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Case Report

Dermatofibrosarcoma Protuberance of Male Breast: A Rare Case Presentation

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ABSTRACT

We report a case of a 50-year-old man who is a chronic smoker, referred to our IBCC (Integrated Breast Care Centre) department, in AIIMS, Rishikesh with complaints of a lump in his left breast for 6 years. A 3 × 2 cm, ill-defined, firm to hard, non-mobile lump located in the upper and outer quadrant of the left breast which is beneath the previous surgical scar with skin infiltration is noted. Histological characteristics of DFSP are the arrangement of monomorphic spindle cells in a storiform pattern. It was staged as T4b N0 M0 and left wide local excisional biopsy of the tumor with a macroscopically free margin of 3 cm and sent for frozen section which revealed a dendritic differentiation with free margins and further confirmed on final histopathological examination. The postoperative recovery of the patient was uneventful. No loco-regional recurrence or distant metastasis was noted for 6 months of regular follow-up.

Keywords: Breast cancer; Dermatofibrosarcoma; Lump

INTRODUCTION

TIn the literature, Taylor first described Dermatofibrosarcoma protuberans (DFSP) as a keloid sarcoma in 1890 [1]. “Ferrand later identified it as a recurrent dermatofibroma” [2]. It is a rare soft tissue sarcoma accounting <5% out of all soft tissue tumors identified in early or middle age [3]. Most commonly seen in 20-50 year age groups but rarely reported in young and elderly people. The term "Dermatofibrosarcoma protuberance" (DFSP) was coined by Hoffman et al in the year 1925, after modifying the previous existing term dermatofibroma [3]. DFSP is an intermediate grade mesenchymal tumor of fibroblastic origin arising from dermis involving subcutaneous tissue and musculature [4]. The majority of this tumor is observed in the trunk region (47.4%) and extremities (38.1%) [5]. Breast involvement of DFSP is a rare entity and even rarer in men. In literature, Some case reports of breast involvement in females are noted, but the involvement of the breast in the male are extremely rare. If involved, they present clinically as erythematous indurated firm subcutaneous nodules in the breast region [4].

On clinical examination, small nodule noted on the skin surface, which is firm and irregular inconsistency, varying from flesh to reddish-brown. "Platelet-derived Growth factor (PDGF) increase response leads to increased formation of tumor cells by reciprocal translocations of chromosomes 17 and 22, t (17;22), and supernumerary ring chromosomes composed of interspersed sequences from bands 17 (17q22) and 22 (22q12)" [6,7]. Histological characteristics of DFSP are the arrangement of monomorphic spindle cells in a storiform pattern. Distant metastasis is noted but the incidence is rare. Treatment is primarily surgical excision with wide margins, with or without adjuvant chemotherapy and radiation. In this article, we report a rare case presentation of a 50 year old male patient with Dermatofibrosarcoma Protuberance tumor in his left breast.

CASE PRESENTATION

We report a case of a 50-year-old man who is a chronic smoker, referred to our IBCC (Integrated Breast Care Centre) department, in AIIMS, Rishikesh with complaints of a lump in his left breast for 6 years. The history revealed similar left breast lump 14 years back for which excision was done in a private hospital. The pre and post-operative details of the patient condition at that time are not available. Later on, he developed breast soreness at the operative site 6 years back which progressively increased in size. No family history significant of breast cancer. This patient is considered for further examination.

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Clinical examination

A 3 × 2 cm, ill-defined, firm to hard, non-mobile lump located in the upper and outer quadrant of the left breast which is beneath the previous surgical scar with skin infiltration is noted. There is a healed scar noted in the upper/outer quadrant of the left breast (Figure 1). No axillary lymph nodes are palpable on examination. Opposite breast and axillary examination findings are insignificant. Then the patient is subjected to further radiological evaluation (Figure 1).

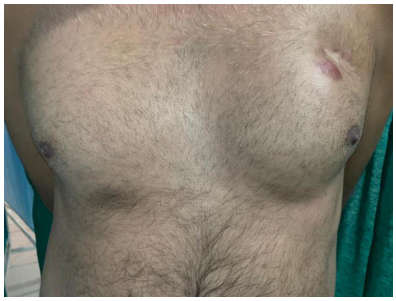

Figure 1: Clinical picture of Dermatofibrosarfoma Protuberance in the left male breast.

Imaging

On ultrasonogram, it was revealed that a relatively well defined 2.5 × 2.3 cm, wider than taller oval-shaped hetero-echoic lesion noted at 11-12 o clock position which is 0.2 cm deep to the skin, 4 cm away from the nipple and noted beneath the scar mark and mammography, demonstrated a well-defined, 2.8 × 2.5 cm, round to oval, a high-density lesion in the upper central region of left breast, within 2 cm from the nipple, with mild architectural distortion noted in the vicinity of the lesion which is categorized as BI-RADS (Breast Imaging Reporting and Data System) 4B lesion with moderate suspicion of malignancy (10%-50%) (Figures 2 and 3).

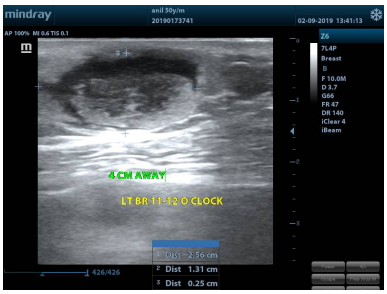

Figure 2: Imaging features of Dermatofibrosarfoma Protuberance of Ultrasound.

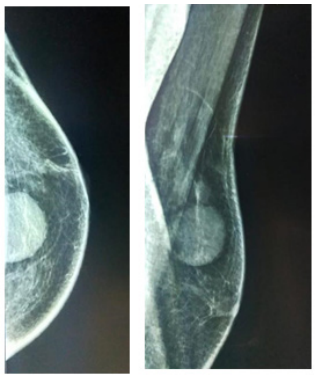

Figure 3: Imaging features of Dermatofibrosarfoma Protuberance Mammogram.

Ultrasound grayscale image shows a well-defined, oval, heteroechoic lesion, free from underlying muscles and overlying skin. Overlying skin is thickened.

(i) Craniocaudal and (ii) mediolateral oblique view of left breast shows a well-defined, round to oval high-density lesion in the upper central quadrant, with mild surrounding architectural distortion. Diffuse skin thickening noted. No calcification noted within the lesion.

Pathological examination

The patient had a Fine needle aspiration (FNAC) of the lesion which revealed as singly cellular exhibiting tumor cell fragments of irregular plump ovoid to spindle cells scanty eosinophilic cytoplasm and bland nuclear chromatin with prominent nucleoli. Tumor cell fragments are elongated with collagenous fibrous stroma which is suggestive of spindle cell sarcoma, malignant phyllodes, and metastatic carcinoma. On Core needle biopsy, it revealed as uniform spindle cells arranged in fascicles and exhibiting prominent storiform pattern with an area of tumor-infiltrating fat also seen which are in favor of dermatofibrosarcoma protuberance (Figure 4). On Immunohistochemistry CD 34 is positive and all other markers are negative such as CK, DESMIN, S-100, SMA, P63, ER, Bcl2, CD99, TLE-1, STAT6, EMA (Figure 5).

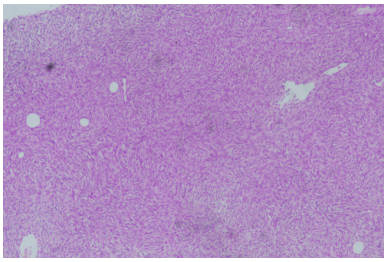

Figure 4: Histopathological features of Dermatofibrosarcoma Protuberance slide showing spindle cells with storiform to whorled pattern.

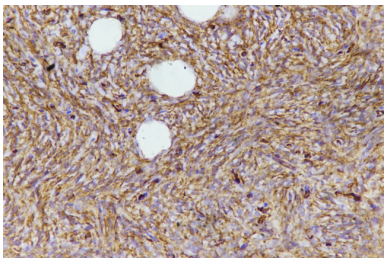

Figure 5: Dermatofibrosarcoma Protuberance immunohistochemistry showing Diffuse CD34 staining.

Metastatic Workup

Chest X-ray and ultrasound of the abdomen were performed, which are normal in the study and we further proceeded for CECT (Thorax and Abdomen) in which there was no evidence of distant metastases.

Management

Provisional diagnosis as Dermatofibrosarcoma protuberance involving left breast is confirmed and was staged as T4b N0 M0. We performed left wide local excisional biopsy of the tumor with a macroscopically free margin of 3 cm and sent for frozen section

which revealed a dendritic differentiation with free margins. The postoperative recovery of the patient was uneventful. On the final histological examination, the surgical margins were 3 cm which is free from the tumor, and the diagnosis of DFSP of the left breast was confirmed. No loco-regional recurrence or distant metastasis was noted for 6 months of regular follow-up.

RESULTS AND DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a soft tissue tumor of low grade involving the dermis and further extends into subcutaneous tissues, underlying musculature [8]. It accounts for about <0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas. It was previously considered that DFSP does not present in any specific ethnic, community or geographic areas, but after analyzing cases by Criscione and Weinstock from the year 1973 to 2002, it was found that the annual incidence of DFSP in the United States is 4.2 per million/yr, and was predominant in black males with a DFSP pigmentary variant (Bednar tumor) [9,10].

The average age of presentation is 38.5 years with equally distributed in both genders [11]. In a study conducted by Rutgers et al., out of total sample size 514 patients, 57% were males [12]. A recent case report by Cavusoglu and colleagues reported tumor presentation and breast involvement are even rare in females [13]. The most common site of this tumor presentation are trunk and extremities and breast involvement is even rare [14]. A study by Karcnik et al. reported less involvement of the breast region [15]. We reviewed the literature regarding this tumor case report and found only 11 cases of DFSP involving breast in male patients in PubMed and Google Scholar from 1994 to 2019 [16-24]. The mean age of these patients was 33.6 years ranging from 9 to 55 years, in our case report, the age of presentation is 50 years and the mean tumor size is 6.2 cm ranging from 3-13 cm similar to our case which has max diameter size of 2.5 cm.

The ethio-pathogenesis of DFSP remains unclear and mainly observed to be associated with post-traumatic areas like tattoos, burn scars, vaccination sites, surgical scars, and radiotherapy [22]. Trauma may initiate a chronic inflammatory process that leads to disorder in the immune system and then malignant transformation of the dermal component is noted [22]. "This hypothesis is supported by genetic findings such as chromosomal translocation of t (17; 22) (q22; q13) between chromosomes 17 and 22, the fusion of gene COL6A3-PDGFD, and the fusion of gene COL1A1-PDGFB" [23,24].

The most common presentation of DFSP in the male breast is an irregular mass with extensive nodules over the surface, with size measuring 1-5 cm in diameter [16-24]. This presentation could imitate various other conditions like carcinoma, phyllodes tumor, and myoepithelioma [22]. In our case, the clinical appearance was the appearance of a small lump at a previously operated site which progressively increased in size. In mammography, it appears as an irregular dense mass without fat or calcification [25]. Ultrasound shows a heterogeneous or hypoechoic mass, irregular with hypervascularity of the affected areas in color Doppler [22].

Due to these nonspecific imaging findings, pathological assessment is required for confirming the diagnosis [4]. Histological characteristics of DFSP are the arrangement of monomorphic spindle cells in a storiform pattern (Figure 4). The intense staining for CD34 differentiates DFSP from myxoid liposarcoma [4,16,17]. Our case showed diffuse strong positivity for CD34 and negative for all other markers like CK, DESMIN, S-100, SMA, P63, ER, Bcl2, CD99, TLE-1, STAT6, EMA (Figure 5).

Surgical excision of the tumor remains the cornerstone for managing these tumors considering oncological outcomes and cosmetic issues related to male breasts. Wide Local Excision technique with or without plastic reconstruction as the preferred surgery in DFSP [19]. The resection margins of at least 2-3 cm are recommended for treatment because of local recurrence rate is 20% to 50% [4]. In a pooled analysis of published series of DFSP treated with MMS or WLE, the local recurrence rate averages about 1.5% [26-31].

Adjuvant therapy like chemotherapy or radiation to the tumor bed is needed for an unresectable lesion or resected lesion with positive surgical margins which will decrease the rate of local recurrence and prevent the mutilation, functional deficits resulting from repeated surgeries [26]. "Imatinib used as an immunotherapy agent for unresectable, recurrent, and metastatic DFSP who are not eligible for surgical excision showed a response rate between 5.2% and 55.2%" [27]. Although distant metastasis rates of DFSP are rare (1%-4%), but they are associated with high rates of local recurrence and mortality rate in 2 yrs. A small subset of patients presents with the variation of DFSP like fibrosarcomatous progression which will have more aggressive features and associated with poor prognosis [31]. Poor prognostic factors associated with DFSP are elderly age groups, a high mitotic index, and increased cellularity [28].

Long-term follow-up is required in these tumors due to high recurrence rates.

CONCLUSION

Our case has been treated with wide local excision with a 3 cm margin status with negative margins on the frozen section and confirmed by histopathology followed by no adjuvant treatment.

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