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Dermatofibrosarcoma Protuberans—An Atypical Breast Tumor

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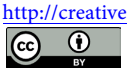

Abstract

A 49-year-old woman was referred to the Department of Plastic and Breast Surgery under suspicion of breast cancer after a mammogram revealed a self-discovered tumor in the lower part of her left breast. Clinical examination, mammography, and histopathological examination revealed that the original tumor in the left breast was benign, and an incidental malignant tumor, a dermatofibrosarcoma protuberans (DFSP), was found in the contralateral breast. DFSP is a rare and highly malignant entity that is often silent and difficult to diagnose, making a biopsy essential. Surgical treatment must be aggressive due to the high risk of recurrence, which constitutes a technical challenge. The patient underwent surgery using an oncoplastic approach with a volume-reducing technique to achieve the best possible therapeutic and aesthetic results. Therapeutic breast reduction was performed on the right breast and the tumor was removed within the resected tissue. A contralateral symmetrizing mammoplasty was also performed simultaneously. The patient was discharged without major complications, and no recurrence of the tumor was seen during the 30-month follow-up period. The surgical approach included alternative solutions in addition to conventional lumpectomy or mastectomy. A multidisciplinary, open-minded, and creative approach resulted in a satisfying outcome for this patient.

Keywords

Dermatofibrosarcoma Protuberans, Breast Tumor, Oncoplastic Breast Surgery

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive cutaneous soft tissue sarcoma. DFSP located in the breast is extremely unusual, and only a

few cases are described in the literature [1]. Approximately 85%-90% of DFSPs are low-grade, while the remaining cases have a high-grade sarcomatous component, which is typically identified as fibrosarcoma and considered an intermediate-grade sarcoma. Though fewer than 5% metastasize, all DFSP variants have a tendency to recur locally [2]. The etiology of DFSP is not yet clearly understood, although studies have suggested that a chromosomal translocation produces a fusion protein that promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF) [3]. The diagnosis is made through a biopsy, and treatment involves wide surgical excision of the tumor with margins of 2 - 4 cm [4].

This case report aims to introduce and describe this rare condition, increase awareness of its management when found in the breast, and highlight the importance of oncoplastic breast surgery when planning a single-stage surgical treatment with satisfying therapeutic and aesthetic results.

2. Case Report

A 49-year-old healthy woman contacts her family doctor due to newly debuted palpatory findings in her left breast. The practitioner orders clinical mammography on a cancer course, which refutes malignant findings on the left breast (fibroadenoma), but raises suspicion of a malignant tumor on the right breast (**Figure 1**). Therefore, the patient was referred to the Department of Plastic and Breast Surgery on suspicion of breast cancer in the right breast. The mammography showed a 13 mm tumor located at the lower part of the right breast. By the time of mammography, a thick needle biopsy was taken, which revealed atypical cells suspicious of sarcoma—a dermatofibrosarcoma protuberans.

The patient was discussed with the sarcoma center at Rigshospitalet, Copenhagen, where an extensive investigation was set, arising with indication for an MRI scan of the thorax and a PET-CT scan of the thorax and abdomen. The PET-CT scan did not show signs of malignancy, while the MRI scan showed a process in the dermis and subcutis in the right breast, without concrete signs of malignancy. As a result, an unclear finding of BIRADS 3 was noted. No signs of metastasis were preoperatively found.

Following the atypical case, a multidisciplinary conference between the Department of Plastic and Breast Surgery at SUH, the Sarcoma Center at Rigshospitalet, Copenhagen, and the Department of Pathology at SUH was held, where it was decided to operate the patient with wide-excision surgery at the Department of Plastic and Breast Surgery at SUH.

The patient was offered two surgical options. The first option was a two-stage surgical procedure consisting of tumor excision with a 3 - 4 cm margin, secondary closure with vacuum treatment until final histology response with later reconstruction of the defect, performing muscle sparring latissimus dorsi flap. Alternatively, the second option was oncoplastic breast surgery using volume reducing technique with a contralateral symmetrizing surgery. The patient chose

oncoplastic surgery.

Surgery was performed as a right-side therapeutic mammoplasty using a volume reducing technique (Wise-pattern) including the tumor in the resection with the intention of wide-excision with a 3 cm margin, and simultaneous contralateral mammoplasty to create symmetry (**Figure 2**, **Figure 3**). There were removed 110 g and 170 g of breast tissue from the right and left breast, respectively. Drains were not applied during this surgery. The patient was discharged with no postoperative complications within the first postoperative day.

The histopathological analysis confirmed the diagnosis finding an 11 mm large dermatofibrosarcoma protuberans, radically excised with over 10 mm of ohealthy tissue around.

The patient is currently followed with clinical checks every 4 months up to 3 years, where the recurrence of DFSP is highest [5], as well as MRI scans at 6, 12, and 24 postoperative months. The patient states complete well-being and today she is satisfied with the result (**Figure 4**).

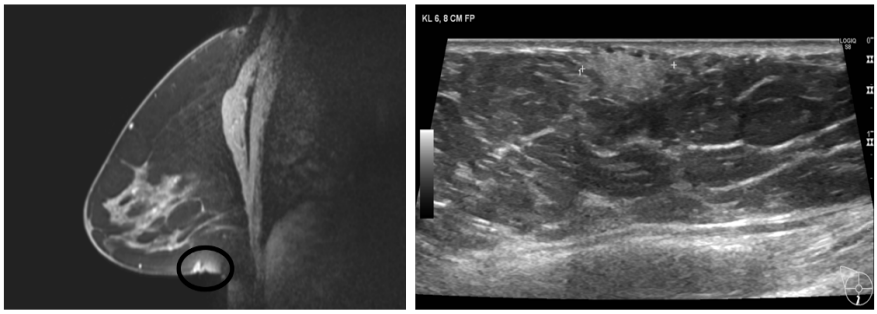

Figure 1. Diagnostic imaging with MRI scanning in MLO view of the right breast, such as ultrasonographic projection of the tumor. Circle: location of the tumor.

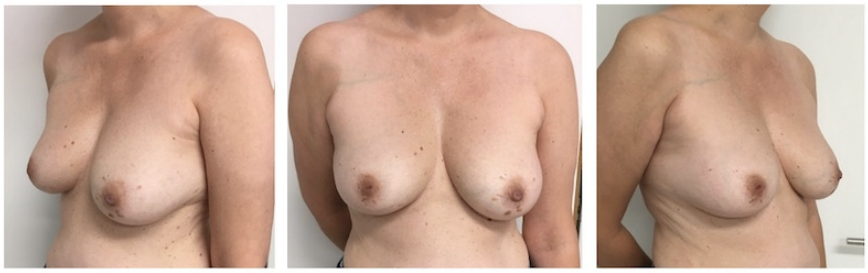

Figure 2. Preoperative clinical photos.

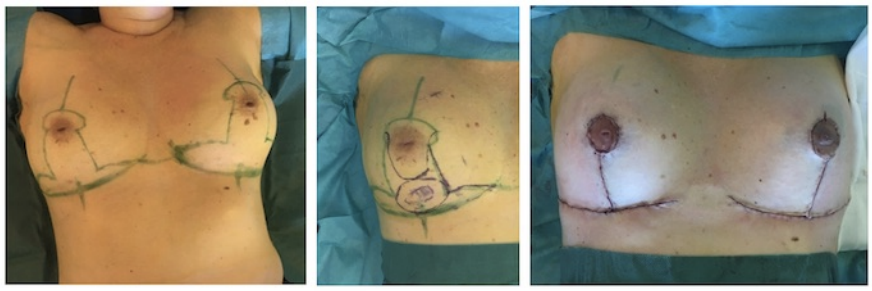

Figure 3. Perioperative clinical photos.

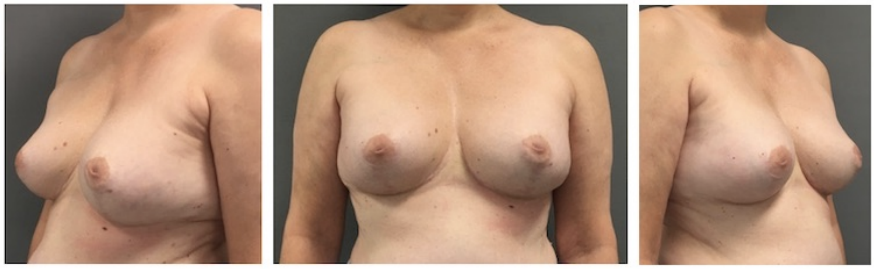

Figure 4. Postoperative result by 24-month follow-up.

3. Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare but locally aggressive low-malignant form of sarcoma that originates from the cells in the dermis or subcutaneous tissue with an incidence of 0.8 - 5/1,000,000 patients per year [2] [4] [6]. DFSP is very rare located in the breast and only a few cases are reported in the literature, mostly as case reports although a single review was published by Wang in 2020 [7]. DFSP located in the breast can lead to diagnostic as well as surgical challenges since the tumor can be misdiagnosed as breast cancer [8].

The diagnosis is suspected clinically and must be confirmed by further histopathological examination, supplemented with fluorescence in situ hybridization (FISH, a cytogenetic technique that uses fluorescent DNA probes to target specific chromosomal locations within the nucleus, resulting in colored signals that can be detected using a fluorescent microscope) and PCR techniques [9]. Genetically, DFSP is characterized by a reciprocal translocation t (17; 22) (q22; q13), or more often, as a supernumerary ring chromosome involving chromosomes 17 and 22 [3] [10]. Initially, the element may be presented in the skin as a small reddish or black nodule, thus resembling a minor hematoma or scar. As the change grows, bulging structures (protuberances) may also develop at the skin surface [11]. Due to a lack of pathognomonic clinical findings, DFSP can be mistaken for a keloid, hypertrophic scar, sebaceous cyst or lipoma and is often referred late for specialized evaluation [8].

The incidence of DFSP is higher in women than men, and higher in African-American than white patients [3]. It most frequently occurs in young and middle-aged patients, between 25 and 45 years of age, with a mean age between 40 to 43 years. However, DFSP has been diagnosed widely from infancy to the elderly [12]. This type of sarcoma is most commonly located on the truncus (50%) and the extremities (35% - 40%) and only approximately 10% - 15% is located in head and neck area [4] [13].

DFSP tends to grow slowly and has a high recurrence rate at 50% - 75%. However, fortunately, it is rare to find metastases (1% - 5%) and this usually happens haematologically to the lungs and bones [14]. In these cases, the mortality rate is high. Lymph node involvement is seen in less than 1% of cases, therefore sentinel node biopsy is not indicated [13].

Surgery is the gold standard treatment, where a local wide excision (LWE) is

judged to be sufficient, classically with a 2 - 3 cm margin of healthy tissue. The prognosis is good when a radical surgical removal has been performed, with a survival rate of 93% - 100% [3] [4] [15].

In this case, the tumor was incidentally detected and examined as the patient was completely asymptomatic concerning the right breast and consulted the family doctor due to a minor fibroadenoma in the left breast.

4. Conclusion

DFSP is a rare and locally aggressive tumor that can occur in uncommon locations, as demonstrated by this case where it was found in the breast. Although the incidence rate is low, it highlights the importance of a multidisciplinary approach, demonstrating that collaboration is pivotal, and that innovative surgical solutions may be necessary when dealing with such exceptional cases, in order to achieve the best possible outcomes for the patients.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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