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Review Article

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DERMATOFIBROSARCOMA OF DARIER AND FERRAND: ABOUT 18 CASES AND REVIEW OF THE LITERATURE

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SUMMARY

Darier and Ferrand dermatofibrosarcoma (DFS) is a fibrous skin tumor of intermediate malignancy. It is a rare but not exceptional tumor, infiltrating the dermis and hypodermis representing 0.1% to 1% of malignant skin tumors. Characterized by a very polymorphic clinical appearance, slow growth, a very high risk of local recurrence, a low metastatic potential. The treatment is surgical and well codified (Chemotherapy: Imatinib). Its term "recurrent" is linked to insufficient excisions, hence the importance of well-conducted primary surgery and regular clinical and radiological monitoring.

We present a retrospective study of 18 cases of DFS diagnosed over a period of 4 years (2020 to 2024) (6 cases de novo - 12 cases of recurrence) and compare it to literature data. This study makes it possible to establish, in addition to the anatomopathological and immunohistochemical characteristics, an epidemiological, clinical and progressive study of this sarcoma. the average age of our patients is 42 years old with a male predominance. The trunk is the preferred location affected in 70% of cases. The tumor size reached 18 cm and measured on average 6.2 cm. The diagnosis was suggested by standard histological examination and confirmed by the intense and diffuse expression of CD34. The evolution was marked by the transformation into a high-grade pleomorphic sarcoma in a case operated initially outside our structure and by a case of local recurrence. Our results are classic and comparable to other series in the literature. Histological examination is essential for diagnosis. Wide surgical excision is the standard treatment. The prognosis is conditioned by mainly local malignancy and a high potential for recurrence. The frankly malignant sarcomatous transformation is exceptional.

KEYWORDS: Cutaneous tumor, dermatofibrosarcoma of Darier and Ferrand, histopathology, CD34, recurrence.

INTRODUCTION

Darier and Ferrand dermatofibrosarcoma (DFS) is a rare but not exceptional malignant cutaneous mesenchymal tumor, representing 0.1% to 1% of malignant skin tumors and less than 5% of soft tissue sarcomas in adults.[1–3] It was described by Jean Darier and Marcel Ferrand in 1924.[4] The preferred sites are the trunk, followed by the proximal extremities then the head and neck.[3] DFS often affects patients in their 3rd-4th decades with a slight male predominance and presents clinically as a firm reddish plaque or nodule.[5,6] Despite its distinct histological presentation, its histogenesis remains undefined.[3] It is a tumor said to have "intermediate malignancy potential", with a good prognosis after complete resection, slow growth, a very high risk of local recurrence, but a low metastatic potential.[1,2,6,7] Due to its rarity, very few epidemiological studies have been devoted to it. We present a retrospective study of 18 cases of Darier-Ferrand dermatofibrosarcoma. We study the different characteristics of our population and we compare our data with those in the literature.

METHODS

We carried out a retrospective study of 18 cases of histologically proven Darier Ferrand dermatofibrosarcoma, over a period of 4 years (2020 to 2024). We analyzed the epidemiological, clinical, paraclinical characteristics, the anatomopathological aspect, the therapeutic management and the follow-up of the patients. Data collection is carried out from patient clinical files and clinical information accompanying the

samples. The diagnosis of Darier Ferrand dermatofibrosarcoma was made by histological examination.

RESULTS

Of the 18 patients studied, we found 7 men (38%) and 11 women (62%), i.e. a sex ratio of 0.6 men to onewomen. The average age of the patients was 42 years with extremes of 17 and 65 years. The notion of previous trauma was found in one case, or 5% of cases. The diagnosis time was long. It varied between 7 months and 9 years with an average of 5 years. There was a recurrence in 12 cases: a first recurrence in 11 cases and a second recurrence in one case.

The trunk is the preferential location, affected in 13 cases (72%), followed by members in 3 cases (16%). The head and neck were affected in 2 cases (11%). Tumor size ranged from 0.6cm to 18cm with an average of 6.2cm. The tumor was nodular in 1 case, multinodular in 16 cases (Figure 3), and in the form of a plaque in 1 case. Ulceration was noted in 1 case. The lesion was painful in 4 cases (22%). The lymph node areas were free in all cases and the general condition was preserved in all cases.

The pathological examination confirmed the diagnosis of DFS by showing a dermo-hypodermal proliferation made of short crisscrossed bundles of storiform architecture, composed of uniform spindle cells that were slightly or moderately a typical.

The positivity of the CD34 immunostaining haspermitto confirm the diagnosis in the majority of cases.

As part of the locoregional and distant extension assessment, a chest X-ray, an abdominal ultrasound, a thoraco-abdominopelvic CT and an MRI of the soft tissues were performed. They showed tumor infiltration of the underlying muscles in two cases without distant metastases. Surgical treatment consisted of a 3 to 5 cm wide excision of the tumor with excision of the superficial aponeurosis. A case of para-umbilical DFS where the excision was 3 cm to preserve the umbilicus had benefited from Slow-Mohs micrographic surgery (Figure 1). A revision was necessary in 2 cases following tumor resection limits. During patient follow-up, we noted no case of local recurrence and, moreover, no case of lymph node or distant metastases or death.a case initially operated outside our structure, returned to the department after recurrence, the pathological study of which revealed a high-grade pleomorphic sarcoma but without metastases.

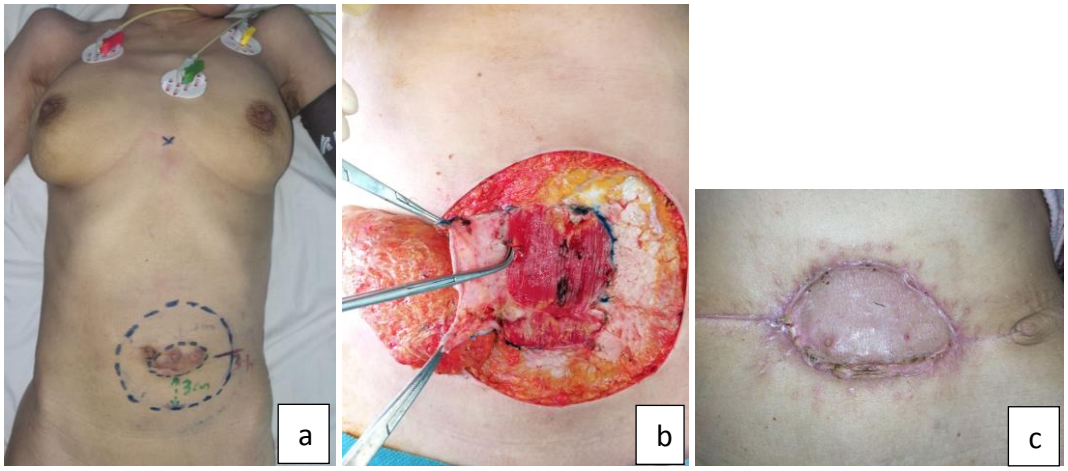

Figure 1: Case of Darrier ferrrand in the abdomen having benefited from Slow-Mohs microscopic surgery (a): preop (b): in perop. (c): on day 20 after coverage by skin graft.

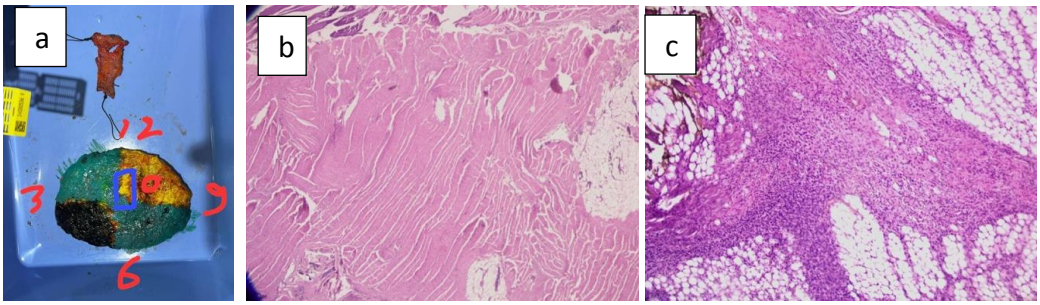

Figure 2: Histological image of the Darrier Ferrand in Figure 1 obtained after preparation and coloring of the part (Slow-Mohs microscopic surgery)
(a): coloring and orientation of the part in the anatomo-pathology laboratory
(b): histological image
(c): histological image

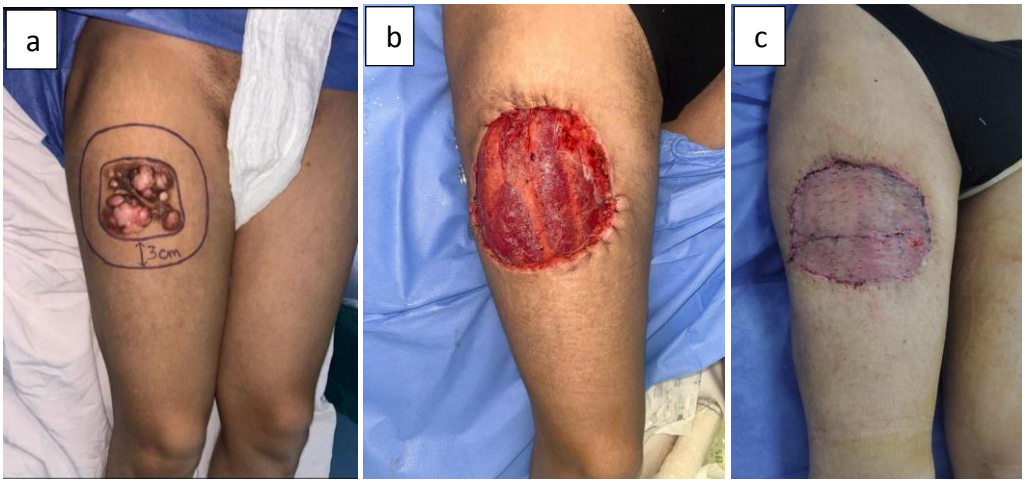

Figure 3: Dermato fibrosarcoma in the thigh (a), excision with anapath study (b), coverage by skin graft (c)

DISCUSSION

DFS is a mesenchymal dermal tumor of intermediate malignancy.[1] It is a rare but not exceptional tumor, representing between 0.1% and 1% of malignant skin tumors.[7] It was first described in 1890 by Taylor[8] as a sarcomatous tumor resembling a keloid scar, dermatofibrosarcoma was subsequently reported by Kuznitzky and Grabish in 1921, Darier and Ferrand in 1924.[4]

Our study makes it possible to establish, in addition to the anatomopathological and immunohistochemical characteristics, an epidemiological and clinical study of this sarcoma. Our data are classic and comparable to other series in the literature. The female predominance that we noted is not consistent with the data of the majority of authors.[2,7]

As described by several authors, this tumor can occur at any age with average ages at diagnosis ranging between 28 and 47 years.[7,10,11] DFS is rare in children under 15 years of age and the congenital form is exceptional.[12] In our series, the average age is 42 years and we did not find any congenital cases or localization in children, the youngest patient was 17 years old.

DFS can affect any part of the body. According to literature data, there is a predilection for the trunk which is affected in 50 to 60% of cases. The limbs represent 20 to 30% of locations and 15 to 20% are attributed to the head and neck.[9.11] In our series, the topography corresponds to the literature data with preferential involvement of the trunk in52% and members in 33% of cases. The location at the level of the head and neck is15%. We did not note any localization in the hands or feet.

Some authors have mentioned the occurrence of DFS after local trauma.[13] This notion is found in 10 to 20% of cases.[11,14,15] Taylor and Helwig, in a series including 115 cases, found a history of trauma in 16.5% of cases.[11] In our series, the occurrence of an initial trauma was noted in 2 patients or 7% of cases.

The diagnostic delay is comparable to that observed in other studies.[2,7,10,11] The time between the appearance of the lesion and the first request for care in our series is 5 years. This delay is explained by the slow evolution of the lesion and the absence of functional signs and general disorders.

Clinical diagnosis is difficult. In the infiltrative stage, the lesion appears as an indurated plaque. At a more advanced stage (nodular stage), the lesion spreads, creating a multinodular mass after a few months to a few years. This evolution in two stages is not constant because certain forms are immediately uninodular or multinodular. Untreated, these lesions can become very large, or ulcerate to become painful and hemorrhagic.[2,7] In our series, the lesion was most often multinodular.

The painful nature was found in4 patients (22% of cases).This rate is comparable to data from the literature where pain is noted in 10 to 25% of cases.[8] As in our series, the general condition of the patients remained preserved for a long time.[7]

According to publications, the tumor measures on average 1 to 5 cm.[7] Cases of "monstrous tumors" reaching 6.5 or even 7 kg have been described.[12] In our series, the average size is 6.2cm. The maximum size is18cm, greater than that of the Taylor series (12cm) and Bédix-Hansen (7cm).

Histological examination is essential for diagnosis. The tumor is made of a dense cellular proliferation, poorly limited, not encapsulated, occupying the dermis, most often in its entirety. It sends fine extensions sometimes very deep into the hypodermis, which would explain the occurrence of recurrences even with wide resection margins. The epidermis is respected. The cells are elongated, spindle-shaped, with more or less abundant cytoplasm, and with an oval, regular nucleus. Mitoses are variable with rare atypia. The stroma varies from one area to another.

Architecturally, the cells are arranged in radiating beams ("wheel spoke" or swirling appearance). Necrotic areas are rarely observed.[5,7,11] In our study, we noted the same histological aspects described in the literature. Necrosis was noted in only one case.

In general, the histological appearance helps guide the diagnosis. In doubtful cases, immunohistochemistry can distinguish DFS from other spindle cell tumors. It shows intense and diffuse positivity of CD34, focal positivity of AML (smooth muscle actin) and constant negativity of desmin and PS100.[7] Areas undergoing sarcomatous transformation only exceptionally and very weakly express CD34.[7] In our series, CD34 immunostaining was intense and diffuse. The negativity of immunostaining with other antibodies (AML, H-caldesmone, desmin, PS100 and CD68) made it possible to eliminate other differential diagnoses.

The differential diagnosis is made with diffuse neurofibroma, nodular fasciitis, malignant histiocytofibroma, myxoid liposarcoma and dermatofibroma. Malignant histiocytofibroma is characterized by marked pleomorphism, high mitotic activity and intratumoral necrosis.[7]

Treatment is difficult due to the subclinical extension of the tumor, which can cause recurrence. Wide surgical excision is therefore the standard treatment with safety margins of 4 to 5 cm and ablation of the superficial fascia.[5, 17] Postoperative radiotherapy is recommended by some, from the second recurrence.[17] Systemic chemotherapy is not recommended.[7] Rigorous clinical monitoring must be maintained, due to the slow progression and high recurrence potential of this tumor.[17] Treatment was surgical in our series and consisted of wide excision, with removal of the underlying aponeurosis or fascia. Chemotherapy was indicated in one patient: the case of the second recurrence.

The prognosis is conditioned by mainly local malignancy.[5] DFS almost never produces metastases and lymph node invasion occurs in less than 1% of cases.[7,10] Its high potential for recurrence, despite often large surgical excisions, transforms this lesion into an entity that is difficult to control clinically.[7] For many authors, the tendency for local recurrence is 20 to 40% of cases.[10] Death is exceptional and occurs late due to local complications.[7] In our series, one case of recurrence was noted during follow-up and, moreover, no case of metastasis or death.

The frankly malignant metastasizing sarcomatous transformation is exceptional and is seen at a very late stage.[7] in this study, a case, initially operated outside our structure, returned to the department after recurrence, the pathological study of which found a high-grade pleomorphic sarcoma but without metastases.

CONCLUSION

Between the harmless fibroma and the more agressif sarcoma, the dermatofibrosarcoma of Darier and Ferrand is a rare fibrous tumor of the skin which is distinguished by the need for a wide (5cm) and deep excision (sacrifice of a healthy barrier), a prognosis which depends on the first excision (respect of the margins or not), its diagnostic difficulty, its very slow local evolution, with a tendency to local recurrence with rare metastases. It requires clinical monitoring due to its high recurrence potential. Its diagnostic and therapeutic problems require a sure histological diagnosis confirmed by an immunohistochemical study.

Microscopic Mohs surgery (MMS) is associated with a low risk of recurrence and could reduce the amount of tissue resected.

The series studied presents clinical, histological and evolutionary similarities with the data in the literature.

Conflicts of interest

The authors declare no conflict of interest.

Author contributions

All authors contributed to the conduct of this research and read and approved the final version of the manuscript.

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