CASE REPORT

Dermatofibrosarcoma Protuberans of the Breast—a Rare Entity

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Abstract Dermatofibrosarcoma protuberans (DFSP) represents about 1% of soft-tissue sarcomas with an estimated incidence of 0.8 to 5.0 cases per million per year. This lesion may occur anywhere in the body but more than 50% occur on the trunk, 20% on the head and neck and 30% on the extremities. DFSP of the breast is an extremely uncommon site of presentation. Data regarding DFSP of the breast is limited and mostly in the form of case reports. Clinical presentation is not uniform and may mimic benign skin lesions [1]. However, it typically presents as a nodular cutaneous mass in early or mid-adult life. We herein report a case of DFSP of the breast in a 33-year-old lady who was managed successfully in our institute and review the literature associated with it.

Keywords Dermatofibrosarcoma protuberans · Lesion · Breast

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an extremely rare neoplasm of the breast [2, 3]. Its exact incidence in Indian population is not known perhaps due to its rarity [4, 5]. It is best considered as a low-grade sarcoma of the skin and subcutaneous tissue. It can recur locally but rarely metastasises. It often affects young and middle-aged adults. The pattern of growth is usually slow and persistent, and as the lesion enlarges over many years, it becomes protuberant. Large lesions often are associated with satellite nodules. Immunohistochemistry is an important tool for diagnosis [1, 6]. Most of them are CD4 positive. Surgical excision is the mainstay of treatment. Up to 50% of them recur after simple excision. However, with aggressive resection and careful attention to radial margins, recurrence rates can be reduced to 5% or less [7]. Follow-up at regular interval is essential to detect cases of local recurrence.

Case Report

A 33-year-old lady presented to us with the chief complaints of painful nodules over the breast for the last 2 years. The nodules were present in the areolar region of the left breast at one to six o'clock position. The nodules were static in size. There were no associated complaints of nipple discharge or lump in the same or in the contralateral breast. There were no complaints of pruritus of the nipple areolar region of the left breast. There were no complaints suggestive of metastatic symptoms. She gave a past history of incision in the site for undiagnosed infection of the skin. Her personal and family history was unremarkable. On physical examination, her general survey was essentially normal. Local examination of the left breast revealed presence of four nodular lesions over the left areola each about 1.8 cm in size. They were mobile and

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Fig. 1 The excised specimen

not fixed to underlying breast parenchyma. They were mildly tender to palpation and eczematous in nature. The skin of the nipple and areola was edematous and appeared thickened. There was no palpable lump in the left breast. No axillary nodes were palpable. The opposite breast was normal. Skin biopsy of the nodules was consistent with a diagnosis of dermatofibrosarcoma protuberans. All her routine biochemical parameters and chest X-ray were normal. Ultrasonography of the left breast showed four irregular hypoechoic nodular masses located beneath the skin. On mammography, four well-circumscribed mass were present in the nipple areolar region without any underlying calcification. She underwent wide local excision with 1-cm margin of normal tissue followed by primary closure under general anaesthesia. Gross examination of the pathological

specimen (Fig. 1) showed skin from the nipple and areola, the subepidermal region of which contained parts of an ill circumscribed tumour which is composed of short haphazardly arranged fascicles and sheets of spindle-shaped, fibroblast-like cells containing oval to elongated, bland nuclei, fine granular chromatin, inconspicuous nucleoli and moderate to abundant amount of eosinophilic cytoplasm. The tumour entraps dermal nerves and the erector piloris muscle and invades into subcutis. There is no necrosis. Definite mitotic activity was not seen. A grenz zone was seen between the tumour and the overlying epidermis. The adjacent breast tissue showed changes of lactation. Final pathology was consistent with DFSP. The maximum size of the tumour was 1.6 cm. The cells stained positive for CD34 and negative for ER, PR, Desmin, S100 and beta catenin (Figs. 2, 3, 4 and 5). The resection margins were negative. Postoperative recovery was fine. Patient is on regular follow-up at six monthly intervals without any signs of local recurrence or distant metastasis.

Discussion

DFSP is a rare soft-tissue neoplasm originally described in 1924 by Darier and Ferrand [8] as a progressive recurrent dermatofibroma and later named dermatofibrosarcoma protuberans by Hoffmann [9] in 1925. The aetiology of DFSP is not completely known but previous trauma has been

Fig. 2 Spindle cells positive for CD34 (in a), negative for S100, desmin and beta catenin. (Areolar smooth muscle positive for desmin in c)

Fig. 3 Photomicrograph shows spindle cells negative for ER (in e) and PR (in f)

implicated as a predisposing factor in the development of malignancy [10]. It is a locally aggressive tumour which may undergo malignant transformation to malignant fibrous histiocytoma [11]. The diagnosis of DFSP is clinically difficult because it can simulate a wide range of benign lesions. DFSP typically presents during early or middle adult life in all parts of the body, although more frequently on the trunk, the extremities and the head and neck. DFSP of the breast is extremely uncommon and a surgeon rarely encounters it during his clinical practice. DFSP has also been described in male breast. Several variants of DFSP have been described depending upon the cell types. Melanin-containing cells are called as Bednar's tumour and cells containing myxoid tissue are called myxoid DFSP. Juvenile DFSP occurring in children are called as giant fibroblastoma. Dermatofibrosarcoma protuberans may appear histologically similar to benign fibrous histiocytoma but grows in a more infiltrative pattern, spreading along connective tissue septa and often having unpredictable radial extensions of tumour permeating through the subcutaneous tissue large distances from the

primary nodule. The central portion of the tumour consists of a uniform population of plump fibroblasts arranged in a distinct ordered pattern. Other differential diagnoses include neurofibroma and liposarcoma. Increased age, a high mitotic index and increased cellularity are predictors of poor clinical outcome [12]. DFSP cells show immunoreactivity to CD 34, Factor xiii a and apo-d. However, CD 34 forms a better indicator. More than 75% of these tumours have a ring chromosome, composed of translocated portions of chromosomes 17 and 22. This fusion gene creates an apparent platelet-derived growth factor auto-crine loop, which is the chief reason for the tumour's exquisite sensitivity to imatinib. Clinically, the tumour may be diagnosed from its characteristic reddish brown to bluish nodular appearance. However, diagnosis may not be always clinically apparent. DFSP may be present as an intramammary mass without a hint. A wide array of diagnostic modalities has been described in literature to ascertain the diagnosis of DFSP in such situations. However, no consensus exists over a particular modality of choice. Use of FNAC to clinch diagnosis has been

Fig. 4 Photomicrograph showing a spindle cell lesion (right half) adjacent to breast acini (left half)

Fig. 5 Photomicrograph showing spindle cells infiltrating between fibroadipose tissues

controversial with some reports showing its use in suspecting the diagnosis. Core biopsy has been described as one time presurgical of diagnostic modality as it provides tissue for both pathological examination and immunohistochemistry. The use of skin biopsy may be a good alternative in cases where the lesion is superficial without any underlying palpable abnormality in the lump as was evident in our case. There was 100% concordance between preoperative biopsy and the final pathological specimen. Mammographically, it may appear as a soft-tissue subcutaneous well-defined mass with or without calcification. Intramammary DFSP may be picked up easily and can be taken when the mass is impalpable. The use of Doppler may show area of hypervascularisation in the tumour [13]. The use of MRI in detecting the depth of tumour infiltration and preoperative planning has also been described by some authors. Mammography subcutaneous variants of DFSP are easily evident on USG of breast [14]. Additionally, USG guided core cut biopsy. Finally, there is no consensus on the treatment of patients with DFSP. The ideal resection margin is still unknown. Although wide local excision has been the standard treatment of DFSP, recurrence rates range from 11 to 53%. Mohs micrographic surgery on the other hand allows the extent of excision to be tailored to the microscopic extent of tumour. It results in extremely low local recurrence rates and a cure rate of up to 98.5% [15]. However, such an expertise may not be always available leading to adoption of WLE as a primary mode of treatment. Complementary radiation therapy or chemotherapy seems not to bring any benefit [16]. Imatinib is approved for treatment of adult patients with unresectable, recurrent and metastatic DFSP who are not suitable for surgery. TKIs have also been used in a neoadjuvant setting. Long-term follow-up requires strict monitoring every 6 to 12 months with ultrasound and biopsy in cases of suspected recurrence. The 5-year survival rate of patients with DFSP is over 99% [17].

Conclusion

This case has been reported due to its rarity. A high index of suspicion is required for diagnosis. Though WLE is the present standard of treatment, the patients should be followed up at regular interval for recurrence. Also this report illustrates the use of skin biopsy in pinpointing diagnosis before surgery.

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