ISR Journal of Applied Medical Science (ISRJAMS)
Homepage: https://isrpublisher.com/isrjams/
Volume 01, Issue 02, August 2025 ISSN: XXXX-XXXX (Online)

Darier and Ferrand's Dermatofibrosarcoma: About 10 Cases and Review of the Literature

S. Charboub[1]*, I. Khaled[1], Z. Berjaou[1], J. Hafidi[1], N. Gharib[1], A. Abbassi[1], S. El Mazouz[1]

[1]Department of Reconstructive and Plastic Surgery, Ibn Sina Hospital of Rabat, Morocco

***Corresponding author:** S. Charboub
Department of Reconstructive and Plastic Surgery, Ibn Sina Hospital of Rabat, Morocco

Article History

Received: 29-06-2025

Accepted: 24-07-2025

Published: 02-08-2025

Summary: Review Article

Darier-Ferrand dermatofibrosarcoma (DFS) is a rare, locally aggressive mesenchymal skin tumor characterized by slow growth and a high potential for local recurrence. Its diagnosis is based on histology and immunohistochemistry, including CD34 positivity. We report a descriptive retrospective study conducted in the plastic surgery department of Ibn Sina University Hospital in Rabat, including ten patients operated on for DFS between January 2019 and December 2022. Epidemiological, clinical, histological, therapeutic, and evolutionary data were analyzed. The majority of patients had a tumor located on the trunk or limbs. All underwent wide surgical excision with appropriate reconstruction. The local recurrence rate was low (1 case). These results are consistent with data from the literature and highlight the importance of safety margins. Wide excision remains the cornerstone of DFS treatment. Early diagnosis, multidisciplinary care, and prolonged follow-up are essential. Access to modern techniques such as Mohs surgery or targeted therapies, as well as molecular screening, could significantly improve prognosis.

Keywords: Darier and Ferrand dermatofibrosarcoma, Dermatofibrosarcoma protuberans (DFSP), Cutaneous soft tissue sarcoma, Oncological wide excision.

INTRODUCTION

Darier-Ferrand dermatofibrosarcoma (DFSP) is a rare, low-grade mesenchymal skin tumor characterized by slow growth but notable local aggressiveness. It primarily affects young adults and is preferentially located on the trunk and extremities. Diagnosis is based on histology and immunohistochemistry, particularly CD34 positivity.

Treatment is based on wide surgical excision to minimize the high risk of local recurrence. Despite its low metastatic potential, DFSP requires rigorous management and prolonged follow-up.

In this work, we report a series of 10 cases treated in our department between 2019 and 2024, highlighting the clinical, therapeutic and evolutionary aspects of this tumor, in light of recent data from the literature.

Materials and Methods

We conducted a descriptive retrospective study within the Plastic and Reconstructive Surgery Department of the Ibn Sina University Hospital in Rabat, over a period of four years, between January 2019 and December 2022. The objective was to analyzethe epidemiological, clinical, therapeutic and evolutionary characteristics of Darier and Ferrand dermatofibrosarcoma (DFS) treated in our structure.

We included all patients with histologically confirmed DFS diagnosis who underwent surgical treatment in our department during the study period. Only complete and usable medical records,

including sufficient postoperative follow-up, were retained.

Data were extracted from archived medical records. For each patient, we collected demographic information (age, sex, history), clinical characteristics of the tumor (location, size, appearance, diagnostic delay), as well as the results of paraclinical explorations, including imaging examinations such as MRI or CT scan, performed to assess local and distant extension.

Histological and immunohistochemical analysis confirmed the diagnosis in all cases. Regarding therapeutic management, all patients underwent wide surgical excision with safety margins varying from two to five centimeters depending on anatomical constraints and tumor location. When necessary, a healthy deep tissue barrier was also resected, including the fascia, muscle, or periosteum.

The coverage of substance losses was ensured by different reconstruction techniques (skin graft, flaps, etc.)

Patients underwent regular outpatient follow-up, including a systematic clinical examination and, where appropriate, radiological monitoring. Any adjuvant treatments received, postoperative complications, and cases of local or distant recurrence observed during the monitoring period were also recorded.

RESULTS

Our series includes ten patients with Darier-Ferrand dermatofibrosarcoma, treated surgically between January 2019 and December 2022.

The mean age of the patients was 41.6 years, with extremes ranging from 25 to 68 years. Male predominance was marked, with a sex ratio of 2.3. The mean time between the onset of the lesion and the first consultation was 6.7 years, highlighting a frequent diagnostic delay.

Clinically, the typical presentation was a nodular or localized mass, firm, slow-growing, sometimes ulcerated or painful. The tumors were located in a variety of locations: four cases were located on the trunk, three on the root of the upper limb, two on the scalp, and one on the abdominal wall. Lesion sizes ranged from 4 to 18 cm, with a mean of 8.6 cm.

The diagnosis of dermatofibrosarcoma was confirmed in all cases by histological study, supplemented by immunohistochemistry highlighting strong positivity for the CD34 marker. An MRI was performed in eight cases, allowing a better assessment of the depth extension and anatomical relationships. A thoraco-abdomino-pelvic CT scan was performed in all patients to search for metastases; no secondary locations were detected.

All patients underwent wide excision with lateral margins of 2 to 5 cm. In four cases, the excision included a deep anatomical structure: aponeurosis in three cases and muscle in one case. The defects were reconstructed, depending on the case, by thin skin graft (eight cases), local flap (one case), and micro-anastomosed free flap (one case).

The postoperative course was uneventful for the majority of patients. No cases of immediate serious complications were observed. Postoperative follow-up, with a mean follow-up of 24 months, revealed only one case of local recurrence, occurring 13 months after the operation, requiring surgical revision. No cases of distant metastasis were observed during the surveillance period.

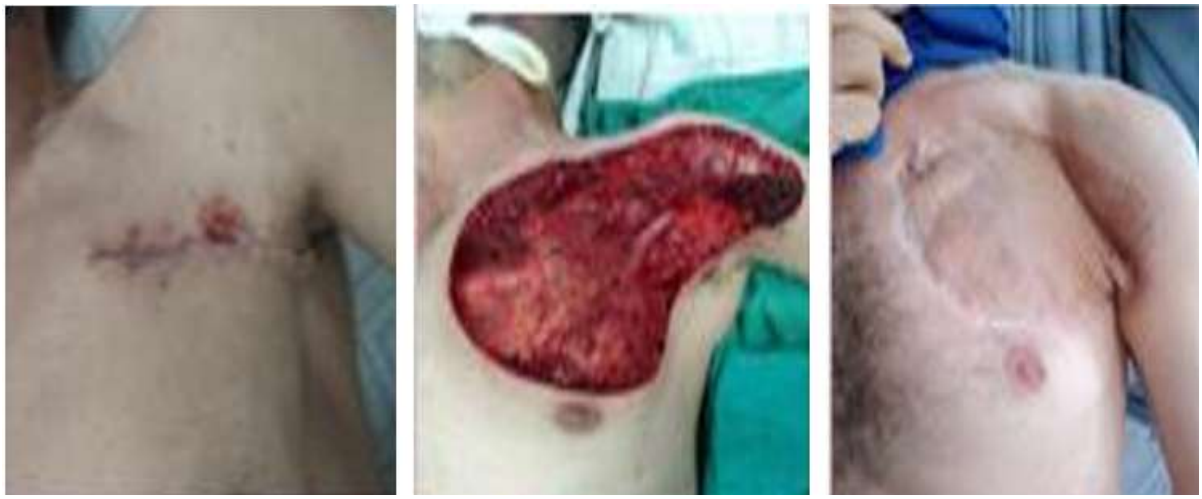

Figure 1: Case 1: Complementary excision of a DFSP with resection of the pectoralis major muscle

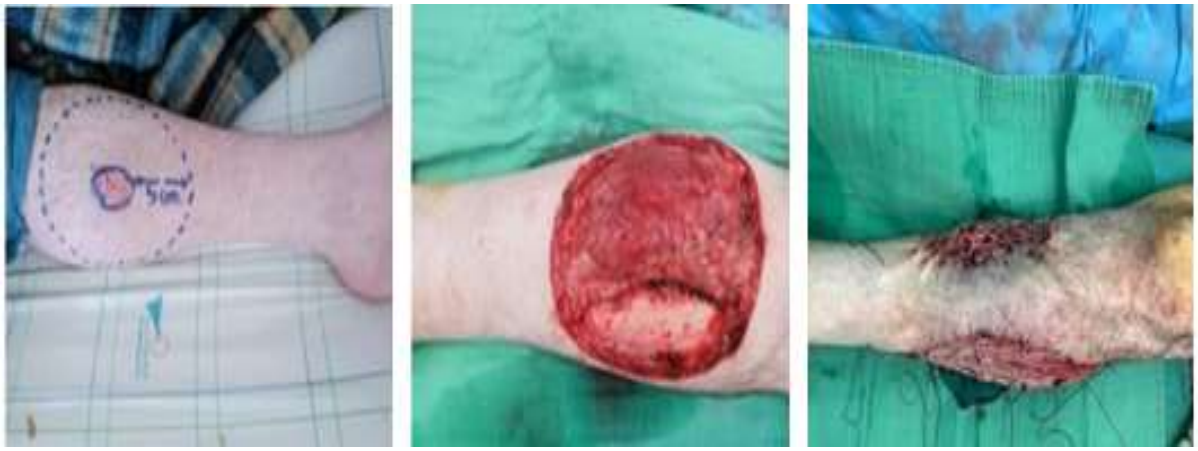

Figure 2: Case 2: Excision of a DFSP with resection of the tibial periosteum and coverage using a bipedicled flap

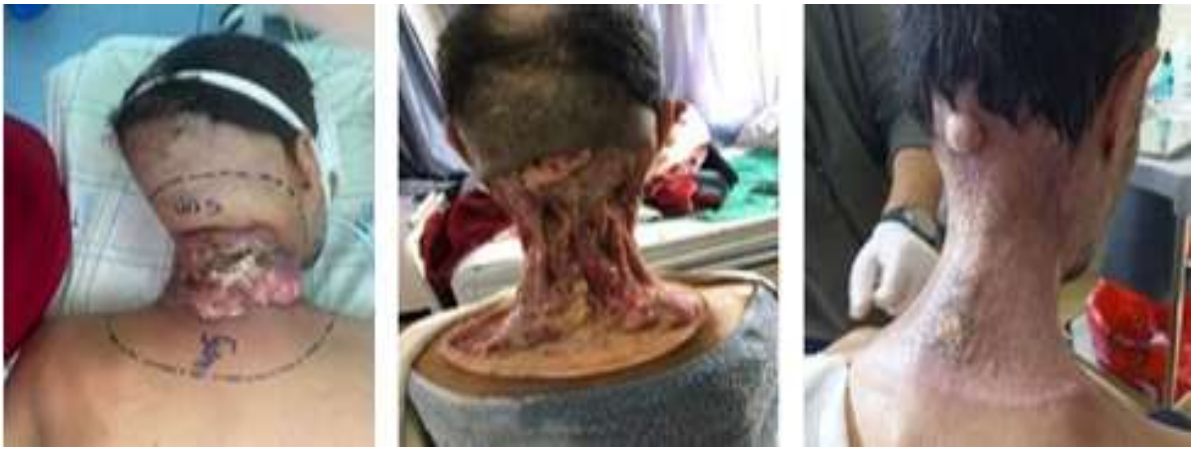

Figure 3: Case 3: Excision of a DFSP and its malignant transformation

DISCUSSION

Darier-Ferrand dermatofibrosarcoma (DFS), also known as dermatofibrosarcoma protuberans (DFSP), is a rare, locally aggressive, intermediate-grade cutaneous tumor. It develops from fibroblasts of the dermis and hypodermis. Although it rarely metastasizes, its potential for local recurrence is high, due to its ability to infiltrate deeply and spread laterally. It represents a therapeutic

challenge due to its insidious progression and sometimes difficult localization [1].

Epidemiologically, DFS represents approximately 1% of all skin tumors and 6% of soft tissue sarcomas. It most often occurs in young adults aged 20 to 50 years, with a slight male predominance according to some series. However, it can occur in children or the elderly. Its topographic predilection is the trunk (50%), followed by the limbs (30%) and the head and neck (10%). In Morocco, the literature remains limited, but several hospital series suggest an incidence comparable to international data [2, 3].

The etiopathogenesis of DFS is now well established. In more than 90% of cases, a specific chromosomal translocation t(17;22)(q22;q13) is found, resulting in the fusion of the COL1A1 and PDGFB genes. This anomaly leads to autocrine activation of the PDGFB signaling pathway, promoting tumor proliferation. This discovery paved the way for targeted treatments with tyrosine kinase inhibitors, including imatinib [4].

Known risk factors include skin trauma, old scars, including keloid scars, burns, and vaccinations, although no definitive causal relationship has been demonstrated. Cases have been reported after surgical scars or following chronic trauma, suggesting a link between chronic tissue repair and the occurrence of DFS [5].

Clinically, DFS manifests as a firm, painless, slow-growing dermal or subcutaneous nodule that may remain stable for several years before becoming protruding. It is often confused with a benign lesion (lipoma, cyst, keloid), which delays diagnosis. The surrounding skin is often fixed or infiltrated. The course is purely local, except in the case of sarcomatous transformation [6].

Histopathologically, the tumor consists of a proliferation of spindle-shaped cells arranged in crisscrossing bundles in a storiform pattern. Wheel-like invasion of adipose tissue is characteristic. Immunohistochemistry shows marked and diffuse expression of CD34, which distinguishes DFS from other fibrous tumors such as dermatofibroma (CD34 negative) [7].

Treatment is essentially based on wide surgical excision with safety margins of 2 to 3 cm. This strategy significantly reduces local recurrences, the frequency of which varies between 20 and 50% according to the literature. However, in certain functional or aesthetic locations, this approach can be difficult to apply [8].

Mohs micrographic surgery is an alternative allowing precise excision with maximum tissue preservation, particularly in cephalic locations. It is recommended in recurrences or complex anatomical areas. However, it remains little practiced in countries with limited resources due to its technicality and cost [9].

In cases of unresectable local recurrence, unresectable positive margins, or advanced form, adjuvant radiotherapy may be proposed to improve local control. Since DFS is a radiosensitive tumor, this approach is beneficial in addition to an incomplete procedure. However, exclusive radiotherapy is rarely curative [10].

Medical treatment is reserved for locally advanced, inoperable or metastatic forms. Imatinib, a tyrosine kinase inhibitor, is effective in cases with COL1A1–PDGFB translocation, with variable response rates. This treatment can allow preoperative reduction of the tumor and facilitate secondary excision [11].

Complications of DFS are dominated by local recurrences, the frequency of which depends directly on the quality of the initial excision. The recurrence rate is less than 10% in series having applied margins ≥2 cm. The risk of metastasis is low (<5%) but higher in forms transformed into fibrosarcoma, which present a more aggressive behavior [12].

In our series of 10 cases of Darier and Ferrand dermatofibrosarcoma, the mean age of patients was 41 years, with extremes ranging from 23 to 68 years. This distribution is generally consistent with that reported by Bowne *et al*., who observed a median age of 39 years in a series of 159 cases [13]. Other studies confirm that DFS mainly affects young to middle-aged adults, although it can also occur in children or the elderly [14, 15].

We noted a slight male predominance (sex ratio M/F = 1.5), which is in agreement with the observations made by DuBay *et al*., in their multicenter cohort [16]. However, some larger series report a relatively equivalent incidence between the two sexes [17].

The trunk was the most common location in our study (60%), followed by the limbs (30%) and the head and neck (10%). This anatomical distribution is also described in the majority of international studies. In particular, Reimann and Fletcher found a predilection for the trunk in almost 50% of cases [18]. Loghdey *et al*., also confirm this dominant location, but point out that the cephalic region represents an area at high risk of recurrence if the excision is not adapted [6].

Clinically, the majority of our patients presented with a painless, slowly growing mass, which corresponds to the typical presentation of DFS. This diagnostic latency is well known and has been highlighted by several authors, notably Takayama *et al*., in the specific case of a giant DFS developed on an old keloid scar [5].

Histological examination confirmed in all cases the diagnosis of classic DFS, with spindle cells in bundles in a storiform arrangement and diffuse expression of CD34. No transformation into fibrosarcoma was noted in our series. This observation is similar to that of DuBay *et al*., who observed this transformation in less than 10% of cases [16].

Surgical treatment in our study was based on wide excision with safety margins of 2 to 3 cm in 90% of cases. This strategy is in line with current recommendations and was associated with a low recurrence rate in our cohort (1 case). This result is comparable to that reported by Bowne *et al*., with a recurrence rate of 7% after initial wide excision [13]. On the other hand, narrow margins or incomplete excisions are recognized as a high recurrence factor, as highlighted by Ashindoitiang *et al*., in their African series [3].

In one case, we resorted to adjuvant radiotherapy due to insufficient margins. This approach is also documented in the literature, notably in the recommendations of Tan *et al*., which suggest the indication of radiotherapy when surgical revision is not possible [9].

None of our patients were treated with imatinib, the treatment being reserved for inoperable or advanced cases, as demonstrated by the studies of Gutierrez *et al*., on targeted therapies based on the COL1A1–PDGFB fusion [4].

Finally, our overall rate of local recurrence (10%) remains below that reported by several historical series, particularly those where surgical margins were less than 2 cm [7,16]. This underlines the importance of a wide and well-planned excision.

Moreover, in our series, we mainly used wide safety margins, respecting an interval of 2 to 3 cm around the tumor, following current recommendations to limit the risks of local recurrence. This approach allows complete excision of the DFSP, known for its subclinical infiltration of peripheral tissues [7]. Wide excision, associated with immediate reconstruction by flap or graft, made it possible to ensure anatomical and functional continuity without delaying management. The use of local fasciocutaneous flaps has shown good efficacy in covering substance losses, particularly in thoracic or scapular locations [14].

However, this strategy has notable drawbacks. Wide excision can lead to significant surgical morbidity, with extensive scarring, a risk of postoperative dehiscence or hematoma, and sometimes functional impairment in articulation or weight-bearing areas [6]. In addition, the use of complex reconstructions, although effective, requires microsurgical expertise that is not available in all centers [12].

With that said, our study has several limitations that should be highlighted. First, the sample size is limited to ten patients, which restricts the statistical scope and generalizability of the results. Second, the lack of long-term follow-up is a significant constraint to fully assess recurrence rates, which are often late in the case of DFSP [7]. Furthermore, our work does not include a comparison between narrow versus wide resection margins, nor a comparative analysis between different surgical reconstruction modalities. Finally, complete histological data, including assessment of CD34 expression or the presence of COL1A1-PDGFB translocation, were not available for all cases, which limits molecular interpretation [18].

With the aim of improving the management of dermatofibrosarcoma protuberans in our context, multiple perspectives can be adopted. On the diagnostic level, wider access to immunohistochemistry and molecular study of the COL1A1-PDGFB translocation would allow better targeting of high-risk tumors and consideration of adjuvant treatment with imatinib in locally advanced or inoperable forms [11]. From a surgical perspective, the progressive introduction of micrographic Mohs surgery, already employed in several international centers, would enable a significant reduction in margins while ensuring optimal tumor control, particularly for locations with functional or aesthetic concerns [4]. Although technically demanding, it is increasingly recommended in specialized centers.

Furthermore, the establishment of a national DFSP monitoring registry would allow the collection of robust multicenter data, particularly on recurrence rates and reconstruction strategies, while promoting the standardization of practices. Finally, improving patient information and the development of postoperative quality of life scores would contribute to further integrating the psychological dimension into the overall management of these rare skin tumors [1].

CONCLUSION

Darier-Ferrand dermatofibrosarcoma, although rare, represents a therapeutic challenge due to its high potential for local recurrence despite low malignancy. Our study confirms the importance of early diagnosis, often delayed due to its misleading clinical presentation, as well as the effectiveness of wide surgical excision adapted to each anatomical location. The low incidence of recurrence in our series reflects the rigorous surgical approach applied.

However, optimal management of this tumor requires advanced diagnostic methods, surgical techniques adapted to local constraints, and prolonged monitoring. Wider access to targeted therapies, the introduction of Mohs surgery in complex locations, and better structured follow-up through a national registry are promising avenues for improving care. The integration of quality of life indicators and long-term functional outcomes also represents an essential perspective in the personalized treatment of these rare skin tumors.

Consent:

Written Informed consent was obtained from the patient for the publication of her case as a report and was documented in the patient's medical notes. A copy of the written informed consent would be available for review by the editor-in-chief of the journal on request.

Conflicts of Interest: The authors declare that there are no conflicts of interest regarding the publication of this case report.

BIBLIOGRAPHY

1. Mitteldorf C, Bertolotto C, Verneuil L. Review of dermatofibrosarcoma protuberans: Pathogenesis, diagnosis and treatment. Cancers (Basel). 2024;16(6):3124.
2. Benlemlih A, Khalloufi M, Belhiba O, *et al*., Darier and Ferrand dermatofibrosarcoma: a report of 11 cases. WJARR. 2024;21(3):202–206.
3. Ashindoitiang JA, Agbo PS, Abah VO, *et al*., Dermatofibrosarcoma Protuberans: A Case Series in a Tropical Setting and Review of the Literature. WJARR. 2024;21(2):100–105.
4. Gutierrez M, Marcu LG. Dermatofibrosarcoma Protuberans: Molecular Pathogenesis and Targeted Therapy. J Clin Med. 2024;13(5):1798.
5. Takayama O, Sakai Y, Shiohara T. Giant dermatofibrosarcoma protuberans originating from a keloid scar: A case report and review. Dermatol Ther. 2022;35(4):e15317.
6. Loghdey MS, Grobler N, George R. Review of dermatofibrosarcoma protuberans. S Afr J Radiol. 2014;18(1):1–4.
7. DuBay D, Cimmino V, Lowe L, *et al*., Low recurrence rate after surgery for dermatofibrosarcoma protuberans: A multidisciplinary approach from a single institution. Cancer. 2004;100(5):1008–1016.
8. Bowne WB, Antonescu CR, Leung DHY, *et al*., Dermatofibrosarcoma protuberans: A clinicopathologic analysis of patients treated and followed at a single institution. Cancer. 2000;88(12):2711–2720.
9. Tan MC, Brennan MF, Kuk D, *et al*., Histology-based classification predicts pattern of recurrence and improves risk stratification in primary dermatofibrosarcoma protuberans. Ann Surg Oncol. 2016;23(12):4014–4021.
10. Reimann JD, Fletcher CD. Imatinib response in DFSP: CD34 expression and translocation status. Mod Pathol. 2007;20(6):645–652.
11. Gloster HM. Dermatofibrosarcoma protuberans. J Am Acad Dermatol. 1996;35(3):355–374.
12. Dubay DA, *et al*., Surgical management of dermatofibrosarcoma protuberans: Multicenter analysis. Cancer. 2004;100(5):1008–1016.
13. Bowne WB, Antonescu CR, Leung DHY, *et al*., Dermatofibrosarcoma protuberans: A clinicopathologic analysis of patients treated and followed at a single institution. Cancer. 2000;88(12):2711–2720.
14. Tan MC, Brennan MF, Kuk D, *et al*., Histology-based classification predicts pattern of recurrence and improves risk stratification in primary dermatofibrosarcoma protuberans. Ann Surg Oncol. 2016;23(12):4014–4021.
15. DuBay D, Cimmino V, Lowe L, *et al*., Low recurrence rate after surgery for dermatofibrosarcoma protuberans: A multidisciplinary approach from a single institution. Cancer. 2004;100(5):1008–1016.
16. Ashindoitiang JA, *et al*., Dermatofibrosarcoma Protuberans: A Case Series in a Tropical Setting and Review of the Literature. WJARR. 2024;21(2):100–105.
17. Reimann JD, Fletcher CD. Imatinib response in DFSP: CD34 expression and translocation status. Mod Pathol. 2007;20(6):645–652.
18. Gloster HM. Dermatofibrosarcoma protuberans. J Am Acad Dermatol. 1996;35(3):355–374.