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Darier Ferrand Mammary Dermatofibrosarcoma Simulating a Breast-Type Myofibroblastoma: A Case Report

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Abstract

Myofibroblastoma and Darier Ferrand's dermatofibrosarcoma are rare entities that are similar both in terms of clinical morphological characteristics and histological characteristics. We report the case of a 49-year-old non-menopausal woman with a history of right breast lumpectomy. Supported by the Chompret criteria, an oncogenetic consultation was performed. Clinical examination revealed a firm 25 mm mass on the medial part of the left breast with skin involvement. A biopsy was performed and analysis result came back in favor of a cellular type myofibroblastoma showing a fibrous component consisting of spindle cells with a herringbone arrangement. Anatomopathological results concluded to a dermatofibrosarcoma of Darier Ferrand while immunohistochemistry stated a tumor population strongly positive for CD34 expression. The search for a rearrangement of the collagen type I alpha 1 gene (COL1A1) by fluorescence in situ hybridization (FISH) was positive. The diagnosis of Dermatofibrosarcoma of Darier Ferrand can be suspected on imaging and confirmed by histology. Surgical treatment of Darier Ferrand dermatofibrosarcoma consists of wide excision of the lesions with margins greater than 2 cm, on which the prognosis mainly depends. Micrographic surgery and oncoplastic breast surgery are of major interest in this location.

Keywords

Myofibroblastoma, Dermatofibrosarcoma of Darier Ferrand, Breast

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) or Darier and Ferrand tumor is an

uncommon mesenchymal skin tumor first described by Darier and Ferrand in 1924 [1]. It can occur all over the body, most commonly affecting the trunk and extremities [2]. It is characterized mainly by its local aggressive potential [2]. Breast localization remains rare. Its diagnosis is essentially histological [3]. The dermatofibrosarcoma of Darier and Ferrand is considered to be of intermediate malignancy, metastases are rare and are favored by multiple recurrences [4]. Because of the rarity of this case, the diagnosis can be misleading. We report the case of a dermatofibrosarcoma of Darier Ferrand with mammary localization simulating a mammary myofibroblastoma and we will discuss through a review of the literature, the diagnostic and therapeutic difficulties imposed by this entity.

2. Case Report

A 49-year-old primiparous female patient presented with a history of a right breast lumpectomy performed at the age of 35 years. The anatomopathological results concluded in a benign tumor. The family history revealed a history of osteosarcoma of the jaw in first-degree relatives, including her brother. She was referred to our senology unit for the management of an old tissue breast mass located in the medial region of the left breast. The clinical examination revealed a firm mass of 25 mm on the medial part of the left breast with skin involvement. The lymph nodes were free. The breast examination revealed a heterogeneous oval formation of 28 mm in the medial aspect. Magnetic resonance imaging (MRI) concluded that the clinical and imaging features could be consistent with a dermoid cyst (see **Figure 1**).

The left breast biopsy found fibro-adipose tissue in the samples collected; with a fibrous component consisting of spindle cells with a herringbone arrangement. The oblong nuclei were regular, without mitosis. Immunostaining revealed CD34 expression, without expression of desmin, smooth muscle actin, PS100, pan-cytokeratin, beta-catenin, or STAT6. The immunohistochemical and morphological appearance was in favor of a myofibroblastoma. Cytopunction of the left breast nodule was negative for neoplastic cells.

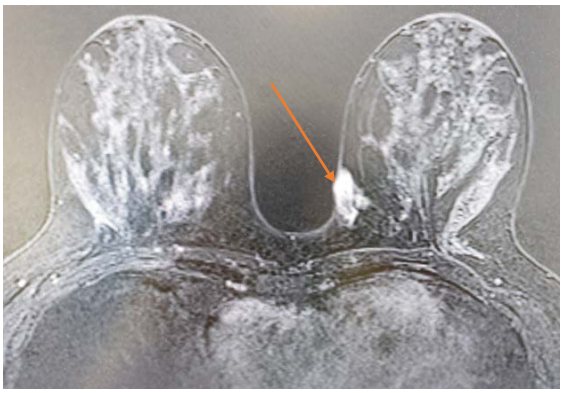

Figure 1. MRI of the patient's breast showing a mass opposite the internal quadrant of the left breast (oriented by an orange arrow). [Medical Imaging Department, RHENA Clinic STRASBOURG]

We performed a lumpectomy of the left breast. The anatomopathological study concluded to a Dermatofibrosarcoma of Darier Ferrand by showing a dermo-hypodermic spindle cell proliferation, made up of monomorphic spindle cells with little atypical nuclei arranged in short bundles or according to a storiform architecture, infiltrating the adipose tissue with a honeycomb aspect. Two mitoses were found in ten fields at high magnification. Immunohistochemistry showed a tumor population strongly positive for CD34 (transmembrane glycoprotein expressed by the hematopoietic precursor cell and capillary endothelial cells). A fluorescence in situ hybridization (FISH) test for collagen type I alpha 1 (COL1A1) rearrangement was positive, confirming the diagnosis. The tumor cells came in contact with all the margins (the excision margins were not healthy).

We performed a surgical resection of the bed with a wide excision in the medial-internal region, which resulted in satisfactory margins.

3. Discussion

Dermatofibrosarcoma of Darier Ferrand is a rare cutaneous sarcomatous tumor. Most cases occur in adulthood between 20 and 50 years of age [2] [4]. The most frequent localizations are the trunk and the extremities [4]. Mammary localization remains exceptional [5].

3.1. Histological and Molecular Aspects

3.1.1. Histological Aspects

The clinical signs are poor and consist of a localized mass associated with moderate pain and sometimes mild pruritus. Histological study provides diagnostic certainty. It is a tumor proliferation located in the dermis and hypodermis, composed of spindle-shaped cells with elongated nuclei with few atypia and mitoses (see **Figure 2**).

These cells are grouped in small flexuous bundles creating a "wheel spoke" or "woven basket" or "honeycomb" appearance [6]. Fibrosarcomatous transformation is reflected by the higher degree of cellularity, cytological atypia and mitotic activity (>5/10 HPF (High-power Fields) fields at high magnification [7] (see **Figure 3**).

Mammary-type myofibroblastoma is a rare benign connective tissue tumor with myofibroblastic differentiation. Clinically, most patients present with a solitary, well-circumscribed, slowly growing nodule in the breast, with lesions occurring most often in postmenopausal women [8]. According to Wargotz *et al.* [9], the average age at diagnosis is 63 years. Histologically, it is a tumor composed of a proliferation of spindle cells arranged in short irregular bundles dissociated by hyalinized collagen clusters.

3.1.2. Molecular Aspects

There are some cases of myofibroblastoma with high cellularity, atypical cells, and infiltrative margins [10]. Mitosis patterns are usually less than 2 mitoses per 10 fields at high magnification (see **Figure 4**). Spindle cells are negative

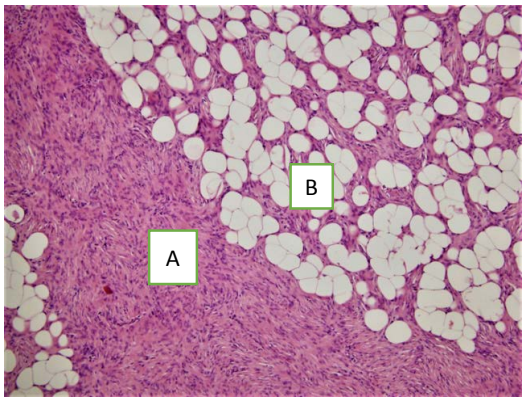

Figure 2. Haematoxylin-eosin stain, ×10 magnification: monomorphic fusocellular proliferation (A), of storiform architecture, infiltrating the adipose tissue (B), creating a "honeycomb" appearance. [Laboratory of Anatomopathology of the University Hospital of Strasbourg]

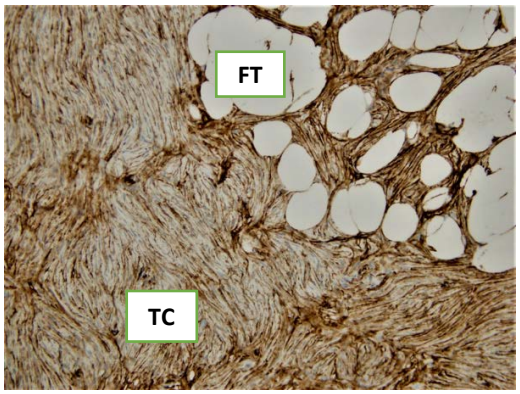

Figure 3. Immunohistochemical analysis performed with an anti-CD34 antibody of the histological section of a Darier Ferrand-type breast dermatofibrosarcoma. Intense labeling of the tumor cells is observed (brown coloration) Magnification ×10. TC: Tumor cells, FT: Fatty tissue. [Laboratory of Anatomopathology of the University Hospital of Strasbourg]

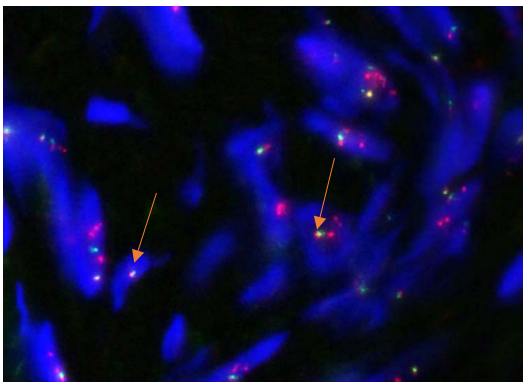

Figure 4. Fluorescent in situ hybridization (FISH) analysis of cell nuclei from adult dermatofibrosarcoma of Darier Ferrand. The red signals correspond to the PDGFB gene (22q13) and the green signals to the COL1A1 gene (17q22). Several fused signals (orange arrows) are observed per cell, corresponding to several copies of the COL1A1-PDGFB fusion gene. [Laboratory of Anatomopathology of the University Hospital of Strasbourg]

for cytokeratins, EMA and S100 protein [10]. However, the cells express CD34 as in Darier Ferrand's dermatofibrosarcoma, smooth muscle actin, desmin and CD10.

The positivity of the PS100 directs rather towards a nerve tumor [5] [6]. At the breast level, this entity can be confused with a wide range of mesenchymal neoplasms in particular Darier Ferrand type dermatofibrosarcoma of the breast. Breast-like myofibroblastoma has features that overlap with that of Darier and Ferrand's dermatofibrosarcoma. Regardless of histological appearance and anatomical location, myofibroblastoma has virtually no potential for recurrence or metastasis, even with positive excision margins [5].

The pathogenesis of Dermatofibrosarcoma of Darier and Ferrand tends rather towards genetic abnormalities present in 90% of cases with a true molecular signature offering a molecular diagnostic possibility [3]. An unbalanced chromosomal translocation between chromosomes 17 and 22 will lead to the fusion of the PDGFB (Platelet-Derived Growth factor group B) gene on chromosome 22 with the COL1A1 (Collagen type I alpha 1) factor on chromosome 17. Contrary to myofibroblastoma, it shares the same chromosomal rearrangements [11].

4. Treatment

4.1. Surgical Management

The treatment of these entities is essentially surgical. The reference for Darier Ferrand dermatofibrosarcoma is a wide excision of the lesions with margins greater than 2 cm [10] (see **Figure 5**).

Six months after the operation, the patient was in good clinical and radiological remission. We recommended continuing the close surveillance with senological assessment and clinical examination, prescription of a surveillance breast MRI, to be performed in six months. Recent guidelines from the National

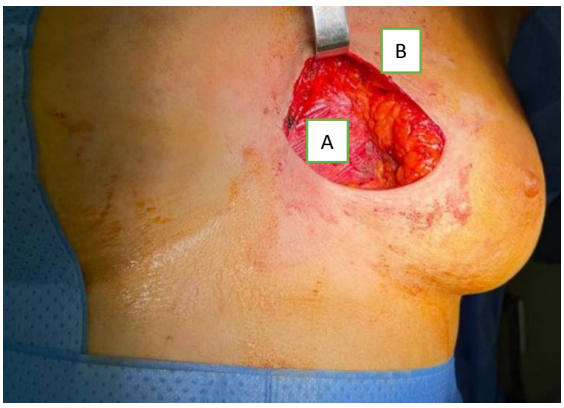

Figure 5. Surgical resection of the left medial tumor bed of a Darier Ferrand mammary-type Dermatofibrosarcoma (in whom the initial biopsy had concluded to be a mammary-type myofibroblastoma and in whom the first resection found a Darier Ferrand DFSP with non-healthy resection margins). (A) corresponds to the deep plane (pectoralis major muscle), (B) corresponds to the superficial plane. [ICANS Operating Room, breast surgery]

Comprehensive Cancer Network (NCCN) recommend margins greater than 2 cm [7]. Oncoplastic breast surgery would be the preferred type of surgery given the large area of tissue resected. Radiation therapy remains a therapeutic modality and is indicated in case of unhealthy margins after re-excision or in case of recurrence. Dermatofibrosarcoma protuberans is a radiosensitive disease with excellent local control after conservative surgery and radiotherapy [12].

4.2. Adjuvant Therapy

Adjuvant radiotherapy should be considered for patients with large or recurrent tumors, or when attempts at wide surgical margins would result in significant morbidity [12]. According to recent National Comprehensive Cancer Network guidelines for adjuvant radiation therapy for positive margins/macroscopic tumor, 50 - 60 Gy is recommended for indeterminate or positive margins, and up to 66 Gy for positive margin or macroscopic tumor (2 Gy fractions per day) and to extend the radiation field well beyond the surgical margin when clinically possible [7].

Chemotherapy is reserved for metastatic stages [13]. Immunotherapy is based on a tyrosine kinase inhibitor: imatinib mesylate, which essentially targets the PDGFB receptor, is indicated for unresectable tumors, recurrence and metastases [13].

The risk of metastasis is 15% - 20% [7]. Approximately 5% of these are primarily pulmonary [14]. The patient should be referred to a center with expertise in the management of soft tissue sarcoma.

5. Conclusion

Dermatofibrosarcoma of Darier Ferrand of the breast type is rare. The degree of morphologic overlap between myofibroblastoma and dermatofibrosarcoma of Darier Ferrand, in combination with shared genetics and a slightly overlapping anatomic distribution, raises the question of whether or not these tumors are truly distinct entities or rather represent a single spectrum of genetically related tumors. The diagnosis can be suspected on imaging and confirmed by histology. Treatment consists of wide surgery of the lesions with excision margins greater than 2 cm, on which the prognosis mainly depends. Breast oncoplastic surgery could be of major interest in this location.

Conflicts of Interest

The authors declare that they have no conflict of interest in relation to this article.

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