An Uncommon Malignancy of the Breast; Dermatofibrosarcoma Protruberans- A Case Report

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Introduction

Dermatofibrosarcoma protruberans (DFSP) is rare, morphologically distinct dermal/ subcutaneous neoplasm [1]. It typically presents during early or mid adult life. It is most common in the trunk and extremities and it accounts for approximately 0.1 % of all cutaneous malignancies and 6 % of all soft tissue sarcomas [2–4]. DFSP is highly aggressive, invades locally and rarely metastasizes [4]. Surgical scars, old burns, contractures and sites of vaccinations have all been reported as DFSP sites [5, 6]. DFSP occurring in the breast is very unusual. A review of literature reveals very few cases of DFSP breast. DFSP in the breast is usually characterized by smooth mobile raised reddish mass with brownish plaque like cutaneous thickening [6]. Treatment is primarily surgical, with chemotherapy and radiation therapy sometimes being used [6, 7].

Case Report

A 62-year old woman presented with progressively enlarging right breast lump since three months. Physical examination showed a firm, well defined, non tender mobile 6×6 cm mass in the upper outer quadrant of the right breast. Fine needle aspiration cytology showed scant material with few dispersed spindle cells, hence lumpectomy was performed. Grossly cut surface showed 6×5 cm pale white to tan well

defined homogenous mass with tiny cystic spaces (Fig. 1). Histology revealed cellular tumour with proliferation of spindle to ovoid cells arranged diffusely and in fascicles with numerous vascular channels and irregular extension into adipose tissue (Figs. 2, 3 and 4). Mitosis was 1–2/10 hpf. Based on histology, differential diagnosis of spindle cell sarcoma, myoepithelioma and Phyllodes tumor were given. The immunohistochemistry showed positivity for CD34 (Fig. 5), vimentin (Fig. 6), Ki-67 and negativity for S-100, bcl-2, pancytokeratin, smooth muscle actin, Factor VIII and calponin. The mass was eventually diagnosed as DFSP of the breast. All the resected margins were free from tumour. Complete evaluation revealed no residual tumour or evidence of metastasis. The patient was referred for local radiotherapy and followed up for 6 months with no evidence of recurrence noted (Fig. 7).

Discussion

Dermatofibrosarcoma protruberans was first described by Darier and Ferrand in 1924 and was referred to as a progressive and recurrent dermatofibroma [1, 5, 6]. DFSP represents about 1 % of soft tissue sarcomas with an estimated incidence of 0.8 to 5.0 cases per million per year [2, 4]. It has strong tendency towards local recurrence after excision [1, 5]. Most recurrences of DFSP are detected within 3 years of primary excision. Most cases of DFSP shows a specific translocation of chromosomes 17 and 22, which results in constitutive production of platelet-derived growth factor B chain and stimulation of DFSP growth. This chromosome translocation t(17, 22) is detected in more than 90 % of DFSP tumors [1, 2, 5]. Metastasis of DFSP is approximately 1–4 % [5]. Apart from female breast, rare case of male breast DFSP is also documented [8].

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Fig. 1 Lumpectomy specimen—grossly cut surface showing pale white to tan well defined homogenous mass with tiny cystic spaces

Ultrasonography and mammography are important tool for the differentiation of DFSP from a primary breast lesion [1, 6]. In most cases, mammography reveals a dense lesion without fat or calcification. Ultrasound exploration identifies the lesion in the dermis or subcutaneous tissue and the use of doppler shows hypervascularization of the area [3]. MRI may be helpful to define the depth of infiltration of the tumor [5]. In our case, mammography showed circumscribed, dense lesion with no evidence of calcification.

Histologically, DFSP is characterized by proliferation of plump spindle cells arranged in monotonous storiform pattern. The cells have little nuclear pleomorphism and low mitoses can be present. The main histologic differential is metaplastic carcinoma, fibromatosis, myoepithelioma and Phyllodes tumor [1, 6–8]. Panel of immunohistochemical markers are needed to differentiate between these lesions. Our case showed immunoreactivity for CD 34, vimentin, Ki-67 and negative for S-100, bcl-2, pancytokeratin, smooth

Fig. 2 Microscopy showing proliferation of spindle cells with entrapped fat (H&E, X100)

Fig. 3 Microscopy showing proliferation of spindle and ovoid cells arranged diffusely with blood vessels and irregular extension into adipose tissue (H & E, X 100)

muscle actin, Factor VIII and calponin. Negative for pancytokeratin and smooth muscle actin excluded metaplastic carcinoma and myoepithelioma. CD 34 immunoreactivity is against fibromatosis. Phyllodes tumor show biphasic pattern composed of spindle cells around ducts that was not present in this case.

The complete surgical excision with wide, pathologically negative margins of 3 cms is the optimal treatment for primary or recurrent DFSP [7, 8]. Since wide excision usually causes distortion and cosmetic problems, reconstructive flap should be performed. Despite optimal surgical treatment, local recurrence of 10–17 % was noted by some studies [8]. Moh's microscopic surgery has emerged as alternative approach allowing immediate microscopic examinations of the margins. However it needs considerable training, specialized team and multiple stages [6–8]. Adjuvant radiation therapy is needed if margins are positive or very close to the tumour after maximal resection or if lesion

Fig. 4 Spindly tumour cells entrapping the subcutaneous fat (H&E, X100)

Fig. 5 Microscopy with ovoid cells invading the fat and entrapped blood vessels (H&E, X100)

Fig. 7 Immunohistochemical stain for vimentin—tumor stained positive for vimentin (X100)

is large. Recently tyrosine kinase inhibitor Imatinib, was shown to induce regression of advanced DFSP when it is impossible to obtain surgical margins [6–8].

Although metastasis of DFSP is rare (only 1–4 % reported), almost all metastatic cases have been associated with local recurrence and poor prognosis. Most patients with metastatic DFSP die within two years. A small subset of DFSP patients present with fibrosarcomatous progression. This fibrosarcomatous progression DFSP variant is more aggressive in nature and the clinical outcome usually is poor [6, 7]. Histologic subtype, high mitotic index, cellularity, size, location on the head and neck, and recurrent lesions are factors reportedly associated with higher recurrence rates [5, 6].

Long-term follow-up requires strict monitoring every six to twelve months with ultrasound and biopsy in cases of suspected recurrence. The five-year survival rate of patients with DFSP is over 99 % [8].

Fig. 6 Immunohistochemical stain for CD34—diffuse, strong staining of the tumor cells (X400)

In conclusion, DFSP of the breast, though uncommon, it does exist. The correct diagnosis by routine histopathology is difficult; hence immunohistochemistry is needed for proper diagnosis and appropriate management. With awareness of this entity, the prompt diagnosis can be made and the disease can be properly managed.

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Conflict of interest nil

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