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Dermato Fibrosarcoma Protuberance of The Breast – A Rare Case And Review of Literature.

KEYWORDS

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ABSTRACT *Background: Dermatofibrosarcoma protuberans (DFSP) is a rare, (incidence one case per million per year) uncommon soft tissue tumor involving the dermis of skin with intermediate to low grade malignant potential. DFS is a locally aggressive tumor with a high recurrence rate and very low metastatic potential (1, 4, 8, 17).*

*Case presentation: We are reporting a 32 years lady, who presented with a lump in the left breast for six months. Swelling gradually increased in size and was not associated with pain or nipple discharge. On local examination, there was 6*5 cm, solitary mass occupying the central quadrant, firm in consistency, and freely mobile. Clinically a diagnosis of fibroadenoma was considered. FNAC showed spindle cells and a core biopsy was done, which was suggestive of DFSP. The Patient underwent wide local excision with oncoplasty followed by adjuvant radiotherapy. The aim of this report is to explore the various treatment options.*

Conclusion: DFSP is a very rare histological variant from the breast (2).There are very few reports available in the literature. Wide local excision with 2-3 cm margin is recommended irrespective of site (6). Adjuvant radiotherapy has an important role in the management of DFSP. Keywords: DFSP, Breast, adjuvant RT.

INTRODUCTION:

DFSP is a relatively rare, slow growing locally aggressive tumor and have a very low to intermediate malignant potential (2-5%) (1,2,4,17). This tumor contributes very small percentage among all soft tissue sarcomas. The young and middle aged adults are the most commonly involved age groups (2). Usually these tumors presents like a bluish or brownish erythematous lesion without any symptoms, later to invades the sub cutaneous tissue. The trunk, extremities and head and neck are the most commonly involved sites (2, 17). Herein, we are presenting a case of DFSP arising from the breast parenchyma, which is a rare entity, commonly misleads diagnosis as benign breast tumors like fibroadenoma.

Case presentation: We are reporting a 32 years lady who presented with a lump in the left side of the breast for six months. Swelling increased in size gradually and was not associated with pain, tenderness, and nipple discharge. On local examination, there was a 6*5 cm, solitary mass occupying the central quadrant that was firm in consistency, and freely mobile. Given the young age, a clinical diagnosis of giant fibroadenoma was considered. Bilateral x-ray mammograms showed well delineated smooth walled dense breast mass 5cm in size in the supero medial aspect of the left breast (Fig 1). Fine needle aspiration cytology revealed spindle cells and hence a trucut biopsy was considered. Biopsy showed spindle cells arranged in fascicles and storiform pattern, with pleomorphism and mitotic figures 3/10 HPF (Fig 2a). There were areas of hyalinization with lymphocytic infiltration without necrosis. The features were suggestive of Demato-fibrosarcoma protuberance. Immunohistochemistry showed strong positivity for CD34 confirming the diagnosis of DFSP (fig 2b).

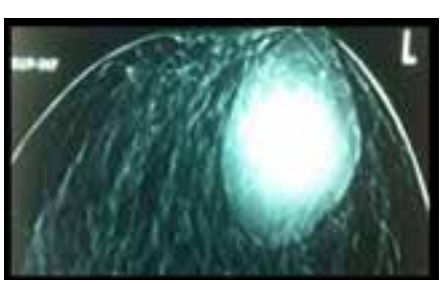

Figure 1 –X-ray mammogram of the left breast shows a well delineated smooth walled dense breast mass – BI-RADS 3.

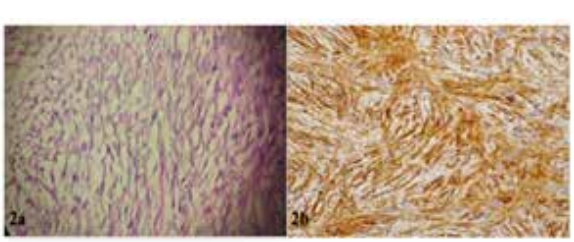

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Figure 2a - Tumor nodules composed of proliferative neoplastic spindle cells arranged in a vague storiform pattern (hematoxylin & eosin stain). (2b) Immunohistochemical staining revealed neoplastic spindle cells, strongly positive for CD34, which is consistent with a diagnosis of dermatofibrosarcoma protuberans.

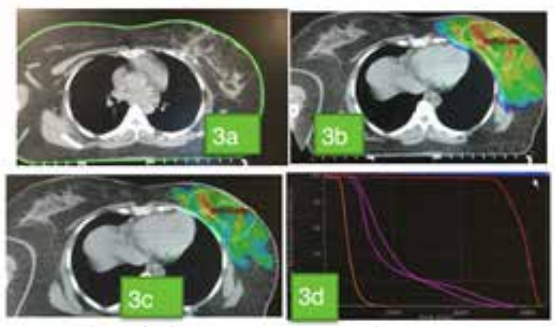

Figure 3a,b,c&d: showing post op CT showing seroma cavity. b, c &d: radiotherapy planning including target deleniations and dose to PTV 50 and PTV 60 Gy, DVH of lungs and heart.

She underwent wide local excision with overlying nipple areola complex and type II oncoplasty. She received adjuvant radiotherapy (60Gy/30 #/@2Gy/# over six weeks under 6MV linear accelerator (Fig 3). Patient completed treatment without any significant adverse effects except for grade 1 skin reactions. She is on regular follow up since six months without any recurrence.

Discussion:

The term DFSP was first coined by Hoffman in 1925 (3,5). It contributes a tiny percentage, around 2 to 6%, of all soft tissue sarcomas (14, 17). It is the most frequently reported sarcoma arising from the skin (14, 17). Though etiological factors are unknown, an injury to the affected skin may be a predisposing factor. Abnormal chromosomes within the tumor cells t (17; 22) (q22; q13) results in a fusion gene COL1A1-PDGFB. This fusion encodes a protein that helps tumor cells proliferate (4, 5).

These tumors are commonly reported to arise from the trunk, followed by proximal extremities and head and neck especially scalp (3, 5). Breast as the primary site of origin is uncommon, and a very few case reports were available in the literature (5, 6, 7). In our case, the lump started as a parenchymal swelling without any skin involvement. DFSP presents initially as a skin lesion, red to bluish in color, and firm in consistency. As the lesion progresses, it can infiltrate underlying structures like fascia, muscle or bone (8, 9). The overlying skin is atrophic and covered by a brown-yellow, red-tinged, scleroderma form or telangiectatic (5, 10). Most of DFSP (90%) are benign in nature and the rest behaves like malignant tumors (11).

Histologically, DFSP shows a pattern of monomorphous proliferation of cytological bland spindle cells with a visible storiform or whorled (rush mat-like) architecture and low mitotic activity. The expression of CD34 on immunohistochemistry is diagnostic of DFSP, and it is useful in differentiating from benign histological variants like fibrous histiocytoma and dermatofibroma (12).

DFSP arising from breast parenchyma is a relatively uncommon, clinical and radiological features are not very definitive for diagnosis. The skin infiltration may be the only differentiating feature between the benign breast tumors and DFSP (13). Surgery is the primary treatment modality (Mohs micrographic surgery or Excision) (5, 14). There was no clear consensus regarding margin status and the role of axillary nodal dissection, however a 2-3 cm margins are considered as adequate (6, 14). Axillary dissection is not routinely recommended in DFSP because the incidence of regional and distant metastasis is extremely rare (5%) (15). Radiation therapy (RT) plays a significant role as adjuvant therapy in DFSP. Radiation therapy is indicated for margins involvement or where wide excision leads to extensive surgical and cosmetic defects. Postoperative adjuvant radiotherapy reduces the risk of recurrence when surgical margins are not attained (2, 5).The recommended adjuvant radiation dose is 50-60Gy for DFSP(16). However, there is difference of opinions regarding the role of radiotherapy (6, 10). Some authors reported that adjuvant radiotherapy improves the cure rates up to 85% (2).

PDGFB-PDGFR-beta signaling pathway plays a dominant role in the proliferation of DFSP tumor cells and targeted therapy has a definitive role in recurrent and metastatic cases (16). Imatinib is a potent and specific inhibitor of several protein-tyrosine kinases, including the platelet-derived growth factor (PDGF) receptors. It inhibits the growth of tumor cells as well as transformation of fibroblasts. Imatinib has shown to improves the clinical response rates (65%) (2,5). Sorafenib is Braf, and VEGF receptor inhibitor and has a definitive role in the treatment of angiosarcoma and MPNST. The role of sorafenib in DFSP is not very clear. However, this drug has shown unremarkable results in some cases (17).

Conclusion:

DFSP is a benign tumor and diagnosis is always by histopathological examination. DFSP arising from the breast parenchyma is an uncommon presentation. Surgery with 2-3 cm margins considered as adequate and is usually the treatment of choice. Radiotherapy has definitive role in the management of DFSP. Targeted therapies like Imatinib and sorafenib has a role in the management of recurrence and metastatic DFSP.

Abbreviations:

DFSP: Dermato Fibro Sarcoma Protruberance.
COL1A1-PDGFB: collagen type I alpha 1-platelet-derived growth factor beta
CD34: Cluster Differentiation 34
PTV: Planning target volume
DVH: Dose volume histograms.
VEGF: Vascular endothelial growth factor
MPNST: Malignant peripheral nerve sheath tumor.

References

1. Darier J:Dermatofibromes progressifs et recidivants ou fibrosarcomes de la peu. Ann Dermatol Syphiligr (Paris) 1924, 5:542-562.
2. I Lym Chan, Sueli Carneiro,Marcia Ramos. Dermatofibrosarcoma Protuberans: A Case Report.Case Rep Dermatol 2014;6:134–139.
3. Laskin WB. Dermatofibrosarcoma protuberans:CA Cancer J Clin. 1992;42:116–25.
4.Simon MP, Pedeutour F, Sirvent N, et al: Deregulation of the plateletderived growth factor B-chain gene via fusion with collagen gene COL1A1 in dermatofibrosarcoma protuberans and giant-cell fibroblastoma: Nat Genet 1997, 15:95–98.
5.Tahir Iqbal Mirza, Kaleem Akhtar, Hussan Burair Abbas et al. Dermatofibrosarcoma Protuberans Male Breast: Oman Medical Journal (2011) Vol. 26, No. 5.
6. Rutgers EJ, Kroon BB, Albus-Lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis: Eur J Surg Oncol 1992 Jun;18(3):241- 248.
7.Cavusoglu T, Yavuzer R, Tuncer S. Dermatofibrosarcoma protuberans of the breast:Aesthetic Plast Surg 2003 Mar-Apr;27(2):104-106.
8. Brabant B, Revol M, Vergote T, Servant JM, Banzet P. Dermatofibrosarcoma protuberans of

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the chest and the shoulder: wide and deep excisions with immediate reconstruction: Plast Reconstr Surg 1993, 92:459–462.

9. Arnaud EJ, Perrault M, Revol M, Servant JM, Banzet P: Surgical treatment of dermatofibrosarcoma protuberans: Plast Reconstr Surg 1997, 100:884–895.

10.Mendenhall WM, Zlotecki RA, Scarborough MT. Dermatofibrosarcoma protuberans:Cancer 2004, 101:2503–2508.

11. Bowne WB, Antonescu CR, Leung DH, et al. Dermatofibrosarcoma protuberans: a clinicopathologic analysis of patients treated and followed at a single institution: Cancer 2000, 88:2711–2720.

12. Weiss SW, Nickoloff BJ.CD-34 is expressed by a distinctive cell population in peripheral nerve, nerve sheath tumors, and related lesions:Am J Surg Pathol 1993, 17:1039–1045.

13.Jin -Qun Jiang, Zhan Huang,Lin-Hui Wang,San-Di Shen,And Hai Lu. Dermatofibrosarcoma protuberans of the breast: A case report: Oncol Lett. 2014 Sep; 8(3): 1202–1204.

14.Chang CK, Jacobs IA, Salti GI.Outcomes of surgery for dermatofibrosarcoma protuberans:Eur J Surg Oncol 2004;30:341-345.

15.Olivier Cottier1*, Maryse Fiche2, Jean-Yves Meuwly3 and Jean-François Delaloye1et alDermatofibrosarcoma presenting as a nodule in the breast of a 75-year-old woman: a case report. Journal of Medical Case Reports 2011 5:503.

16.Matthew T Ballo,Gunner K Zagars,Peter Pisters,.Alan Pollack.The Role of Radiation Therapy in the Management of Dermatofibrosarcoma Protuberans:Int J Radiat Oncol Biol Phys. 1998 Mar 1;40(4):823-7.

17.Francois G, Kamar, Victor F Kairouz and Alain N Sabri et al Dermatofibrosarcoma protuberans (DFSP) successfully treated with sorafenib: case report.clinical sarcoma research 2013.