CASE REPORT

Dermatofibrosarcoma Protuberans of the Breast Skin Simulating Mammary Carcinoma

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Abstract Dermatofibrosarcoma protuberans is a rare soft tissue sarcoma that arises from the dermis and invades deeper tissues. Usually, it behaves as an intermediate grade malignancy but in rare instances, it can metastasize. Frequent occurrences have been observed in trunk and extremities but involvement of the breast has rarely been reported. Therefore, dermatofibrosarcoma protuberans (DFSP) breast often masquerades primary breast malignancy on clinical and radiological grounds. Histomorphology and immunohistochemistry are helpful in making a definitive diagnosis. In the present case, monomorphic spindle cells were seen arranged in classical storiform pattern with large areas of myxoid change. Many times, such myxoid areas pose a diagnostic challenge and necessitate differentiation from other myxoid neoplasms. Herein, immunohistochemistry offers an advantage where tumor cells show strong reactivity for CD34 antibody. Wide surgical excision remains the treatment of choice. The salient clinical features, histogenesis, histomorphology, and immunohistochemical findings have been described with strong emphasis on the diagnostic and therapeutic management of this rare entity.

Keywords Dermatofibrosarcoma protuberans · Breast · Morphology · Immunohistochemistry

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, slowly growing spindle cell neoplasm of soft tissues with intermediate malignant potential. It presents as an indurated plaque on which multiple reddish purple, firm nodules arise, sometimes in association with ulceration [1]. It usually appears on trunk and extremities of young adults; however, involvement of the breast has been reported in rare instances [2]. Thus, DFSP of the breast is often misdiagnosed as primary breast lesion, and histopathology is essential for accurate diagnosis.

Case Report

A 50-year-old woman presented with a painless left breast mass since the last year. She was hypertensive and anxious and was suffering with somatoform disorder. Recently, progressive enlargement of the mass was noted. Physical examination revealed a firm to hard, well-circumscribed, non tender, mobile, 4 × 3 cm mass in the upper inner quadrant of the left breast. No axillary lymph nodes were palpable. Chest X-ray showed a lobulated opacity due to calcified breast lump. Mammography revealed a well-circumscribed, lobulated, high density mass located in subcutaneous plane of upper inner quadrant. The mass showed internal vascularity on color Doppler, and therefore, a grade of BIRADS 5 was given. FNAC was attempted twice and the report was inconclusive. With high index of suspicion on clinical and radiological features, wide excision lumpectomy of the left breast was performed.

Gross examination showed a nodular protruding mass measuring 5 × 4.5 × 3.5 cm. Cut section of the tumor was gray-white, firm, and homogenous. Large areas of myxoid change with foci of hemorrhages were noted (Fig. 1). Histologically,

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Fig. 1 Gross photograph of the left breast lump showing a protruding firm grayish white mass with a large area of myxoid change (marked with black arrows)

the tumor was composed of monomorphic looking fibroblastic cells loosely arranged in vague storiform pattern underlying the epidermis (Fig. 2a, b). The tumor cells were oval to spindle with pale cytoplasm and indistinct borders. The nuclei showed fine chromatin with inconspicuous nucleoli. Dispersed within the tumor, several mast cells were noted that were highlighted by toluidine blue stain. The tumor showed myxoid areas with loss of storiform pattern, prominent vasculature, and mild to moderate anisonucleosis (Fig. 2c). This feature was evident in 35–40% of the tumor. Mitotic activity (mitotic count $<1-2/\text{hpf}$) was not significant. The tumor showed infiltrative borders with extension into the subcutaneous fat of breast tissue. However, all the resected surgical margins were free of tumor. CD34 and vimentin

immunostaining showed diffuse reactivity (Fig. 3a, b) while SMA was strongly expressed in classical areas (Fig. 3c). Immunohistochemical markers for pancytokeratin, epithelial membrane antigen, and CD68 were negative. Ki67 proliferative index in myxoid areas ($\text{Ki-67} = 3-4\%$) (Fig. 3d) was comparatively higher than that in cellular areas ($\text{Ki67} < 1\%$). Thus, final diagnosis of dermatofibrosarcoma protuberans of the breast was formed. Postoperative recovery of the patient was uneventful, and even after 10 months of follow-up, she did not exhibit any signs of recurrence.

Discussion

Primary mesenchymal tumors of the breast are rare where sarcomas arising in the breast parenchyma account for 0.2 to 1.0% of all breast malignancies [3]. The reported incidence of DFSP is approximately five cases per one million individuals per year [1]. Dermatofibrosarcoma protuberans, first described by Darrier and Ferrand in 1924 [4] and termed by Hoffmann in 1925 [5], is an uncommon tumor originating in the skin and subcutaneous tissue [4]. Molecular phenotypic studies to characterize the cellular origin in DFSP have proposed the possible phenotype to be similar to immature dermal reserve cells. It has an indolent growth and long preclinical duration with most cases becoming apparent in young adulthood [6].

DFSP arising in a breast is rare, and a comprehensive search on PubMed and Google Scholar displayed less than 90 such cases from 1990 to 2016. Previous case reports were reviewed with the observation that DFSP breast is more

Fig. 2 **a** Photomicrograph showing uninvolved epidermis with underlying spindle cell tumor showing a distinct demarcation between cellular and myxoid areas (marked with black arrow) (H&E, $\times 4$). **b** Photomicrograph showing tumor cells arranged in characteristic storiform pattern. (H&E, $\times 20$). **c** High power view of hypocellular myxoid area exhibiting prominent and dilated vascular channels (H&E, $\times 40$)

Fig. 3 **a** Immunostain for CD34 antibody showing strong positive reactivity in both cellular and myxoid areas ($\times 20$). **b** Vimentin stain demonstrating diffusely positive tumor cells arranged in storiform pattern ($\times 20$). **c** Immunostain for SMA showing positivity in tumor cells with prominent expression in classic areas than that in myxoid areas ($\times 20$). **d** Immunostain for Ki-67 from myxoid area showing positivity in few tumors cells ($\times 40$)

common in females between 20 to 30 years of age (Table 1). However, it has been described in extremes of ages also with one case reported in a young girl of 2 years age and another one in a centenarian female aged 102 years [12, 16].

The clinical presentation is usually in the form of a firm, plaque-like lesion of the skin surrounded by blue discoloration. Many times, it may enter a rapid growth phase giving rise to multiple nodules. DFSP breast has been observed in patients taking estrogen replacement therapy and radiation therapy, during pregnancy and on preexisting surgical scars [6].

The present case is unique due to its unusual location in the upper inner quadrant of the breast. The tumor had areas of calcification and increased internal vascularity, and was therefore clinically and radiologically misinterpreted as primary malignancy of the breast.

DFSP arising in the breast needs to be differentiated from other spindle cell lesions of the breast such as phyllodes tumor, fibromatosis, myoepithelioma, or metaplastic carcinoma [13]. Phyllodes tumors are biphasic by nature and exhibit hypercellular mesenchymal component surrounding benign breast ducts. In the present case, although multiple sections were taken from the surgical specimen, proliferating breast ducts were not identified. DFSP can be indubitably separated from other spindle cell tumors of the breast by the application of immunohistochemical markers.

Histopathology in the present case revealed largely areas showing classic storiform pattern of DFSP; however, myxoid changes were noted in 35–40% of the tumor. Due to rich vascularity in the latter, the lesion was misdiagnosed on color Doppler as malignant. Myxoid change can be observed in other mesenchymal spindle cell tumors such as myxoid liposarcoma, myxoid benign

fibrous histiocytoma and myxoid neurofibroma [17]. The superficial location, gross configuration, complete lack of lipoblasts, and CD34 positivity help in differentiating DFSP from myxoid liposarcoma. Areas of myxoid degeneration in DFSP breast have been described in isolated case reports by Saikia BK et al., Roy AD et al., and Ahmed AA et al. [8, 12, 16]. Neurofibroma usually contains tactoid structures and other neural elements with presence of S-100-positive tumor cells. Benign fibrous histiocytoma can be ruled out by its site, growth pattern of the tumor cells, and accompanying cell population like inflammatory cells, giant cells, and siderophages with focal CD34 immunostaining [17]. The present case showed diffuse strong positivity for CD34 and vimentin antibody and focal immunoreactivity for SMA. In addition to the typical case, many morphologic variants have been described such as pigmented, fibrosarcomatous, granular cell type, myxoid, and myofibroblastic.

The myxoid subtype of DFSP is diagnosed when more than 50% of the stroma is myxoid [18, 19]. The present case showed only 35–40% of the myxoid areas. Myxoid changes in DFSP is characterized by interstitial accumulation of hyaluronic acid ground substance rendering the usual storiform pattern, less pronounced, and a non patterned array of vessels, more apparent [17]. Moreover, a peculiar finding that needs special mention is that the mitosis was comparatively higher in these areas than that in the cellular area.

It was observed in a study by Kim M et al. [20] that intratumoral mast cells are negatively correlated with mitosis and tumor size implying that mast cells do not have a causative primary role in tumorigenesis but rather play a secondary

Table 1 Clinicopathological and immunohistochemical features of dermatofibrosarcoma protuberans breast in isolated case reports

Sn	Author	Age/ sex	Location	Size	Clinical presentation	Additional histopathology	Follow-up	IHC
1.	Kinney MH et al. (2016) [7]	26/F	UIQ left breast	3 × 2 cm	Purple-hued, protuberant, irregular	—	—	—
2.	Saikia BK et al. (2016) [8]	40/M	Central part of left breast	6.5 × 4 cm	Firm, mobile, non tender, nodular, ulcerated	Fibrosarcomatous and myxoid change	—	CD34+, SMA–, S100–, desmin–, CK–, EMA–
3.	Rahbi SA et al. (2015) [9]	28/F	UOQ right breast	5 × 6 cm	Firm, well–circumscribed, tender, mobile, brownish	—	6 months	CD34+, vimentin+, SMA–, desmin–, S-100–, CD31–, pancytokeratin–
4.	Tarakji MA et al. (2015) [10]	27/M	LOQ right breast	4 × 3 cm	Bluish, lumpy, mobile swelling with telangiectatic patches	—	12 months	CD34+, bcl 2+, CD99+, β-catenin+, vimentin+
5.	Jiang JQ et al. (2014) [11]	26/F	LOQ left breast	3.2 × 0.9 cm	Brown-red, firm, fixed breast mass with an ill-defined border and smooth margins	—	12 months	CD34+
6.	Roy AD et al. (2013) [12]	102/F	UOQ right breast	8 × 6 cm	Firm, well-defined, tender, mobile, erythematous mass	Necrosis and myxoid degeneration	—	Vimentin+, CD34+
7.	Rao C et al. (2012) [13]	62/F	UOQ right breast	6 × 6 cm	Firm, well-defined, non tender, mobile mass	—	6 months	CD34+, vimentin+, S-100–, bcl-2–, pancytokeratin–, SMA–, Factor VIII–, calponin–
8.	Cottier O et al. (2011) [14]	75/F	UIQ left breast	1.4 × 0.8 cm	Dish-shaped skin nodule	—	12 months	CD34+, CD31–, S-100–
9.	Park TH et al. (2011) [15]	28/F	Medial lower quadrant of left breast	2 × 1 cm	Slow-growing pigmented protruding smooth mobile mass in the	—	12 months	CD34+ and focally positive for SMA, and CD68– and S-100–
10.	Ahmed AA et al. (2010) [16]	2/F	Left breast	—	Rapidly enlarging mass	Large hypercellular fibrosarcoma-like areas, few myxoid areas, and other areas with multi-nucleated giant cells	—	CD34+
11.	Present case	50/F	UIQ left breast	4 × 3 cm	Firm to hard, well–circumscribed, non tender, mobile	Myxoid areas with loss of storiform pattern	10 months	CD34+, Vimentin+, focally SMA positivity, CK–, EMA–, CD68–

UIQ upper inner quadrant, UOQ upper outer quadrant, LOQ lower outer quadrant

role. In the present case also, there were several intratumoral mast cells but mitotic activity was not significant, which supports the above hypothesis.

Although, DFSP usually does not metastasize, it may frequently recur if not extirpated completely [6]. In the present case, the tumor was completely excised with wide surgical margins, and the patient is free of disease after 10 months of follow-up.

Conclusion

It is being emphasized that though localization of DFSP in the breast is less heard of, its awareness is essential as on several occasions, its clinical and radiological resemblance to primary breast malignancy leads to misdiagnosis. Histopathological examination and immunohistochemistry aid in establishing the definitive diagnosis especially when it undergoes myxoid

changes where its differentiation is needed from other spindle cell lesions of the breast.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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