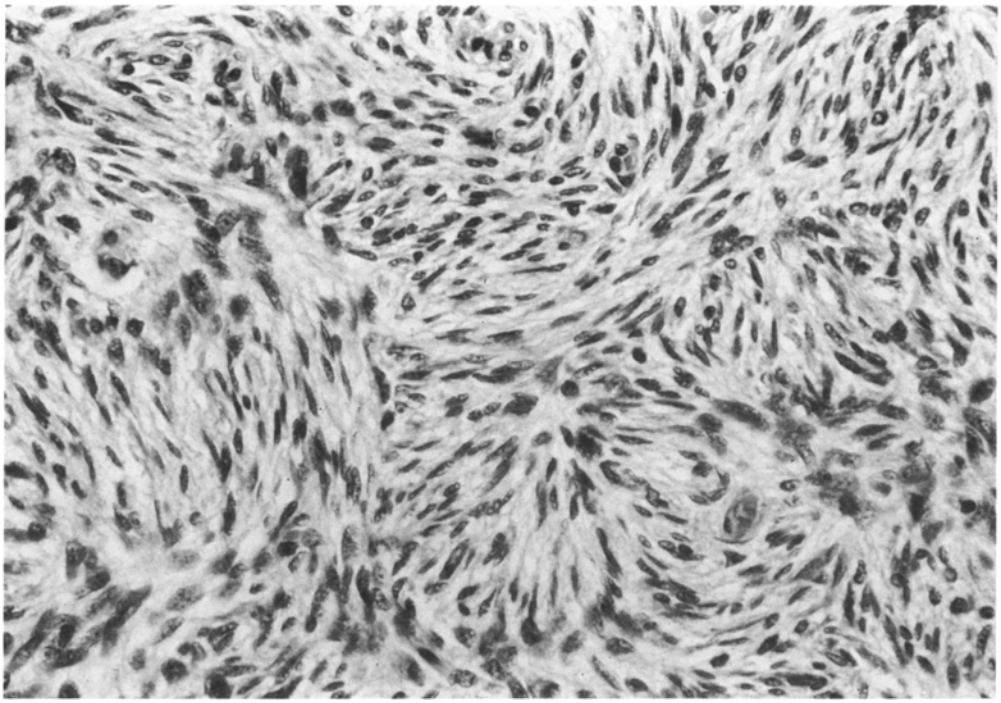

Fig 4. Microscopic findings of the tumor showing a uniform population of fibroblasts, which revealed a distinct storiform pattern. These findings were compatible with dermatofibrosarcoma protuberans.

inadequate removal. It can rarely give rise to metastasis after several years, or multiple recurrence. Statistics of 267 reported cases in Japan revealed that the male-to-female ratio is 1.2:1, a peak incidence is in the third and fourth decades and that 38% of the lesions occur on the chest and abdomen[10].

Clinically, the neoplasm is a firm, protuberant, multinodular tumor colored deep red or bluish-red with a luster. Histologically, it is characterized by a storiform pattern that shows quite definite bands which interweave or radiate like spokes of a wheel, and has fairly uniform spindle cells with elongated nuclei. It extends right up to the dermo-epidermal junction and down into the subcutaneous fat or deeper, and widely infiltrates laterally between collagen bundles of the deeper dermis and blends into the normal dermis. Small, early lesions which lack these clinical features have been misdiagnosed as dermatofibromas; others are clinically confused with keloid or morphea[4]. Tumors in the advanced stages have been misinterpreted clinically as cutaneous metastasis, malignant lymphoma and Kaposi sarcoma[11].

This tumor is commonly treated by wide resection with a macroscopic surgical margin of about 3 cm[12]. McPeak *et al* concluded that a 3-cm lateral margin should be excised through the deep fascia, and even with this margin the recurrence rate was 10% (8 of 82)[8]. The virtue of this policy has been confirmed by immunohistochemical margin control applied to Mohs micrographic surgical excision[12,13]. Parker and Zitelli reported that a 2.5-cm surgical margin through the deep fascia (nonscalp) or periosteum (scalp) cleared all of the tumors, and tumors smaller than 2 cm were completely cleared with a 1.5-cm surgical margin[12]. In addition, none of their 20 patients have experienced recurrence of the tumor and primary repairs were possible in 16 of 20 patients. Considering the pathological diagnosis in the tumor margin, we performed wide resection at salvage surgery. We found three other cases of DFSP of the anterior chest wall resected in our hospital, and they were all males. DFSP is not derived from the mammary gland. Since breast surgeons rarely encounter DFSP of the breast[14] and the findings of imaging diagnosis are of little value, it can be difficult to make a correct diagnosis. Considering the spread of the tumor and cosmetic results, the extent of the resection may be limited, as in this case. However meticulous resection is recommended for DFSP in the skin over the breast.

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