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(CASE REPORT)

Facial darier-ferrand dermatofibrosarcoma: A case report with review of the literature

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Abstract

Darier-Ferrand Dermatofibrosarcoma (DFS) is a rare mesenchymal tumor of intermediate malignancy, characterized by the proliferation of spindle cells within the deep dermis and subcutaneous tissues. This neoplasm is defined by slow growth but is notable for its marked local aggressiveness and high potential for recurrence without appropriate treatment. Diagnosis is based on histopathological examination, and surgical excision with margins of at least 5 cm remains the standard treatment to ensure complete tumor removal and minimize the risk of local recurrence.

The aim of this study is to describe the management of a patient with Darier and Ferrand dermatofibrosarcoma of the face and to review the literature

Keywords: Darier-Ferrand Dermatofibrosarcoma 1; Face 2; Surgery 4; Mohs micrographic surgery 5; Targeted therapy 6

1. Introduction

Darier-Ferrand dermatofibrosarcoma (DFS) is a rare cutaneous mesenchymal tumor characterized by the proliferation of spindle cells. It is predominantly locally aggressive, with a significant capacity for tissue destruction, while transformation into a frankly malignant sarcomatous form with metastases remains extremely rare. Although it progresses slowly, DFS is associated with a high risk of recurrence.

The gold standard of treatment is wide surgical excision, with safety margins of at least 5 cm and the sacrifice of a healthy anatomical barrier at depth. The occurrence of recurrences is frequently attributed to overly conservative initial procedures, underscoring the crucial importance of rigorous surgical management from the outset. However, when surgery is not possible, it is essential to resort to therapeutic alternatives such as radiotherapy and targeted therapies like imatinib, which can effectively control the disease by targeting its specific molecular characteristics. These approaches offer promising solutions for limiting local progression and improving the management of unresectable forms.

2. Case Presentation

We report the case of a 45-year-old patient, with no notable pathological history, admitted to a plastic surgery consultation for a left jugal swelling that had been progressively evolving for 15 months.

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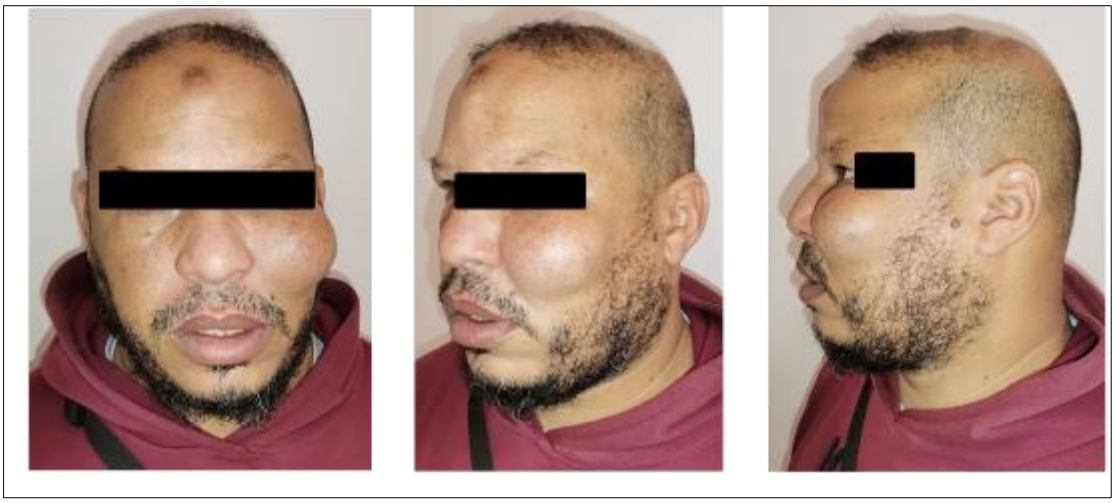


Figure 1 Patient's three-angle examination upon admission

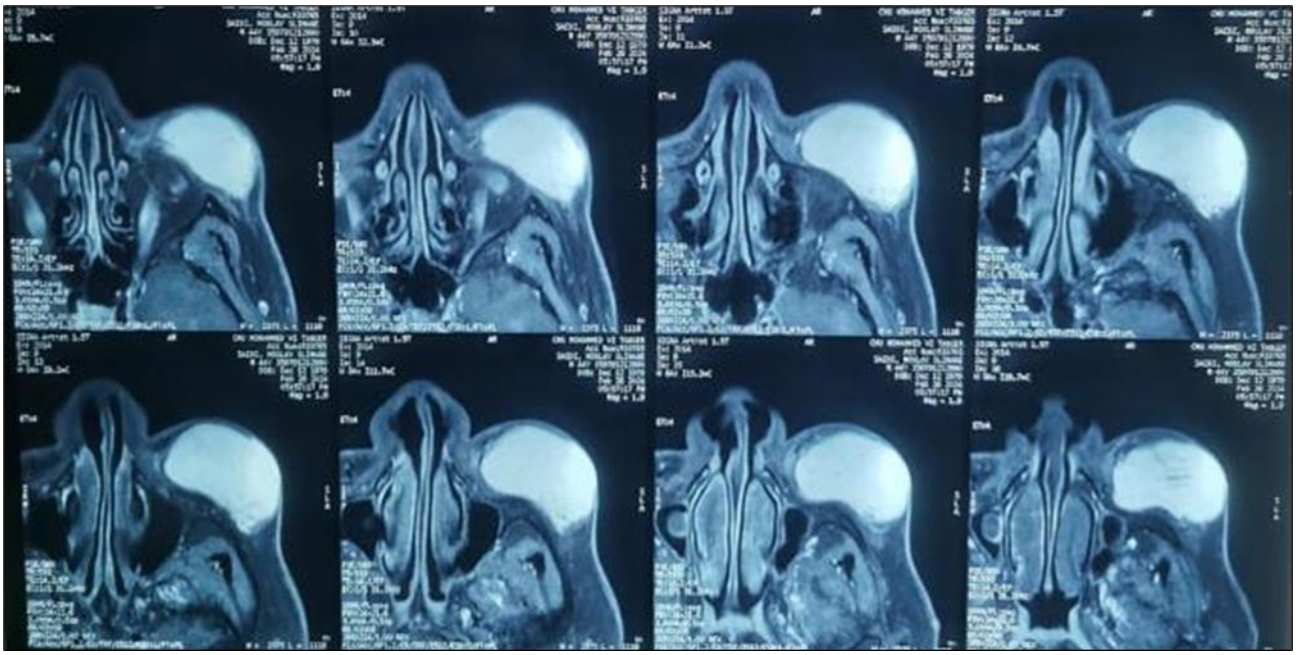
General examination revealed a conscious patient, hemodynamically and respiratorily stable.

Locally, a round mass was noted in the left jugal region, mobile in relation to the superficial and deep planes, and measuring 4 cm along its long axis.

The rest of the mucocutaneous assessment was unremarkable.

Ultrasonography of the soft tissue revealed a subcutaneous lesion with lobulated contours, hypoechoic, vascularized on Doppler, and measuring 42 × 32 × 42 mm. The lesion was in contact with the muscular and bony planes.

MRI of the facial mass showed a well-defined, oval, subcutaneous lesion in the left jugal area. It appeared isointense on T1-weighted imaging, with discrete hyperintensity on T2-weighted imaging and small serpiginous vascular-like structures crossing through it. The lesion measured 42 × 49 × 28 mm, corresponding to a volume of 28 ml, with no associated periosteal reaction. These findings were suggestive of a protuberant dermatofibrosarcoma.



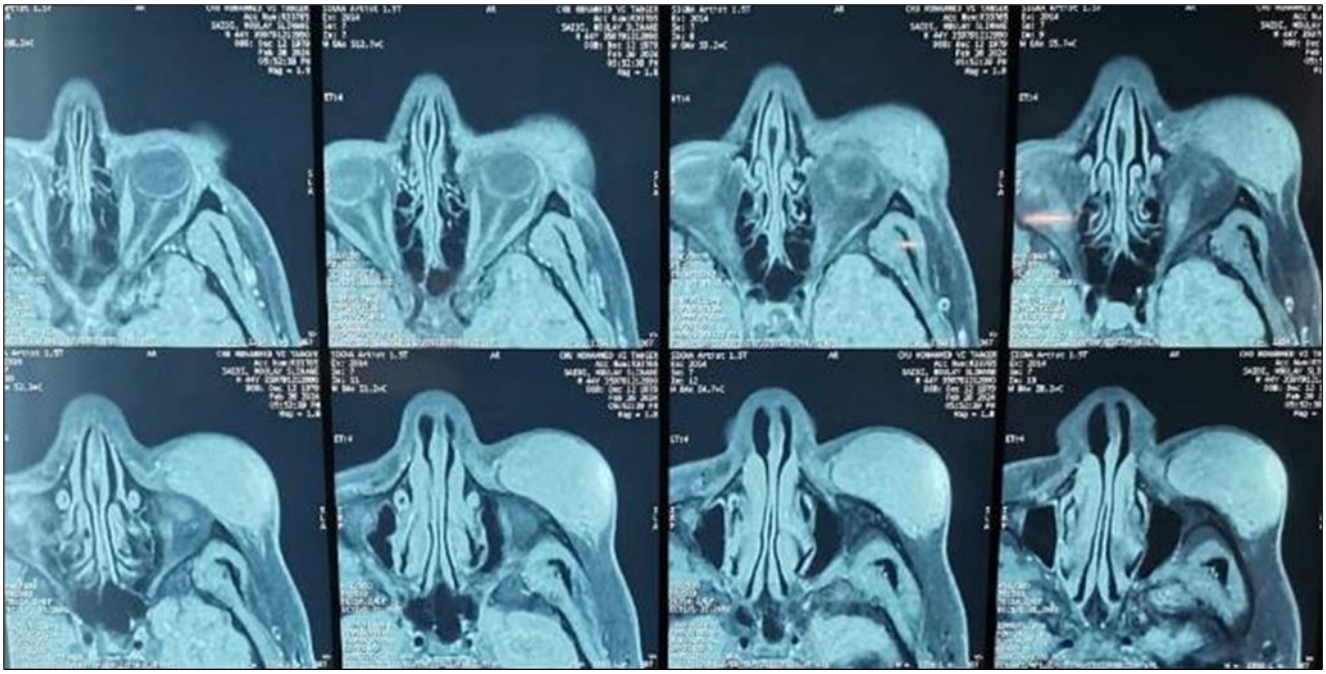


Figure 2 Facial MRI results

The patient was presented at a multidisciplinary consultation meeting (RCP) to manage a suspicious lesion. The initial decision was in favor of surgical excision with extemporaneous examination for histological confirmation.

During a preoperative consultation, the patient was informed of the potential extent of excision required in the event of a confirmed diagnosis of Darier-Ferrand dermatofibrosarcoma (DFS). Despite detailed explanations regarding the importance of wide surgical margins to reduce the risk of recurrence, the patient expressed his refusal to undergo an extensive procedure that could result in significant deformity. He clearly stated his preference for a simple excision, fully aware of the associated risks.

In the operating room, a simple excision without safety margins was performed in accordance with the patient's decision. Final pathological analysis confirmed the diagnosis of DFS.

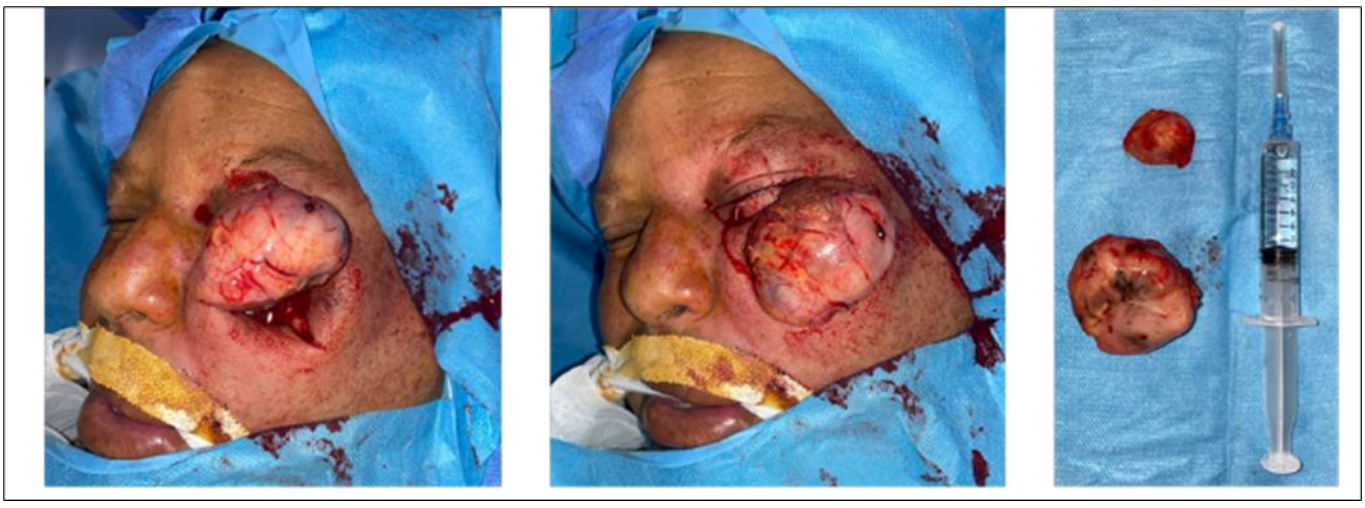


Figure 3 Intraoperative images of the surgical procedure

The case was presented at a post-operative multidisciplinary consultation (PCR). Given the absence of surgical margins and the associated risk of local recurrence, several therapeutic options were discussed. The radiotherapists recommended preserving radiotherapy as a potential option in the event of recurrence, to avoid compromising future therapeutic strategies.

The final decision was to initiate targeted therapy, accompanied by close clinical and radiological monitoring.

Figure 4 Patient 6 months after surgery

3. Discussion

Dermatofibrosarcoma is a rare but not exceptional mesenchymal tumor, accounting for between 0.1% and 1% of malignant skin tumors [1,2]. It is an infiltrative tumor that develops in the dermis and is characterized by slow growth. Its main clinical challenge lies in its very high risk of local recurrence, especially in the event of inadequate initial treatment [1,3]. However, its metastatic potential is low, contributing to a generally favorable prognosis when properly managed. Although exceptional, late transformation into a frankly malignant form, with metastases, remains possible, underlining the need for prolonged follow-up. This particular profile enables us to classify this tumor as a lesion with "intermediate malignant potential", requiring rigorous management to ensure a good prognosis [4,5,6,7].

Dermatofibrosarcoma, which is relatively common in African countries [2,8], presents several clinical challenges. Its misleading clinical appearance, often mistaken for keloid scarring, frequently leads to delayed diagnosis.

As described by several authors, this tumor can occur at any age, although its onset is most frequent in adults, generally between the ages of 20 and 50, with mean ages ranging from 28 to 47 depending on the study [2,3,9]. It is rare in children under 15, and congenital forms are exceptional [10]. The tumor shows a slight male predominance [2,6], although some studies report the opposite [11].

Although no causal relationship has been established, a history of trauma is found in 10-20% of cases [3,13,14]. Some authors even report the occurrence of DFSP after local trauma, as observed by Taylor and Helwig, who found a history of trauma in 16.5% of cases [3]. However, in our case, no specific triggering factor was identified. Other studies mention possible exogenous factors, such as burn scars, radiotherapy treatments, or lesions linked to microtrauma [15, 16, 17].

Clinically, DFSP typically appears as firm, indurated, pinkish, reddish, or purple plaques, or as dermal nodules [6,12]. It can develop on any part of the body, although certain areas are more frequently affected. According to the literature, the trunk is the predominant site, accounting for between 50 and 60% of cases. The limbs are involved in 20-30% of cases, while the head and neck account for 15-20% [3,16,18].

Histological examination is crucial for diagnosis. It reveals a dense, poorly delineated cellular proliferation invading the dermis and sometimes the hypodermis, which may explain recurrences despite wide resection margins. Immunohistochemistry confirms the diagnosis by showing positivity for CD34, a marker characteristic of DFSP, and distinguishing this tumor from other spindle cell lesions [7,12,18,19].

The surgical management of dermatofibrosarcomas is essentially based on wide surgical excision, which is considered the reference treatment due to the frequent subclinical extension of the tumor, which can be the cause of recurrence [11,18,19,20]. This treatment requires peripheral safety margins of 5 cm and the removal of a healthy anatomical barrier at depth [21,22,23]. However, this strategy may prove difficult to apply in certain complex anatomical locations, notably the face, where extensive resection could result in major functional and aesthetic deficits [20,24]. For these

reasons, some authors propose a systematic reduction of lateral margins to 3 cm [25,26]. Although these results are promising, they remain preliminary, with limited hindsight, and still require in-depth evaluation by larger studies and over longer follow-up periods.

A promising alternative to conventional wide excision is Mohs micrographic surgery (MMS) [27,28]. This technique is based on sequential horizontal resections, followed by precise extemporaneous histological control, enabling detailed histological mapping of the lateral and deep margins. Unlike the standard vertical histological section, MMS enables the identification and complete excision of pseudopodia peripheral projections of neoplastic cells that might escape conventional histological examination, thus reducing the risk of false negatives. Studies using this technique have shown that lateral excision margins of 3 cm, or even 2.5 cm, can be sufficient to achieve effective tumor control, with a particularly low overall recurrence rate, estimated at around 3% in some series [20,29,30,31].

Mohs micrographic surgery offers major advantages, particularly for difficult locations such as the head and neck, where it enables maximum preservation of uninvolved tissue. This is particularly important when conventional 5 cm resection margins cannot be respected for anatomical or aesthetic reasons. By reducing the size of the margins while ensuring rigorous histological control, MMS makes it possible to limit the extent of loss of substance, thus facilitating reconstruction while maintaining optimal tumor control [30,31].

However, despite its many advantages, Mohs micrographic surgery has certain limitations. It remains a cumbersome and demanding technique, requiring specific expertise and a suitable infrastructure. Moreover, its application is less obvious for large tumors or in cases of recurrence, where its efficacy may be more questionable due to the aggressive and difficult-to-control nature of these lesions.

The management of loss of substance resulting from surgical excision is based on a variety of reconstructive plastic surgery techniques, ranging from skin grafts to more complex microsurgical methods. In the majority of cases, reconstruction is performed after confirmation of the carcinological nature of the excision. Systematic lymph node dissection is of no interest, as DFSP presents an exceptional metastatic risk, with lymph node invasion observed in less than 1% of cases [3,6,9,32].

Radiotherapy is often considered an essential strategy in the management of recurrent lesions. Several authors emphasize its usefulness in cases of multiple recurrences, others in situations of excision with insufficient or invasive margins, or for bulky tumors and complex localizations limiting a broad surgical approach [33, 34,35]. Although these lesions are frequently described as radioresistant [3, 16, 21, 36], recent data suggest that radiotherapy can significantly reduce local recurrence and improve surgical results by enabling more conservative interventions [37,38,39]. However, chemotherapy, although associated with radiotherapy in certain palliative situations, has shown no significant efficacy in terms of overall survival, despite the various protocols tested [23, 40].

In the case of inoperable tumors, where surgery is rendered impossible due to localization, tumor size, or associated functional and aesthetic consequences, targeted therapies offer a valuable therapeutic option [41]. These approaches are based on the specific molecular features of DFSP, notably the presence, in over 90% of cases, of a chromosomal rearrangement responsible for aberrant activation of cellular signaling pathways, leading to sustained cell proliferation [42]. Imatinib mesylate, a tyrosine kinase inhibitor, has demonstrated significant clinical efficacy in the treatment of unresectable DFSP [43]. Both in vitro and in vivo studies have shown that imatinib blocks tumor cell proliferation and induces apoptosis [44]. These results led to its approval by the FDA and the European Medicines Agency for this indication. In addition, imatinib can be considered as a preoperative or adjuvant treatment after incomplete surgical resection [41,42]. These advances reinforce the importance of therapies targeting specific molecular abnormalities in the management of human malignancies.

According to the recommendations of the French National Cancer Institute, rigorous surveillance is essential after surgery, given the slow evolution and high recurrence rate of this tumor [19]. Clinical follow-up is recommended every 3 to 6 months for the first three years, when the majority of recurrences occur, and on an annual basis thereafter. This approach enables early detection of recurrences and rapid adaptation of management, thus guaranteeing better disease control.

4. Conclusion

Darier et Ferrand dermatofibrosarcoma is a rare mesenchymal tumor characterized by a high potential for local recurrence. Surgery, with wide excision margins, remains the central pillar of treatment, offering the best rates of disease control. However, in complex localizations such as the head and neck region, complete excision may be difficult

to achieve due to functional and aesthetic constraints. Inoperable cases can be controlled with targeted therapies and radiotherapy, which are therapeutic alternatives that can contribute to local disease control. The prognosis of DFSP depends on the earliness of diagnosis and the quality of initial treatment, hence the importance of prolonged clinical monitoring to prevent recurrence.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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