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Dermatofibrosarcoma protuberans in the breast: Case report

Hwansoo Kim [1]

Department of Surgery, Kangwon National University Hospital, Republic of Korea

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Dermatofibrosarcoma protuberans (DFSP) represents about 1% of soft tissue sarcomas with an estimated incidence of 0.8–5.0 cases per million per year. The involvement of DFSP in breast is very rare and very few cases have been reported in the literature. DFSP was recurred in situ, not spread to distant site. The complete surgical excision with wide, pathologically negative margins of 3 cms is the optimal treatment for primary or recurrent tumor.

PRESENTATION OF CASE: A 46-year-old woman presented with palpable lump in the in the right breast. On ultrasonography, a lesion appeared as hypoechoic, circumscribed mass of approximately 37 mm × 30 mm in diameter in the upper central part of the right breast. The mass of right breast was demonstrated DFPS by pathologic examination. Chest computerized tomography (CT) scan and 2-[18F]-fluoro-2-deoxy-D-glucose (18F-FDG) positron-emission tomography (PET) showed only primary lesion in subcutaneous layer and no enlarged lymph node. The patient underwent excision of the tumor widely. There was no evidence of DFSP local recurrence after five years of follow-up of the patient.

DISCUSSION: DFSP is a rare tumor arising from dermis and subcutaneous mesenchymal tissue. Whereas, characteristic imaging feature of DFSP in the breast are not well-defined. The primary treatment for DFSP is considered to be surgical excision.

CONCLUSION: DFSP in breast is extremely uncommon and can mimic a primary breast tumor. Surgical excision with adequate resection margins is recommended to ensure local control of the disease.

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1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon tumor. Its annual incidence rate is about 0.8–4.5 per million population, and the age of patients ranges from 20 to 50 years [1]. Although metastasis from DFSP is rare, it is locally aggressive and local recurrence after resection is observed in 50–70% of DFSP patients [2,3]. The primary sites affected by DFSP are extremities and trunk, and it is rarely found in the breasts [4,5]. The five-year survival rate of patients is 88.9%. Since the therapeutic outcomes of chemotherapy and radiotherapy are not satisfactory, surgery is considered the primary treatment option for DFSP [6]. Patients diagnosed with DFSP who underwent a complete excision have an excellent prognosis with a 5-year-survival rate of 99% [7]. We emphasize the importance of complete resection by reporting a 45-year-old woman with subcutaneous DFSP on the right breast.

The work has been reported in line with the SCARE 2020 criteria [8].

2. Case report

In May 2015, a 45-year-old woman diagnosed with DFSP was referred to our clinic. The patient has not been using drug, and also she denied the presence of family and hereditary diseases. She had not past surgical history and any other chronic diseases. On physical examination of the patient, a movable, hard, and subcutaneous lump of 4 cm diameter was noted in the upper part of the right breast. There was no palpable lymph node in right axilla. Breast ultrasonography (US) revealed that an oval-shaped hypoechoic mass (3.7 × 1.2 × 3.0 cm in size) with hyperechoic rim was present in the subcutaneous layer of the right breast. This mass showed slightly increased vascularity and several micro-calcifications (Fig. 1), and it was identified to have mildly increased ^{18}F-fluorodeoxyglucose activity at the same site that was detected by ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography (^{18}FDG PET-CT) (Fig. 2C). Breast magnetic resonance imaging showed a subcutaneous mass with well-defined margins and irregular lobulations (Fig. 2A, B). The patient underwent wide resection of the tumor along with the skin and subcutaneous tissue with a 2 cm resection margin. By frozen section intraoperative consultation, and it was confirmed that there were no remaining tumors in the margin. However ipsilateral axillary lymph node dissection was not performed. Pathologic examination revealed that the size of the tumor was 3.7 cm with

[1] 156, Baengnyeong-ro, Chuncheon-Si, Gangwon-Do 24289, Republic of Korea.

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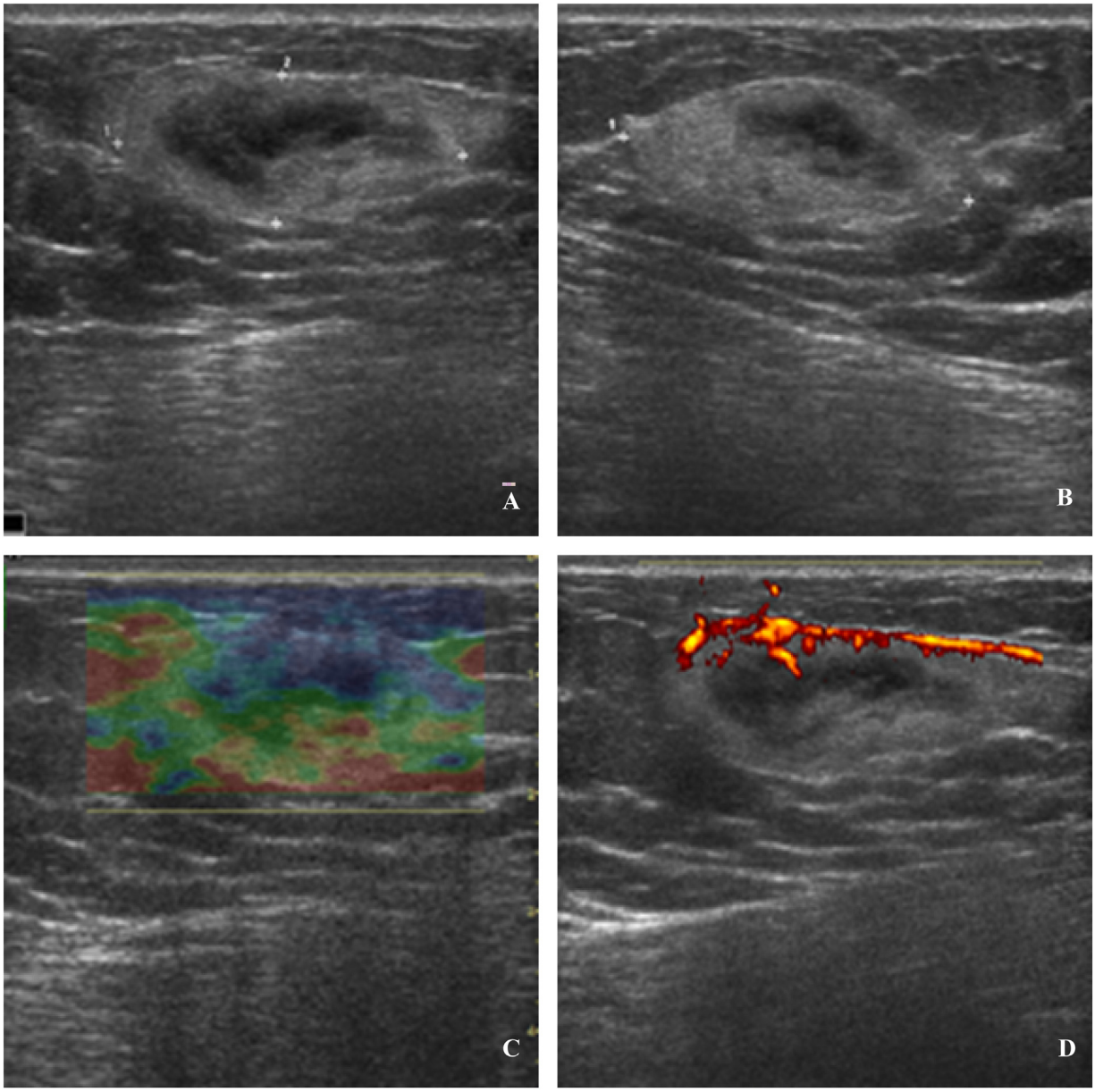

Fig. 1. Breast ultrasonography. A hypoechoic lesion (measuring 3.7 × 1.2 × 3.0 cm) with hyperechoic rim is seen in the subcutaneous layer of the right breast at five o'clock position, 12 cm from the nipple (A, B). Elastography showing the presence of a relatively hard mass (C) that exhibited slightly increased vascularity and several microcalcifications on power Doppler evaluation (D).

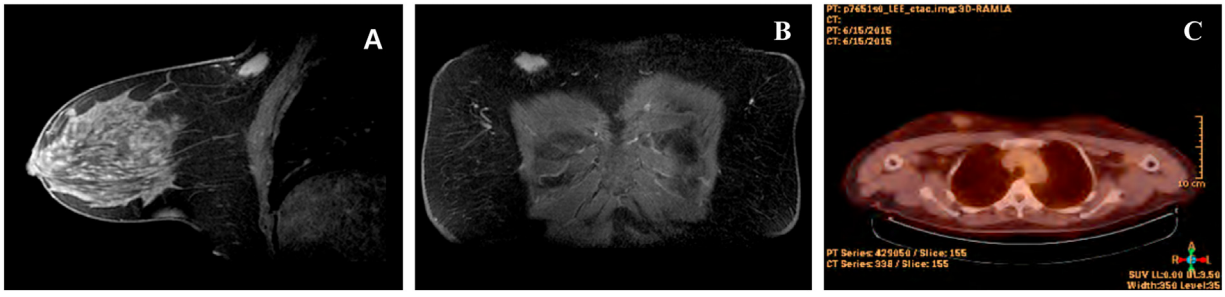

Fig. 2. Breast magnetic resonance imaging shows a subcutaneous mass with well-defined margins and irregular lobulations. (A, B). ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography shows a mild hypermetabolic lesion in the upper part of the right breast (C).

a free resection margin. Histopathological examination showed lace-like infiltration of the tumor cells into adjacent fat and lack of circumscription (Fig. 3A). The tumor cells were uniform in size and morphology, comprising spindle cells arranged in a storiform pattern (Fig. 3B). Immunohistochemical staining of the tumor cells presented a diffuse, positive reaction for CD34 protein (Fig. 3C).

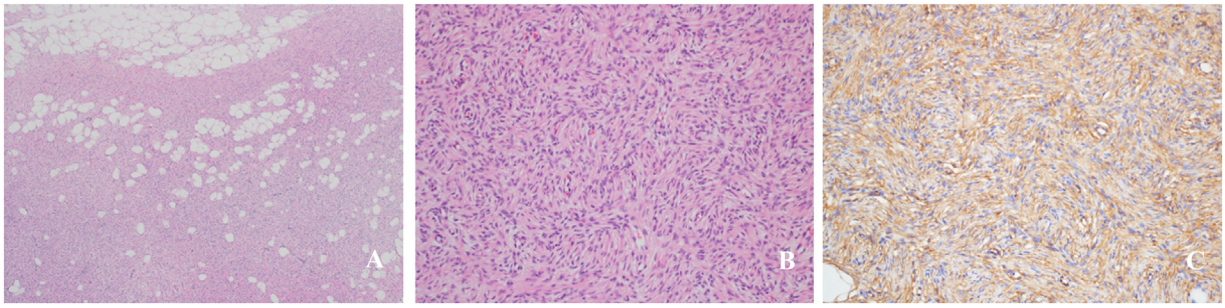

Fig. 3. Hematoxylin and eosin staining of the tumor tissue shows lace-like infiltration into the adjacent fat, with a lack of circumscription (A, ×200) and the proliferation of uniform and bland spindle cells arranged in a monotonous storiform pattern (B, ×40). Immunostained tumor cells show diffuse CD34 expression (C, ×40).

Pathological findings confirmed the diagnosis of DFSP in the left breast. There was no evidence of DFSP local recurrence after five years of follow-up of the patient.

3. Discussion

DFSP is a rare tumor arising from dermis and subcutaneous mesenchymal tissue.In 1924, Darier and Ferrand were the first ones to describe the progressive recurrent dermatofibroma, and DFSP was later described by Hoffmann in 1925 [9,10]. DFSP grows slowly and has a low malignancy potential; however, if the tumor is not adequately resected, local recurrence may occur, resulting in aggressive sarcoma [11,12]. It has been reported that DFSP is mostly diagnosed between 20–59 years for most patients [13,14]. In DFSP patients, the most commonly affected site is trunk, but neck, head, and extremities are also reported to be frequently affected [15–17]. However, the occurrence of DFSP in the breasts is rare [18].

The characteristic imaging feature of DFSP in the breast are not well-defined [19]. The screening of breast tumor usually includes mammography, US, computed tomography, and MRI. Mammography of the breast reveals an oval-shaped high-density lesion without fat content and calcification [20]. US shows the presence of a round or oval mass in the dermis or subcutaneous tissue, and this mass is hypoechoic or heterogeneous echoic with a circumscribed or microlobulated margin. In addition, hypervascularity is often present around the mass [21,22]. In six case series, CT scan revealed a hypodense mass with a well-defined margin and diverse patterns of enhancement [23]. Further, breast MRI demonstrated that DFSP has diverse intensity and patterns of enhancement [24].

In histological examination, the DFSP tumor cells represent a unique appearance, showing proliferation of uniformly shaped and sized spindle cells in a storiform arrangement. Moreover, immunostaining of the cytoplasm with CD34 antibody has also been helpful in the diagnosis of DFSP [25].

The primary treatment for DFSP is considered to be surgical excision. On account of the local recurrence, the tumor should be removed along with a margin of at least 2–3 cm [26]. Radiation therapy can be used as a primary treatment for unresectable tumors, but it is usually performed after surgery on patients with positive margins. Previous studies have shown that adjuvant radiation therapy reduces the local recurrence rate and prevents repeated surgery [27]. Furthermore, imatinib mesylate can be administered when a patient cannot be treated with radiation due to unresectable tumors or when additional resection is expected to cause serious functional impairment in the patient with local recurrence [28].

4. Conclusions

The occurrence of DFSP in the breast is a rare phenomenon. Imaging studies are critical to determining the location and size of the tumor before surgery and detecting the postoperative recurrence; however, based on imaging findings, the criteria distinguishing DFSP from other tumors are not well-established. Histological examination is helpful for identifying the characteristic features of DFSP. Thus, surgical resection with adequate margin should be given the first priority during the treatment of DFSP, and radiotherapy or imatinib mesylate could be considered for adjuvant therapy.

Conflicts of interest

The author reports no declarations of interest.

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Ethical approval

The ethical committee approval was not required given the article type (case report).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

HS Kim: data collection, data analysis or interpretation, writing the paper, references, paper writing and revision, figures revision.

Registration of research studies

Not applicable.

Guarantor

Hwansoo Kim is the guarantor of the study and accept full responsibility for the work and/or the conduct of the study, had access to the data and controlled the decision to publish.

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Hwansoo Kim, MD(HS Kim) had been performing breast and thyroid surgery from 2013 to 2017 at Korea University Medical Center, and has been at Kangwon National University Hospital from 2018 to present.

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