

Dermatofibrosarcoma Protuberans: A Rare Cause of Giant Breast Mass

Dermatofibrosarkoma Protuberans: Dev Meme Kitesinin Nadir Bir Sebebi

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor of dermis and subcutaneous tissue, mostly seen in trunk and extremities. Here we present a case of DFSP in the breast of a young women. A 26 years old female admitted to hospital with a 7 cm painless mass at right breast. Trucut biopsy reported spindle cell tumor. PET/CT and thorax CT revealed no metastases. Breast conserving surgery and sentinel lymph node biopsy was done. Pathological examination report was DFSP and reactive lymphadenopathy. Due to high recurrence rates in DFSP cases, wide local excision for acceptable oncologic margins and follow up is necessary.

Key Words: Dermatofibrosarcoma protuberans, spindle cell, breast tumor

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ÖZET

Dermatofibrosarkoma protuberans (DFSP) genellikle gövde ve extremitelerde görülen, dermis ve cilt altı dokulardan kaynaklanan nadir bir yumuşak doku tümörüdür. Burada genç bir kadının memesinde DFSP vakası sunulmaktadır. Sağ memede ağrısız 7 cm lik kitle ile 26 yaşında kadın hastanemize başvurdu. Trucut biyopsi sonucu iğsi hücreli tümör olarak raporlandı. PET/BT ve toraks BT' de metastaz saptanmadı. Meme koruyucu cerrahi ve sentinel lenf nodu biyopsisi yapıldı. Patoloji sonucu DFSP ve reaktif lenf nodları olarak bildirildi. DFSP vakalarında yüksek rekürens riski nedeni ile onkolojik olarak kabul edilir geniş sınırlarla rezeksiyon ve yakın takip gereklidir.

Anahtar Sözcükler: Dermatofibrosarkoma protuberans, iğsi hücre, meme tümörü

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), first reported by Darier and Ferrand in 1924 (1), is a rare soft tissue tumor of dermis and subcutaneous tissue. It is seen in all ages but particularly between the second and fifth decades. Although the lesion can be seen at any part of the body, mostly trunk and extremities are involved (2). Breast involvement is rare and here we present DFSP in the breast of a young woman.

CASE REPORT

A 26 years old woman admitted to our hospital with a 7 cm painless mass at her right breast just under the incision scar of a previous segmental mastectomy (patient mentioned) 6 years ago. The pathological evaluation of the biopsy material was benign according to the patient has mentioned. Breast USG, mammography and MRI (Figure 1) depicted a 7x6x5 cm well circumscribed mass at right breast and reactive lymphadenopathies at the right axilla.

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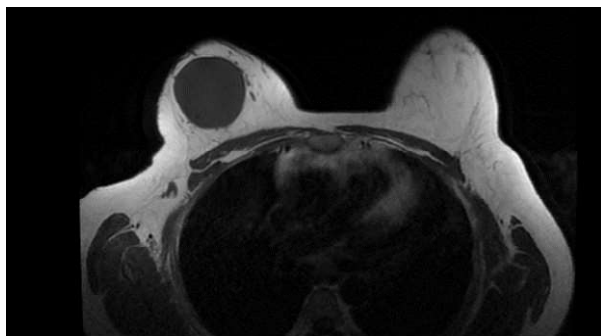


Figure 1. MRI view of 7x6x5 cm well circumscribed mass of right breast and reactive lymphadenopathies.

Trucut biopsy reported a spindle cell tumor. In FDG-PET/CT and thorax CT, there was no metastases. Therefore level 1 oncoplastic breast conserving surgery and sentinel lymph node biopsy were done. Intraoperative frozen section analysis reported acceptable surgical margins (2 cm) were achieved with no lymph node metastases. Final pathology report was DFSP with achieved acceptable surgical margins and reactive lymphadenopathies (Figure 2-3-4). No tumor recurrence was seen during sonographic follow-up over 20 months after surgery.

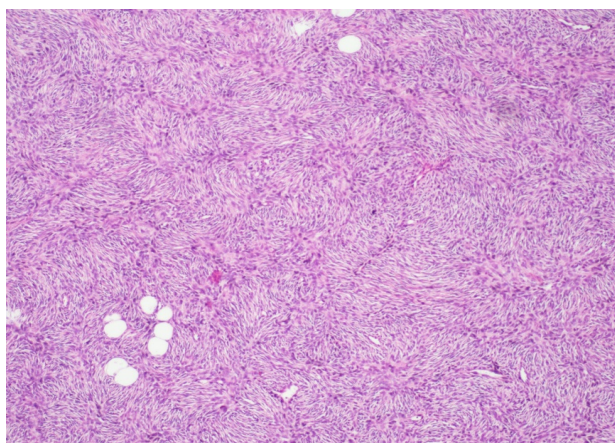


Figure 2. DFSP, adipose tissue in the storiform pattern of spindle cells.

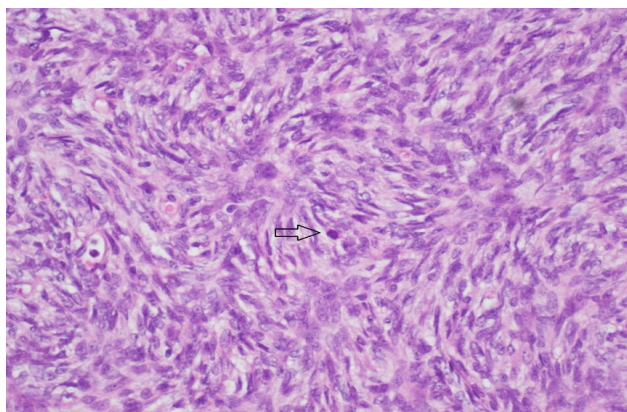


Figure 3. DFSP, storiform pattern of spindle cells with a few mitotic figures.

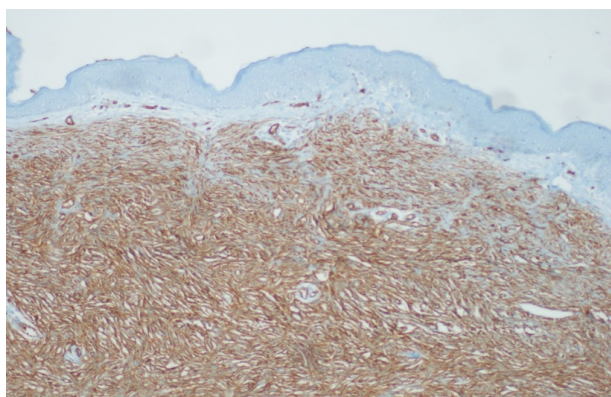


Figure 4. DFSP, diffuse and strong positivity with CD 34 staining.

DISCUSSION

Up to date few breast DFSP cases have been reported and differential diagnoses of these lesions are difficult (2). In the evaluation of breast lesions, trucut biopsy has one of the top priority. As in presented case, the spindle cell lesions of breast, fibroadenoma, cystosarcoma phylloides, hamartoma, adenomyoepithelioma, pseudoangiomatous stromal hyperplasia, metaplastic carcinoma, sarcomas should be kept in mind (3) and intraoperative frozen section assessment should be done for the achievement of accurate surgical margins and examination of lymph node involvement.

In histological examination of DFSP, spindle cells arranged in storiform pattern, honeycomb infiltration of lipomatous tissue can be seen. Mitose rates are low and in 84-100 % of lesions diffuse CD 34 staining are seen (4).

DFSP is a locally aggressive tumor which invades subcutaneous tissue and adjacent structures. Recurrence rate is high whereas metastasis is rare but if it occurs lungs and lymph nodes are usually affected (5). In our case, the patient had a history of segmental mastectomy 6 years ago because of a lesion at the same localization. Probably she had DFSP, and this is the local recurrence of the lesion.

Surgical treatment for DSFP is wide local excision and Mohs micrographic surgery. Different recurrence rates are reported for both procedures (2,6). Surgical excision with a margin of at least 2 to 3 cm is recommended for treatment because the local recurrence rate is 20% to 50% in cases of incomplete resection (7). Wide local excision with clear margins was performed for this case. For selected patients with DSFP in breast, who likely to default follow-up modified radical mastectomy can be performed. Ismail A. S. Burud et al. recommend mastectomy with axillary clearance for the patients who have the involvement of the resected margins, high mitotic figures, risk of recurrence, previous breast surgeries and strong family history of breast cancer (8).

In 90 % of cases, there is translocation between 17. and 22. chromosomes causing overproduction of platelet derived growth factor β (PDGF β) and stimulating DFSP cell growth (9). This started the use of imatinib, C-kit and PDGF-R inhibitor adjuvant and neoadjuvant therapies for DFSP (6). We vigorously recommended the patient medical oncology consultation, but she did not admit.

CONCLUSION

Breast involvement of DFSP is very rare however it should be kept in mind in the differential diagnosis. In this report DFSP in the breast of a young woman is presented. Due to high recurrence rates, wide local excision for acceptable oncologic margins and follow up is necessary. Keeping in mind modified radical mastectomy can be an option for selected patients.

Conflict of interest

No conflict of interest was declared by the authors.

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