Case Report

A case report on dermatofibrosarcoma protuberans with extensive myoid differentiation in breast: A rare presentation at an unusual site

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ABSTRACT

Dermatofibrosarcoma protuberance (DFSP) is a rare, locally aggressive, cutaneous soft-tissue sarcoma characterized by a high propensity for local recurrence but a low metastatic potential. Its occurrence in the breast is extremely uncommon and may mimic benign or malignant breast lesions clinically and radiologically, posing a diagnostic challenge. Here, we present a case of a 42-year-old female, who presented with a slow-enlarging, firm, and painless swelling in the upper outer quadrant of the right breast for 1 month. Clinical and radiological evaluation suggested a benign spindle cell lesion. Histopathological examination revealed a spindle cell neoplasm with a storiform pattern arising from the dermis and infiltrating the surrounding breast tissue. Immunohistochemistry showed strong CD34 and h-caldesmon positivity and negativity for S-100, desmin, cytokeratin, and signal transducer and activator of transcription 6, confirming the diagnosis of DFSP with extensive myoid differentiation.

Key words: Breast, Dermatofibrosarcoma protuberance, Histology, Rare, Storiform

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive, fibroblastic neoplasm affecting deep dermis and subcutaneous tissues. It was first reported by Darier and Ferrand in 1924 [1]. Histologically, DFSP is composed of uniform spindle cells, arranged in storiform pattern and typically shows diffuse CD34 positivity. Although the classic morphology is well recognized, several histologic variants have been described, including fibrosarcomatous, plaque, atrophic, myxoid, pigmented (Bednar tumor), and myoid types [2,3]. Myoid differentiation within DFSP is exceedingly uncommon and may pose a diagnostic challenge due to its unusual morphology and potential overlap with other spindle cell tumors showing smooth muscle differentiation. Recognition of this variant is important to avoid misdiagnosis and to understand the spectrum of differentiation possible in DFSP.

We report a case of DFSP with myoid differentiation, highlighting its histopathological and immunohistochemical features.

CASE REPORT

A 42-year-old female presented with palpable swelling in the right breast for 1 month. The patient was not on any drug, and there was no relevant family history. There was no associated pain or nipple discharge.

On examination, a mobile, hard, non-tender lump was palpated in the right upper outer quadrant of the breast. There was no significant axillary lymphadenopathy. Mammogram showed an irregular high-density subcutaneous lesion measuring 5 cm × 4 cm, with spiculated margins in the upper outer quadrant of the right breast (Fig. 1).

The patient underwent a wide local excision with a 2 cm margin clearance. Ipsilateral lymph node dissection was not performed. On macroscopic examination, the skin overlying the breast appeared nodular, and cut section showed a firm, white, and homogenous lesion arising from the overlying skin and extending to the breast (Fig. 2).

Microscopy shows an infiltrating neoplasm composed of spindle cells arranged in a storiform pattern. Cells were plump and elongated with indistinct nuclei and a moderate amount of cytoplasm. Entrapped mature adipocytes and bilayered ducts are seen (Fig. 3).

Immunohistochemistry showed spindle cells positive for CD34, h-caldesmon, and CD10. Desmin, estrogen receptor, progesterone receptor, smooth muscle actin (SMA), cytokeratin, S-100, and signal transducer and activator of transcription 6 were negative. Hence, the possibility of other spindle cell neoplasms such as cellular myofibroblastoma and solitary fibrous tumor

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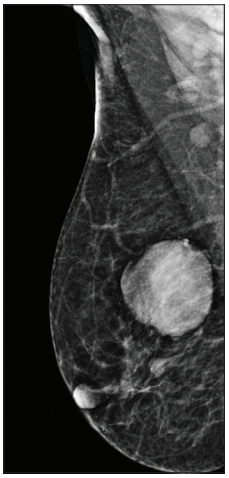

Figure 1: Mammogram showing an irregular high-density subcutaneous lesion measuring 5 cm x 4 cm, with spiculated margins in the upper outer quadrant of the right breast

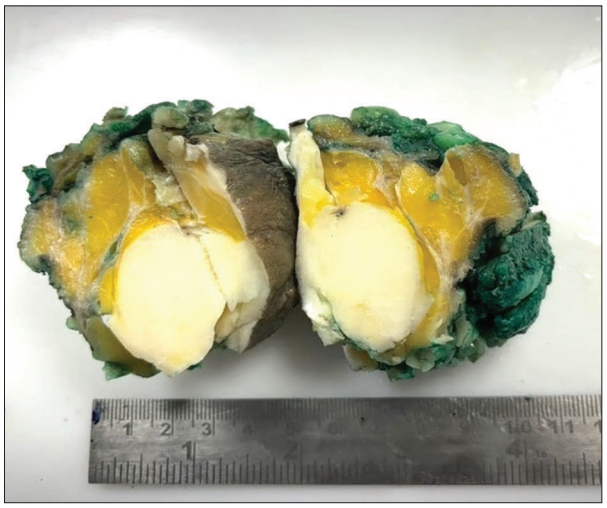

Figure 2: Gross image showing breast tissue with a firm, white, homogenous lesion arising from the overlying skin

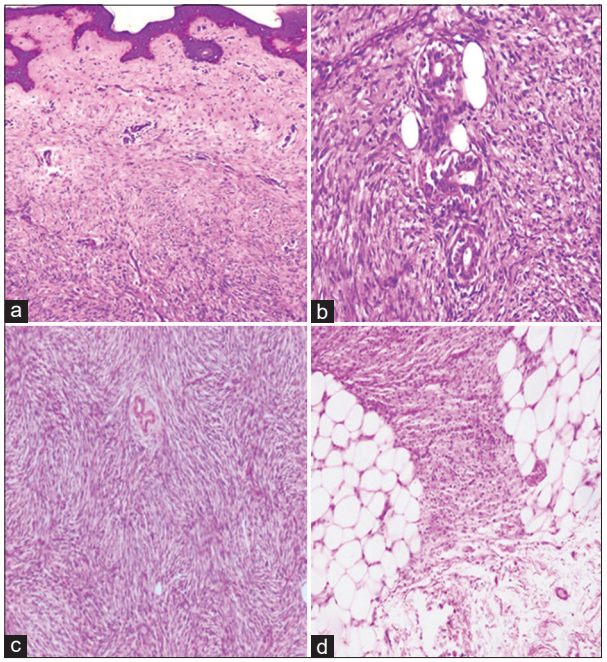

Figure 3: Microscopy (a) shows spindle cell neoplasm arising from the dermis of the overlying skin of the breast. (b) Entrapped bilayered mammary ducts and adipocytes within the neoplasm. (c) Spindle cells are arranged in a storiform and whorled pattern. (d) Neoplasm infiltrating into the surrounding fatty tissue

(SFT), neural tumors, epithelial, and myoepithelial tumors was ruled out. Ki-67 proliferation index in this case was 20% (Fig. 4).

DISCUSSION

DFSP of the breast is an uncommon, low to intermediate-grade dermal soft-tissue tumor known for its locally aggressive nature and high recurrence rate. Although the tumor usually involves the trunk and extremities, its occurrence in the breast is extremely rare [4]. Because of this unusual site, DFSP of the breast can be easily confused with more common benign or malignant spindle cell lesions of the breast, leading to diagnostic difficulty. In our case, the additional presence of myoid differentiation further contributed to the diagnostic challenge.

Myoid differentiation in DFSP is recognized as an uncommon histological variant. It is characterized by areas showing spindle cells with more eosinophilic cytoplasm and perivascular accentuation, resembling smooth muscle or myofibroblastic tumors. This variant becomes significant in the breast, where the differential diagnosis includes myofibroblastoma, leiomyoma/leiomyosarcoma, fibromatosis, SFT, and low-grade sarcomas. Hence, careful histopathological evaluation combined with immunohistochemistry is essential.

In usual DFSP, myoid areas intermingle with the surrounding areas and involve <5% of the total tumor. In the fibrosarcomatous variant, the myoid area is seen as confluent nodules in 10% of the tumor area [5]. Myoid areas usually show positive staining for smooth muscle markers. Staining for CD34, in those areas, is weak or negative. However, our present case was associated with extensive myoid differentiation. Myoid differentiation is said to be due to vascular smooth muscle hyperplasia/proliferation of pericytes [3].

In most cases, DFSP shows diffuse and strong CD34 positivity, which remains the key diagnostic marker [6]. Although SMA was negative in this case, h-caldesmon was positive. Myoid areas may show focal positivity for SMA. Demonstration of *COL1A1-PDGFB* fusion gene, when available, provides additional confirmation, though it is not routinely done in many centers in India [7,8].

The standard treatment for DFSP remains complete surgical excision with adequate margins. Incomplete excision is associated with a high risk of recurrence. Mohs micrographic surgery is preferred where available [9]. Targeted therapy with imatinib may be reserved for recurrent, unresectable, or metastatic DFSP harboring the fusion gene. The overall prognosis remains good with adequate local control.

This case is presented due to the rarity of DFSP involving the breast and the additional uncommon feature of myoid differentiation. Awareness of this variant is important to avoid misdiagnosis and to ensure appropriate management. Early recognition helps to prevent inadequate excision and reduces recurrence rates.

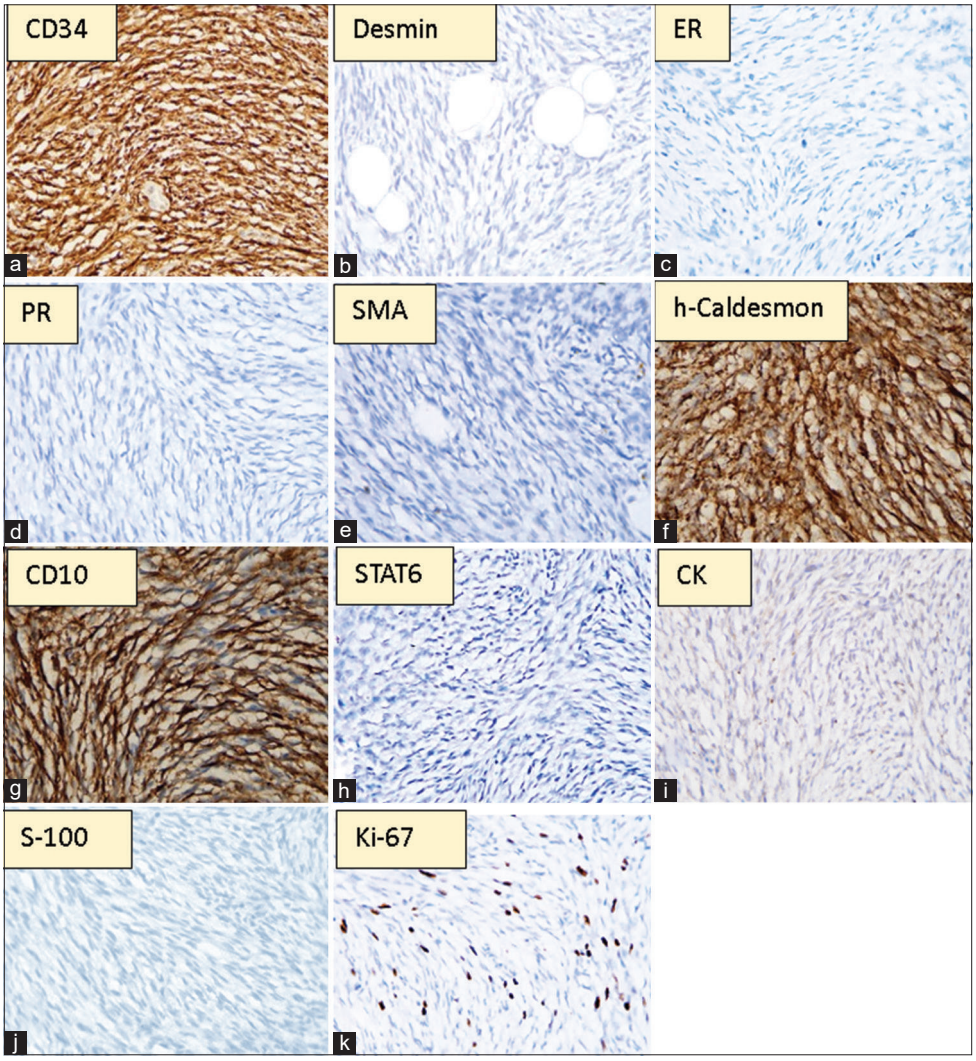

Figure 4: (a) CD34 is strongly positive in neoplastic spindle cells; (b-e) desmin, estrogen receptor, progesterone receptor and smooth muscle actin are negative in the neoplastic spindle cells; (f and g) h-caldesmon and CD10 are positive in the neoplastic spindle cells; (h-j) STAT-6, CK, and S-100 are negative in the neoplastic spindle cells; and (k) ki-67 proliferation index is 20%

CONCLUSION

This case highlights the importance of recognizing myoid differentiation in DFSP of the breast, an uncommon variant that may pose diagnostic challenges. Accurate histopathological evaluation and immunohistochemistry remain essential for diagnosis, ensuring appropriate surgical management and long-term follow-up.

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