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Wide Local Excision for Dermatofibrosarcoma Protuberans: A Surgical Approach

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ABSTRACT

Introduction: Darier and Ferrand dermatofibrosarcoma is a locally malignant soft-tissue sarcoma with a high recurrence rate and rare metastasis, first described in 1924.

Results: This retrospective study analyzes 20 cases of chest wall dermatofibrosarcoma treated at Hassan II University Hospital in Fez from 2009 to 2023. The median patient age was 44.95 years, with a slight female predominance. Tumors varied in size and presentation, commonly appearing as multi-nodular or ulcerating masses. Histological analysis showed spindle-shaped cells with CD34 expression. Surgical treatment involved wide excision with a 5 cm safety margin, followed by reconstruction using skin grafting or directed healing.

Conclusion: Over an average follow-up of 23 months, no recurrences were observed, highlighting the importance of early diagnosis and multidisciplinary management to improve outcomes and minimize the need for complex surgical repairs.

INTRODUCTION

Dermatofibrosarcoma of Darier and Ferrand (DFS), also known as dermatofibrosarcoma protuberans (DFSP), is a rare cutaneous/subcutaneous mesenchymal tumor with an intermediate-grade malignancy [1]. Dermatofibrosarcoma of Darier and Ferrand is characterized by its indolent growth, and a notable propensity for local recurrence, whereas metastasis remains a rare occurrence. The standard treatment for Dermatofibrosarcoma of Darier and Ferrand's is wide excision. In certain cases, Mohs micrographic surgery is recommended to minimize tissue resection, especially in more visible areas such as head and neck tumors. The aim of this study was to further elucidate the clinical and pathological characteristics of Dermatofibrosarcoma of Darier and Ferrand, and to assess the cure rates of patients with primary and recurrent Dermatofibrosarcoma of Darier and Ferrand who were treated with wide local excision at our department.

MATERIALS AND METHODS

The medical records of 20 consecutive patients with Dermatofibrosarcoma of Darier and Ferrand, treated at the Department of Thoracic Surgery of Hassan II University Hospital in Fez between January 2010 and January 2023 were retrieved from our retrospective database.

Clinical and pathological variables collected included patient age, sex, presentation status (primary vs. recurrent), tumor characteristics (location, size, grade, histology), treatment information (surgery, chemotherapy, radiotherapy), date of last follow-up, recurrence, and vital status. These variables were compared to the results reported in the literature.

RESULTS

Of the 20 patients included in the study, 11 were female and 9 were male. Age at the time was 22–79 years (Of the 20 patients included in the study, 11 were female and 9 were male. The age at diagnosis ranged from 22 to 79 years (median: 44.9 years), with

the majority of patients being between 41 and 50 years old. The diagnostic delay was considerable, spanning from 6 months to 8 years. Among the cases, 12 were primary tumors and 8 were recurrent, with the latter having undergone 1 to 4 prior excisions (initially treated at other institutions). Tumor presentation was multinodular in 8 cases (39%), nodular in 5 cases (28%), as an indurated plaque in 1 case (5%), and as an ulcerobudding tumor in 6 cases (28%). Tumor size ranged from 3 cm to 20 cm, with a mean size of 7.3 cm. In all patients, the general health condition was well-preserved. The diagnosis of Dermatofibrosarcoma of Darier and Ferrand was confirmed histologically prior to surgery.

Surgical treatment involved wide local excision with a 5cm margin, including the underlying deep fascia, subjacent muscle, and overlying skin. In two cases, additional wide local excision was performed following the analysis of the operative specimen, as the initial resection margins were found to be insufficient.

Postoperatively, microscopically negative margins (R0 resection) were achieved in all our patients.Following surgery, re-epithelialization was required in 8 cases, while secondary wound closure with a skin graft or flap was performed to achieve optimal cosmetic outcomes. The clinical follow-up of the patients was updated to January 2023, with a median follow-up duration of 41.5 months; however 4 patients were lost to follow-up. Patients were regularly seen every 6 months during the initial years and subsequently annually for a minimum of five years.

TABLE I: Patient demographics and clinicopathologic features.

CHARACTERISTICS	N
OVERALL	20
GENDER	
M	9
F	11
AGE (YEARS)	
<40	7
40–60	11
+60	2
SITE:	
TRUNK	20
TUMOR:	
PRIMARY	12
RECURRENT	8
RECONSTRUCTION	
R-E	8
SG	20
RECURRENCES	
YES	0
NO UNKNOWN	16
	4

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP), a rare, low-grade mesenchymal tumor of the skin, was first described in 1924 as an invasive and progressive dermatofibroma [4]. The incidence is estimated to be 4.1 per million person-years. Dermatofibrosarcoma of Darier and Ferrand is known to be most commonly reported between 20 and 59 years, but in our series, it predominates between 41 and 50 years, with a median age of 45.7 [5]. Although recent studies have shown no gender preference in its prevalence. Dermatofibrosarcoma of Darier and Ferrand presents as a plaque-like area of cutaneous thickening that can be skin-colored, red-brown, or violaceous, with violet-red or blue margins. As it slowly enlarges over the years, it becomes a raised, firm nodule with overlying telangiectasia [6]. Size varies considerably and is generally related to the duration of evolution. Most tumors are diagnosed at a size of less than 5 cm and are adherent to the dermis. According to Vitiello et al, Dermatofibrosarcoma of Darier and Ferrand most often affects the trunk (50-60%), followed by the extremities (20-30%) and head and neck (10-15%) [7-11]. However, our study included only cases of trunk tumors.

Dermatofibrosarcoma of Darier and Ferrand is a locally aggressive tumor that rarely metastasizes. Due to its infiltrative nature, Dermatofibrosarcoma of Darier and Ferrand has a tendency to recur if clear surgical margins are not achieved, with an average recurrence rate of 43%, underscoring the importance of wide excision margins. Microscopically, Dermatofibrosarcoma of Darier and Ferrand is characterized by monomorphous spindle cells arranged in a storiform pattern, embedded in a sparse to moderately dense fibrous stroma, and occasionally extending into the subcutaneous fat. Diagnostic markers include CD34 antigen in most instances, which, although non-specific, is still the most valuable immunohistochemical marker [8-9].

According to the National Comprehensive Cancer Network (NCCN) guidelines, surgery is the primary treatment modality for Dermatofibrosarcoma of Darier and Ferrand, with the main objective is to achieve clear margins [10]. However, obtaining cancer-free margins in this type of tumor can sometimes be challenging. There is no consensus on the standard technique for its excision, and a successful surgical approach depends on a complete assessment of the margins. The National Comprehensive Cancer Network currently recommends wide local excision (WLE) with 4 cm margins, including the fascia. According to the study by Molina et al, All recurrences occur in patients who underwent wide local excision with margins of 2 cm or less. No recurrences were observed when margins of at least 3 cm were used [10-14].

Mohs micrographic surgery (MMS) is an alternative to wide local excision (WLE) for treating Dermatofibrosarcoma of Darier and Ferrand. This technique involves tangential and horizontal sectioning of the tumor, enabling close examination of deep and peripheral margins. Mohs micrographic surgery allows surgeons to identify tumor projections and guide residual tumor resection until clear margins are achieved [12]. However, there is controversy over the use of one or the other procedure. One contributing factor is the absence of randomized controlled trials and the limited availability of comparative studies.

Wide local excision (WLE) is typically preferred for lesions on the trunk and limbs, where excision is relatively straightforward. In contrast, Mohs micrographic surgery (MMS) is often employed for head and neck lesions, where anatomical constraints, as well as functional and cosmetic considerations, may necessitate its use. The Mohs micrographic surgery procedure requires a specialized surgical team, and multiple stages may be required to achieve complete tumor removal, resulting in higher costs compared to wide local excision, which is generally a more expedient procedure. Postoperative defects on the trunk and extremities can usually be closed primarily, obviating the need for subsequent reconstructive interventions [13]. Radiation therapy (RT) has been suggested as an adjuvant treatment option for cases with positive surgical margins or as a primary therapeutic modality in advanced cases where surgical intervention is not feasible. The mean follow-up period for these patients ranged from 26 to 127 months.

Javed et al. demonstrated that the local recurrence rate is known to be the highest, with up to 80% of recurrences occurring in the first 3 years after excision. Our follow-up lasted from 23 to 60 months, and no cases of recurrence were observed.

CONCLUSION

The primary goal in the management of Dermatofibrosarcoma of Darier and Ferrand is to ensure complete surgical clearance. Wide local excision (WLE) with adequate lateral and deep margins is an effective approach for preventing local recurrence. Additionally, continuous and long-term clinical surveillance is crucial for the optimal management and early detection of any potential recurrence in patients with this malignancy.

DECLARATION OF INTERESTS: The authors declare that they have no conflicts of interest related to this article.

PATIENT CONSENT: All patients gave written informed consent to collect, analyze, and publish their medical data in this study.

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