Ann Med Res 2020;27(11):3041-3

Annals of Medical Research

DOI: 10.5455/annalsmedres.2020.01.040

CASE REPORT

Dermatofibrosarcoma protuberans: An unusual case of breast

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare tumor of the breast. It is locally aggressive tumor with high rate of local recurrences. A 64 years-old female patient had recurrent large two DFSP lesions in left breast 14 months after the segmental mastectomy. No distant metastasis was detected. The patient underwent mastectomy. She refused adjuvant treatment. Despite wide resection was performed in the second surgery, the disease recurred again. As a conclusion, unless effective adjuvant treatments and medications are not present, surgical excision is crucial to prevent recurrence.

Keywords: Breast; dermatofibrosarcoma protuberans; recurrence; surgery

INTRODUCTION

DFSP is a rare and aggressive soft tissue tumor. Most occur on the trunk (42%),followed by the upper extremities (23%), lower extremities (18%), and head and neck (16%) (1). It is characterized by infiltrative growth and marked tendency to local recurrence after surgical resection (2). Although DFSP behaves as a benign tumor but metastatic cases have been reported in 2–5% of patients, and it should be considered to have malignant potential (3). The incidence of DFSP is predicted to be 0.8 to 5 cases per million per year (4). DFSP is usually resistant to chemotherapy and radiotherapy, thus, complete surgical resection or wide resection is the crucial approach in treatment of these patients (5). We aimed to present an unusual case with infiltrative recurrent DFSP in breast.

CASE REPORT

A 64-year-old female patient was admitted with a history of recurrent lesionsin left breast. The tumor held all the dials of the left breast. She had 23x17 cm and 8x6 cm breast lesions (Figure 1). Examinations and imaging findings of the left axillary region, right breast and right axillary region were evaluated as normal. Computed tomography and magnetic resonance imaging are used for radiological examination (Figure 2). Two years ago, a mass was detected in the 17 cm abdominal wall adjacent to the lower border of the left breast, the mass was reported to be 20 years old and rapidly growing in the last year. Biopsy taken two years ago reported as DFSP. Patient underwent wide resection with 1 cm wide clear surgical margin, pathological diagnosis reported as DFSP. The patient did not receive radiotherapy after the first operation. She had recurrent lesions in left breast 14 months after the wide resection. Tru cut biopsy was taken from the mass, and the operation was planned for the patient, since dermatofibrosarcoma protuberans containing 12% fibrosarcomatous changes in pathology. No distant metastasis was detected. During the operation, wide surgical clear margin was obtained macroscopically on the supeior, medial, inferior and lateral sides. Since the pectoral muscle in the posterior is completely invasive by mass,resection was performed up to the ribs and intercostal muscles following was confirmed by frozen section. While there were at least 2 cm clear surgical margins in the superior, medial, inferior and lateral, 1 cm clear surgical margin was detected in the posterior. Pathology was reported as DFSP. Macroscopically 23x17 and 8x6 sized poorly circumscribed, gray-white mass that infiltrates the dermis and subcutaneous tissue. Microscopically tumor cells have large nuclei with low pleomorphism and rare mitotic figures. dermatofibrosarcoma protuberans containing 12% fibrosarcomatous changes. CD34 positive was detected in the immunohistochemistry study in tumor cells. According to the result of consultation with medical oncology, we were told that the patient was not suitable for chemotherapy and we sent the patient to radio therapy. She was directed to radiotherapy, but she refused treatment. Unfortunately recurrence developed again 18 months after second surgery. According to the result of

Received: 11.01.2020 **Accepted:** 30.04.2020 **Available online:** 20.11.2020
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the 2nd consultation with medical oncology, imatinib treatment was administered, no treatment response was obtained yet. She has been followed-up for two months after the second recurrence.

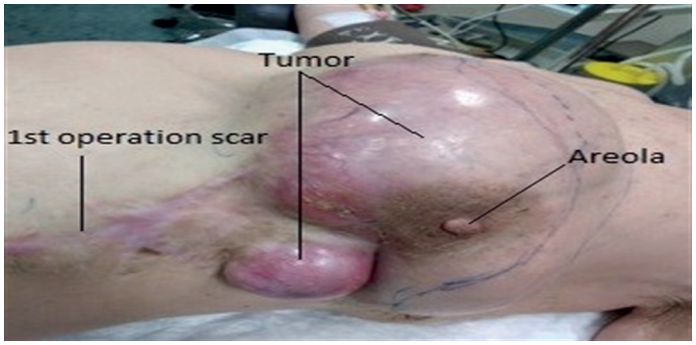

Figure 1. Two DFSPs on the left breast and old operation scar

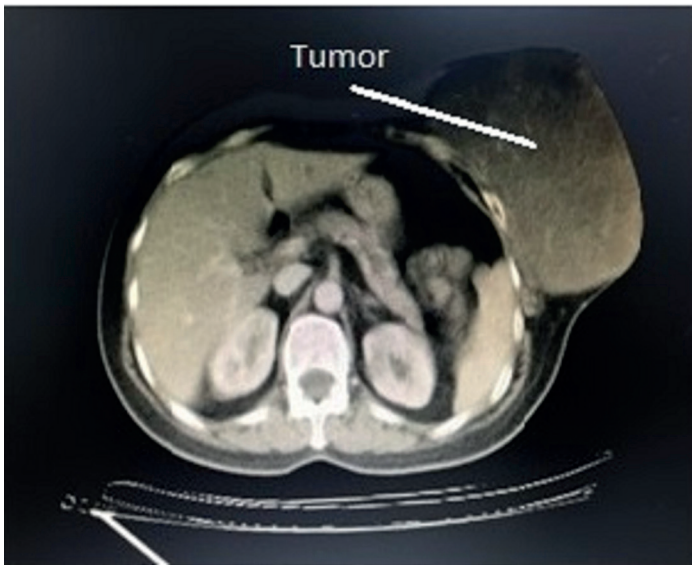

Figure 2. Axial CT image of DFSP in the left breast

DISCUSSION

DFSP was first described by Darier and Ferrand in 1924 and later named by Hoffmann in 1925 (6). DFSP accounts for 0.2% to 1% of all primary breast malignancies (7). The disease is commonly seen in adults over the age of 30 and constitutes 2-6% of all soft tissue sarcomas. Lesions of DFSP emerge as purple and pinkish nodule or plaque on the skin (8). Pigment variations occur in approximately 1% of cases with dermatofibrosarcoma. These are called Bednar tumors and are 7.5 times more in the white race (9). It may easily be misdiagnosed as a benign neoplasm (10). Pathological diagnosis of DFSP was obtained with histopathological and immunohistochemical studies. Local trauma is reported to be responsible in 10-20% of the disease (11). Although lesions may occur on the trunk (42%), upper extremities (23%), lower extremities (18%), head and neck (16%), it is very rare seen in the breast (1). Computed tomography and magnetic resonance imaging may be performed to evaluate the local spreading of tumor. Although there is no standard staging system, some authors use staging according to the American Joint Commission on Cancer Table 1 (12).

Table 1. DFSP Staging System (AJCC)

		DFSP STAGING SYSTEM (AJCC)	
T1	Tumor size < 5 cm		
T2	Tumor size > 5 cm	STAGE 1	Any T, N0, M0, G1
NO	No nodal metastases	STAGE 2	T1, N1, M0, G2-3 T2, N0, M0, G2
N1	Nodal metastases presence	STAGE 3	T2, N0, M0, G3 Any T, N1, M0
G1	Lowe grade	STAGE 1	Any T, any N, M1
G2	Intermediate grade		
G3	High grade		
M0	No distant metastasis		
M1	Distant metastasis presence		

We could not find a review in the literature about breast DFSP cases. When we look at the case reports about breast DFSP; There was not enough information about surgical clear margin. In these cases, the chest wall is seen as the most important factor that limits the size of the surgery. Among the cases of breast DFSP published in Pup med, we did not find a larger tumor size than our case.

Although Mohs micrographic surgery (MMS) emerges as an alternative approach, mainstay of treatment is wide local excision (13). Surgical treatment of DFSP is wide resection of the tumor with 2-5 cm margins and 1-1.3 cm lateral intact borders in MMS (14). Similar survival rates have been reported between wide excision and MMS (15). Adding lymph node dissection to surgery is not be required. Radiotherapy may be administered in cases where the tumor is close to the surgical border and in unresectable cases (16). Imatinib mesylate may be administered for metastatic, recurrent, or inoperable tumors (17). The risk of recurrence was between 1.1-29.8% and mortality rate was between 0.8-14.7% with surgery in DFSP (18). In one of the largest studies on this subject, the mortality associated with the disease was stated as less than 2% for five years and less than 3% for 10 years. (19). Positive surgical magrin, high Ki67-index and fibrosarcomatous transformation were found to be related to local recurrence rate (4). When there is no histologically negative surgical margin, the recurrence rate is about 70% (20). In some studies, no residual tumor has been reported in cases with a intact margin above 5 cm (21). Unfortunately, distance of intact margin has not been reported in approximately 60% of cases with breast DFSP. Clear surgical margin was at least 2 cm in all dimensions other than on thorax wall (10 mm) in our case, no additional surgical intervention was considered due to unwillingness of the patient. We think that some potential reasons such as aggressive tumor histology, not to obtain wide surgical margin in the primary surgery, avoiding rib resection in the secondary surgery and absence of effective neoadjuvant or adjuvant

treatment may have caused recurrence of disease. DFSP tends to spread into subcutaneous and muscle tissue, and local recurrence rates are high especially in the first 3 years after resection (22). Although the development of metastatic disease is rare (2-5%), median time to local recurrence of around 32 months; long-term follow-up is therefore mandatory (2).

CONCLUSION

DFSP may have highly aggressive manner. However, due to the close proximity of the breast tissue to the thorax wall, the width of the surgical procedure may be limited. This surgical disadvantage narrows the safe surgical margin, which naturally leads to early recurrences. Unless there is a more effective treatment modality before or after surgical treatment, wide excision margin during surgery should be kept as far as possible in all dimensions of lesion. As a conclusion, wide excision and resection of adjacent ribs may be considered in patients with aggressive breast DFSP lesions depending on the general condition of patient.

Competing interests: The authors declare that they have no competing interest.

Financial Disclosure: There are no financial supports.

Patient informed consent: Written informed consent was obtained from patient who participated in this study.

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