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Breast Dermatofibrosarcoma Protuberans: Report of a Rare Case

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ABSTRACT

Introduction: Dermatofibrosarcoma protuberans (DFSP) is a rare tumor and has a slow-growing behavior. Presentation is nonspecific, and diagnosis is made after the operation and pathologic examination. The most common sites of this tumor are the trunk, head and neck, and extremities. **Case Presentation**: We report a 42-year-old woman presenting with a breast mass and positive history of breast cancer in her mother. Pre-operation biopsy showed phyllodes tumor. After wide excision of the mass, the pathology report confirmed the DFSP diagnosis. Apart from surgery, she received no adjuvant treatment (chemotherapy, radiotherapy, or hormone therapy). The patient has not experienced recurrence since then. **Conclusion**: According to the rarity and high rate of recurrence of breast DFSP, it should not be missed as a differential diagnosis of breast masses. Taking a multidisciplinary approach and using proper diagnosis and treatment methods can improve the outcome.

Keywords: Sarcoma, Dermatofibrosarcoma Protuberans, Radiotherapy, Case Reports

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing tumor with an incidence rate of 5 cases per million each year, representing about 6% of human sarcomas (1). This malignancy usually occurs between the second and fourth decades of life, and males and females are almost equally vulnerable.

Although it can affect any part of the body, it most commonly occurs in the trunk, extremities, head, and neck (2). DFSP usually presents as a soft superficial skin lesion with a little chance for metastasis but high potential for local recurrence. Preoperative diagnosis is rather difficult and is usually revealed by pathology (3). A wide margin of normal tissue must be removed to reduce the chance of recurrence, which is often associated with some cosmetic problems.

Breast DFSP is extremely rare and shares certain histological and cytomorphological features with some other benign and malignant tumors. Therefore, the diagnosis and treatment are challenging, and the outcome is unpredictable. Herein we shared a case of this uncommon sarcoma and provided a literature review on the previous cases available in the literature.

CASE PRESENTATION

The patient was a 42-year-old woman who presented with a new breast mass in April 2018. Her family history was significant as her mother had a history of breast cancer. Initial mammography showed an oval-shaped lesion with halo (Fig. 1). In addition, ultrasound showed an echo-complex, non-homogenous 44×24 mm mass which was in favor of phyllodes tumor.

In July 2018, she underwent wide local excision (WLE) with a margin of 2cm. Sufficient sections were taken from the tumor for further evaluation (at least one per cm of diameter). Immunohistochemistry (IHC) of the specimen showed to be diffusely and strongly positive for CD34 and negative for Cytokeratins (CK). Hematoxylin and Eosin (H&E) study showed a tumor mainly located in the dermis and subcutaneous tissue. Hypercellularity and monomorphism of the cells with small cartwheel

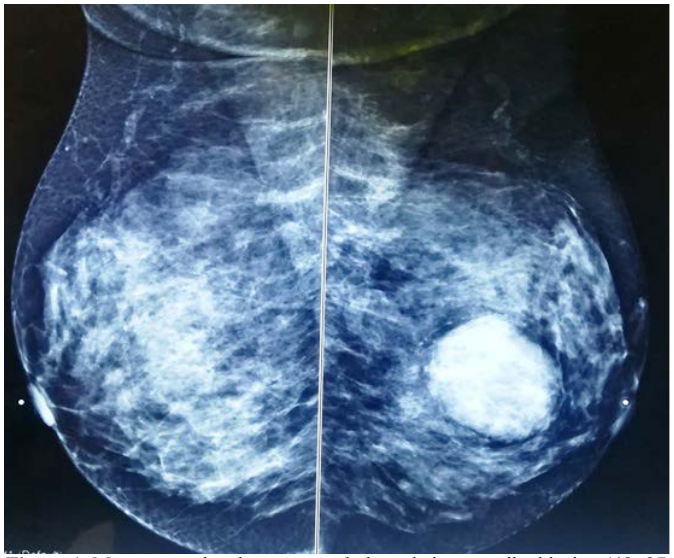

Figure 1. Mammography shows an oval-shaped circumscribed lesion (48×37 mm) with halo in favor of benign lesion.

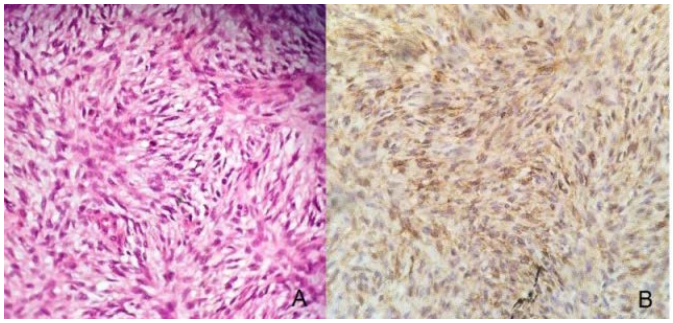

Fig 2: A: H&E show monomorphism of the cells and cartwheel formation, B: IHC was diffusely and strongly positive for CD34.

patterns as well as diffused elongated fibroblastic-like proliferation were noticeable. Adnexa was almost free of tumor, and fat entrapment was significant throughout the sections. Mitosis could rarely be seen at high power. The pathology confirmed the diagnosis of DFSP (Fig. 2). The patient has received no adjuvant treatment and is currently under observation. She has been in remission and in good health for 2 years now.

DISCUSSION

DFSP was first described by Darier and Ferman in 1924 and named by Hoffman in 1925. It comprises 1% of human sarcomas, and black people seem to be more vulnerable than other ethnic groups (3). This malignancy is seen in a wide range of age groups and is most common in the third decade of life (4-8). Despite being a rare sarcoma, DFSP is the most common skin sarcoma (18%) after Kaposi (9-11) and is characterized by a high recurrence rate. Most cases are low grade (85-90%), and the remaining cases are classified as intermediate with higher potentiality for recurrence(4). Although metastasis is rarely seen, recurrence is clearly not unexpected(12-14)..

As to DFSP management, the best treatment to date is surgery (15). Radiotherapy does not seem to be an effective alternative to surgical excision and is usually reserved for inoperable cases or those with margins (3). Since margin status plays a central role in recurrence rate, performing WLE is of great importance. The recurrence rate after simple excision of DFSP with clear margin is up to 60% while with WLE is generally less than 41% (3, 4, 15-17). Despite the predilection for local recurrence, metastasis, including regional nodal involvement, is rare and lymph node (LN) dissection is not indicated (18).

Regarding prognosis, DFSP is a non-fatal sarcoma; however, repeated surgeries for managing local recurrence may lead to some morbidity. (4) Age is also an important factor in DFSP prognosis since patients older than 50 have a higher chance of recurrence. Interestingly, tumor size does not have a significant role in recurrence (4, 15-17). Although the chances for recurrence is higher in the first 3 years, the tumor may recur after 5 years, and regular follow-up must continue (4, 10, 19-21). Late recurrence even after 20 years has also been reported (22)

Less than 1% of breast cancers are sarcomas, and among sarcomas, only a few cases are DFSP (3). Because of the rarity of breast DFSP, few reports of these cases are available. We searched for breast DFSP cases available in the literature and provided information on the cases we had access to in Table 1.

Twenty-five breast DFSP cases were found in the literature (Table 1). The majority of the patients were female (18 versus 7), and the mean age was 36.87 (2-102) years. All patients noticed a palpable mass, and none of them was found in the screening. Three patients had painful or tender mass, and 2 of them had ulcerated mass. The mean duration of the symptoms before seeking treatment were 55.6 (1-240) months. MRI, mammography, and sonography were performed in 7, 9, and 11 patients, respectively. For 6 patients, no diagnostic tool was used. All patients had surgery, and most had WLE with no further treatment. Two patients received adjuvant radiotherapy, and four had surgery with a negative margin. Five of the twenty-five patients experienced recurrence (6-312 months). However, in patients without WLE, follow-up was not long enough to report recurrence. Overall, like DFSP management in other sites, it seems that WLE is the most effective way known for breast DFSP management.

As mentioned above, our patient was approached by a multidisciplinary team and managed with WLE with a 2cm margin without adjuvant radiotherapy. After 24 months of follow-up, there was no sign of recurrence.

CONCLUSION

Breast DFSP rarity and its morphological resemblance to other spindle cell malignancies make its diagnosis and treatment rather difficult. At the same time, the relatively high rate of recurrence attests to the need to develop and employ reliable diagnostic and treatment techniques. We believe taking a multidisciplinary approach and using proper diagnostic and treatment procedures, when feasible, can remarkably improve the management of this challenging sarcoma.

ETHICAL CONSIDERATION

Ethical principles were observed and followed based on the ethical code and infromed consent was gathered.

CONFLICT OF INTRESETS

There are no conflicts of interest in terms of the present manuscript.

REFRENCES

1. Cavuşoğlu T, Yavuzer R, Tuncer S. Dermatofibrosarcoma protuberans of the breast. Aesthetic plastic surgery. 2003;27(2):104-6.
2. Liu SZ, Ho TL, Hsu SM, Zhan HL, Chou CP. Imaging of dermatofibrosarcoma protuberans of breast. The breast journal. 2010;16(5):541-3.
3. Al Tarakji M, Toro A, Di Carlo I, Junejo K. Unusual presentation of dermatofibrosarcoma protuberans in a male patient's breast: a case report and review of the literature. World journal of surgical oncology. 2015;13(1):158.
4. Bowne WB, Antonescu CR, Leung DH, Katz SC, Hawkins WG, Woodruff JM, et al. Dermatofibrosarcoma protuberans: a clinicopathologic analysis of patients treated and followed at a single institution. Cancer. 2000;88(12):2711-20.
5. Marcus JR, Few JW, Senger C, Reynolds M. Dermatofibrosarcoma protuberans and the Bednar tumor: treatment in the pediatric population. Journal of pediatric surgery. 1998;33(12):1811-4.
6. Thornton SL, Reid J, Papay FA, Vidimos AT. Childhood dermatofibrosarcoma protuberans: role of preoperative imaging. Journal of the American Academy of Dermatology. 2005;53(1):76-83.
7. Pappo AS, Rao BN, Cain A, Bodner S, Pratt CB. Dermatofibrosarcoma protuberans: the pediatric experience at St. Jude Children's Research Hospital. Pediatric hematology and oncology. 1997;14(6):563-8.
8. Love WE, Keiler SA, Tamburro JE, Honda K, Gosain AK, Bordeaux JS. Surgical management of congenital dermatofibrosarcoma protuberans. Journal of the American Academy of Dermatology. 2009;61(6):1014-23.
9. 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 International Journal of the American Cancer Society. 2008;113(3):616-27.
10. Chang C, Jacobs I, Salti G. Outcomes of surgery for dermatofibrosarcoma protuberans. European Journal of Surgical Oncology (EJSO). 2004;30(3):341-5.

11. **Table 1.** Information of 25 patients with breast DFSP. (WLE: Wide local excision, FNA: Fine needle biopsy, NA: Not available, RT: Radiotherapy, CDS: clinical decision support , MRM: Modified radical mastectomy, ALND: Axillary lymph node dissection)

Case number (reference)	Age (years)	Sex	Clinical findings	Time from presentation to surgery (months)	Imaging technique	Diagnostic technique	Treatment	Recurrence (months)	Follow-up (months)
1 (3)	27	M	Mass, blue	18	Sonography, Mammography, MRI	Core needle	Excision	No	12
2 (23)	23	F	Mass	1	Sonography	Core needle	WLE	NA	NA
3 (24)	22	M	Mass	36	NA	FNA	WLE	No	18
4 (25)	Young age	M	Mass	NA	Sonography	FNA	WLE	9	NA
5 (26)	43	F	Mass	NA	Mammography	NA	WLE and sentinel dissection	No	6
6 (27)	26	F	Mass on previous ablation scar	NA	Mammography	NA	WLE	No	12
7 (1)	26	F	Red mass	120	NA	NA	WLE	No	12
8 (2)	45	F	Mass	NA	Sonography, Mammography, MRI	NA	WLE	6	NA
9 (28)	26	F	Mass	24	MRI	NA	WLE	No	NA
10 (29)	40	F	Mass	NA	Sonography	Core needle biopsy	WLE	No	NA
11 (30)	14	F	Mass	NA	Sonography, Mammography, CDS	Core needle biopsy	WLE	No	NA
12 (31)	26	F	Mass	180	Sonography	NA	WLE and RT (50Gy)	No	NA
13 (32)	40	M	Ulcerated mass	3	NA	FNA	MRM and ALND	No	NA
14 (33)	102	F	Painful mass on previous surgery scar	60	NA	NA	WLE	300	NA
15 (34)	44	F	Ulcerated mass	NA	Sonography, Mammography	NA	Simple mastectomy	No	NA
16 (35)	41	F	Tender mass	1	Mammography, CDS	NA	R0 mass excision	No	48
17 (36)	26	F	Mass	96	Sonography	NA	WLE	No	NA
18 (37)	41	M	Non mobile mass	12	Mammography, MRI	NA	NA	NA	NA
19 (38)	9	M	Red mass	12	MRI	NA	WLE	No	10
20 (39)	48	F	Mass on previous surgery scar	6	Sonography	Core needle biopsy	WLE, RT refused by the patient	No	18
21 (40)	44	F	Mass on previous surgery scar	120	Sonography, Mammography	Core needle biopsy	WLE	120	NA
22 (41)	2	F	Mass	1	MRI, CDS	NA	R0 resection	No	NA
23 (42)	57	F	Mass	72	NA	NA	R0 resection	No	NA
24 (22)	39	M	Mass on previous surgery scar	During recent pregnancy	NA	Excisional biopsy	WLE	312	NA
25 (43)	74	F	Acute painful mass on previous surgery scar	240	Sonography	FNA, Core needle biopsy	Surgical biopsy+ RT	No	12

Kransdorf M. Malignant soft-tissue tumors in a large referral population: distribution of diagnoses by age, sex, and location. AJR American journal of roentgenology. 1995;164(1):129-34.
12. Arnaud EJ, Perrault M, Revol M, Servant J-M, Banzet P. Surgical treatment of dermatofibrosarcoma protuberans. Plastic and reconstructive Surgery. 1997;100(4):884-95.
13. Khatri VP, Galante JM, Bold RJ, Schneider PD, Ramsamooj R, Goodnight JE. Dermatofibrosarcoma protuberans: reappraisal of wide local excision and impact of inadequate initial treatment. Annals of surgical oncology. 2003;10(9):1118-22.
14. Mentzel T, Beham A, Katenkamp D, Dei Tos AP, Fletcher CD. Fibrosarcomatous (" high-grade") dermatofibrosarcoma protuberans: clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance. The American journal of surgical pathology. 1998;22(5):576-87.
15. Suit H, Spiro I, Mankin HJ, Efird J, Rosenberg A. Radiation in management of patients with dermatofibrosarcoma protuberans. Journal of clinical oncology. 1996;14(8):2365-9.
16. Rutgers E, Kroon B, Albus-Lutter C, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. European journal of surgical oncology: the journal of the European Society of Surgical Oncology and the British Association of Surgical Oncology. 1992;18(3):241-8.
17. Terrier-Lacombe M-J, Guillou L, Maire G, Terrier P, Vince DR, Somerhausen NdSA, et al. Dermatofibrosarcoma protuberans, giant cell fibroblastoma, and hybrid lesions in children: clinicopathologic comparative analysis of 28 cases with molecular data: a study from the French Federation of Cancer Centers Sarcoma Group. The American journal of surgical pathology. 2003;27(1):27-39.
18. Mavili ME, Gursu KG, Gokoz A. Dermatofibrosarcoma with lymph node involvement. Annals of plastic surgery. 1994;32(4):438-40.
19. Snow SN, Gordon EM, Larson PO, Bagheri MM, Bentz ML, Sable DB. Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohs micrographic surgery with long-term follow-up and review of the literature. Cancer: Interdisciplinary International Journal of the American Cancer Society. 2004;101(1):28-38.
20. Kabumoto T, Fujiwara H, Kariya N, Tsujimoto T, Ito K, Ito M, et al. Skin metastasis of dermatofibrosarcoma protuberans with distinct morphological features, confirmed by COL1A1-PDGFB fusion gene analysis. Journal of the American Academy of Dermatology. 2009;61(1):130-2.
21. Monnier D, Vidal C, Martin L, Danzon A, Pelletier F, Puzenat E, et al. Dermatofibrosarcoma protuberans: a population-based cancer registry descriptive study of 66 consecutive cases diagnosed between 1982 and 2002. Journal of the European Academy of Dermatology and Venereology. 2006;20(10):1237-42.
22. Swan M, Banwell P, Hollowood K, Goodacre T. Late recurrence of dermatofibrosarcoma protuberans in the female breast: a case report. British journal of plastic surgery. 2005;58(1):84-7.
23. Ramakrishnan V, Shoher A, Ehrlich M, Powell S, Lucci Jr A. Atypical dermatofibrosarcoma protuberans in the breast. The breast journal. 2005;11(3):217-8.
24. Akhtar K, Sherwani RK, Ray PS. Dermatofibrosarcoma protuberans of male breast: An unusual presentation. Oman medical journal. 2012;27(4).
25. Mirza TI, Akhtar K, Abbas HB, Sameena M, Tahir F, Khan S. Dermatofibrosarcoma Protuberans Male Breast: A Case report. Oman Medical Journal. 2011;26(5).
26. Pohlodek K, Mečiarová I, Grossmann P, Kinkor Z. Dermatofibrosarcoma protuberans of the breast: A case report. Oncology letters. 2017;14(1):993-8.
27. Jiang JQ, Huang Z, Wang LH, Shen SD, Lu H. Dermatofibrosarcoma protuberans of the breast: A case report. Oncology letters. 2014;8(3):1202-4.
28. Lee HJ, Kim MJ, Choi J, Kim EK, Park BW, Park S. Dermatofibrosarcoma protuberans arising on the skin of the breast. The breast journal. 2011;17(1):93-5.
29. Koh T, Kato S, Hirajima S, Nishio M, Soga K, Oguro A, et al. A Case of Dermatofibrosarcoma Protuberans Arising in Subcutaneous Tissue of the Breast. The breast journal. 2016;22(5):578-9.
30. Kim MK, Chang ED, Kim JS, Whang IY. Dermatofibrosarcoma protuberans of the breast. The breast journal. 2013;19(4):442-3.
31. Kim T, Choi YL, Park HY, Song JY, Shin JH, Nam SJ, et al. Dermatofibrosarcoma protuberans of the breast skin. Pathology international. 2010;60(12):784-6.
32. Saikia BK, Das I, Mandal GK. Fibrosarcomatous change in the background of dermatofibrosarcoma protuberans in male breast: Study of a rare case and review of the entity. Journal of mid-life health. 2016;7(1):45.
33. Roy AD, Nishant K, Joshi D. Painful late recurrence of dermatofibrosarcoma protuberans of breast in a centurian female. Journal of clinical and diagnostic research: JCDR. 2013;7(10):2278.
34. Özcan TB, Hacıhasanoğlu E, Nazlı MA, Aksoy Ş, Leblebici C, Talu CK. A rare breast tumor: dermatofibrosarcoma protuberans. The journal of breast health. 2016;12(1):44.
35. Bae SH, Lee JY. Imaging features of breast dermatofibrosarcoma protuberans in various modalities including FDG-PET CT. Iranian Journal of Radiology. 2016;13(2).
36. Kinney MH, Knox SM, editors. Operative management of dermatofibrosarcoma protuberans of the breast. Baylor University Medical Center Proceedings; 2016: Taylor & Francis.
37. Chen X, Chen YH, Zhang Y-l, Guo Y-m, Bai Z-l, Zhao X. Magnetic resonance imaging and mammographic appearance of dermatofibrosarcoma protuberans in a male breast: a case report and literature review. Journal of medical case reports. 2009;3(1):8246.
38. Murase Y, Takeichi T, Matsumoto T, Sakakibara A, Akiyama M. A juvenile male case of dermatofibrosarcoma protuberans on the breast. Clinical and experimental dermatology. 2019.
39. Dragoumis DM, Katsohi L-AK, Amplianitis IK, Tsiftsoglou AP. Late local recurrence of dermatofibrosarcoma protuberans in the skin of female breast. World Journal of surgical oncology. 2010;8(1):48.
40. Sin FN-y, Wong K-w. Dermatofibrosarcoma protuberans of the breast: a case report. Clinical imaging. 2011;35(5):398-400.
41. Ahmed AA, Ostlie D, Fraser JD, Newell B, Cooley L. Dermatofibrosarcoma protuberans in the breast of a 2-year-old girl. Annals of diagnostic pathology. 2010;14(4):279-83.
42. Sandberg AA, Anderson WD, Fredenberg C, Hashimoto H. Dermatofibrosarcoma protuberans of breast. Cancer genetics and cytogenetics. 2003;142(1):56-9.
43. Kim MS, Kim KS, Han HY, Kang YJ, Choi YJ, Kang DW, et al. Fibrosarcomatous transformation in dermatofibrosarcoma protuberans of the breast—a case report. Journal of Clinical Ultrasound. 2009;37(7):420-3.

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