Case Report

DERMATOFIBROSARCOMA PROTUBERANS OF BREAST- HISTOPATHOLOGIST'S PERSPECTIVE

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare spindle cell skin tumor mostly located in the dermis and infiltrating into the subcutaneous fat. Breast is a rare site of involvement although trunk and extremities represent the most frequently involved areas. Differentiation from other entities such as a leiomyoma is important as this lesion requires adequate margin clearance to prevent recurrence. We present case of a young lady that was clinically considered a fibroadenoma and was initially excised accordingly. Histopathology revealed a spindle cell tumor arranged in storiform pattern. Immunohistochemistry revealed a strong positivity for CD34 immunomarker while negative for other markers applied. This report stresses on the diagnostic aspect of DFSP.

Key Words: Dermatofibrosarcoma Protuberans, Immunohistochemistry, Storiform pattern.

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare spindle cell skin tumor located in dermis or subcutaneous tissue. It shows a high rate of local recurrence but shows a low metastatic potential. Common sites of involvement include the trunk and proximal extremities, although it may occur at any part of the body [1]. Initially it presents as a clinically benign appearing lesion, but if left untreated infiltrates deeply into subcutaneous fat and muscle and so may show fixation to underlying tissues raising the suspicion of malignancy [2]. This may be a reason why sometimes the treatment is initially delayed. DFSP in the region of breast is even rarer and the gender-wise distribution is fairly balanced with a slight predominance amongst females [3]. Radiology does not play a very important role in the pre-operative diagnosis as in many cases the lesion is excised considering it a benign tumor, a fibroadenoma in many circumstances without the patient undergoing a radiological investigation. Atypical cases may undergo an MRI or a CT scan to ascertain the extent of invasion. It should be noted however, that the appropriate surgical treatment is a wide local excision, with a 2-4 cm margin clearance generally required to prevent recurrence [4].

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CASE REPORT

A twenty-six-year-old lady presented to the surgical clinic of Shifa International hospital, Islamabad with a breast lump. Due to the apparently benign nature of the lesion an excision was performed without the patient undergoing any radiological investigation, and the specimen was submitted for histopathologic analysis. The specimen consisted of a lobulated mass measuring 4.5 x 4.5 x 3.2 cm with attached skin measuring 3 x 0.9 cm, connected to the mass by thin strands of fat. Small amount of fat was also identified attached to the tumor. Outer surface was inked and then the tumor was serially sectioned to reveal a solid tan white cut surface with tumor grossly reaching the surgical margins. Representative sections were submitted in multiple blocks and stained with routine Hematoxylin and Eosin (H&E) stain. The tumor on histology comprised of bland appearing spindle shaped cells arranged in a storiform pattern (figure-1). There was no significant mitotic activity and adipocytes were found trapped within the tumor at places (figure-1, inset), in other areas there were admixed vascular channels lined by endothelial cells. The tumor was found reaching the inked margins on histology as well. No connection was found with the skin epidermis. Upon immunohistochemistry, tumor cells were strongly positive for CD34 immunomarker (figure 2) while negative for ASMA (figure 2, inset), S-100 and CKAE1/AE3 (to rule out metaplastic carcinoma). After immunohistochemistry, a final diagnosis of DFSP was rendered.

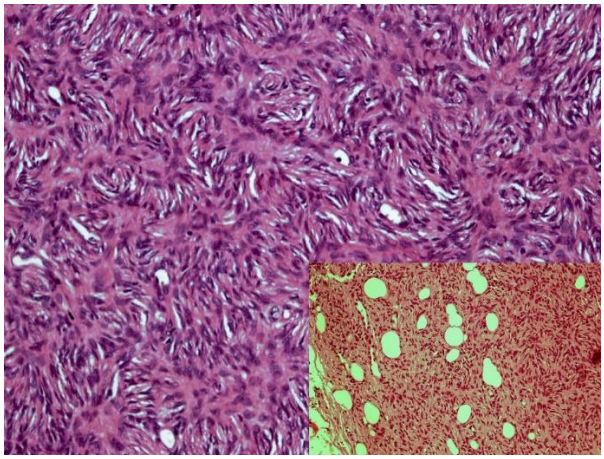

Figure-1: DFSP, H & E stain, 40X. Inset shows infiltration in fat.

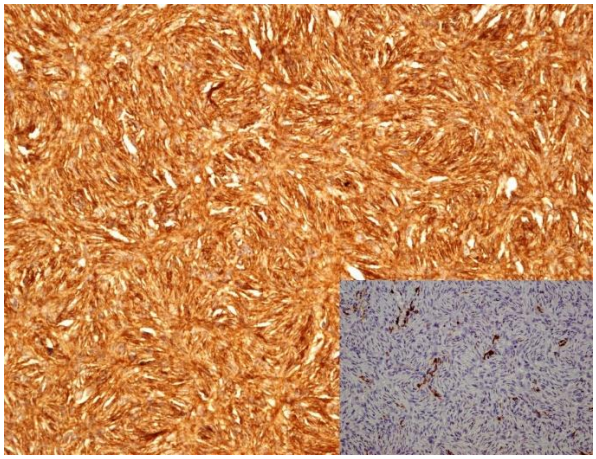

Figure-2: CD34 positivity in DFSP. Inset shows negative ASMA.

DISCUSSION

Sarcomas as a primary tumor are rare in the breast making up to 0.2- 1% of all breast primaries and less than 5% of all soft tissue tumors [5]. DFSP was first mentioned in the literature in 1924 by Darier and Ferrand as "progressive recurrent dermatofibroma" and by Hoffmann in 1925 as 'Dermatofibrosarcoma protuberans' [9]. It is an uncommon but locally aggressive tumor, partly due to inadequate resection margin clearance. In a review of 64 patients, cases that underwent a >1cm margin clearance showed the best results in terms of negative recurrence rates. This study does indicate cases on the trunk; however, no case was specifically mentioned in the breast [7]. At the chromosomal level this group of tumors is characterized by abnormalities of chromosomes 22 and 17 including supernumerary ring chromosomes and amplifications or unbalanced 17; 22 translocations. Recent studies have identified specific fusion of *COL1A1* with *PDGFB* in most of the cases [8]. It most commonly presents as a blue-brown erythematous area with protuberance on skin surface. Recurrent dermatofibroma, keloid, skin manifestation of myofibroblastoma, metaplastic carcinoma and fibromatosis are the entities which enter the clinical differential diagnosis. The tumors which may enter the histologic differential diagnosis include metaplastic carcinoma, fibromatosis, myoepithelioma, and Phyllodes tumor [9]. These findings have also been described in one of the only two cases retrieved from national journals, mentioning DFSP specifically in the breast [9, 10]. According to a recent international publication, many of the cases described in literature have been tumors of subcutaneous tissue secondarily invading the breast. Applying this criterion, only five cases of DFSP breast have been identified in the international literature [11].

Despite the specific genetic abnormalities described above, most cases are diagnosed by routine histopathology combined with immunohistochemistry. Histologically, the tumor shows a diffuse proliferation of plump spindle shaped cells arranged in a storiform pattern and in the form of short fascicles. Individual cells are bland and marked mitotic activity is not necessary. Our case was also devoid of any mitotic activity and a storiform pattern was clearly identified. Our case did not morphologically mimic at least any of the aggressive histopathological entities. Most of the cases are positive for CD34 and vimentin immunomarkers, while negative for Pancytokeratin, S-100 and Smooth muscle actin. Fairly recent advanced studies have also identified the positive expression of newer immunomarkers like CD105 in DFSP [12]. Considering our morphological differentials, we applied the available immunomarkers CD34 and ASMA with S-100 later included in the panel. CD34 was strongly positive with ASMA and S-100 being negative in the tumor cells.

CONCLUSION

DFSP is a rare tumor and even rarer in the breast. Recognizing it here in this region is important as it requires wide local excision and a close follow-up, without lymph node dissection in most cases. However, it needs aggressive therapy as compared to benign tumors of this area.

AUTHORS CONTRIBUTION

Muhammad Bilal: Conception and design of work, acquisition and analysis of data and literature review
Zafar Ali: Analysis of data and literature review
Nadira Mamoon: Overall supervision and final approval

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