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(CASE REPORT)

Dermatofibrosarcoma protuberans: Case report

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Abstract

Dermatofibrosarcoma Protuberans (DFSP) is a fibrous skin tumor characterized by slow growth, a very high risk of local recurrence, but low metastatic potential.

In this article, we report on the case of a middle-aged patient at the Department of Plastic, Reconstructive and Aesthetic Surgery, CHU Tangier, presented with a dermatofibrosarcoma protuberans located at the anterior abdominal wall.

The patient was treated by surgical excision with a 3 cm lateral margin, including in-depth resection down to a healthy anatomical barrier. Reconstruction of the tissue defect was performed at the same time, using a large skin flap elevated upward onto the inframammary fold. After 9 months of follow-up, no recurrence has been noted, and the aesthetic and functional outcomes were considered satisfactory.

Darier and Ferrand's DFSP is a tumor whose prognosis and risk of progression are mainly linked to the diagnostic delay and the quality of the initial excision. A late diagnosis complicates both excision and reconstruction surgery.

Improving prognosis requires early and standardized multidisciplinary management, highlighting the importance of raising awareness and informing general practitioners for early diagnosis and appropriate referral of these patients to specialized centers.

Keywords: Dermatofibrosarcoma protuberans; Wide local excision; Recurrence; Reconstruction; Abdominal wall

1 Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare fibrous superficial cutaneous tumor.

Young to middle-aged adult patients are the most affected by this condition.

It is considered a tumor with a medium grade of malignancy, generally of good prognosis after complete resection, with a high risk of local recurrence, but it has a low metastatic potential. certain cases with sarcomatous transformation have been reported.

Earlier diagnosis may improve overall prognosis and outcomes.

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The management of these tumors requires multidisciplinary expertise.

While surgery remains the cornerstone of treatment in these tumors, imatinib may be an alternative in advanced stages or metastatic DFSP.

2 Patient and observation

54 years old female, with no notable medical history, presented with a mass of the anterior abdominal wall developing for 6 years prior to the first consultation.

Clinically, it is about a huge mass of 15 cm long axis, oval protuberant with no pain, easily bleeding at light touching and firmly fixed to the deep plane; no other physical signs were noted. A biopsy was performed by our team to determine the nature of the mass which came in favor of dermatofibrosarcoma protuberans.

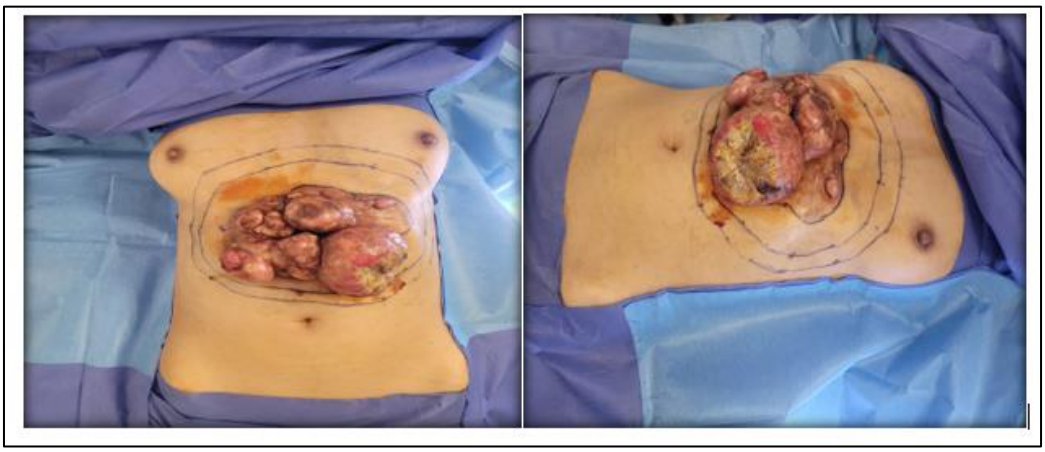

Figure 1 Pre-operative image of the tumor mass

In order to precise the degree of extension and boundaries of the tumor, a CT scan was required and This latter showed a cutaneous and subcutaneous tissue mass of the anterior abdominal wall, measuring 17*12*9 cm, oval, well limited, containing several confluent nodular lesions, without individualization of the central necrosis nor calcification, enhanced diffusely and homogeneously after contrast, without invasion of the anterior rectus sheet muscle nor adjacent costovertebral bone lysis or endothoracic or retroperitoneal extension.

The patient has undergone a wide excision with a safety margin of 3 cm laterally and in depth. The anterior rectus muscle sheet was taken away in a single block with the tumor mass and the cutaneous, subcutaneous plane.

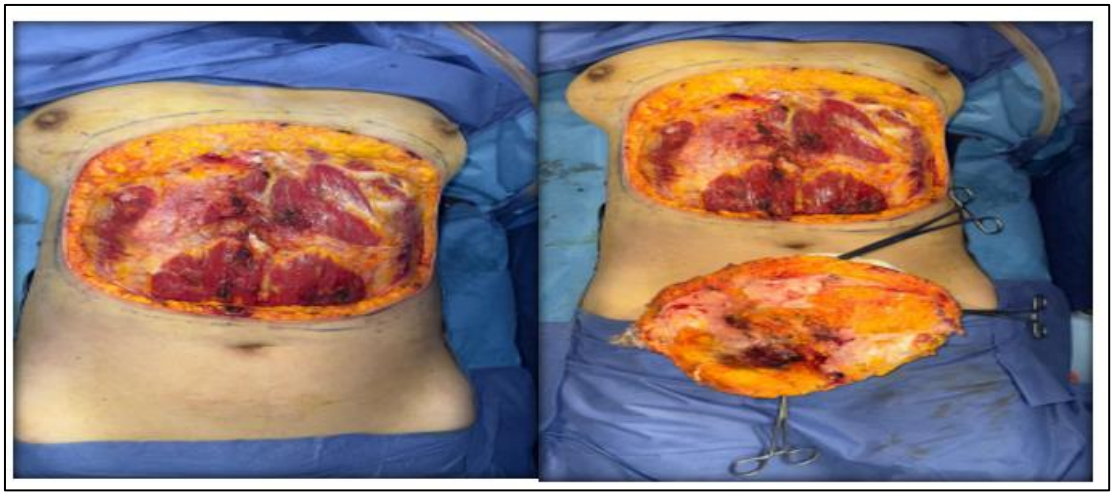

Figure 2 large resection of the tumor mass with a safety margin of 3 cm exposing the rectus abdominal muscle underneath

The reconstruction of the anterior abdominal wall was carried out on the same surgical procedure. It consisted of the mobilization of a large cutaneous flap upward to close the abdominal wall defect. No prosthetic material was fitted in to reinforce the muscle.

The histological study of the surgical specimen showed a mesenchymal proliferation in a cellular spindle of storiform architecture and dissociation of adipose lobules in depth suggestive of DFSP, and the surgical resected margin was disease-free.

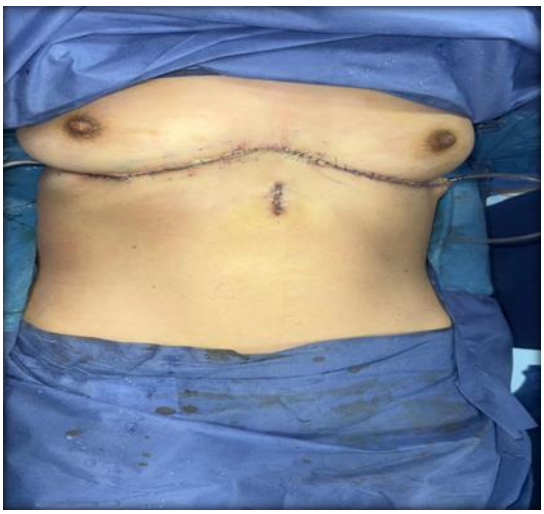

Figure 3 Closure of the abdominal wall defect with a large skin flap sutured to the inframammary fold

The patient was discharged after 10 days and addressed to receive a postoperative adjuvant treatment by a course of radiotherapy (25 sessions) on the recommendation of the multidisciplinary consultation meeting held prior to surgery. A clinical check-out at 3, 6 and 9 months showed no signs of local recurrence or sarcomatous transformation.

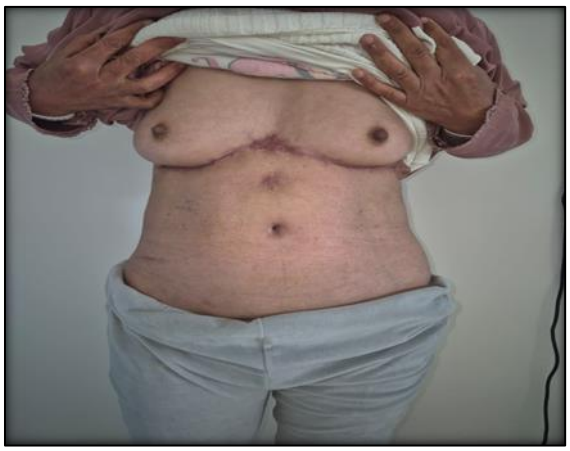

Figure 4 9 months check-up, no signs of local recurrence

3 Discussion

DEGOS [1] defines this tumor as: "It is a dermal connective tissue tumor composed of spindle-shaped cells, more or less similar in its histological structure to sarcomatous tumors, but it differs from true primary fibrosarcoma by its consistently cutaneous origin and by its very slow progression. It rarely, and at a very late stage, undergoes a clearly malignant, metastasizing sarcomatous transformation."

According to the literature, the tumor affects mainly young adults aged 20 to 50 years, some authors reported a slight male predominance [2,3], It affects equally both white and black subjects [4], on the contrary some studies have found that african patients with dark skin to be more concerned with this disease [2,5].

Unlike some findings of previous studies [6,7], in our case, the tumor occurred on healthy skin without pre-existing dermatosis, local trauma, burns or radiotherapy. Some cases in children and newborns were found suggesting heredity as a risk factor by some authors.

Our patient had consulted after 6 years from the first appearance of the tumor mass, this delay in diagnosis and therapeutic management can be explained by the slow progression of the disease, its painless nature, the absence of associated symptoms, and the preservation of the patient's general condition. In the literature, this delay is frequently reported and for the same reasons [8,9].

The data in the literature suggest that there is a preferential predilection for the trunk, which is affected in 60% of cases. The limbs account for 20 to 30% of tumor locations, and 15 to 20% are found on the head and neck [10,11,12].

In our case study, since the tumor size was extremely large, we performed partial biopsies, followed by histological examination which confirmed the diagnosis.

Histological analysis reveals a mesenchymal tumor proliferation originating in the dermis, with spindle-shaped cells of fibroblastic appearance arranged in a spoke-wheel pattern. The tumor strands infiltrate the dermis and separate the hypodermal fat lobules, following vascular pathways. Mitoses are variable, with rare, atypical forms. The stroma varies from one area to another. Collagen and reticulin fibers are more or less abundant, while elastic fibers are pushed to the periphery of the tumor. The absence of nuclear abnormalities and low mitotic index differentiates Darier and Ferrand's dermatofibrosarcoma from fibrosarcoma.

Wide local excision with a 3 to 5 cm safety margin remains the cornerstone of the treatment of DFSP; and in depth we tend to sacrifice a healthy anatomical barrier [13,14]. However, the response from patients remains variable. for some authors, Mohs micrographic surgery with incremental excision until normal tissue is obtained may be an alternative to avoid large excisions and complicate reconstruction [15].

A significant correlation between a wide excision and a low recurrence rate (9,13) has been clearly outlined by some studies [15]. Making the superiority and effectiveness of wide excision surgery as unequivocal.

No recurrence has been observed in our case so far after 9 months of follow-up. Meanwhile, recurrence can occur in 20% of cases (Jones et al., 2019) [16]. We hypothesize that this high rate of recurrence reported in many studies can be linked to some factors such as size, age, rapid progression, and especially the quality of initial excision.

The coverage of tissue loss resulting from the excision involves various methods, ranging from skin grafts to complex microsurgical techniques using free micro-anastomosed musculo-cutaneous flap transfers. In our case, we mobilized a large skin flap to cover the abdominal wall defect.

Due to its low mitotic activity, DFS is not radiosensitive [6,17]. Chemotherapy is not an effective method; however, there is some hope with Imatinib, and currently, several clinical studies on the neoadjuvant role of this molecule are underway [18].

There is no consensus regarding the rate of surveillance which must be focused on clinical examination and maintained over time due to the slow evolution and recurrent potential of this tumor. Monitoring can be done every 3 to 6 months for the first 5 years and then annually. Sometimes MRI will be used in a few selected cases [19].

4 Conclusion

DFSP fits between the benign pole of the cutaneous fibroma and the malignancy pole of the true cutaneous fibrosarcoma. Frequent delay in diagnosis is often observed due to its slow progression and its lack of awareness by most of the medical profession.

wide surgical excision with clear margins remains the cornerstone of effective treatment. Advances in reconstructive surgery enable optimal functional and aesthetic outcomes following large resections. While radiotherapy and conventional chemotherapy have limited roles, targeted therapies such as Imatinib have shown promise in selected cases, particularly for unresectable or recurrent tumors. Continued research and multidisciplinary collaboration are essential to improve long-term outcomes and minimize recurrence.

Compliance with ethical standards

Disclosure of conflict of interest

Authors declare no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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