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Dermatofibrosarcoma of Darrier and Ferrand: Recurrency Character: A Case-Report (Experience of the University Hospital Tangier-Tetouan-Al Hoceima)

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Abstract **Case Report**

Introduction: Dermatofibrosarcoma of Darier and Ferrand (DFSP) is a rare skin tumor characterized by its local aggressiveness and high potential. Its prognosis depends essentially on the quality of its management. ***Observation and Results***: In this article, we report the case of a young patient treated at the Department of Plastic, Reconstructive and Aesthetic Surgery, Tangier, for a Darier and Ferrand dermatofibrosarcoma that recurred five times after surgical excision, with a 5-cm margin on the surface and a healthy anatomical barrier at depth. Coverage of the loss of substance (LOS) was performed after anatomopathological confirmation of the carcinological nature of the excision, and essentially involved skin grafting. Despite a course of radiotherapy prescribed at the 4th recurrence, a 5th rapid recurrence was noted. ***Discussion***: The great clinical, histological and evolutionary polymorphism of this type of sarcoma results in an underestimated incidence and explains the delay in diagnosis. Recurrences are frequent, and usually local. Initial wide surgical excision is the reference treatment, and directly conditions the prognosis of DFSP. According to several studies, recurrence may depend on the quality of surgical excision, but could also be attributed to genetic or environmental factors, or to variations in the histological subtype of the sarcoma. ***Conclusion***: Darier Ferrand is an uncommon tumor with local malignancy, whose evolutionary risk depends mainly on the quality of the initial excision. The chances of cure in the case of well performed primary surgery are significantly higher than in the case of salvage surgery. Tumor excision must the refore be wide and deep, with sacrifice of a healthy barrier at depth.

Keywords: Demofibrosarcoma, Excision, Recurrence, Skin Graft.

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INTRODUCTION

- Darier and Ferrand dermatofibrosarcoma (DFS) is an uncommon fibrous skin tumor
- It is a tumor of local malignancy, with a very low metastatic potential but is characterized by an incredible tendency to recurrence.
- The treatment is essentially surgical and the therapeutic modalities are well defined: wide excision on the surface (5cm) and deep with sacrifice of a healthy anatomical barrier in depth
- Although it is considered a low grade malignancy, its designation as “recurrent” and therefore its prognosis is a matter of concern among clinicians.

PATIENT AND OBSERVATION

We report the case of a 40-year-old patient, without any notable pathological history, who consulted for a mass at the level of the left laterodorsal wall which recurred 4 times over the past few months, all these recurrences being spread over a period of 20 years ago, the first two excisions were without histological proof; the last one came back in favor of a dermofibrosarcoma of Darier and Ferrand.

On examination, we find an oblong protuberant mass of 8cm long axis, which is painful and fixed in relation to the deep plane, a peri-tumoral scar from three previous resections. The rest of the exam is unremarkable.

A first morphological exploration by ultrasound showed a poorly delimited mass of 8cm long axis, of iso-echoic, heterogeneous echo-structure, with color Doppler vascularization fitting closely with the sides, without intra-thoracic extension. To better specify this tumor and to study these relationships, additional exploration by magnetic resonance imaging (MRI) of the basi-thoracic soft tissues was requested. This MRI shows a cutaneous and subcutaneous tissue mass of the posterior basi-thoracic and left lumbar wall, 8.5 cm long axis, oval, very limited, containing several confluent nodular and pseudo-nodular lesions, without individualization of the central necrosis nor calcification, presents in discreet T1 hyposignal, T2 hypersignal, enhanced diffusely and homogeneously after contrast, with invasion of the left latissimus dorsi opposite without adjacent costovertebral bone lysis or endothoracic or retroperitoneal extension. (Figure 1).

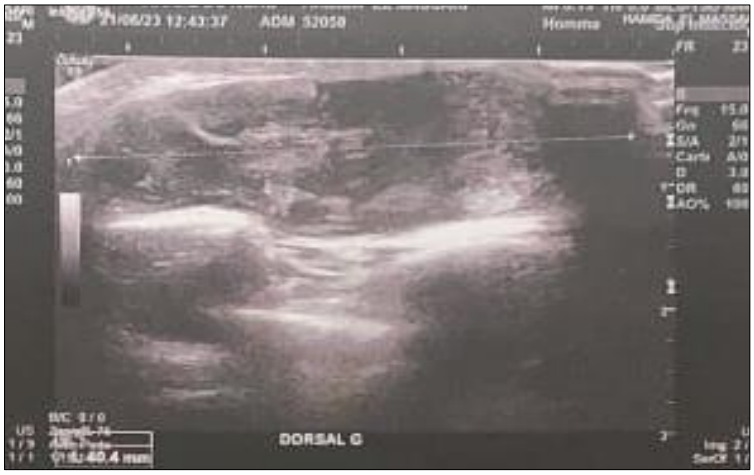

Figure 1: Sagittal view, hypointense on T1 and hyperintense on T2, enhancement of the mass without signs of intrathoracic extension on Sagittal T1 with fat saturation and gadolinium injection

The patient was operated on with a wide excision (WLE: Wide local excision) with a safety margin of 3 to 5cm laterally and in depth, removing in a single block the tumor mass comprising the cutaneous, subcutaneous plane.

The histological study of the surgical specimen showed a mesenchymal proliferation in a cellular spindle of storiform architecture (in the shape of a wheel) (Figure 2) and dissociation of adipose lobules in depth suggestive of DFS, with healthy microscopic resection margin. The file was presented to a multidisciplinary consultation meeting and postoperative adjuvant treatment was indicated by a course of radiotherapy (25 sessions). A clinical and ultrasound check at 9 months showed no signs of local recurrence.

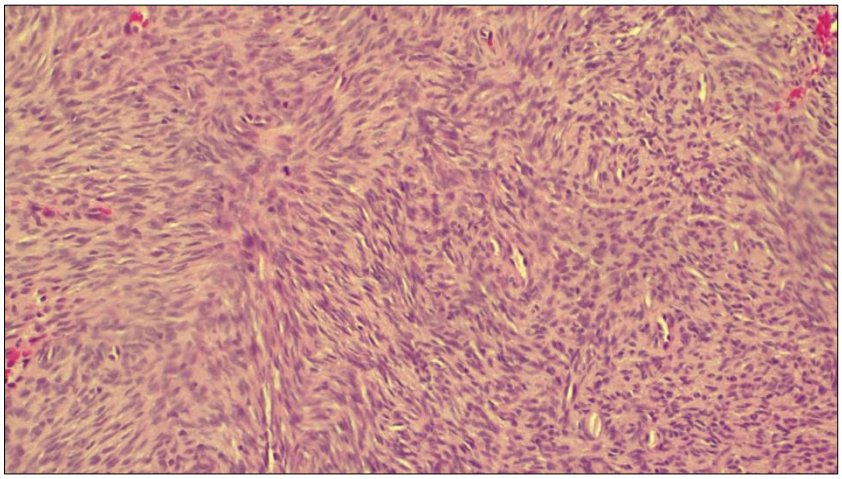

Figure 2: Mesenchymal proliferation in cell spindle of storiform architecture (wheel spoke), dissociation of adipose lobules in depth (pathognomonic)

A local recurrence 8 months after the radiotherapy treatment evident clinically by an isolated nodule with a major axis of 4 cm, and radiologically by an ultrasound of the soft tissues demonstrated a tumor recurrence by three pathological implants (figure 3). For which we carried out a wide excision with a 5cm margin and an anapathological study which confirmed that the lateral and deep excision limits were free from tumor

elements, the patient subsequently had coverage of her loss of substance after adequate preparation by a semi-thick skin graft. With closer follow-up consultation (figure 4).

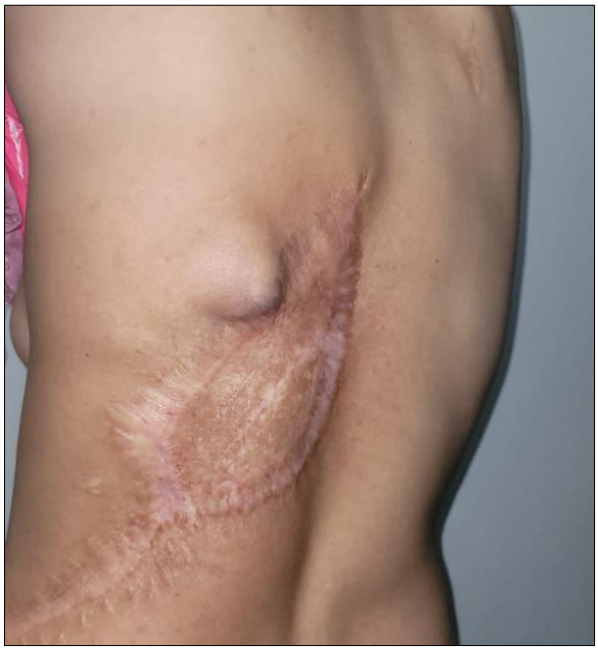

Figure 3: Recurrent nodular DFS (the fifth recurrence) after primary excision and coverage by skin graft.

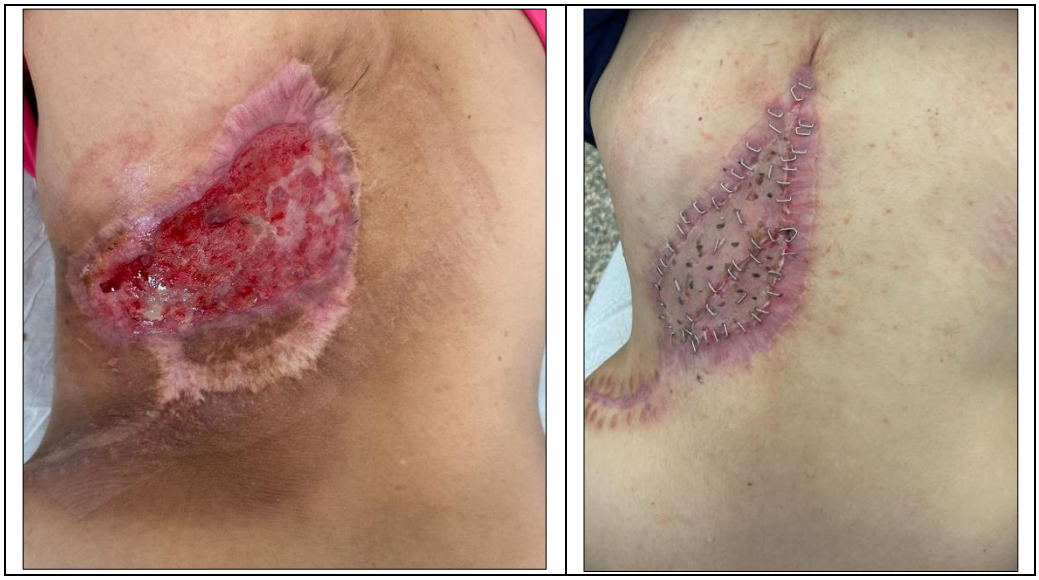

Figure 4: Reconstruction of the resulting loss of substance by a skin graft of semi-thick skin.

DISCUSSION

The strong point of this publication is that it reports a rare case of a skin tumor with a deceptive appearance. The cases reported in the literature are not numerous and the management does not follow a consensus. It is a low-grade cutaneous mesenchymal tumor with intradermal development, which mainly affects young adults aged 20 to 50 years, with a slight male predominance reported by some authors. It affects both white and black subjects [1, 2]. The etiopathogenesis remains a mystery. It occurs after local trauma in 10 to 20% of cases which could aggravate a pre-existing disorder and can also occur on healthy skin without pre-existing dermatosis or local trauma as in our case.

Some authors raised the theme of heredity, and reported cases in children and congenital cases which would be related to the COL 1A1 - PDGFβ fusion gene linked to the translocation of chromosomes 17 and 22 (t (17; 22) (q22; q13) [1-4].

DFS can affect the entire human body. The truncal seat is the most common in 50 to 60% of cases. The size varies according to the publications between 1

to 5cm, reaching 30cm in the Hammas series [5, 6]. This lesion is painful in 10 to 25% of cases, due to the mass effect when it increases in volume and the presence of hemorrhagic ulcerations. Most authors report the clinical evolution in two stages. Initially, at the infiltrative stage, the lesion appears as an indurated plaque, covered with normal skin, pink or purplish in color, well defined and mobile in relation to the deeper planes. More rarely, it presents as a firm nodule, an atrophic plaque or a sclerodermiform lesion. In the nodular stage, this initial lesion spreads and becomes irregular, producing after a few months to a few years, a multinodular mass, often polychrome, hard and mobile [5, 6]. The transformation into a malignant fibrosarcomatous form can be either de Novo or after a long progressive history of the disease.

The ultrasound mainly shows a hypoechoic mass richly vascularized on Doppler. The CT scan shows a solitary, well-defined, isodense, cutaneous or subcutaneous mass without calcifications and the MRI shows a well-defined mass, isointense with the muscle on T1, and hypointense on T2. Radiological extension assessment is only recommended for patients whose clinical examination raises suspicion of metastases, in cases of recurrent DFS and in cases of sarcomatous transformation [7].

The treatment of dermatofibrosarcoma is surgical, the superiority and effectiveness of surgery unequivocal. The difficulty of surgery lies in the subclinical extension of the tumor. The most widespread technique is that of wide excision (WLE: Wide local excision) with a safety margin of 3 to 5cm lateral and in depth with a sacrifice of a healthy anatomical barrier [1-9], without lymph node dissection [2-10].

Histologically, the tumor is considered a low-grade sarcoma of malignancy made of a dense cellular proliferation, poorly limited, not encapsulated, occupying the dermis most often in its entirety, and sending fine extensions sometimes very deep into the hypodermis., which explains the recurrences, the epidermis is respected and the cells are arranged in radiating bundles (Wheel Spoke appearance) very evocative [1-6].

For radiotherapy, DFS having low mitotic activity is not radiosensitive. However, it finds its place for recurrences, multiple locations, large tumors or inoperable primary, locations preventing wide excision and especially after an insufficient or invaded excision margin. Chemotherapy is not an effective method, however a certain protocol with imatinib (Glivec), pazorpenib or anthracycline can be used [1-3].

The recommendations of the National Cancer Institute can guide management according to the evolution of the tumor [1]. 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 [8].

The Recurrent Character

The patient in question presents a particularly refractory case of DFSP. The literature shows that although surgery with wide margins is the gold standard of treatment, recurrence can occur in 20% of cases (Jones *et al*., 2019) [11]. This patient's multiple recurrences raise concerns that some strains of DFSP may be resistant to standard treatment protocols.

Other studies, such as that of Martin *et al*., (2020) [12], suggest that aggression and propensity to recidivate could be linked to specific genetic mutations. These findings could explain the recurrent nature observed in this patient.

However in other research by Smith and colleagues (2020), it was noted that excision margins are crucial to avoid recurrence.

Age can also play a role. According to a study by Martin and Chen (2019)**,** older patients appear to have a slightly higher risk of recurrence, although the exact reason remains unclear.

Comparing with the literature, the recurrence observed in our patient is notably higher. This could be attributed to genetic, environmental factors or variations in the histological subtype of the sarcoma.

Furthermore, a study conducted by Park *et al*., (2021) [13], showed that some patients with DFSP could benefit from postsurgical targeted therapy to reduce the risk of recurrence. Given the number of recurrences the patient has experienced, such an approach could be considered for her future treatment.

The most commonly cited predictive factors for recurrence are: size, rapid progression and especially the quality of initial excision [2-6].

CONCLUSION

This is a case of a young patient with no previous history who presents with a rare skin tumor (DFS) which is located between the benign pole of the cutaneous fibroma and the malignancy pole of the true cutaneous fibrosarcoma. This tumor often presents a delay in diagnosis given its slow progression and its lack of awareness by most of the medical profession. Surgical excision, immediately wide and deep, is the treatment of choice. The prognosis is assessed based on recurrence factors. Regular and prolonged postoperative monitoring is necessary for the detection of recurrences and exceptional sarcomatous transformation.

This patient's case highlights the need for innovative therapeutic approaches for refractory DFSP. A better understanding of the underlying genetic mechanisms and the exploration of adjuvant treatments could offer solutions for patients facing frequent recurrences.

Abreviation

DFSP: Dermatofibrosarcoma of Darier and Ferrand.

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