

## Case Report

# Dermatofibrosarcoma protuberans in breast: a case report and review of literature

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**Abstract:** Dermatofibrosarcoma protuberans (DFSP) is an uncommon neoplasm of skin and subcutaneous tissue of low malignant potential. It rarely occurs in breast. In this case report, we described the comprehensive imaging features, including mammography, ultrasound, CT and MRI of a case of DFSP involving the left breast of a 29 year old woman.

**Keywords:** Dermatofibrosarcoma protuberans, breast, mammography, ultrasound, mr imaging, computed tomography

### Introduction

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive skin mesenchymal tumor with a low malignant potential, but with a characteristic tendency for local recurrence [1]. Local recurrence rates of 50% to 70% have been reported after lumpectomy [2]. DFSP accounts for 1% of all soft tissue sarcomas, and for <0.1% of all malignant tumors. The annual incidence is about 0.8-4.5/1000000 [3]. The disease can occur at any age, but 20-50 years age-group is most commonly affected. Early detection and treatment is usually possible due to its superficial location. Imaging findings for DFSP tend to be non-specific [4]; however, comprehensive preoperative imaging may play an important role in the correct diagnosis, help avoid postoperative recurrence and preclude the need for a second surgery.

We reported a case of a 29-year-old woman who had subcutaneous DFSP in the left breast. We assessed the diagnostic value of mammography, ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI).

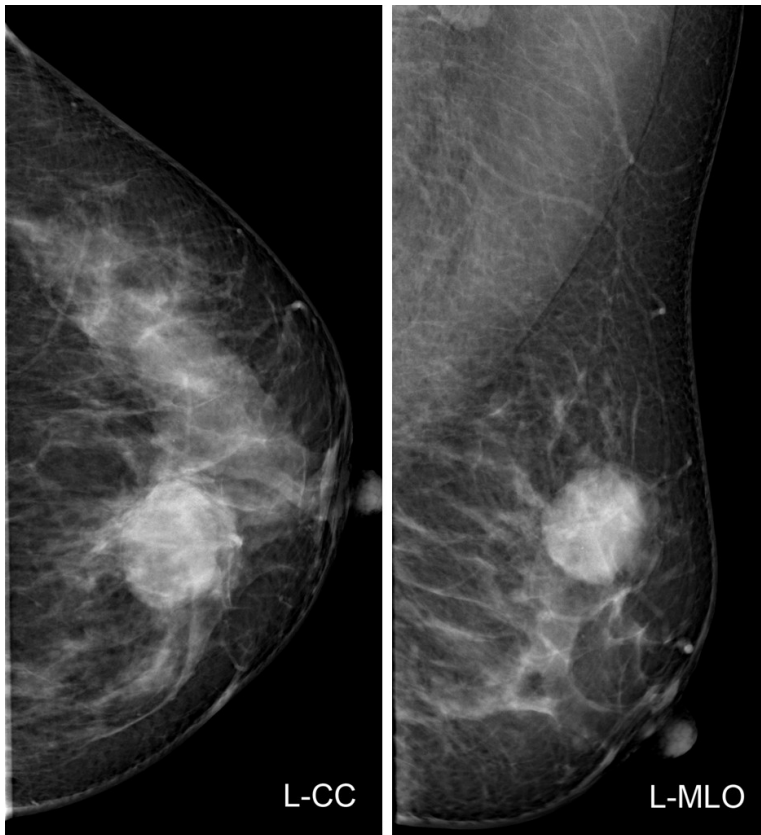
### Case report

A 29-year-old female patient found a lump in her left breast five years ago; however, she did not receive any treatment for the same. Two months ago, she noticed an increase in the size

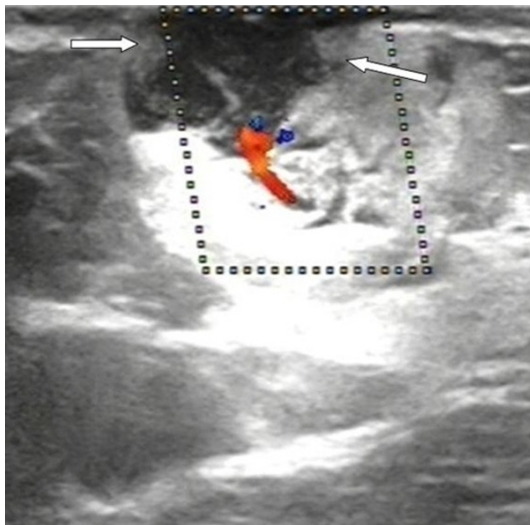
of the lump, but without any associated pain. On physical examination, a lump of 3 cm diameter was noted subcutaneously in the upper inner quadrant of her left breast. The lump was mobile, hard in consistency, and had irregular margins. There were no tenderness or skin changes. No lump was noted in the right breast. There were no palpable axillary lymph nodes.

Mammography revealed a high density oval mass (2.3 cm × 2.8 cm) in the upper inner quadrant of her left breast, with well-defined lobulated margins (**Figure 1**). Ultrasound showed a mixed echogenic mass of about 1.6 cm × 2.8 cm in the left breast at 9 to 11 o'clock position, with an partial unclear local boundary and slightly irregular morphology, spotty hypoechoic areas near the surface, and abundant blood flow signal at the margins (**Figure 2**).

A routine preoperative chest CT was performed to exclude lung lesions and metastasis. CT examination also revealed the subcutaneous mass, slightly low density, oval, 1.8 cm × 2.7 cm in size, clear boundary, with a CT value of about 28 HU (**Figure 3A**). MRI showed a subcutaneous mass with well-defined margins and irregular lobulations. It was slightly hypointense on T1WI (**Figure 3B**), homogeneously hyperintense on T2WI (**Figure 3C**) and showed restricted diffusion (**Figure 3D**). The lesion measured 2.3 cm × 2.4 cm × 2.7 cm in size.



**Figure 1.** Mammogram shows a well-delineated oval high density mass located in the inner upper quadrant of the left breast.



**Figure 2.** Ultrasound shows a mass with mixed echogenic signal in the subcutaneous fat layer of the left breast, with unclear local boundary (arrow), irregular form, and abundant blood flow signal at the margins.

Contrast enhanced T1WI MRI showed obvious homogeneous enhancement of the mass and

its satellite lesions; slight thickening of skin with marked enhancement (**Figure 3E**). On dynamic enhanced scan, the time-signal intensity curve was of a rapid-rise and slow-decline type (**Figure 3F**). Some small lymph nodes were seen in the left axilla; the largest of these measured 0.3 cm × 0.7 cm in size. Based on the results of mammography, ultrasound and MRI, the tumor was graded as BI-RADS 4.

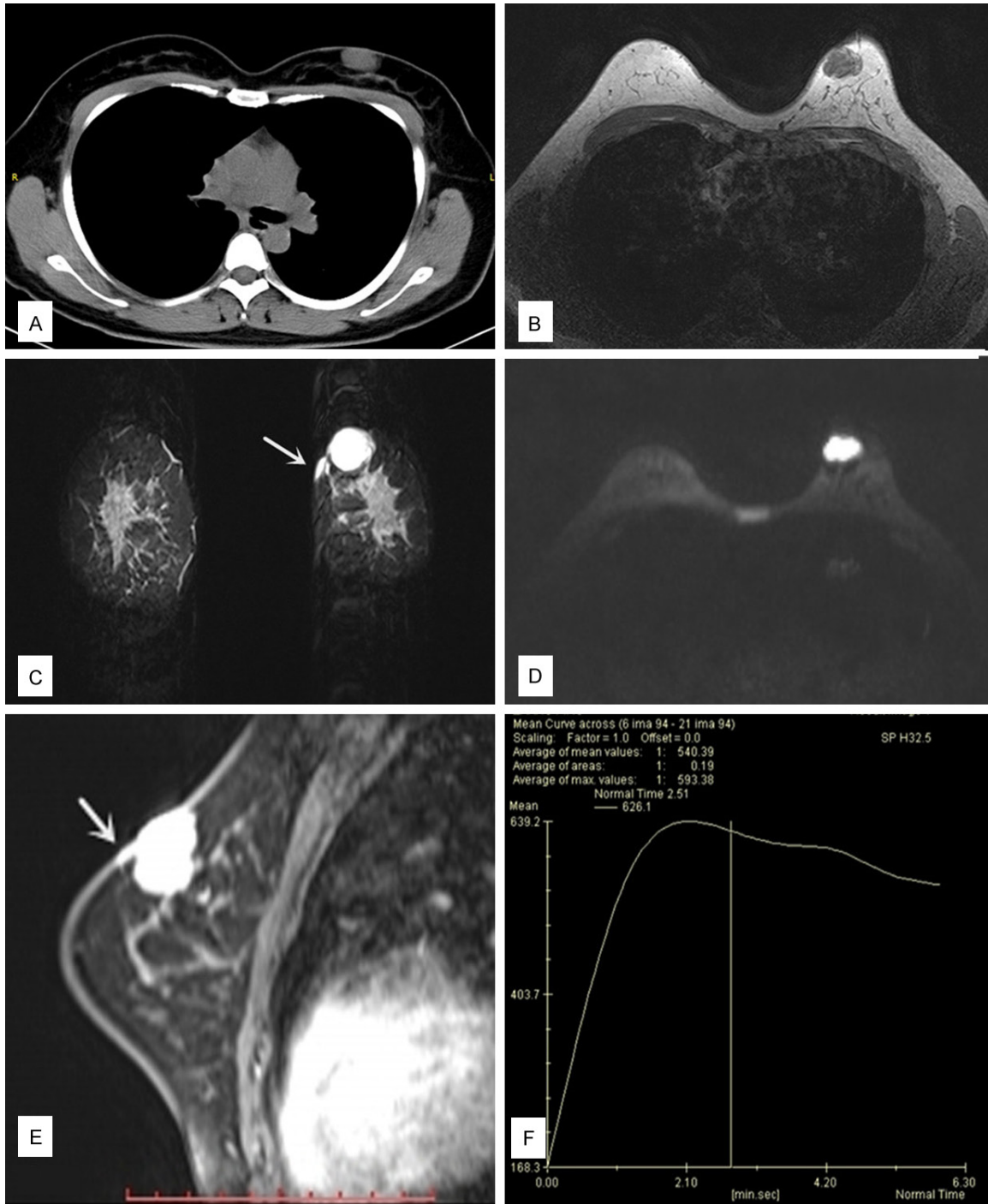
The patient underwent wide resection of the tumor (The largest distance from resection margin to the tumor was about 3 cm), removal of the skin, subcutaneous tissue and glands under the mass, and removal of the pectoralis major muscle fascia. The tumor size was about 2.0 cm × 2.5 cm × 3.0 cm; five large lymph nodes in the left armpit were also removed. Histopathological examination showed diffuse infiltration of

spindle cells in the adipose tissue under the dermis; tumor cells were uniform in size and morphology, comprising of spindle cells with storiform arrangement; no tumor cells were found around the margins (**Figure 4A, 4B**). Immunohistochemistry showed CD34 positivity (**Figure 4C**). Pathological findings confirmed the diagnosis of DFSP in the left breast.

#### Discussion

DFSP occurs in the dermis and subcutaneous mesenchymal tissue and has a tendency for local recurrence. However, distant metastasis is rare [5, 6]. The incidence in men is slightly higher than that in women. It usually presents as painless skin nodules, and grows slowly over a span of several years to several decades. This case was a 29-year-old female, and she had a 5-year-long history. DFSP occurs mostly in the superficial skin and subcutaneous tissue of the trunk (50%-60% of all DFSP), followed by that in the limbs, head and neck. It rarely occurs in the breast [7]; only a few such cases are on record. Imaging findings of DFSP in the breast are not

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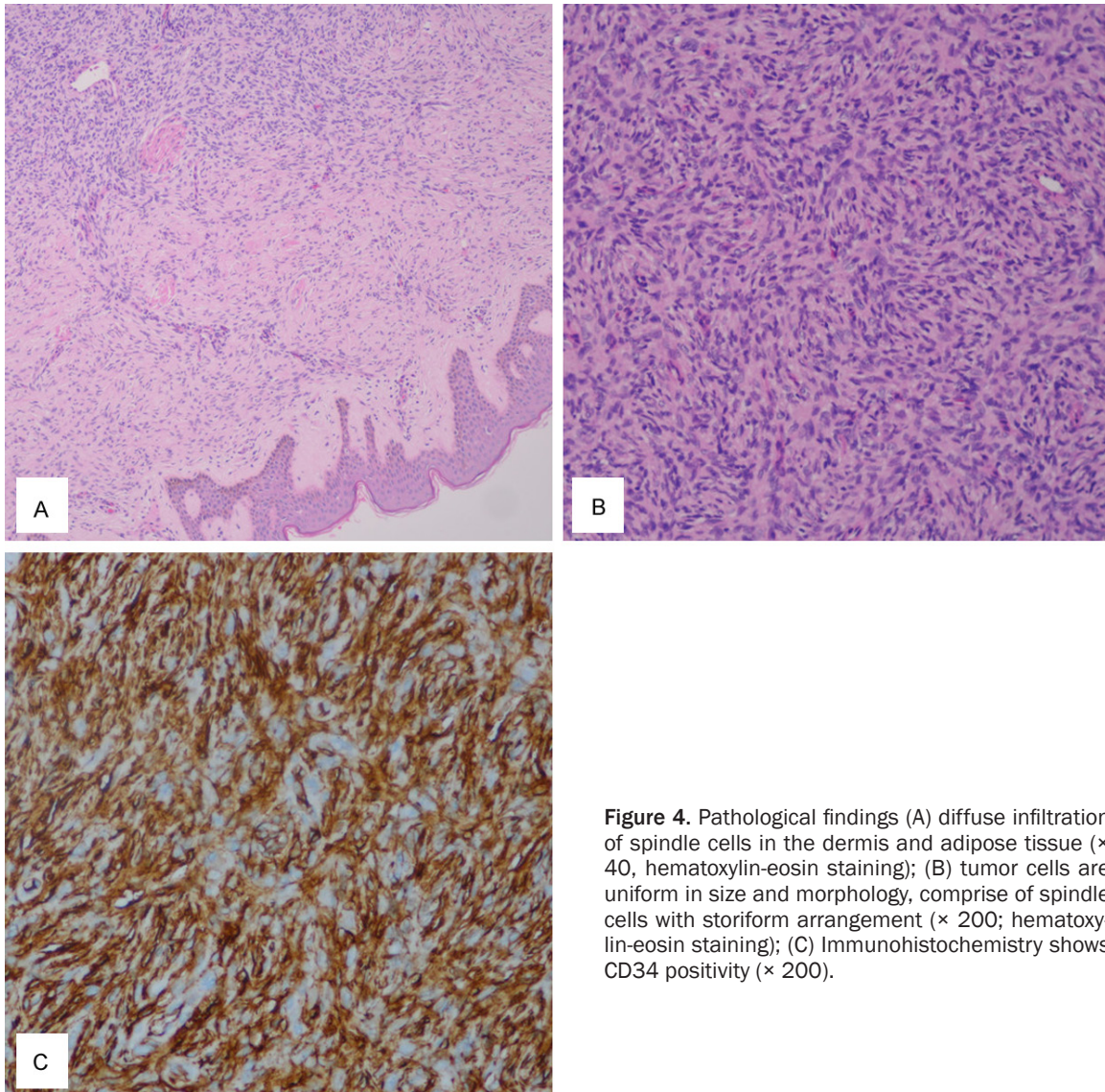
**Figure 3.** Computed tomography (CT) and magnetic resonance imaging (MRI) findings. A: Chest CT shows a well-delineated subcutaneous oval mass of homogeneous density; B: Axial T1WI shows a slightly lobulated mass with inhomogeneous low signal; C: Coronal fat suppression T2WI shows a mass with high signal, and visible satellite lesions in the skin around the lesion (arrow); D: DWI shows lesions with high signal; E: Sagittal enhanced fat suppression T1WI shows irregular slightly lobulated mass with obvious homogeneous enhancement; the surrounding skin is slightly thickened and shows marked enhancement (arrow). F: Time-signal curve of the dynamic enhanced MRI is rapid-rise slow-decline type.

well-characterized [8]. In this case, comprehensive preoperative imaging data, including mam-

mography, ultrasound, CT and MRI were recorded.



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**Figure 4.** Pathological findings (A) diffuse infiltration of spindle cells in the dermis and adipose tissue ( $\times 40$ , hematoxylin-eosin staining); (B) tumor cells are uniform in size and morphology, comprise of spindle cells with storiform arrangement ( $\times 200$ ; hematoxylin-eosin staining); (C) Immunohistochemistry shows CD34 positivity ( $\times 200$ ).

Mammography and ultrasound are the preferred imaging modalities for screening of breast cancer. In previous reports [9-11], DFSP presented with a high-density mass with a clear oval boundary on mammography was regarded as a benign tumor. No calcification and fat content is observed [12]. The mammography features in the present case are consistent with those reported elsewhere [9-11].

Shin *et al* [13] reported that ultrasound findings of round or oval subcutaneous mass adjacent to the skin, with lobulated hypoechoic lesions or irregular mixed echoes are typical of breast DFSP. Our ultrasound results showed mixed echogenic mass in the subcutaneous fat layer, with unclear margins, irregular shape, and blood flow signals at the margins. It was diag-

nosed as BI-RADS 4 tumor. On the whole, however, mammography and ultrasound have limited value in preoperative diagnosis of breast DFSP.

Kransdorf *et al* [6] reported CT findings of six DFSP cases, in which equal or slightly low-density mass with clear boundary and heterogeneous marked enhancement were observed. Lee *et al* [14] reported a case of DFSP with irregular mixed low-density mass suspended in the outer skin.

In the published literature, typical MRI features of breast DFSP have included solid mass with heterogeneous enhancement; heterogeneous hemorrhage, necrosis, and mucoid degeneration [3, 15]. Our results are similar to those

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reported by Lee [12]. The satellite lesions also showed features similar to those reported by Xin [16] and Kim [17]. Variable enhancement patterns were noted on CT and MR imaging, T2-hyperintense and marked enhancement are often observed [18].

Mammary DFSP located in the subcutaneous mesenchymal tissue, can invade the adjacent skin and make it thicker. When the breast lesion is <5 cm, imaging results show the mass to be oval, with a clear boundary, equal or slightly low density than the adjoining normal tissue, low T1WI signal, high T2WI signal, with homogeneous signal density (similar to that in benign lesions), and marked homogenous enhancement. For lesions >5 cm in size, tumor stroma contains a lot of collagen fibers and thin dense connective tissue, the degree of malignancy tends to increase and often present as intratumoral bleeding, necrosis and mucoid degeneration. On imaging, the mass has irregular shape, shows mixed low-density with unclear boundary, low T1WI signal (high signal in the event of intratumoral bleeding), slightly high signal on T2WI and T2WI fat-suppression images mixed with patchy low signal or small patchy high signal, and remarkably heterogeneous enhancement. Considering the histopathological findings, the heterogeneous enhancement is likely attributable to the collagen fibers or dense connective tissue in the tumor stroma, while the lack of enhancement is linked to the areas of necrosis or mucoid degeneration.

On histological examination, breast DFSP tumor cells are uniform in size and morphology, comprise of spindle cells with storiform arrangement. Immunohistochemical finding of strongly positive CD34 expression is helpful in the diagnosis [19]. The recommended treatment is local wide excision [20]; a 3-5 cm wide margin from the tumor can significantly reduce the rate of local recurrence. In this case, the margin of surgical resection from the tumor was about 3 cm, and no recurrence was observed over the 2-year follow up period.

DFSP should be differentiated from fibroadenoma and phyllodes tumors. The former is usually round or oval, with smooth and sharp edges, sometimes calcification, good mobility, and no skin adhesion; in some cases, T2WI shows characteristics of separated low or mod-

erate signal; enhanced MRI shows slow gradual homogenous enhancement and the time-signal curve is of a rising type. The latter is usually lobulated, smooth-edged high-density mass, but the surface of the skin and subcutaneous fat is intact; enhanced MRI shows marked progressive enhancement, time-signal curve is rapid climbing-platform type.

In summary, breast DFSP is very rare, its characteristic location in the subcutaneous mesenchymal tissue, and imaging findings may suggest the diagnosis, while the final diagnosis should be made based on pathological examination. Comprehensive imaging, especially CT and MRI can help delineate the tumor location, size, shape, margins, internal structure, skin infiltration and associations of the surrounding tissue, for preoperative assessment as well as for evaluation of post-operative recurrence.

### Disclosure of conflict of interest

None.

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