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The Evolutionary Profile of Dermatofibrosarcoma of Darier and Ferrand: Study of 23 Cases in Bamako

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Abstract **Original Research Article**

Introduction: Darier and Ferrant dermatofibrosarcoma is a rare malignant cutaneous mesenchymal tumour, representing less than 0.1% of malignant tumours and less than 5% of soft tissue sarcomas in adults. The aim of this study was to describe the evolutionary profile of cases of dermato fibrosarcoma of Darier et Ferrant reported in the dermato-venereology department of the National Centre for Disease Control (CNAM). ***Patients and methods:*** This was a descriptive cross-sectional study conducted in the dermatology department of the National Centre for Disease Control Support between 1991 and 2016, a period of 25 years. Was included, all patient with a histologically confirmed dermatofibrosarcoma notified in the registers of the CNAM histology laboratory or in the patient's medical file during the study period. ***Results:*** In total we collected 23 cases out of 5520 biopsied patients, i.e. a proportion of 0.41%. Males represented 60.8% with an average age of 40 years. The lesions were simple nodular in 12 of the 23 patients (52.2%), multi-nodular in 7 of the 23 patients (30.4%), nodulo-ulcerous and in patches in 4 of the 23 patients (17.4%). The lesions were located on the trunk in 60.8% of cases (image 1), on the head in 21.7% of cases and on the limbs in 17.5% of cases. The mean size of the lesions was 10×8cm with extremes of 3cm×3cm to 25cm×20cm. Management was exclusively surgical. ***Conclusion:*** DFSP is a rare tumour with a slow evolution, characterised by its rare metastasis but above all by its strong tendency to recurrence. The diagnosis is often evoked clinically and confirmed by histological study. The treatment of DFSP is surgical based on a large and deep lesion removal. Clinical monitoring allows early detection of recurrence, which is frequent in this disease.

Keywords: evolutionary profile, dermatofibroma of Darier and Ferrand, Bamako, HDB.

1. INTRODUCTION

Darier and Ferrant dermatofibrosarcoma is a rare malignant cutaneous mesenchymal tumour, representing less than 0.1% of malignant tumours and less than 5% of soft tissue sarcomas in adults [1, 2]. This tumour was described by Jean Darier and Marcel Ferrand in 1924 [3]. The condition occurs in the 3rd and 4th decades of life and is predominantly male. Clinically, the tumour presents as firm reddish plaques or nodules [4, 5]. The preferred sites are the trunk followed by the extremities and then the head and neck. It is a tumour of intermediate malignancy potential, its etiopathogenic mechanism remains undefined despite its distinct histological presentation, the prognosis is good after complete resection, because the metastatic potential is low however the risk of local recurrence is very high [2].

Nowadays, it is established that the diagnosis of this condition is histological, sometimes coupled with immunohistochemistry, and management is based on wide resection with minimum safety margins of 3 cm from the first operation as well as regular annual surveillance to detect recurrence, as incomplete resections are a source of recurrence and can be responsible for frankly malignant and metastatic sarcomatous transformation. In developing countries where there are few medical specialists, histopathology laboratories are rare and not easily accessible by the remote population, these conditions may be mistaken for commonplace conditions such as lipoma and other benign tumours.

The aim of this study was to describe the evolutionary profile of cases of dermato fibrosarcoma of Darier et Ferrant reported in the dermato-

venereology department of the National Centre for Disease Control (CNAM).

2. PATIENTS AND METHODS

2.1- Study Location

Our study was carried out in the dermatology department of the National Centre for Disease Control Support (CNAM), in commune IV of the district of Bamako (Mali). The CNAM was created by ordinance N°036 of 15 August 2001, ratified by law N°02-009 of 4 March 2002. It was born from the restriction of the Marchoux Institute. Its main missions are: continuing medical education (CME), operational vaccinological research, support for the disease control programme. Its field of action covers leprosy, dermato-venereology, sexually transmitted infections (STI, HIV/AIDS), tuberculosis, malaria and other endemic diseases.

2.2. Type and Period of Study

This was a descriptive cross-sectional study conducted in the dermatology department of the National Centre for Disease Control Support between 1991 and 2016, a period of 25 years.

2.3 Population

2.3.1. Study Population

The study population consisted of patients seen during this period and having undergone a histological examination and recorded in the register of the histopathology laboratory of the CNAM.

2.3.2. Case Definition

Any patient with a histologically confirmed dermatofibrosarcoma notified in the registers of the CNAM histology laboratory or in the patient's medical file during the study period was included.

2.3.3. Inclusion Criteria: Patients meeting the case definition.

2.3.4: Non-Inclusion Criterion: Patients not meeting the case definition.

2.4. Data Collection

Data were collected from histopathology laboratory records and/or patient medical records. We collected the information on a pre-established survey form containing the socio-demographic, clinical and histological characteristics of the patients and the evolutionary characteristics of the disease.

2.5. Analysis and Data Entry

We entered the data on epi data software and then transferred to Stat version 2013 for analysis. The statistical test used was Chi-square with an alpha risk of 0.05%.

The variables studied were epidemiological characteristics (age, sex), clinical characteristics (personal history, lesion appearance, lesion size, location, duration of evolution), histological result.

2.6. Ethical Considerations

The anonymity of the patients was guaranteed, the inclusion was safe for the patients.

3-RESULTS

In total we collected 23 cases of dermato fibrosarcoma of Darier et Ferrand out of a sample of 5520 patients biopsied for histopathological examination in the histopathology laboratory of the CNAM, i.e. a proportion of 0.41%. They were divided into 14 men and 9 women, i.e. a sex ratio of 1.7. The average age of the patients was 40 years with extremes (17 to 56 years). Clinically, 5 out of 23 patients were recurrent at the time of diagnosis: a first recurrence in three patients and a second recurrence in two patients, the average time to recurrence was two years (1 to 4 years).

The lesions were single nodular in 12 out of 23 patients (52.2%), multi nodular in 7 out of 23 patients (30.4%), nodulo-ulcerous and plaque-like in 4 out of 23 patients (17.4%). They were located on the trunk in 60.8% (image 1), the head in 21.7% and the limbs in 17.5%. The mean size of the lesions was 10×8cm with extremes of 3cm×3cm to 25cm×20cm. Management was exclusively surgical, with 20 out of 23 patients being managed at our facility and 03 patients being referred to other surgical departments. Monobloc excision with a margin of 3 to 5 cm was performed in 15 patients; excisional biopsy was performed in 6 patients, followed by surgical repair after histological confirmation (image 2). Directed wound healing was performed in patients operated on at the CNAM. The postoperative course was simple and healing was achieved in an average of 30 days (20-60 days).

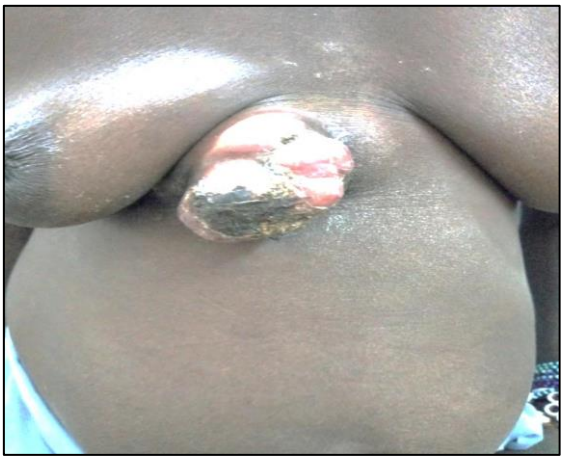

Image 1: Darier et Ferrand dermatofibrosarcoma of the epigastrium

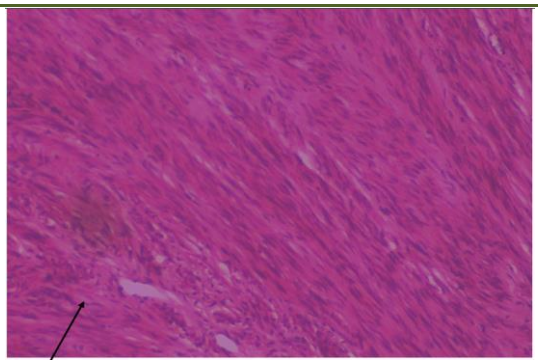

Image 2: Histological appearance of dermatofibrosarcoma of Darier and Ferrand

After the third year of evolution, all patients were lost to follow-up, only seven patients returned after recurrence and two died after a monstrous recurrence. These characteristics are summarised in Table 1.

Table 1

Age in years	Sex	year diagnosis	ATCD	Time limit recidivism	Clinical aspect	Size	Location	Deadline Consultation	Evolution Over 3 years
55	M	2002	NO	0	NODULE	3 ×2cm	Shoulder L	24	healing
55	F	1999	1 recurrence	2	PLATES	12×10cm	back	72	recurrence
31	M	2013	2 recurrences	1	NODULE	10×09cm	head	13	Death after recurrence
32	M	2014	2 recurrences	1	Nodule ++	7 ×5cm	Front	24	lost to view
54	F	2013	NO	0	NODULE	15×10cm	abdomen	120	recurrence
40	M	2002	NO	0	NODULE	06×04cm	3^{e} Finger	12	lost to view
24	M	2013	NO	0	PLATES	04×03cm	Cheek R	4	lost to view
42	F	1996	1 recurrence	4	NODULE	10×07cm	shoulder R	5	lost to view
33	F	1994	NO	0	NODULE	15×09cm	Back	6	Recurrence
20	F	2013	NO	0	Nodule ulcer	03×03cm	Cheek R	12	lost to view
22	M	2013	NO	0	nodule ++	08×06cm	Chest	7	lost to view
53	M	2010	NO	0	NODULES	7 ×5cm	Foot	5	lost to view
25	F	1992	NO	0	NODULE	18×12cm	Bas	6	recurrence
17	M	2015	NO	0	NODULES	9 ×7cm	Foot L	9	lost to view
45	F	1994	NO	0	NODULE	10 ×8cm	Back	24	lost to view
45	M	1992	NO	0	NODULE	6 ×4cm	Tête	11	lost to view
31	M	1998	NO	0	NODULE	8 ×7cm	COU	50	lost to view
55	M	2009	NO	0	NODULE	15×1Ocm	Epaule G	24	lost to view
56	M	2009	1 recurrence	1	NODULE	12 ×9cm	Flanc D	36	lost to view
40	M	2008	NO	0	NODULE	8 ×8cm	Dos	50	lost to view
50	F	1995	HTA	0	NODULES	5 ×7cm	Fesse G	36	lost to view
46	M	2008	NO	0	NODULES	16×14Cm	Cuisse	84	Recurrence
52	F	2015	NEPHRECT	0	MNOD	25×20cm	Ventre	96	Death after recurrence

4- DISCUSSION

The aim of this study was to describe the clinical and evolutionary epidemiological profile of patients with dermatofibrosarcoma in the dermato-venereology department of the national support centre for the fight against the disease over the period 1991 to 2016.

Dermatofibrosarcoma is a spindle cell dermal connective tissue tumour, more or less similar in histological structure to sarcomatous tumours but opposed to true primary fibrosarcomas by its cutaneous origin and by its very slow evolution. We collected 23 cases of Darier Ferrand dermatofibrosarcoma out of a sample of 5,520 patients biopsied for histopathological examination in the histopathology laboratory of the

CNAM, i.e. a proportion of 0.41%. In accordance with the literature, Darier Ferrand dermatofibrosarcoma represents between 0.1% and 1% of cutaneous malignant tumours.

The patients were generally young adults with an average age of 40 years and extremes (17 to 56 years). According to the literature, the age of predilection is from 20 to 50 years with averages ranging from 28 to 47 years. The condition does not spare subjects of extreme ages. Cases have been described at the age of 18 months and at 82 years [6].

We report a male predominance, already reported by other authors [7], while others [8] have found the opposite. A third group reported an equal sex ratio [6, 9]. According to the literature, this variation in the sex ratio is due to fluctuations in sampling over small series. The majority of patients consulted late (31 months on average), which is explained by the painless nature of the lesions.

Our lesions were located preferentially on the trunk, in particular on the back and chest, and rarely on the head and limbs. They were mostly uni-nodular (56.5%), rarely multi-nodular (26.1%) and exceptionally ulcerated or in patches (8.7%), and varied in size, reaching up to 25 cm in diameter with an average size of 10 cm. Clinically, our results are in line with the literature regarding tumour location, size, clinical appearance and delay in treatment [10]. The lesion initially presents as an indurated plaque covered with skin, normal in appearance and colour, sometimes whitish, yellowish white, pinkish, purplish or reddish. It is apparently well delineated and not very mobile in relation to the deeper planes. In an older stage, the plaque spreads, its surface becomes irregular and bumpy, and after a few months or years it becomes a multi-nodular mass, often polychrome, of variable size and hard consistency. This two-stage evolution is not always constant as some forms are initially uni-nodular or multi-nodular with secondary nodule fusion. In the majority of cases, the lesion evolves slowly and progressively, without functional signs or general disorders, which explains the long delays in consultation. In our study, the longest duration was 10 years, while other authors report a delay of 82 years [11]. The aesthetic concern explains the earlier consultation in women. Cases of monstrous tumours of up to 6.5 or 7 kg have been described [12]. The tumours can reach enormous dimensions of up to 25 cm in diameter. The lesion can develop in any part of the body [7]. Histological confirmation was done for all our patients, the appearance of the tumour is a dense, poorly limited, non-encapsulated cell proliferation occupying the dermis, most often in its entirety. It sends extensions into the hypodermis, without destroying the elements of the latter, while the epidermis is respected. The cells are elongated, spindle-shaped, with more or less abundant cytoplasm and regular oval nuclei. Mitoses are variable with rare atypia. The stroma is variable from one area to another. Collagenous and reticulinic fibres are more or less abundant, while elastic fibres are pushed to the periphery of the tumour. Within the clusters of neoplastic cells, a variable number of vascular spaces and peri-nervous cell streams can be distinguished. Over time, there is a progressive decrease in the connective tissue component and an increase in cell density. Architecturally, the cells are arranged in radiating bundles ("wheel spoke" appearance) or swirling bundles [13].

Surgery is the only treatment that has been proven to be effective in eradicating the tumour and preventing recurrence. In our series, three patients were referred to other surgical departments for management, so of the 23 cases, 20 patients were managed at the CNAM. This surgical management consisted of a wide exeresis with margins of 2 or 3 cm on healthy skin and deep down to the underlying aponeurosis in monobloc, some authors recommend [8] a widening of the exeresis margin to 5 cm on the surface with sacrifice of a healthy anatomical barrier in depth, countries with a high technical platform use the Mohs technique with an extemporaneous examination [9]. This involves first removing most of the tumour mass and then making freeze cuts on the underside of the surgical specimen, which allows horizontal tissue slides to be collected. After reading, the invaded areas are treated again in the same way until no tumour tissue is found on the sections. Teams using this technique [9] have shown that lateral excision margins of 3 cm to 2.5 cm are sufficient.

Cure was obtained after directed healing. After the histological result, a revision was carried out with a large exeresis and removal of the aponeurosis in case of incomplete exeresis on histology. The technique as well as the margin of excision in the other structures was not precise.

All patients were lost to follow-up after healing. Seven patients came to us with recurrence (30.4%), this frequency is comparable to the Dakar study (36%) [11] and very high compared to the Moroccan series (13.6%) [7]. Correlations have been reported by authors on the recurrence and the insufficiency of the excision margin, which was in our study 2 to 3 cm, 3.5 cm in the Dakar series [11] against 5 cm in the Moroccan series [8].

Among our recurrences, two patients died, the first one was a 52-year-old female patient, housewife from a rural area, the tumour measured 25×20 cm on the abdomen, a large exeresis was done, however the patient was not seen again at the follow-up examinations after several months. She was readmitted with a larger multi nodular ulcerated recurrence and an altered general condition. Death occurred during resuscitation.

The second patient was a 31-year-old farmer who was in his first recurrence at the time of diagnosis, the tumour was located in the temple. After a second recurrence, the patient would have refused to be consulted in the medical centre in favour of a traditional treatment whose evolution was marked by the extension of the tumour, ulceration, necrosis and death in a context of neurological disorder.

The patients were all lost to follow-up after the healing of the surgical wound, which can be explained by the lack of clear information of the patients on the pathology, on the need for long-term surveillance on the risk of recurrence, but also the ignorance of the patients to the tumours in general, with sometimes a false belief to be able to cure them by the traditional treatment.

5-CONCLUSION

DFSP is a rare tumour with a slow evolution, characterised by its rare metastasis but above all by its strong tendency to recurrence. The diagnosis is often evoked clinically and confirmed by histological study. The treatment of DFSP is surgical based on a large and deep lesion removal. Clinical monitoring allows early detection of recurrence, which is frequent in this disease.

Current state of knowledge on the subject:

- A rare malignant cutaneous mesenchymal tumour, less than 5% of adult soft sarcomas
- Affects young adults
- Predominantly male

Contribution of our study:

- Knowledge of the frequency of this tumour in Mali, a first study
- Multiple recurrence despite the respect of the excision margin of more than 2 to 3 cm.
- Approximately more than 8% of lethality

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

AUTHORS' CONTRIBUTION

Data Collection: Ali Coulibaly, Simon Laho Tamekou, Mamoudou Diakité, Mamadou Gassama.
Data Analysis and Interpretation: Alimata Keita, Bekaye Traoré, Binta Guindo, Omarou Maiga, Youssouf Fofana, Koureissi Tall, Madou Sissoko, Aboubacrine Traoré.
Writing of the Article: Alimata Keita, Bekaye Traoré, Ali Coulibaly, Binta Guindo, Ousmane Sylla.
Review of the Article: Alimata Keita, Bekaye Traoré, Binta Guindo, Mamoudou Diakité, Koureissi Tall, Mariam Diarra, Aissata Samaké Adama Aguissa Dicko, Ousmane Faye. All authors approved the final version of the manuscript

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