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Darier-Ferrand Dermofibrosarcoma: A Case Report of a Cervical Localization

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Abstract

Darier-Ferrand dermatofibrosarcoma (DFS) is a cutaneous mesenchymal tumor of intermediate malignancy. It is a rare but not exceptional tumor, accounting for 0.1% of malignant skin tumors. Histological examination is essential for diagnosis. Wide surgical excision is the standard treatment. DFS is a tumor whose prognosis and evolutionary risk are mainly linked to the delay in diagnosis and the quality of the first excision. Late diagnosis makes excision and reconstruction surgery difficult. The chances of recovery in the case of well-performed primary surgery are significantly greater than in the case of salvage surgery. To improve prognosis, early, codified, multidisciplinary management is essential. In our African context, and especially in the case of patients living in rural areas, errant diagnoses are often found hence the importance of raising awareness and providing information to healthcare personnel. We report a case of an enlarged left supraclavicular Darier-Ferrand dermatofibrosarcoma. The patient had come for a late consultation at a stage when the tumour was large. The CT scan was a great help in the pre-operative phase. The patient underwent complete surgical excision, the postoperative course was straightforward and the histology of the surgical specimen confirmed the diagnosis. The resection margins were healthy. The patient is alive at one year with no recurrence or metastasis.

Keywords

Cervical Localization, Dermatofibrosarcoma, Skin Tumor, Wide Surgery

1. Introduction

DFS is an intradermal mesenchymal skin tumor first described by Taylor as a

sarcomatous tumor [1]. Described by Jean Darier and Marcel Ferrand in 1924 [2] [3].

It lies between the benign pole of the very common and harmless cutaneous fibroma and the malignant pole of the true cutaneous fibrosarcoma. Its frankly malignant sarcomatous transformation with metastasis is exceptional.

This tumor, whose frequency is not negligible in African countries, still poses a number of problems: its misunderstanding by the vast majority of general practitioners and even some specialists; its misleading clinical appearance, evoking above all a keloid scar (common in the African population) and often responsible for a delay in diagnosis; its severity, due to its local aggressiveness and destructive potential; and its recurrent nature if the initial treatment has not respected the rigorous rules required for the management of this particular tumour [1].

The preferred sites are the trunk, followed by the proximal extremities, then the head and rarely the neck. Because of its rarity, very few epidemiological studies have been devoted to it [2] [4].

DFS often affects patients in their 3rd-4th decades, with a slight male predominance, and presents clinically as a firm reddish plaque or nodule [2].

Despite its distinct histological presentation, its histogenesis remains undefined. It is a tumor of "intermediate malignancy potential", with a good prognosis after complete resection, slow growth, very high risk of local recurrence, but low metastatic potential [2].

This clinical case allows us to share our experience of the cervical location of Darier-Ferrand dermatofibrosarcoma and to recall the various epidemiological, clinical, therapeutic and prognostic characteristics of this very particular and rare skin tumor.

2. Clinical Case

We report the case of patient B. D, aged 37, a farmer by profession living in a rural area. He presented with a large supra-clavicular left cervical mass that had been present for 2 years and had been progressively increasing in volume for 3 months. After several visits to traditional healers with no improvement, the patient decided to come to our facility for better care. He had no particular pathological history, and the examination revealed a voluminous left cervical mass, with partial depigmentation of the skin opposite the mass, which was slightly infiltrated (**Figure 1** and **Figure 2**).

The tumour was painless, fixed in relation to the superficial plane and slightly mobile in relation to the deep plane. Cervical examination revealed no associated adenopathy. Clinically, the mass was suggestive of a large trigeminal lymph node, a primary adenopathy with a metastatic appearance, or a lymph node metastasis of an ENT cancer.

A cervicothoracic and abdominopelvic CT scan was ordered. It revealed an 11 cm long, circumscribed, well-limited, homogeneous tissue mass. There was no

abnormality at the thoraco-abdomino-pelvic level (Figure 3 and Figure 4).

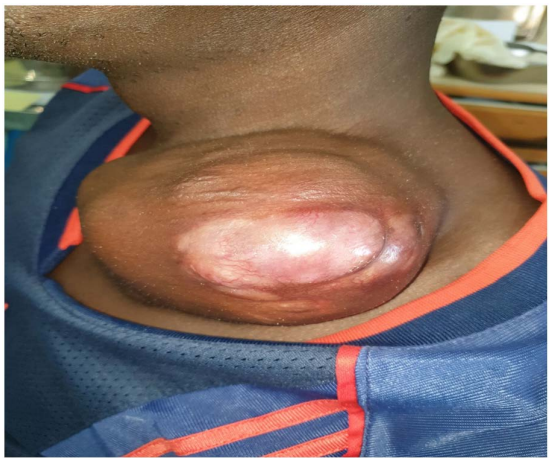

Figure 1. Lateral view of the mass.

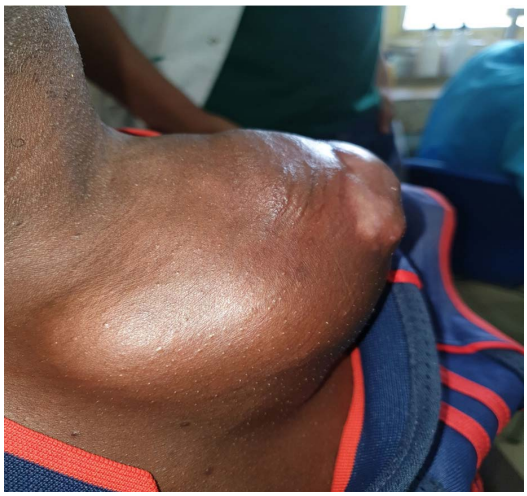

Figure 2. Anterior view of the mass.

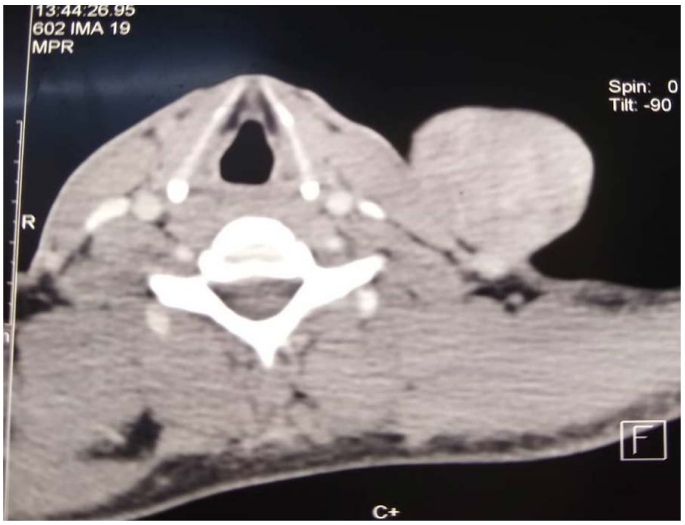

Figure 3. Axial CT section of the tumor.

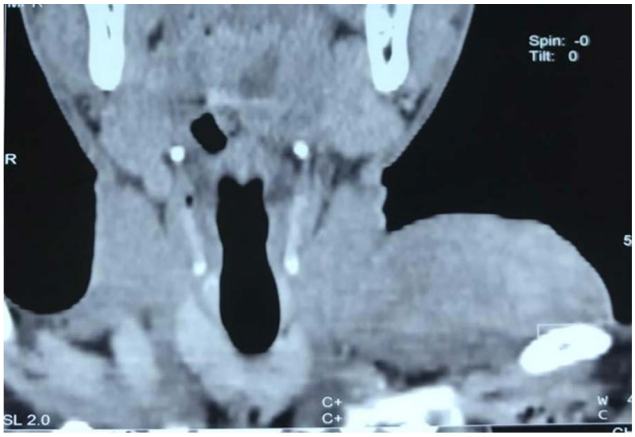

Figure 4. Coronal CT section of the tumor.

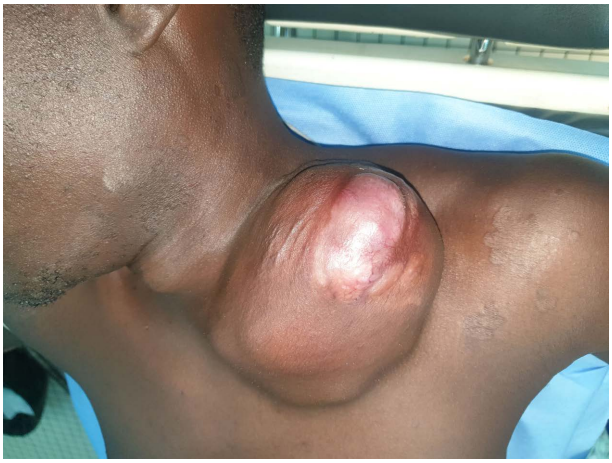

Figure 5. Top view of the mass on the operating table.

Given the context: the delay in consultation, the rapid evolution of the mass in a few weeks, the patient's lack of financial means, the diagnostic aid of the CT scan and the risk of a long wait for histological results in the event of a biopsy being performed, we decided to carry out an exploration in the operating room.

The patient had benefited from surgery. Intraoperatively, the scannographic features of the mass were found, and the exeresis was complete and wide (**Figures 5-10**). The postoperative course was straightforward.

Anatomopathological examination of the surgical specimen showed a Darier-Ferrand dermatofibrosarcoma with healthy resection margins. The patient is being followed up regularly, and at 1 year post-op (**Figure 11**), there has been no recurrence.

3. Discussion

Of all the definitions, DEGOS's is the most comprehensive: DFS "is a spindle-cell dermal connective tissue tumor, more or less similar in histological structure to sarcomatous tumors, but which differs from true primary fibrosarcomas in that it always originates in the skin, and evolves very slowly. Only

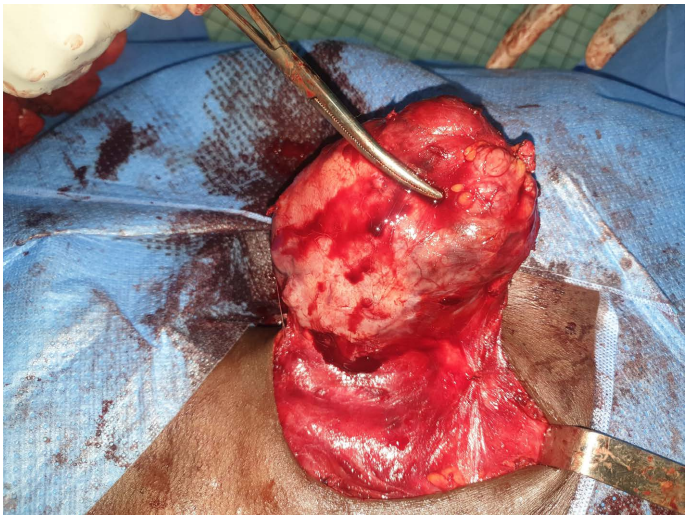

Figure 6. Progressive removal of the tumor following the cleavage planes.

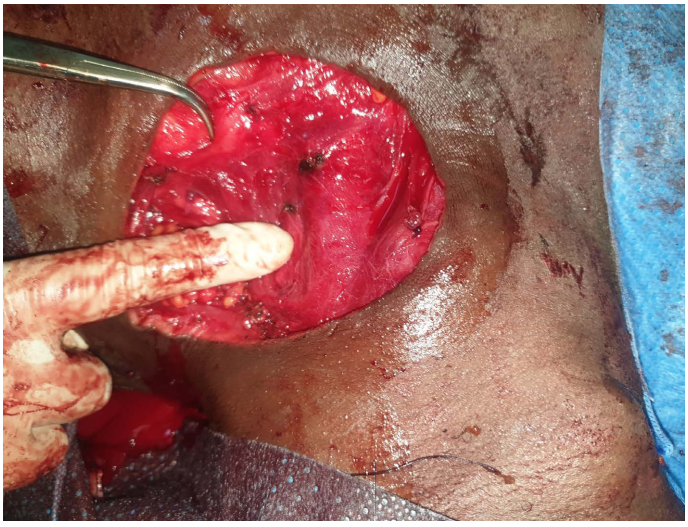

Figure 7. After removal of tumor, which rested on the SCM muscle.

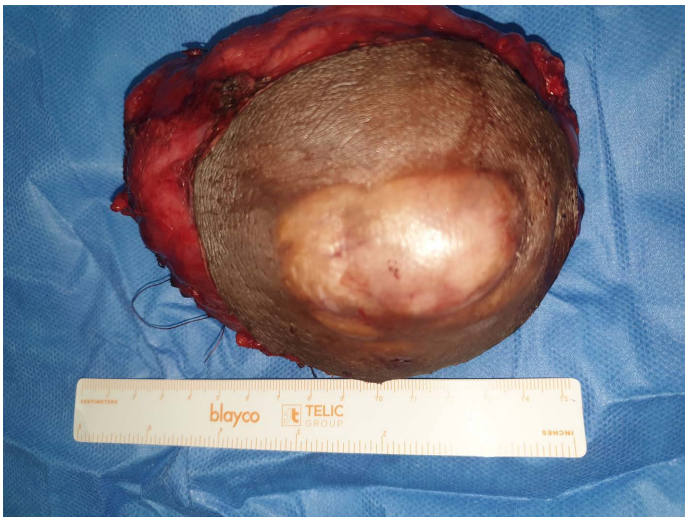

Figure 8. Superior view of tumor after excision.

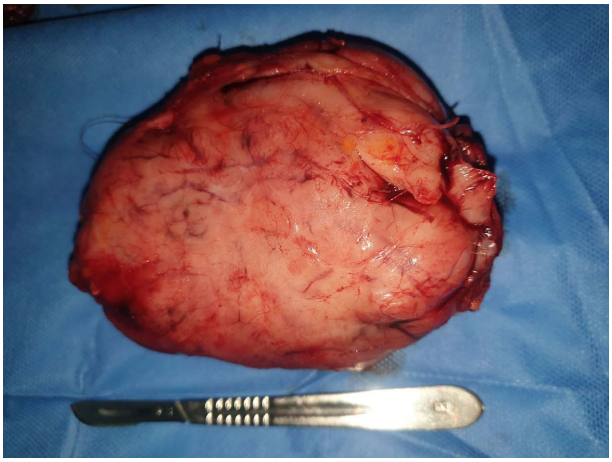

Figure 9. Internal view of tumor after excision.

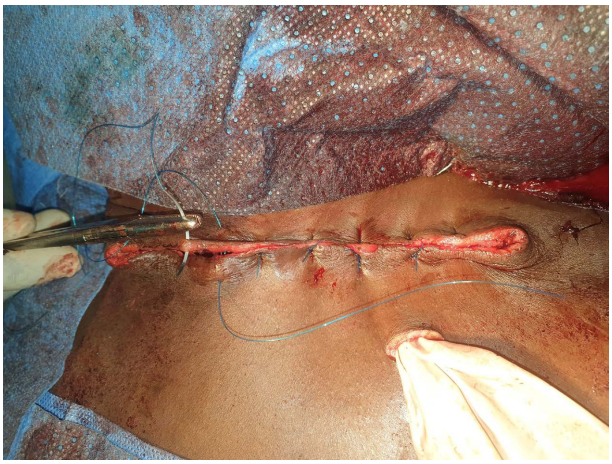

Figure 10. Single stitchskin suture.

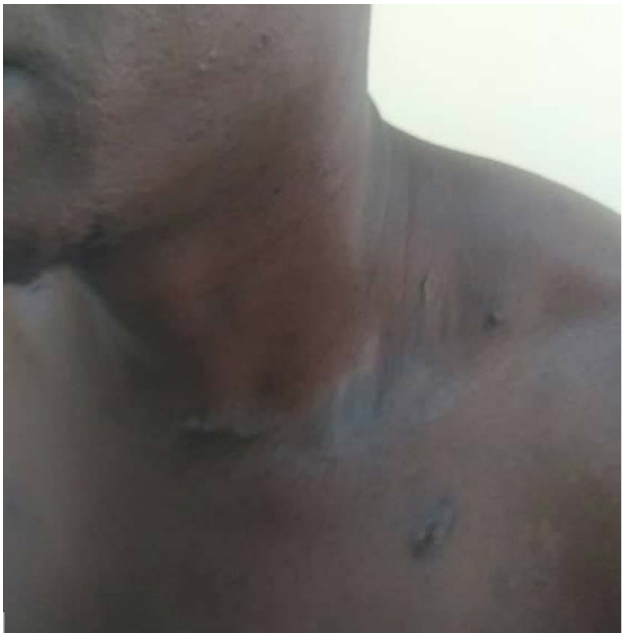

Figure 11. One year after surgery. Healing is good.

exceptionally, and at a very late stage, does it undergo a frankly malignant metastasizing sarcomatous transformation" [1].

According to some authors, this tumor is more frequent in Africans and the black race in general. Age of onset ranges from 20 to 50 years, with averages varying from 28 to 47 years depending on the author [1] [2], which is in line with our clinical case. With regard to sex, some authors, like our case, have found a male predominance [2] [3], while others noted opposite [1] and nor yet other authors, there is no clear predominance of gender [4].

It is often primary, but can also develop on surgical or burn scars. The tumor begins as a single, indurated, brownish or purplish, well-limited, mobile plaque. At this stage, the skin is of normal color, and the lesion may take on the appearance of an atrophic plaque or a sclerodermiform lesion. After several years, one or more nodules appear, embedded in the plaque. The skin then becomes taut, smooth and shiny. This two-stage evolution is not constant, as some forms are initially uninodular or multinodular, with secondary fusion of the nodules. Cases of "monstrous tumors" reaching 6.5 or even 7 kg have been described [1] [3].

The size of the lesion varies from 1 to 5 cm, and can reach over 20 to 25 cm [1] [3]. In our patient, the tumor was 11 cm in size, which is comparable to taylor's series [2]. The tumor then infiltrates the deeper layers, forming a hard, bumpy plastron. The lesion may present an ulcero-necrotic and bleeding appearance. General health is preserved for a long time. Metastases are rare, appearing late in the clinical course or after several recurrences. The route of dissemination is mainly blood-borne, which explains why metastases are more often found in the lungs than in the lymph nodes [3].

DFSP can occur in any part of the body, with a predominance of the trunk and extremities. Cervical localization is rare [1] [2] [3] [4].

The clinical form classically described, protuberant, corresponds to an advanced stage of the tumor. It's a firm, multinodular mass, attached to the surrounding skin but mobile in relation to the underlying layers like our patient. The tumor is painless, except when ulcerated. The diversity of clinical forms is a source of diagnostic delay [4].

In the literature medical imaging plays a minor role in the diagnosis of DFS in contrast to our clinical case, where the CT scan was a great aid to diagnosis and guided our therapeutic attitude. CT and MRI show a tumoral process with cystic, necrotic or hemorrhagic areas. They have the advantage of identifying bone involvement and determining the limits of the lesion [3].

Histological examination is essential for diagnosis. The tumor consists of a dense, poorly defined, non-encapsulated cellular proliferation occupying the dermis, usually 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. Cells are elongated, spindle-shaped, with more or less abundant cytoplasm and regular, oval nuclei. Mitosis is variable, with rare atypia. The stroma varies from one area to another [1]

[2]. In our patient, we had no pre-operative histology, given the rapid evolution of the mass and the patient's lack of means.

The treatment of choice for DFS is surgery. The exeresis margin is 5 cm around the lesion, and must include a healthy aponeurotic plane [1] [3] [4]. However, this is a mutilating surgery, especially for cervicofacial lesions. To make this surgery less disfiguring, while still being carcinological, some authors have achieved 3 cm margins of exeresis with extemporaneous histological study. This technique is similar to Mohs' micrographic surgery, where proof of complete excision of the tumour is confirmed by a pathologist in the operating theatre. In fact, excision only stops when the tumour is in a healthy zone [3] [4]. This technique is suitable for cervico-facial localizations, but obtaining such a technical platform and experienced personnel is far from being within everyone's reach. In our context, we recommend wide excision margins. Radiotherapy is used postoperatively for large, recurrent lesions or when the patient is inoperable. Imatinib mesilate is approved for the treatment of patients with recurrent and metastatic DFS [3] [4]. However, its unavailability and high cost limit its use in our context.

Local recurrence is frequent, occurring between 1 and 25 years of age. Their frequency depends on the initial excision margins: 40% with 2 cm margins, 10% - 20% with 3 cm margins, 1.75% with 4 cm margins [3].

The prognosis is conditioned by a predominantly local malignancy. DFS almost never metastasizes, and lymph node invasion occurs in less than 1% of cases. Frankly malignant metastasizing sarcomatous transformation is exceptional and occurs at a very late stage. Death is exceptional, and occurs late due to local complications [2].

The resection margins were healthy in our patient and at one year follow-up, our patient showed no recurrence or metastasis.

4. Conclusion

Intermediate between the harmless fibroma and the dreaded sarcoma, Darier-Ferrand dermatofibrosarcoma is a rare fibrous tumor of the skin, distinguished by its diagnostic difficulty and very slow local evolution, with a tendency to local recurrence and rare metastases. Cervical localization is rare and may mimic other pathologies of the ENT sphere. Surgical excision is easy if the patient is seen early. In our practice, CT scans are an important pre-therapeutic aid. In our context, delayed diagnosis and lack of resources have a negative impact on patients' prognosis hence the need for good information, communication and awareness-raising among patients and healthcare staff. Regular clinical monitoring is essential postoperatively, given the high recurrence rate.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Elamrani, D., Droussi, H., Boukind, S., Elatiqi, K., Dlimi, M., Benchamkha, Y. and Ettalbi, S. (2014) Le dermatofibrosarcome de Darier et Ferrand, une tumeur cutanée particulière: à propos de 32 cas et revue de la literature. *Pan African Medical Journal*, **19**, 196-203. https://doi.org/10.11604/pamj.2014.19.196.4470

[2] Hammas, N., Badioui, I., Znati, K., Benlemlih, A., Chbani, L., El Fatemi, H., Harmouch, T., Bouyahyaou, Y., Boutayeb, F., Mrini, A., Mesbahi, O., Mernissi, F.Z. and Amarti, A. (2014) Le dermatofibrosarcome de Darier et Ferrand: À propos de 27 cas et revue de la literature. *Pan African Medical Journal*, **18**, 280-286. https://doi.org/10.11604/pamj.2014.18.280.1087

[3] Miloundja, J., Assini Eyogho, S.F., Mandji Lawson, J.M., Makungu, A.P., Bamba, J.S., Nzenze, S.A.M., Nziengui, A., Ell Ondo, T. and N'zouba, L. (2012) Dermatofibrosarcome de Darier-Ferrand: Deux cas à localisation cervicofaciale. *African Journal of Cancer*, **4**, 249-252. https://doi.org/10.1007/s12558-012-0223-z

[4] El Kacemi, H., Aissa, A., Bazine, A., Kebdani, T., Bougtab, A. and Benjaafar, N. (2014) Dermatofibrosarcome de Darier et Ferrand: À propos de 38 cas. *Pan African Medical Journal*, **19**, 274-278. https://doi.org/10.11604/pamj.2014.19.274.3604