Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Dermatology

Analysis of a Series of 27 Cases of Darier-Ferrand Dermatofibrosarcoma at the Mohammed VI University Hospital in Marrakech

Halima Elkhadraoui[1*], Imane Boujgana[1], Hind Rachidi[1], Meryem Khallouki[2], Anas Fakhri[1], Hanane Rais[1]

[1]Anatomical-cytopathology Department of the Mohammed VI University Hospital in Marrakech-Morocco
[2]Dermatology Department of the Mohammed VI University Hospital in Marrakech-Morocco

DOI: https://doi.org/10.36347/sjams.2024.v12i08.018 | **Received:** 14.07.2024 | **Accepted:** 21.08.2024 | **Published:** 24.08.2024

***Corresponding author:** Halima Elkhadraoui
Anatomical-cytopathology Department of the Mohammed VI University Hospital in Marrakech-Morocco

Abstract **Original Research Article**

Darier Ferrand dermatofibrosarcoma is a low-grade superficial fibroblastic tumor with a storiform architecture. It is characterized by its local aggressiveness and low metastatic potential. We report a retrospective study spread over a period from January 2019 to July 2024 and involving 27 cases collected at the Mohammed VI University Hospital in Marrakech. Our series showed a sex ratio M/F of 1.4 with an average age of our patients at the time of diagnosis was 31.7 years with extremes of age between 68 and 21 years. The buttock and thigh were the predominant location observed in 55.6% (15 patients), the thorax 18.5% (5 cases), the leg 14.8% (4 cases), the face 3 ,7% (1 case), the back (1 case) and the vulva (1 case). The aim of this work is to report the experience of the Mohammed VI University Hospital and to discuss the related literature on the epidemiological and anatomopathological characteristics.

Keywords: Darier ferrand dermatofibrosarcoma, epidemiology, anatomopathological profile.

INTRODUCTION

Darier Ferrand dermatofibrosarcoma is a low-grade superficial fibroblastic tumor with a storiform architecture, harboring a characteristic translocation t (17,22) (q21.3; q13.1) that leads to a COL1A1-PDGFB fusion (1,2). It is characterized by its local aggressiveness, and by a low metastatic potential. The aim of this work is to report the experience of the Mohammed VI University Hospital and to discuss the related literature on the epidemiological and anatomopathological characteristics.

MATERIAL AND METHODS

Our work concerns a retrospective study conducted in the anatomic-cytopathology department of the Mohammed VI University Hospital in Marrakech over a period from January 2019 to July 2024. It concerned 27 cases which benefited from surgical excision with an immunohistochemical study.

RESULTS

Our series showed a M/F sex ratio of 1.4. The mean age of our patients at diagnosis was 31.7 years with extremes of age between 68 and 21 years. The buttock and thigh were the predominant location observed in 55.6% (15 patients), the thorax 18.5% (5 cases), the leg 14.8% (4 cases), the face 3 ,7% (1 case), the back (1 case) and the vulva (1 case). The specimens received were surgical excision pieces with wide safety margins ranging from 3 to 5 cm **(Figure 1)**. Morphological analysis revealed a diffuse tumor proliferation, made of spindle cells extending along the connective septa and infiltrating the hypodermal fat, giving way to the adipose vacuoles it encompasses. The tumor cells were monomorphic and regular, without major atypia. They were arranged in wheel spokes with a "storiform pattern" **(Figure 2)**. In immunohistochemistry, tumor cells were positive for anti-CD34 antibody **(Figure 3)** in 16 patients with an intensity that differed between intense and moderate while it was positive on the biopsy and negative on the vulvectomy specimen. The final diagnosis for this patient after elimination of other differential diagnoses was sarcomatous transformation.

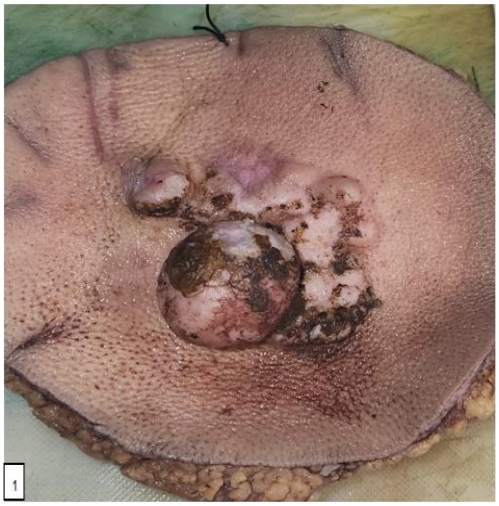

Figure 1: Macroscopic appearance of a surgical biopsy weighing 310g and measuring 14x10x7 cm, the site of a budding neoplasm measuring 5x4x4 cm. The latter is located respectively 5, 3, 3 and 4 cm from the 4 lateral limits and 5 cm from the deep limit

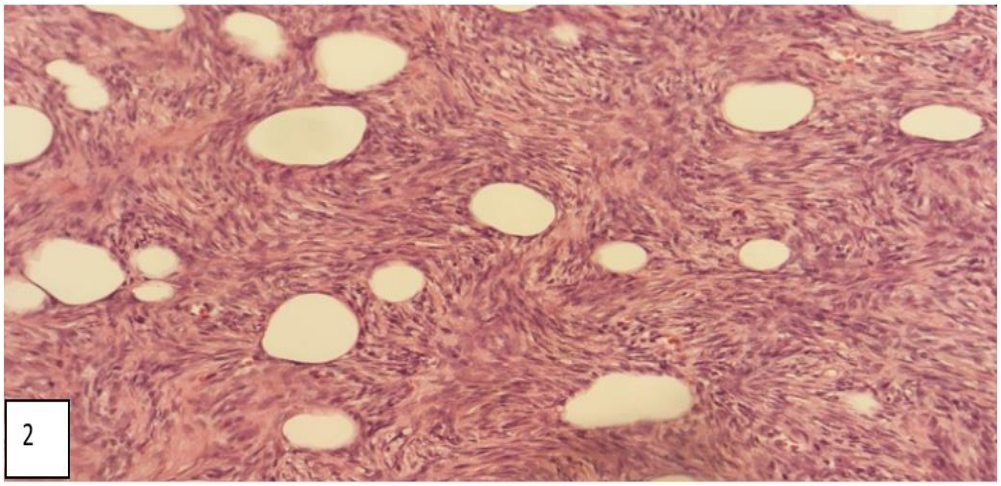

Figure 2: A diffuse tumor proliferation, made of spindle cells arranged in a storiform pattern extending along the connective septa and giving way to the fat vacuoles that it encompasses (HEx200)

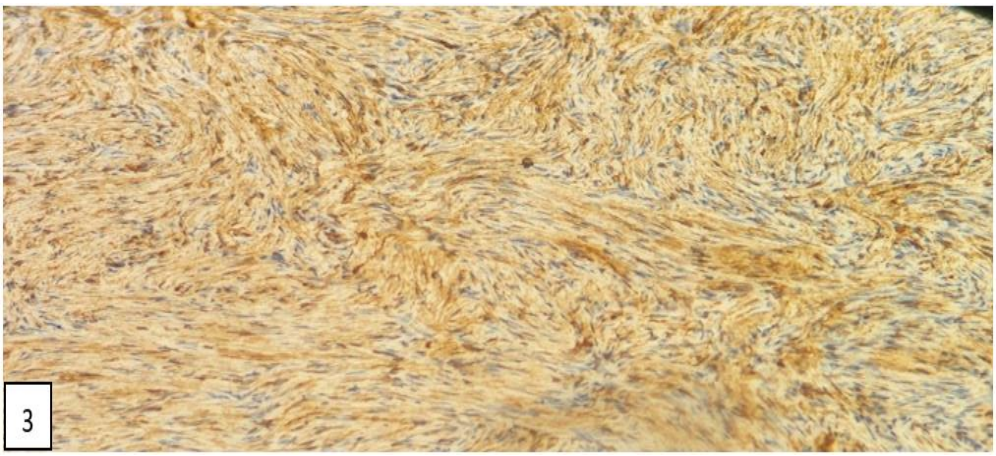

Figure 3: A moderate and diffuse cytoplasmic expression of spindle tumor cells to the anti-CD34 antibody (x200)

DISCUSSION

Dermatofibrosarcoma represents 2% of all sarcomas and 4 to 5% of spindle cell sarcomas [3]. It most often develops in young or middle-aged adults, whose location at the trunk or the root of the limbs is the most common [3], which is consistent with our series. Its diagnosis is based on a microscopic study showing a storiform appearance and a positivity of the CD34 antibody in immunohistochemistry with negativity of

other markers to eliminate other differential diagnoses. (1+2). The reference treatment for DFS remains wide surgery, it is recommended to respect margins of 2 to 4 cm or even 5 cm, given the high rates of local recurrence [4]. DFS are characterized by a translocation between chromosomes 17 and 22 t (17,22) with overexpression of the PDGFRB receptor which can be targeted by Imatinib; Indeed, some studies have shown its interest, it is a tyrosine kinase inhibitor that has proven its effectiveness in localized and metastatic DFS in the presence of t (17,22) [4]. DFS have low mitotic activity and are therefore not radiosensitive. However, radiotherapy has its place in cases of recurrence, multiple locations, large tumors or inoperable locations preventing wide excision and especially after an insufficient or invaded excision margin [5].

CONCLUSION

Darier Ferrand dermatofibrosarcoma is characterized by aggressiveness and frequent local recurrences, often repeated, unless it is widely excised. The fibrosarcomatous transformation observed in 5% of cases presents a more aggressive behavior and distant metastases (1+2).

REFERENCES

1. El Khadraoui, H., Soufi, S., Benallal, C., Benzi, B., CHouaf, A., Rachidi, H., & Rais, H. (2024). Ovarian Localization of a Peripheral Primitive Neuroectodermal Tumor: Definitive Diagnosis is a Real Challenge. *SAS J Surg*, *5*, 626-628.
2. Fred. T. Bosman, MD, PhD, Elaine, S, Jaffe, Sunil R. Lakhani, FRCPath, Hiroko Ohgaki. WHO Classification of Skin Tumours. Published by International Agency for Research on Cancer (IARC).2018
3. Tumeurs Des Tissus Mous –Groupe Sarcome Français-Tome 1-2017-Tumeurs à cellules fusiformes. Page 117.
4. Guerrouaz, M. A., Kharkhach, A., Miry, A., Harroudi, T. E., Sbai, A., & Mezouar, L. (2022). Recurrent Darier-Ferrand dermatofibrosarcoma in the abdominal wall: the role of preoperative radiotherapy (case report). *The Pan African Medical Journal*, *41*, 234-234.
5. Frikha, F., Mseddi, M., Bahloul, E., Chaabouni, R., Masmoudi, A., Boudaya, S., ... & Turki, H. (2020). Dermatofibrosarcoma protuberans (Report of 49 cases). *Our Dermatol Online*, *11*(1), 25-31.