Case Report

Dermatofibrosarcoma protuberans of breast - A case report

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Received - 29 January 2018 Initial Review - 25 February 2018 Published Online - 12 April 2018

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is best regarded as well differentiated, primary fibrosarcoma of skin. It is an uncommon, slow growing, locally aggressive, and reoccurring but rarely metastasizing tumor of the deep dermis and subcutaneous tissue. Clinically they present as firm solid nodules that arise most frequently on trunk and extremities. DFSP of the breast has been rarely reported. Here, we report a rare case of DFSP of the breast in 47-year-old woman.

Key words: *Breast, CD34, Dermatofibrosarcoma protuberans, Low grade*

Dermatofibrosarcoma protuberans (DFSP) is a mesenchymal neoplasm of the dermis and subcutis, which can present as nodular and polypoidal mass [1]. It is a rare soft-tissue neoplasm originally described in 1924 by Darier and Ferrand as a progressive, recurrent dermatofibroma and later named DFSP by Hoffmann in 1925 [2]. It constitutes approximately 6% of all soft tissue sarcoma [3] with an estimated incidence of 0.8–0.5 cases per million per year [4]. DFSP occurs most frequently in the 2nd to 4th decade of life and usually involves the trunk. Other sites involved are limbs, head, and neck. However, the involvement of the breast is rare, and to best of our knowledge, only a few case reports are found in the literature [5]. At least 40 case reports of DFSP in the breast has been reported [6]. DFSP rarely occurs in the breast, and it is difficult to diagnose. The differential diagnosis includes another mesenchymal tumor such as pseudoangiomatous stromal hyperplasia, myofibroblastoma, leiomyoma, and fibroepithelial malignancies such as phyllodes tumor and periductal stromal sarcomas [7]. DFSP typical storiform pattern and CD34 immunoreactivity were also against the diagnosis of a fibromatosis [8]. The histology of the tumor did not support an inflammatory myofibroblastic tumor because of the lack of a conspicuous inflammatory infiltrate and the negative smooth muscle markers and ALK1 immunoreactivity [9]. Both phyllodes tumor and periductal stromal hyperplasia/tumor are spindle cell neoplasms, but they show a biphasic pattern composed of spindle cells around tubular or ductal structures that were not present in the present tumor [8].

CASE REPORT

A 47-year-old woman presented to surgery outpatient department with chief complaints of lump left breast since 5 years. The lump was progressively increasing in size and caused mild discomfort to the patient. On physical examination, a 4×3 cm mobile, well-defined, firm, non-tender, lump was detected in the upper inner quadrant of the left breast. The breast sonography showed a 4.5×3 cm slightly heteroechoic nodule located in the dermal layer, suggesting the possibility of a dermal lesion. Fine needle aspiration cytology procedure was done which suggest the cytological features of the mesenchymal lesion. A left-sided modified radical mastectomy was planned, and mastectomy specimen was send to the pathology department for histopathological examination.

We received a mastectomy specimen with nipple and areola measuring 17×15×7 cm. Cut section showed a gray-white firm growth measuring 4×3×3 cm involving the overlying skin (Fig. 1). 11 lymph nodes were identified ranging in size from 0.5 to 1 cm. On histopathological sections examined showed a hypercellular tumor comprised the monotonous oval to spindle cells (resembling the fibroblast). These cells were arranged in the storiform pattern involving the entire dermis including the subcutis. Mitosis was rare. The overlying epidermis was thinned out. Deep extension from dermis into epidermis was also noted. No terminal ductular lobular units/mammary tissue was identified on the sections examined. These features suggest the possibility of Mesenchymal/Dermal/Fibroepithelial tumor (Fig. 2). All the lymph nodes showed reactive lymphoid hyperplasia.

For confirmation immunohistochemistry (IHC) was done and the tumor cells were strongly immunoreactive for CD34 (76–100% of cells) which confirms the diagnosis of DFSP (Fig. 3).

DISCUSSION

DFSP is a relatively rare cutaneous malignancy that arises from the dermis. The reported incidence of DFSP is approximately five cases per one million individuals per year [10]. While it is a low-grade sarcoma, it is capable of infiltration and local recurrence following inadequate excision.

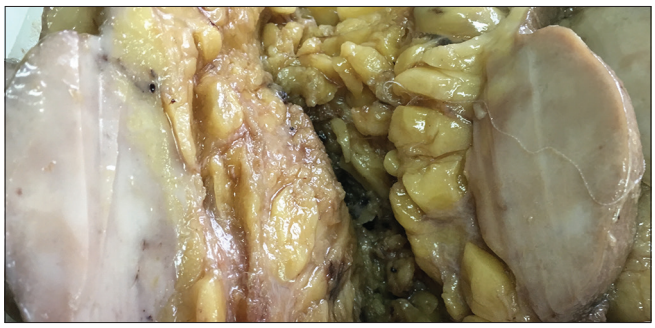

Figure 1: Gross examination of Tumor. Cut section is grey white

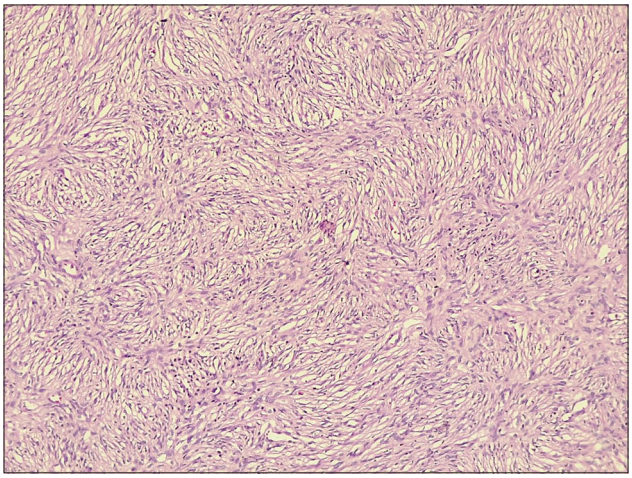

Figure 2: Microscopic examination showing the storiform pattern

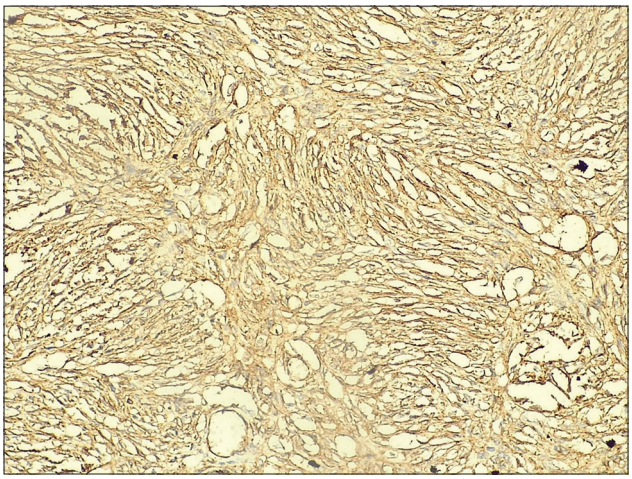

Figure 3: IHC-CD 34 positive which confirms it is dermatofibrosarcoma protubrens

The most commonly reported sites of DFSP are the trunk and extremities [11]. However, it is rarely reported in the breast [12]. The initial presentation of DFSP is a skin lesion described as a single, raised, red to bluish, firm cutaneous nodule, or plaque with surrounding discoloration [13]. The lesion is painless and indurated, but is extremely infiltrative and has a locally destructive growth that can invade the underlying structure such as fascia, muscles, or bones [14].

Large core needle biopsy or excisional biopsy can help to provide a correct diagnosis. The characteristic findings are interlacing spindle-shaped tumor cells in the dermis and subcutaneous fat layer that forms definite bands that interweave or radiate like spokes of a wheel, forming a so-called "cartwheel" or "storiform" pattern [15]. IHC markers are highly sensitive for DFSP. In particular, CD34 is a useful marker that allows differentiation of DFSP tumor cells from normal stromal cells and the other mesenchymal lesions [16].

The standard treatment for localized DFSP tumor is the wide local surgical resection. The recommendations for complete local surgical resection include surgical margins of 2–3 cm and three-dimensional resection including skin, subcutaneous tissue, and underlying fascia [17]. Distant metastasis is rarely observed, with a rate of 4–6%, predominantly to the lungs. DFSP can transform, especially in the recurring forms into fibrosarcomatous DFSP, a tumor with higher invasion and malignancy potent [18].

CONCLUSION

We highlighted a rare case of DFSP of the breast. It is a lowgrade sarcoma, capable of infiltration, and local recurrence. Wide local surgical excision remains the standard treatment for operable cases of DFSP. Due to high recurrence rate, long-term follow-up with clinical and radiological assessment is recommended for earlier detection of any sign of local recurrence.

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Funding: None; Conflict of Interest: None Stated.

How to cite this article: Kaur M, Tiwana KK, Nibhoria S, Singla N. Dermatofibrosarcoma protuberans of breast - A case report. Indian J Case Reports. 2018;4(2):118-120.