

Dermatofibrosarcoma Protuberans of the Breast: A Rare Localization

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Abstract

The dermatofibrosarcoma protuberans (DFP) is a rare skin tumor. It represents 0.1% of the malignant skin tumor. Surgery is its only treatment. The breast involvement is exceptional. We report our cases, 13 years old patient we have dermatofibrosarcoma protuberans in her breast.

Keywords

Breast, Dermatofibrosarcoma, Surgery

1. Introduction

Dermatofibrosarcoma protuberans is a dermal spindle cell tumor of intermediate malignancy characterized by a slow evolution with a major risk of recurrence in case of insufficient resection.

Described by Darier and Ferrand in 1924 under the name of progressive and recurrent dermatofibroma, it is called Darier and Ferrand's tumor by the French.

Of intermediate malignancy, this tumor is neither strictly malignant: it never metastasizes, nor strictly benign: it is characterized by its high propensity for recurrence in the event of insufficient excision. The breast localization is exceptional. Our aim is to report our experience in the management of this pathology and to discuss the clinical and paraclinical diagnostic challenges, therapeutic modalities and prognosis.

2. Case Report

We report the case of a 13-year-old, nulligravida, nullipara patient with a recurring tumor of the left breast that had been evolving for some years and had un-

dergone initial surgery 2 years earlier.

Clinical examination revealed a multiple budding tumor of the right breast without ulceration (**Figure 1**). The nipple was displaced to the right.

Initial biopsy revealed a conjunctival tumor.

The cancer staging workup revealed no signs of other localization.

The multidisciplinary team meeting agreed on resection with immediate reconstruction, followed by radiotherapy.

The patient underwent mastectomy (**Figure 1**) with reconstruction using a pedicled latissimus dorsi flap (**Figures 2-5**).

Anatomopathological study of the surgical specimen with immunological examination revealed a Dermatofibrosarcoma protuberans.

After wound healing, the patient is currently undergoing radiotherapy.



Figure 1. Bulging tumor of the right breast.



Figure 2. Mastectomy with 5 cm margins.



Figure 3. Coverage with latissimus dorsi flap.



Figure 4. Thoracic defect after pectoralis major resection.



Figure 5. Latissimus dorsi flap harvest.

3. Discussion

3.1. Epidemiological Aspects

Dermatofibrosarcoma protuberans represents only 0.1% of all skin tumors.

Sampling fluctuations on small series explain the variations in the sex ratio in the scientific literature. A third group claims equal frequency in both sexes [1] [2] [3].

The most common age range is between 20 and 50, with averages ranging from 28 to 47 depending on the author. The condition does not spare subjects of extreme age groups. DFSP is a mesenchymal tumor that is rare in children but not exceptional, although few series have been published in the literature. It is the most frequently observed skin sarcoma after Kaposi's. The frequency of pediatric cases has been estimated at between 6% and 20%, depending on the studies and age criteria applied [4]. The frequency of congenital cases varies from 3 to 10% in the scientific literature, but their prevalence is very low [5] [6] [7], with cases described at the age of 18 months or 82 years. Some authors consider this tumor more common in Indo-Europeans, while others report a particular predisposition of the black population [8]. Large series in multiracial populations have shown a predominance of Caucasians.

The relatively long progression of 5.3 years may be explained by diagnostic difficulties, repeated resections and recurrence, all of which motivates patient's consults.

3.2. Clinical Aspects

The onset of the condition is usually insidious, with no apparent cause or pre-existing dermatosis. Some authors mention various exogenous factors in the onset of the condition, such as parasitosis, angiomatous lesions [9], burn scars, vaccination scars, human bite scars [10], surgical scars, radiotherapy scars, traumatized nevi, syphilitic lesions, microtrauma on healthy skin, iatrogenic or occupational arsenical keratosis lesions.

From recurrence to recurrence, it can eventually develop into a full-blown sarcoma.

Its "recurrent" label is only linked to initial exereses that are too limited or insufficient.

The most common location is the anterior aspect of the trunk [11], specifically the periumbilical region different of our cases. Breast localizations are not frequently found. Lymph node metastases are only found exceptionally after a long evolution and sarcomatous transformation. No lymph node hypertrophy was observed in our study.

Histological study provides diagnostic certainty. It is a tumoral proliferation located in the dermis and hypodermis, consisting of spindle-shaped cells with elongated nuclei and little atypia or mitosis. These cells are grouped in small, flexuous bundles, entangled in nodular areas of high cell density, creating a storiform "spoke wheel" or "woven basket" appearance [12].

Differential diagnosis is essentially based on biopsies of benign histiocytofibromas, neurofibromas or dermatomyofibromas.

Immunohistochemistry facilitates diagnosis by showing the expression of CD34, a marker that is not very specific but very reliable when matched to the morphological appearance [13].

The tumor is positive for vimentin and inversely negative for HMB-45 and

S-100 proteins. PS-100 positivity points to a nerve tumor, while actin positivity points to a dermatomyofibroma.

3.3. Therapeutic Aspects

Surgery is the only proven therapeutic method for eradicating the tumors and preventing recurrence. Exeresis must comply with certain precise rules: monoblocexeresis, wide skin margins of 2.5 cm or more, extra-compartmental exeresis. Non-monobloc excisions run the risk of seeding the surgical site. Skin excision margins of less than 2 cm are associated with a high probability of recurrence. In our study involving large tumors, the average size of excision margins was 2.5 cm. This suggests that the recommended excision margins for large tumors should be revised upwards.

This conclusion is further confirmed by the findings of Parker [14], who showed that, in order to avoid recurrence, treatment of tumors less than 2 cm in diameter required margins of at least 1.5 cm, whereas treatment of tumors larger than 2 cm required margins of more than 2.5 cm. In our cases, we use large excision with 3 cm of margin

In Kassé's study [15], 5 cm margins on both sides were recommended.

Conventional reconstructive techniques, such as single-stage myocutaneous flaps, offer the possibility of covering vast defects after excision of advanced lesions. We use in our cases latissimusdorsi flap.

The autologous latissimusdorsi flap or latissimusdorsi flap without breast implant is the most recent technique, and the one that has benefited most from scientific advances in recent years. The surgical technique is well codified and allows the harvesting of the various fatty zones adjacent to the latissimusdorsi muscle, for autologous reconstruction. The flap is reliably vascularized via the thoracodorsal pedicle, which allows it to be used in a wide range of indications, including cases where TRAM and DIEP flaps are not indicated.

No adjuvant treatment has proven effective. Chemotherapy is reputed to be ineffective, with only methotrexate providing some objective responses. Radiotherapy, considered by some authors to be able to delay recurrence [16]-[19], is ineffective or even dangerous, according to Agiris [20], as it can lead, even at low doses, to sarcomatous transformation or even secondary cancers.

The use of other modes of irradiation has not yet been explored [21] (hyperthermia, radiosensitizers, heavy particles, high dose-rate brachytherapy, etc.).

4. Conclusion

Dermatofibrosarcoma protuberans is a malignant tumor of varying localization that is essentially managed surgically with wide excision margins to prevent any recurrence.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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