

Dermatofibrosarcoma protuberans of the breast: A case report of a rare breast tumour

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Abstract

Introduction: Dermatofibrosarcoma protuberans (DFSP) is an intermediate and slow-growing sarcoma, arising most commonly in the trunk and extremities and rarely in the breast. It may be recurrent, but metastasis is rare. Excision with good resection margins reduces the rate of recurrence.

Case Presentation: A 28-year-old lady presented with a painless lump in the left breast of 16 years duration. Examination revealed an ulcerated firm mass, well defined, with perilesional nodules on the skin of the breast. Wedge biopsy favoured Borderline Phylloides Tumour over DFSP. Patient was offered mastectomy and the histology of the specimen confirmed Fibrosarcomatous DFSP.

Conclusion: This stresses the importance of histology in diagnosing a rare breast tumour. This is very important because it may show similar appearance to other benign and malignant breast lesions, which makes clear the role of pathological investigation to make a definitive diagnosis.

Keywords: Breast; Dermatofibrosarcoma Protuberans; Mesenchymal Tumour; Fibrosarcomatous DFSP

1. Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare tumour involving the deep dermis and subcutaneous tissues. It accounts for 1.0% of all soft tissue sarcomas, and for less than 0.1% of all malignant tumours [1]. The disease can occur at any age, but 20-50 years age group is most commonly affected. It is a slow-growing sarcomatous tumour involving the trunk and extremities [2]. It was first described by Darier and Ferrand in 1924 [3]. The tumour has a variable degree of malignancy with a high tendency for local invasion and a high rate of recurrence, especially if a negative margin fails to be achieved in the primary resection surgery [4] [5]. Metastasis is rare.

Early detection and treatment is possible because of its superficial location [1]. Imaging findings for DFSP is usually non-specific.

While it is true that the trunk and extremities are most commonly affected, DFSP can affect virtually all parts of the body. However, occurrence in female breast is extremely rare [6]. None has been reported in our environment occurring in female breast; and the only one reported was seen in a male breast [7]. Thus, the purpose of this article is to report a case of DFSP involving the left breast of a 28-year old lady, and to do literature review on the subject matter.

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2. Case Report

A 28-year-old lady presented with a lump in the left breast of 16 years duration. Lump is painless and gradually progressively increased in size. No associated nipple discharge. No swelling in the contralateral breast or axilla. No family history of breast swellings.

Age at menarche was 13 years and she has a regular menstrual cycle. Age at first live birth was 23 years. She has 2 children each of which was breast-fed for one year. No history of use of hormonal contraceptives. Does not smoke. No Family history of breast malignancy. No weight loss, anorexia or bone pain.

Examination revealed a young lady in no obvious distress

Breast examination; There was a 9x7 cm mass at the upper outer quadrant of the left breast, ulcerated. It was firm, attached to skin but not to chest wall. Has well defined edges, but the adjacent areas of the breast skin has involved nodular lesions. No axillary lymphadenopathy. The right breast and axilla were essentially normal.



Figure 1 Clinical pictures shows the 9x7cm mass at the upper outer quadrant of the left breast



Figure 2 Post op picture after mastectomy

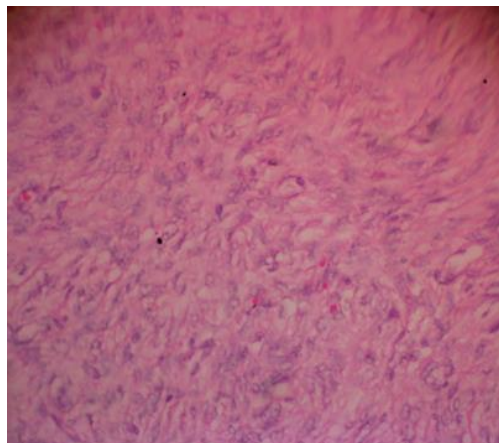


Figure 3 Micrograph of histology shows spindle cells disposed in storiform and honey-comb pattern. (H&E X 400)

She later had wedge biopsy done, the histology of which revealed features of borderline Phyllodes tumour favoured over Dermatofibrosarcoma Protuberans. Patient was counselled for simple mastectomy because of the surrounding nodular lesions. Mastectomy specimen was sent for histology which revealed features of Fibro sarcomatous Dermatofibrosarcoma Protuberans.

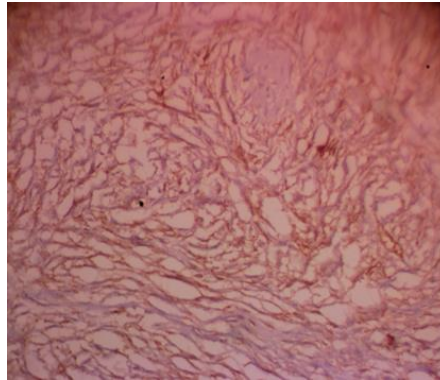


Figure 4 Micrograph of IHC staining for CD34 shows cytoplasmic staining for the lesional cells(x400)

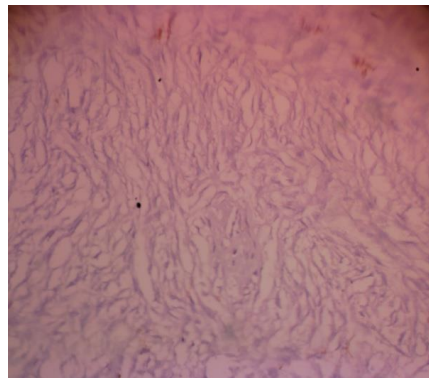


Figure 5 Micrograph of IHC staining for S100 is negative(x400).

Dermatofibrosarcoma protuberance (DFSP) was confirmed by positive reaction to CD34 and negative reaction to S100.

3. Discussion

DFSP is a rare type of mesenchymal tumour. It is said to occur in 40-50% of cases at the trunk, 30-40% of cases at the proximal parts of the extremities, while 10-15% occur in the head and neck [8] [9]. Cases of breast involvement have been rarely reported in literature [10]. It typically develops as deep-red and blue-red lesion and grows slowly, usually exceeding 3cm in size [2] [11].

Some possible events have been discussed as the trigger for DFSP; e.g trauma has been mentioned in 10-20% of cases. Sites of vaccine, surgery scars, radiodermatitis, burns and central veins are less common [8] [10].

Diagnosis of DFSP of the breast at first stage is not accomplished easily since it is a relatively rare breast tumour. A definitive diagnosis is usually established with routine histopathological and immunohistochemical features [12]. In this case, pathology and IHC confirmed the lesion as DFSP. The typical appearance of DFSP is a storiform pattern with uniform spindle cells interdigitating among fatty cells forming a honey-comb pattern in foci. The specimen shows positive reactivity for CD34 at IHC. Conventional mammography usually reveals a subcutaneous oval mass with smooth well-defined borders [11] [13]. Breast ultrasound scan shows a hypoechoic mass lesion with no peripheral or internal blood flow [2]. MRI may be necessary for the localization and differential diagnosis of DFSP [12] [13]. Extensive evaluation with CT scan and laboratory investigations are not usually recommended since metastasis is rare (about 2-5%) [12]. Imaging studies are generally nonspecific.

Histological classification recognizes several subtypes of DFSP: pigmented, atrophic, sclerosing, fibrosarcomatous, and giant cell fibroblast-like, granular cell variant, and myxoid DFSP [12]. Biopsy is needed for a definitive diagnosis. FNAC has low diagnostic accuracy for mesenchymal breast tumours, and so core needle biopsy is considered the optimal procedure for a definitive diagnosis [12]. But a diagnosis from core needle biopsy can be challenging and difficult because the differential diagnosis of solitary fibrous tumour (SFT) may not be appreciable, including alternating cellular and hypocellular architecture, and vascular pattern [12] [14].

Complete surgical excision with a wide resection margin is the optimal treatment for DFSP of the breast [14]. There has been controversy as to the minimum resection margin required to achieve a local control of the tumour. Excision with close margin leads to a local recurrence rate of 26-60% [12]. The optimal resection margin is considered to be 2-3cm² [12] [15]. Farma et al [16] suggested that the use of standardized surgical approach and meticulous pathological evaluation of margins guarantees a low recurrence rate (1%). DFSP is considered to be radiosensitive but the role of adjuvant radiotherapy in the treatment of this neoplasm remains unclear [17].

Imatinib mesylate, a tyrosine kinase inhibitor, may assist in disease control in patients with locally advanced or metastatic disease. However, fibrosarcomatous variant of DFSP may not respond to imatinib [18]. This is because this variant does not exhibit either ring chromosome or an unbalanced chromosomal translocation between chromosomes 17 (Col1A1) and 22 (platelet-derived growth factor B) which is seen in about 90% of DFSP [7] [18]. Fields et al [18] analysed the result of treatment from 244 patients with DFSP following an average follow-up of four years. It was concluded that primary or recurrent treatment of DFSP should be excision with negative margins. Imatinib can also be used in the neoadjuvant period to effectively decrease the tumour size for a successful excision [19]. However, this can only be successful in variants that will respond to imatinib.

4. Conclusion

DFSP is a rare breast tumour. In spite of its classification as a tumour of intermediate malignancy with a limited potential for metastasis, DFSP possesses the potential for aggressive local behavior. Complete surgical excision is the optimal treatment. However, mastectomy can be done if complete excision is not possible. Imatinib may assist in disease control either in adjuvant or neoadjuvant period especially with locally advanced or metastatic disease. But the fibrosarcomatous variant of DFSP do not respond to imatinib. Due to the rare involvement of the breast in patients with DFSP and the rare fibrosarcomatous variant, the present study reports this unique case with the clinical features and histologic findings.

Compliance with ethical standards

Disclosure of conflict of interest

There is no conflict of interest to declare.

Statement of informed consent

Informed consent was obtained from the individual used for the study.

References

- [1] Zhao Z, Cheng S. Dermatofibrosarcoma Protuberans in breast: A case report and review of literature. *Int.J Clin Exp Med.* 2017; 10(2): 3823-3828.
- [2] Jang J, Huang Z, Wang L, Shen S, Lu H. Dermatofibrosarcoma protuberans of the breast: A case report. *Oncol Lett.* 2014; 8(3): 1202-1204.
- [3] Darier S, Ferrard M. Dermatofibrosarcomes progressifs et récidivants en fibrosarcomes de lepeau. *Ann Dermatol Venereol.* 1924; 5: 545-562.
- [4] Cavupoolu T, Yavuzer R, Tuncer S. Dermatofibrosarcoma protuberans of the breast. *Aesthetic Plast Surg.* 2003; 27: 104-106.
- [5] Osime C, Irowa O, Obaseki D, Osaigbovo E. Dermatofibrosarcoma protuberans of the breast in a male: A case report and review of the literature. *Afr J Med Health Sci.* 2014; 13(2): 119-121.

- [6] Zhang L, Liu Q, Cao Y, Zhong J, Zhang W. Dermatofibrosarcoma protuberans: Computed tomography and magnetic resonance imaging findings. *Medicine (Baltimore)*. 2015; 94: e1001.
- [7] Ezejiofor I, Onwukamuche M, Anyiam D, Ugwu J, Ndukwe C, Madubuike K, et al. Breast dermatofibrosarcoma protuberans in an adolescent male: A case report and extensive review of the literature. *Trop J Med Res*. 2017; 20(2): 204-207.
- [8] Llombert B, Serra-Guillen C, Monteagudo C, Lopez J, Senmartin O. Dermatofibrosarcoma protuberans: A comprehensive review and update on diagnosis and management. *Semin Diagn Pathol*. 2013; 30(1): 13-18.
- [9] Maji S, Paul M, Sen S. Dermatofibrosarcoma protuberans of the breast- A Rare Entity. *Indian J Surg Oncol*. 2018; 9(2): 351-354.
- [10] Hoseinpour P, Olfatbakhsh A, Keshavarz M, Beheshtian T, et al. Dermatofibrosarcoma Protuberans; Case Report of a rare Tumour of the Breast. *Multidiscip Cancer Invest*. 2017; 1(1): 26-28.
- [11] Sin F, Wong K. Dermatofibrosarcoma protuberans of the breast: A case Report. *Clin Imaging*. 2011; 35: 398-400.
- [12] Pohlodek K, Meciarova I, Kinkor Z. Dermatofibrosarcoma protuberans of the breast: A case report. *Oncol Lett*. 2017; 14(1): 993-998.
- [13] Chen X, Chen Y, Zhang Y, Guo Y, Bai Z, Zhao X. Magnetic resonance imaging and mammographic appearance of dermatofibrosarcoma protuberans in a male breast: A case report and literature review. *J Med Case Rep*. 2009; 3: 8246.
- [14] Lim S, Ong K, Tee Tan B, Selvarajan S, Tan P. Sarcoma of the breast: An update on a rare entity. *J Clin Pathol*. 2016; 69: 373-381.
- [15] Hameed N, Haque M, Baskovich B. A unique presentation of fibrosarcomatous Dermatofibrosarcoma protuberans of the breast: A case Report. *American J Clin Path*. 2018; 149(1): 105-111.
- [16] Farma J, Ammori J, Zager S, Marzban S, Bui M, Bichakjan C, et al. Dermatofibrosarcoma protuberans: How wide should we respect? *Ann Surg Oncol*. 2010; 17(2): 2112-2118.
- [17] William N, Morris C, Kirwan J, Dagan R, Mendenhall W. Radiotherapy for dermatofibrosarcoma protuberans. *Am J Clin Oncol*. 2014; 37(3): 430-432.
- [18] Fields RC, Hameed M, Qin L, Moraco N, Jia X, Maki R, et al. Dermatofibrosarcoma protuberans: Predictors of recurrence and the use of systemic therapy. *Ann.Surg Oncol*. 2011; 18: 328-336.
- [19] McGee M, Boukhar S, Monga V, Weigel R, Phadke S. Dermatofibrosarcoma protuberans - the use of neoadjuvant imatinib for treatment of an uncommon breast malignancy: A case report. *J Med Case Reports*. 2019; 13: 374-378.