

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Case report: Dermatofibrosarcoma of the breast

Hiba Abduljawad, MBBCH, BAO, NUI (Hons)^{a,*}, Eman Mansour, MD, FEFR, FABR, MSc^a,
Marwah Abdulkader, MD^{a,b}, Mohamed A. Tawab, MBBS, MSc, MD^a

^a Medical Imaging Department, King Hamad University Hospital, Busaiteen, Kingdom of Baharin

^b Department of Pathology, King Hamad University Hospital, Busaiteen, Kingdom of Baharin

ARTICLE INFO

Article history:

Received 20 March 2021

Accepted 24 March 2021

Keywords:

Dermatofibrosarcoma protuberans
breast
MRI
Ultrasound

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is an uncommon neoplasm of the skin and soft tissue, commonly appearing on the trunk and extremities. The occurrence of DFSP in the breast is extremely rare. It has low to intermediate malignant potential with a high rate of local recurrence and invasion. We present a case of a 28-year-old female with a skin lesion on the breast associated with an underlying breast lump. We aim to discuss the sonographic and magnetic resonance findings of breast dermatofibrosarcoma.

© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license
(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Dermatofibrosarcoma protuberans (DFSP) is uncommon neoplasm of the skin and subcutaneous tissue [1]. This tumor commonly involves the trunk, extremities, and the neck [1]. It is a slow growing lesion that may go unnoticed by the patient and more commonly affects females between the ages of 20 to 40 years old [2]. It is classified as a low-grade malignant tumor but can be locally aggressive with a high recurrence rate; especially if it is not excised with negative margins [3]. The breast is a rare location for DFSP [2]. The radiological findings of this lesion are often misleading, indicating benignity. This is a case of a 29-year-old female with a two-year history of a left breast skin lesion with a breast lump that proved to be DFSP on histopathology. We aim to discuss the ultrasound and magnetic resonance findings of DFSP.

Case presentation

A 29-year-old female presented with a two-year history of a nontender slowly growing itchy skin lesion associated with an underlying lump in the right breast. She had been ignoring the mass because a previous ultrasound of the breast performed in a different hospital revealed it to be a benign lesion. However, she felt a recent increase in size and sought medical advice.

Upon examination of the right breast, there was a nodular pigmented lesion with irregular margins in the 12 o'clock position measuring 4 × 4 cm. Palpation of the breast revealed a firm, non-tender breast mass. An ultrasound examination revealed an upper inner quadrant, para-areolar, large cutaneous and subcutaneous soft tissue lesion with heterogeneous echogenicity, central necrosis, surrounding echogenic halo, and posterior acoustic enhancement with evidence of increased internal vascularity. The lesion appeared as a large

* Corresponding author.

E-mail address: hibaabduljawad@gmail.com (H. Abduljawad).

<https://doi.org/10.1016/j.radcr.2021.03.047>

1930-0433/© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

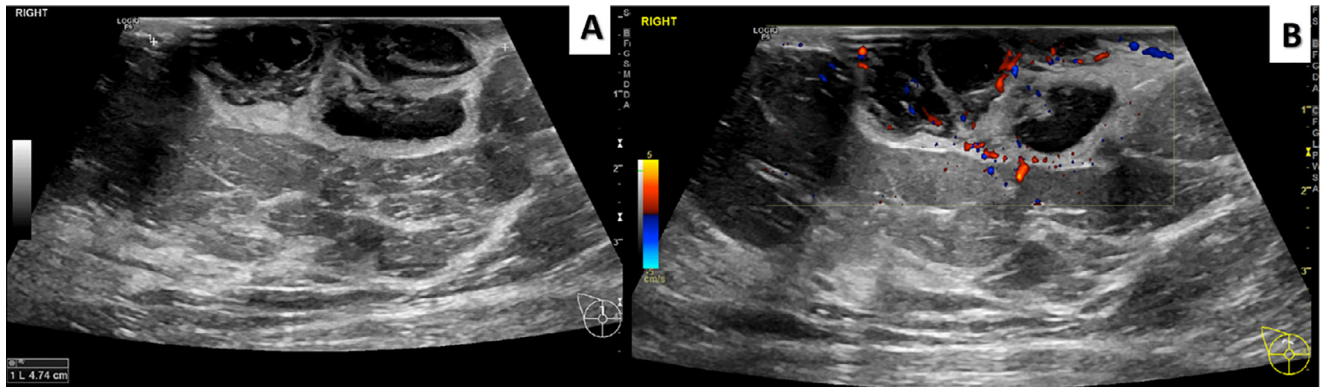


Fig. 1 – A: Grey scale US shows the lesion is seen arising from the skin with echogenic solid component with liquefactions within. Noted well definition from underlying breast parenchyma indicating the cutaneous origin of the lesion. **B:** Colour Doppler images reveals high vascularity of the lesion.

area of skin thickening under which there were multiple focal areas of subcutaneous mass like lesions with the largest measuring about $5 \times 2 \times 4.7$ cm [Fig. 1].

On MRI examination of the breast, the solid part of the lesion was hyperintense on both T1 and T2 images compared to breast parenchyma. In addition, a high signal on STIR and diffusion restriction were present. The lesion showed avid post contrast enhancement and early washout [Fig. 2]. The patient also underwent an incisional biopsy. Histological examination revealed atrophic skin with an underlying spindle cell tumor centered within the dermis and composed of low-grade spindle cells. Cells are arranged in a storiform to whorled pattern, their nuclei are monomorphic and appear to spare adnexal structures. There is no evidence of melanin pigment, sarcomatous transformation or myxoid areas [Figs. 3,4]. The specimen showed CD34 diffuse immunoreactivity and was negative for CD68, CAM 5.2, Melan A, CD31, Desmin and S-100 markers. The overall findings favored the diagnosis of dermatofibrosarcoma protuberans. The patient was referred to another center for resection. A 6 months follow up after surgery was free of recurrence.

Discussion

DFSP is a rare mesenchymal tumor with an estimated incidence of 0.5 cases per million per year. It was first described by Darier and Ferrand in 1924, but the term was later coined by Hoffmann in 1925 [4,5]. The breast is a rare location for mesenchymal tumors in general and an exceedingly rare location for DFS [3], which is usually presented as an indolent slowly growing skin lesion with or without an associated breast lump [3]. It rarely presents solely as an intramammary lesion [6]. DFS is usually ignored by patients due to its slow growth pattern [3]. The skin lesion is usually slightly hyperpigmented with a

reddish to bluish color, and nodular in appearance [7]. Young premenopausal females are commonly affected.

The lesion in our case was multiloculated with a large dominant component as seen on ultrasound imaging. The picture mimicked that of an abscess however, the lack of clinical findings and the long history of symptoms excluded this diagnosis. The typical reported sonographic findings of DFS are an oval hypoechoic or heterogenous lesion, oriented parallel to the skin with post acoustic enhancement [3]. The sonogram can also show well-defined borders such as the lesion in our case or irregular, ill-defined margins. MRI imaging is utilized for clear demarcation of the involved region, accurate size assessment, and to measure its distance to the pectoralis muscle. Most cases were reported to be iso to hypointense signal intensity on T1 and T2 weighted images with intense post contrast enhancement and a washout kinetic curve [3,8,9]. A case report by Bae SH et al, showed mild Fluorodeoxyglucose (FDG) uptake of the DFS lesion on PET-CT, and suggested a correlation between the Standardized uptake value (SUV) and cellularity of the lesion [9]. Radiologically, DFS can mimic a fibroadenoma, phyllodes tumor or inflammatory lesion as in our case [3, 8-10].

Histologically, the DFS is composed of spindle cells on the background of fibrous stroma with strong CD34 immunoreactivity. The differential diagnoses of spindle cell lesions, which are CD34 positive and occurring in the breast region include spindle cell lipoma, solitary fibrous tumor, hemangiopericytoma, and dermatofibrosarcoma protuberans [11].

DFS can rarely metastasize, however, local recurrence can occur in up to 60% of cases. Management is by wide surgical resection with negative margin of at least 2-3 cm [11]. The wider the surgical margins, the lower the chance of local recurrence [11].

In summary, DFS of the breast can mimic primary breast malignancy. Clinical diagnosis alone is difficult, especially if there is no clinically visible dermal component. Ultrasound

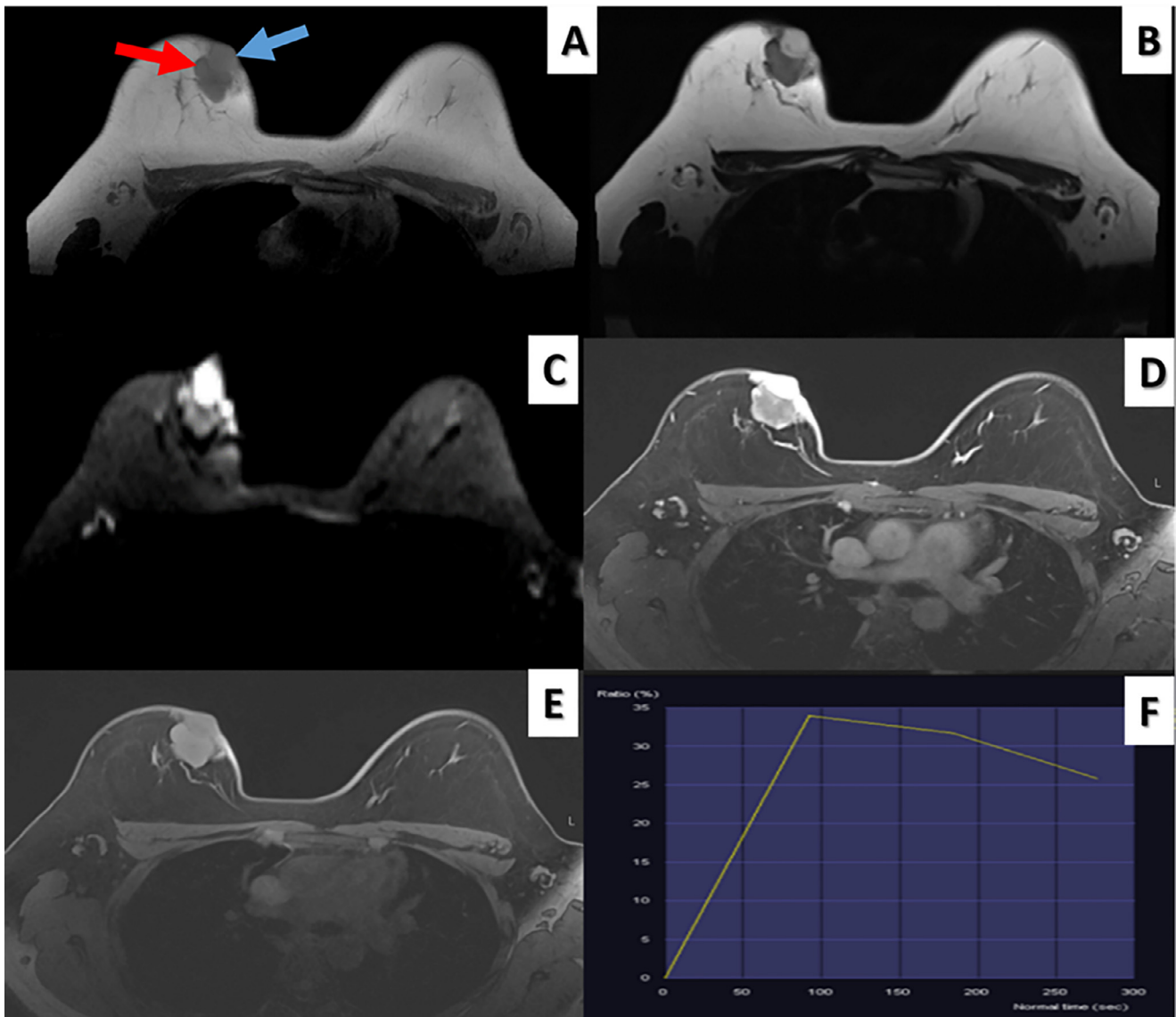


Fig. 2 – A: T1W images showing hyperintense solid component and hypointense cystic component. **B:** In T2 WIs, the lesion appears hyperintense, but liquefied parts show lower T2 signal intensity indicating proteinaceous/hemorrhagic nature. **C:** Early post contrast T1 images showing early enhancement of the solid part of the lesion. **D:** Delayed post contrast T1 images shows wash out of the solid component of the lesion. **E:** Kinetic curve of the solid component shows early intense contrast enhancement with washout.

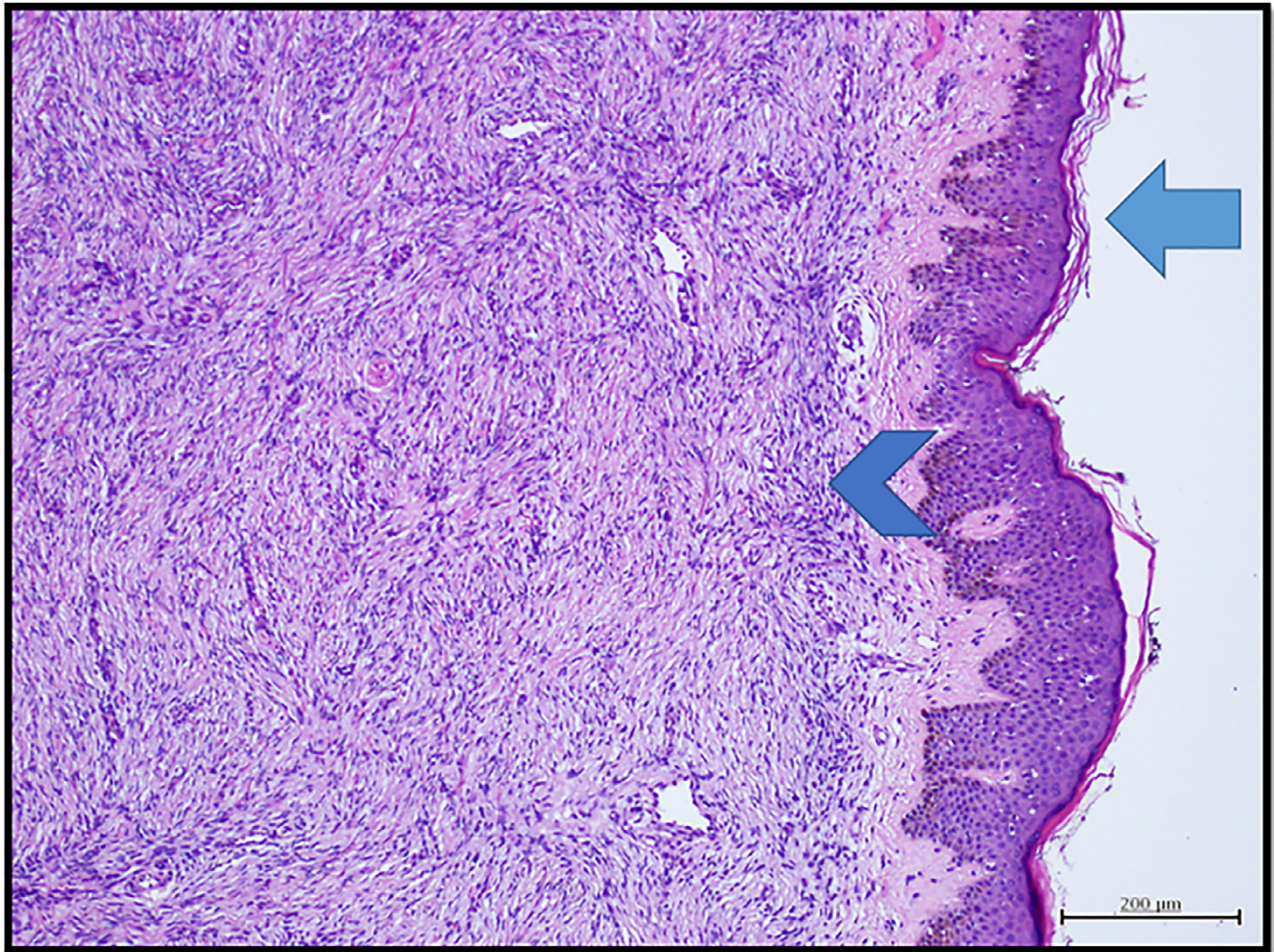


Fig. 3 – Section reveal atrophic skin (Arrow) with an underlying spindle cell tumor that is centered in the Dermis (Arrowhead). The cells are arranged in a storiform to whorled pattern.

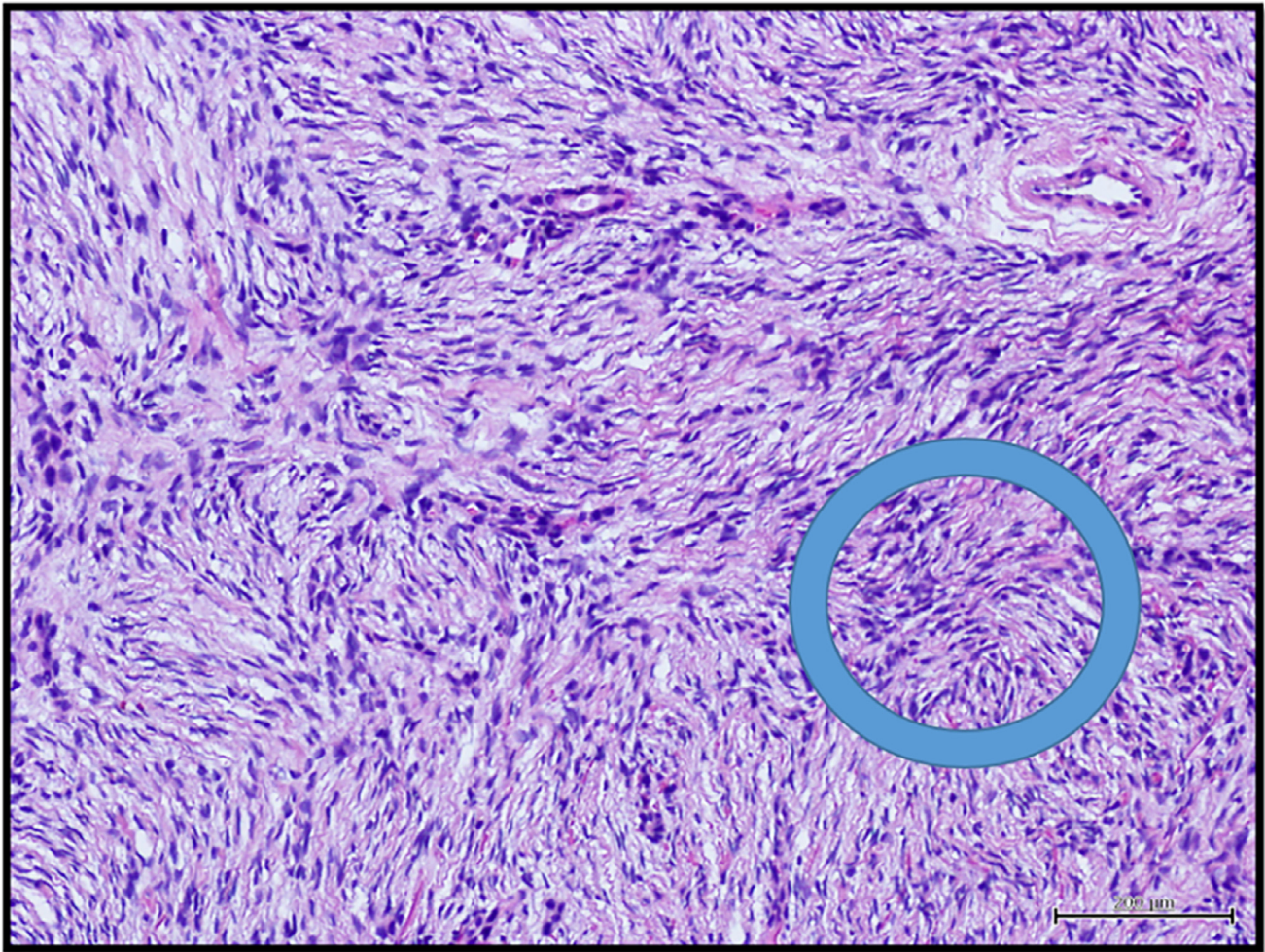


Fig. 4 – On High power, the cells are low-grade spindle cells with a monomorphic nuclei(circle). There is no sarcomatous transformation or myxoid areas. The storiform arrangement of the cells is pronounced at high power

and MRI findings are useful adjuncts. DFS should be included in the differential diagnosis of wide base subcutaneous lesions with high vascularity.

Consent

As per research policy in King Hamad university Hospital:

A written informed consent was obtained from the patient on the date of January 11, 2021, confirming that the patient agrees on using the images of the investigations done for academic purposes only without disclosure of the patient name or personal photo.

REFERENCES

- [1] Kransdorf MJ, Meis-Kindblom JM. Dermatofibrosarcoma protuberans: radiologic appearance. *AJR. Am j roentgenol* 1994;163(2):391–4 Aug.
- [2] Lee SJ, Mahoney MC, Shaughnessy E. Dermatofibrosarcoma protuberans of the breast: imaging features and review of the literature. *Am J Roentgenol* 2009;193(1):W64–9 Jul.
- [3] Kim SM, Rha EY, Jung SN, Lim JS, Yoo G, Byeon JH. Dermatofibrosarcoma protuberans with pulmonary metastasis in the absence of local recurrence. *Archives of plastic surgery* 2012;39(3):265 May.
- [4] Darier J. Dermatofibromes progressifs et recidivants ou fibrosarcomes de la peau. *Ann Dermatol Venereol* 1924;5:545–62.
- [5] Hoffmann EI. Über das knollentreibende Fibrosarkom der Haut (Dermatofibrosarkoma protuberans). *Dermatology* 1925;43(1-2):1–28.
- [6] Ramakrishnan V, Shoher A, Ehrlich M, Powell S, Lucci Jr A. Atypical dermatofibrosarcoma protuberans in the breast. *The Breast J* 2005;11(3):217–18 May.
- [7] Rouhani P, Fletcher CD, Devesa SS, Toro JR. Cutaneous soft tissue sarcoma incidence patterns in the US: an analysis of 12,114 cases. *Cancer: Interdisciplinary Int J Am Cancer Society* 2008;113(3):616–27 Aug 1.
- [8] Zhao Z, Cheng S. Dermatofibrosarcoma protuberans in breast: a case report and review of literature. *Int J Clin Exp Med* 2017;10:3823–8 Jan 1.

-
- [9] Bae SH, Lee JY. Imaging features of breast dermatofibrosarcoma protuberans in various modalities including FDG-PET CT. *Iranian J Radiol* 2016;13(2) Apr.
- [10] Chen HK, Li CF, Uen YH, Tian YF, Chen MJ, Yan CS, et al. Huge dermatofibrosarcoma protuberans mimicking a breast malignant tumor with abscess. *Journal of Cancer Research and Practice* 2014;1(3):267–72 Dec 1.
- [11] Lin JY, Sheen-Chen SM, Hsu W, Huang CC, Ko SF, Eng HL. Dermatofibrosarcoma protuberans of the breast. *Tumori J* 2008;94(6):861–3 Nov.