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Review Article

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SURGICAL TREATMENT OF RECURRENT DERMATOFIBROSARCOMA OF DARRIER ET FERRAND. A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Darrier and Ferrand's dermatofibrosarcoma is a rare skin tumor with spindle cells and intradermal development, representing 0.1% of malignant skin tumors, good prognosis after complete resection, at very high risk of local recurrence but with low metastatic potential. It is distinguished by its diagnostic difficulty, its tendency to recurrence and the rarity of its metastases. it preferentially affects men. Enlarged tumor excision is the treatment of choice. This is a case report of dermatofibrosarcomas operated in the orthopedic-trauma service of the IBN SINA RABAT University Hospital.

KEYWORDS: Dermatofibrosarcoma protuberans, histology, skin tumor histologyrecurrence -Exeresis Large.

INTRODUCTION

Dermatofibrosarcoma (DFS) of Darier and Ferrand is a rare spindle cell skin tumor with intradermal development. Representing 0.1% of cutaneous malignancies, it has a good prognosis after complete resection, a very high risk of local recurrence but a low metastatic potential. First described by Jean Darier and Marcel Ferrand in 1924.[1]

The usual locations are the trunk, limbs, and cephalic extremity, and it usually presents as a firm reddish plaque or nodule.

The pathological diagnosis is primarily morphological. However, dermatofibrosarcoma may pose a differential diagnosis problem with other spindle cell tumors.

Given its relative resistance to chemotherapy and radiotherapy, its treatment remains exclusively surgical with the need for the largest possible excision simulating the requirements of carcinological surgery.

We report in this work the study of a patient, operated in the orthopedic-traumatology department of the CHU IBN SINA RABAT with a review of the literature with the aim of proposing a better therapeutic conduct in order to reduce the risk of recurrence and thus to improve the prognosis.

MATERIAL AND METHOD

We report the case of a 33-year-old patient with no notable antecedent who presented in an orthopedic trauma consultation following a nodular tumor of the right shoulder that appeared 12 years ago and for which he underwent two surgical interventions.

The physical examination revealed a mass of about 10cm by 4cm, mobile, with no inflammatory signs, the peripheral pulses were present, and there was no sensory-motor deficit, and the mobility of the shoulder was maintained. The rest of the examination was unremarkable.

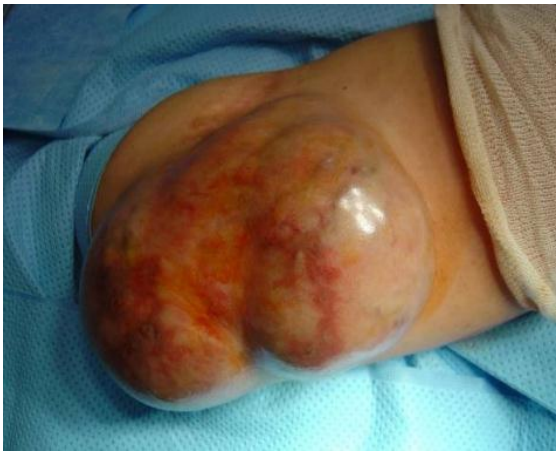

Figure 1: Multinodular mass of the right shoulder stump.

An MRI was indicated and showed a mass on the external aspect of the arm in hyposignal on the T1 sequence (Figure 3) and in hypersignal on the T1 sequence with fat saturation and injection of gadolinium (Figure 4). The mass is in intimate contact with the skin and the muscular aponeurosis.

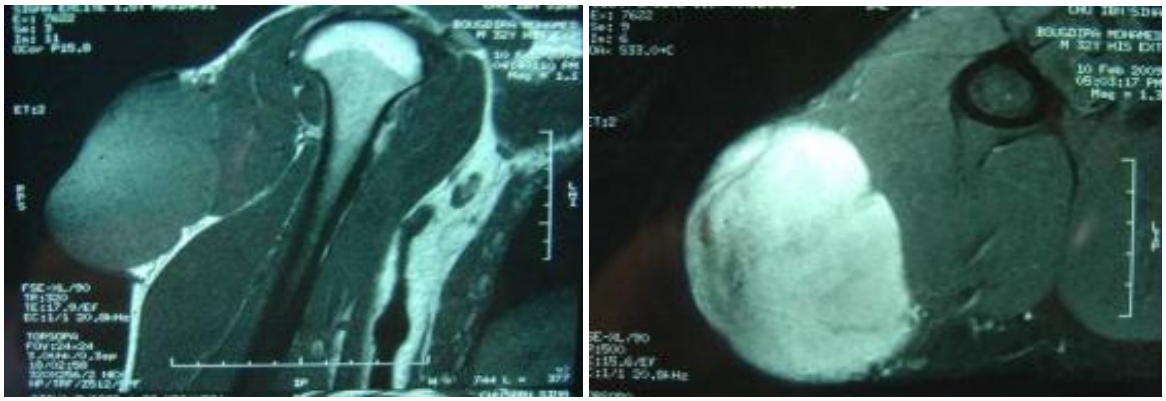

Figure 2: MRI of the right shoulder, T1 sequence with fat saturation and gadolinium injection.

The extension study did not reveal any metastases
A biopsy was performed and the anatomopathological study confirmed the diagnosis of dermatofibrosarcoma of darrier et ferrand

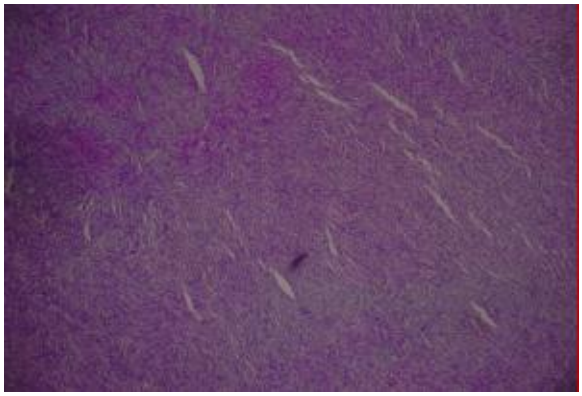

Figure 3: Histological section seen at high magnification: tumor proliferation made of spindle-shaped cells with elongated nuclei adopting a storiform structure.

The patient was operated on under GA in DLG with en bloc resection of the tumor (Figure 6) taking away the skin flap and the muscle aponeurosis adhering to the tumor up to the healthy tissues, then a skin closure proximal and distal leaving an oval loss of skin substance of 7 cm in diameter (Figure 7). Finally, a dressing in fatty tulle was made (directed healing).

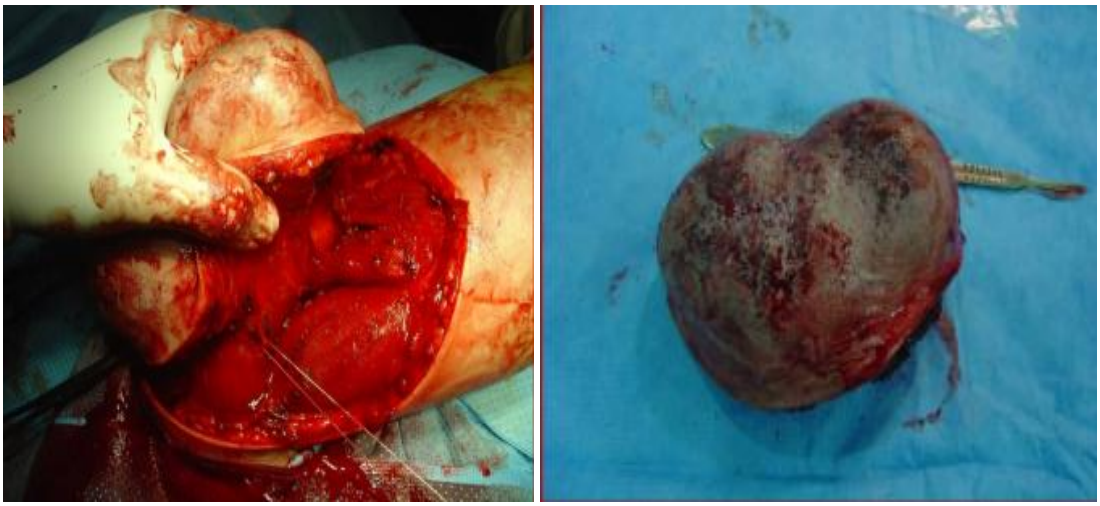

Figure 4: Large tumor removal.

The evolution was marked by: spontaneous healing of the loss of skin substance (Figure 10). The patient did not receive chemotherapy

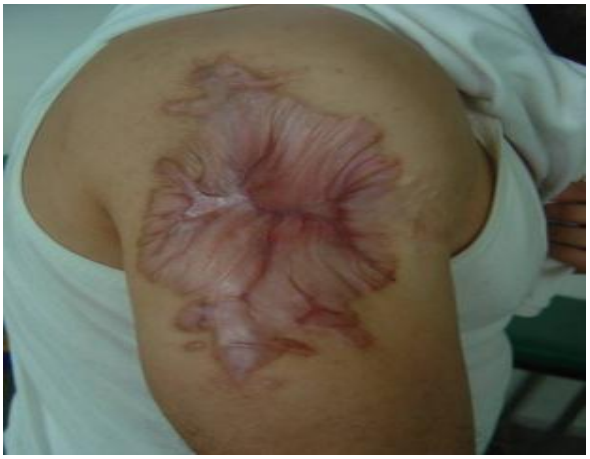

Figure 5: Evolution after healing.

With a 5-year follow-up, the evolution is favorable without recurrence and without any impact on the function of the shoulder.

DISCUSSION

Dermatofibrosarcoma, currently classified as a fibrohistiocytic tumor of intermediate malignancy according to the 2013 WHO classification.[2] constitutes an anatomoclinical entity defined by the association of a nodular skin tumor and a peculiar microscopic appearance. They represent less than 2% of all soft tissue sarcomas, with an estimated incidence of approximately four cases per one million.[3] The majority of authors mention this male predominance.[4,5] With an average age between 20 and 50years.

It is characterized by its slow progression, high local recurrence rate, and low metastatic potential. Frankly malignant sarcomatous transformation is exceptional.

The origin of DFSP remains controversial. Several origins can be evoked: fibroblastic, histiocytic, neural.[6]

DFSP can take on several clinical aspects, which sometimes poses a diagnostic problem, hence the interest of anatomopathological examination, possibly supplemented by immunohistochemistry.

The lesion may take on the appearance of a single plaque described as "nonprotruding" by Martin et al[7] as it may initially manifest as a firm, skin-embedded, well-delineated nodule that is mobile with respect to deep planes.[8,9,10]

The evolution of the tumor is slow: it takes a few months to a few years to result in a painless tumor formation suggestive, taking the appearance of a dermohypodermal plaque, dotted with nodules, polychromatic yellowish-white, brown, or pink, and sometimes telangiectatic, of variable size fixed to the skin opposite but perfectly mobile with respect to the deep planes.[11,9,12] General condition is noted with absence of adenopathy.

The size of the lesion can be extremely variable, usually depending on the time to consultation. It averages 1 to 5 cm ranging up to 40 cm.[9,12,13,14]

The time from lesion onset to the first request for care averages 7.9 years (3months-20years).

Standard radiographs, soft-tissue ultrasound, and CT or even MRI will tell us about the deep invasion of the tumor. The CT scan gives a better analysis of the bone structures and the appearance of the tumor is that of a soft tissue density mass without any particular specificity.[15] PDC injection shows heterogeneous tissue enhancement of the lesion, related to tumor vascularization.[16] MRI accurately determines the tumor process in the soft tissue. It provides information on the size of the tumor, dermal and hypodermal expansions, and deep extension in relation to the fascia[16] in order to guide the surgical procedure.

DFSP is a so-called "intermediate malignancy potential" tumor with a good prognosis after complete resection and a very high risk of local recurrence, but with a low metastatic potential. The preferred metastatic site is the lung.[17] Therefore, a chest X-ray should be requested and even a thoracic CT scan.[15]

Macroscopically, Dermatofibrosarcoma of Darier et Ferrand takes the appearance of a greyish-white, firm or elastic tissue nodule, infiltrating the dermis and hypodermis.[9,18,19] The skin opposite is taut, thinned, smooth, sometimes pigmented, and more rarely ulcerated, especially when the tumor mass is large.[9,31] Conventional surgery necessarily consists of wide and deep excisions in order to reduce the risk of recurrence, for which the margin of exeresis has evolved over the years.[20,21,36] The tumor may infiltrate the aponeurotic plane, but rarely muscle or bone, except when it is large or recurrent.[17]

It is a dermal proliferation organized in short intertwined bundles, creating a "storiform" appearance, very suggestive of dermatofibrosarcoma in the most cellular areas. In the less dense areas, the cells are arranged in parallel or form wavy, flexuous sheets, reminiscent of a neurofibroma.[9]

In doubtful cases, FSD can be distinguished histologically from other spindle cell tumors by immunohistochemistry. In DFSP, the tumor cells express CD34, most often diffusely.

In 1997, Arnaud, with 5 cm margins, observed a recurrence rate of 0% for primary treatment (tumors seen in first intention) and 4.6% for secondary treatment (tumors operated on at recurrence stage).[22] Thus, the efficacy of the primary treatment is of primary importance and remains the main prognostic factor.

Systematic lymph node dissection in the tumor's drainage area is not justified because dissemination is more often by hematogenous route than by lymphatic extension.[23]

Directed wound healing is a method of plastic surgery in its own right. The aim is to obtain epidermalization of the loss of substance after it has budded. It is therefore used for tumors of 3 to 5 cm in apparent diameter and in non-functional areas, i.e. far from the periorificial and articular areas.[24]

Skin graft: The graft is a fragment of skin taken from a donor site and deposited on a recipient site from which it will be revascularized. This technique is simple and allows for the best postoperative surveillance.[25] It is most often performed immediately after excision or at a later time. When a loss of skin substance cannot be sutured, cannot heal spontaneously or cannot be grafted, a flap is necessary.

Several authors report that radiotherapy is not an effective therapeutic modality for DFSP.[26,4] Others affirm the role of radiotherapy in local tumor control.[27,23] According to a recent study of 38 cases, local tumor control was achieved, with a 1 to 22 year follow-up, with adjuvant radiotherapy in 14 of 17 patients with insufficient or invaded excision margins (82%).[28] Thus, the combination of surgery and radiotherapy seems to be effective in preventing recurrence.

Systemic chemotherapy is not recommended for DFSP.[29,30] It is used as palliative therapy in association with radiotherapy.

CONCLUSION

Darier et Ferrand DFSP is a rare cutaneous tumor with a slow local evolution over several years. The diagnosis is often evoked clinically and confirmed histologically; immunohistochemistry is used in case of diagnostic doubt.

The treatment of DFSP is surgical and must meet a double objective: to perform a large exeresis from 3 to 5 cm from the edges depending on the location and on the primary or recurrent nature of the tumor, while striving to take a healthy plane in depth and to cover the loss of substance generated by the exeresis. Finally, DFSP has a good prognosis when the treatment is well carried out, but nevertheless requires lifelong clinical surveillance, as some cases of recurrence have been described very late in life.

Consent: The patient has given their informed consent for the case to be published.
Conflict of interests: None.

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