Author's response to reviews

Title: Dermatofibrosarcoma of the breast: a case report

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare neoplasm of soft tissue described by Darier and Ferrand as “progressive recurrent dermatofibroma” in 1924 and by Hoffmann as “dermatofibrosarcoma protuberans” in 1925. This tumor is a dermal spindle cell tumor of intermediate malignancy characterized by a slow evolution and a greater risk of recurrence if excision is insufficient and a low rate metastatisation [1,2]. DFSP can occur in all parts of the body, but is found more frequently on the trunk, limbs, head and neck. Its location in the breast is extremely rare and very few cases are described in the literature. Confusion is possible with another primary breast lesion [3,4].

Case report

We present here the case of a swiss 75-year-old woman, who underwent twenty-one years ago a right mastectomy and axillary dissection followed by radiotherapy and breast reconstruction by prosthesis for invasive ductal carcinoma of the right breast, had a mass in her left breast. Mammography revealed a dish-shaped skin nodule formation in the upper outer quadrant of the left breast (Figures 1 and 2). Echography confirmed the presence of a lesion measuring 14 x 8 mm referring first as an angiosarcoma (Figures 3 and 4). MRI was not feasible due to a pacemaker for cardiac arrhythmia. No biopsy was taken because of anticoagulation by acenocoumarol for a pulmonary embolism 2 years ago. She has also been diagnosed for hypertension.

We proceed to a quadrantectomy of the left breast after having changed anticoagulation therapy (Figures 5 and 6). The postoperative recovery was rapidly favorable. Final pathological diagnosis revealed a Dermatofibrosarcoma protuberans not excised in toto with a margin at less than 0.1 cm. Tumor cells diffusely and strongly expressed the CD34 antigen, but were negative for CD31 and S-100 protein (Figures 7, 8 and 9). Then we decided to perform a wide surgical excision. This allows for additional safety margins of at least 5 cm, up to the pectoral muscle fascia including some muscle fibers. Sampling of the specimen revealed no residual tumor (confirmed by immunohistochemistry for CD34).

Discussion

Dermatofibrosarcoma protuberans represents less than 0.1% of all tumors, about 1% of soft tissue sarcomas and has an estimated incidence of 0.8-5.0 cases per million per year[5]. In most cases mammography reveals a dense lesion not containing fat or calcification. Ultrasound identifies the lesion in the dermis or subcutaneous tissue without visible connection to the skin and the use of Doppler shows hypervascularisation of the area[3]. Ultrasound-biopsy is essential to obtain a diagnosis before treatment and to facilitate excision with adequate safety margins. MRI is crucial to define the depth and infiltration of the tumor[6].

Pathologic examination reveals spindle-shaped cells arranged in storiform bundles, extending generally asymmetrically in the direction of the hypodermis. Immunohistochemistry is often positive for CD34 antigen but this marker is not specific. Tumor cells can also be positive for factor XIIIa [7]. This tumor has a very low mitotic power and is linked to a genetic anormality in 95% of cases (for exemple : translocation of chromosomes 17 and 22) [8,9]. The fusion of genes COL1A1 and PDGFB leads to the expression of a protein that interacts with the PDGF receptor and plays the role of an oncoprotein. Molecular detection of this complex has become a means of diagnosis [10].
Surgical excision is the treatment of choice. Safety margins should be of 5 cm of healthy tissue on the surface and should have anatomical borders not invaded at depth. The exact distance from the border of the tumor is not established. Risk of recurrence is not clearly identified; Complementary radiation therapy is not very effective[11]. Chemotherapy is not a treatment of choice, although specific inhibitor of tyrosine kinase (for example Imatinib, which inhibits the PDGFB receptor) appears promising [12]. Long-term follow-up requires strict monitoring every 6-12 months with ultrasound and biopsy in cases of suspected recurrence. The 5-year survival rate is over 99%[13,14].

**Conclusion**

Dermatofibrosarcoma protuberans of the breast is extremely uncommon and can mimic a primary breast tumor. Surgical excision with adequate resection margins in case of diagnosis of DFSP is recommended to ensure local control of disease. Otherwise new surgery has to be performed. A plastic surgeon should be present if a difficulty with the wound closure by first intention is to be expected.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Competing interest**

The authors declare that they have no competing interests.

**Authors’ contributions**

OC, JFD analyzed and interpreted the patient data. MF performed histological examination, JYM performed imaging and ultrasonography. OC was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

**References**

Reference List


