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Dermatofibrosarcoma Protuberans: A study of 10 cases

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a skin fibroblastic tumour that is locally aggressive, with a tendency for high local recurrence rate but low metastatic risk. Diagnosis is suspected clinically and confirmed by Histology. The standard treatment is complete surgical excision with wide local margins, in order to reduce the recurrence rate. Methods: Retrospective study of patients with DFSP diagnosed during 2023–2024 in University Hospital Center of Mohammed VI of Tangier. The medical records were examined to collect the following informations: age, sex, tumour location, evolution time, diameter, treatment, and follow-up. Results: 10 patients were included in this study (6 women/4 men, mean age 43.5 years). Tumors were located on the back in 6 cases, the abdomen in 2, the thorax in 1, and the face in 1 case. Surgical treatment was indicated in 9 patients (90%). recurrences were observed on 3 patients (30%). Conclusion: DFSP is a low- to intermediate-grade malignancy that commonly affects young to middle-aged individuals. The treatment of choice is surgery, however, adjuvant therapies, such as radiation and targeted therapy, should be considered for patients who are not candidates for surgical excision.

Keywords: Dermatofibrosarcoma protuberans; Skin; Wide local excision; Recurrence

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is the most common sarcoma of the skin[1], derived from dermal fibroblasts, however, they may also develop in the subcutaneous tissue[2]. Although metastasis only occurs in exceptional cases, morbidity is high due to the high local invasive capacity of the tumor and the high rate of recurrence following surgical removal. The first descriptions of this entity were made in 1924 by Darier and Ferrand[3].

Histopathologically, it is composed of uniform spindle cell fascicles growing in a storiform pattern with multiple variants and with strong and diffuse CD34 immunoreactivity. Cytogenetically, more than 90% of DFSP have a t(17; 22) (q22; q13), leading to the formation of COL1A1-PDGFB fusion transcripts[4].

The standard treatment of resectable DFSPs is complete surgical excision with either wide local excision with tumor free margins or Mohs micrographic surgery. Unresectable DFSPs are treated with radiation therapy. The aim of this article is to study the general characteristics of patients treated for this tumor

2. Material and methods

This is a retrospective study evaluating patients with Dermatofibrosarcoma protuberans diagnosed by surgical specimens from wide excisional biopsies from the Department of Plastic and Reconstructive Surgery of University Hospital Center of Mohammed VI of Tangier, during a period of 1 year, between 2023 and 2024

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Data collected from the medical records, including patients age at diagnosis, sex, tumor volume, anatomical location, modality of treatment, recurrence and follow-up were collected and analyzed.

Informed consent from patients was obtained

3. Results

In total, 10 cases consisting of 4 male and 6 female patients, with an age range of 26–63 years, and a mean age of 43.5 years. All patients presented with primary disease without evidence of metastasis. The characteristics of the DFSP patients were as follows: the anatomical locations of the lesions were as follows: the back in 6 cases, the abdomen in 2, the thorax in 1, and the face in 1 case. The evolution time at diagnosis of DFSP ranged between 4 and 96 months (median 28 months), and tumor diameter ranged between 5–18 cm (mean: 9 ± 3.3 cm). The clinical examination revealed multiple raised, erythematous, and confluent nodules in all cases, with the presence of ulceration and central necrosis in 2 cases (20%). The overall sensitivity for diagnosis was 83.8%. The false-negative initial diagnoses included schwannoma (1 case).

Nine patients were treated by wide surgical excision, aiming to achieve 3–5 cm tumor-free margins. Defects were treated with skin grafts in 8 cases and with a latissimus dorsi flap in 1 case. Adjuvant radiation was indicated for all patients.

One patient was lost to follow-up, and the rest were followed until the last date of follow-up; recurrence was observed in 2 cases, with the back being the most common anatomical location for tumor recurrence.

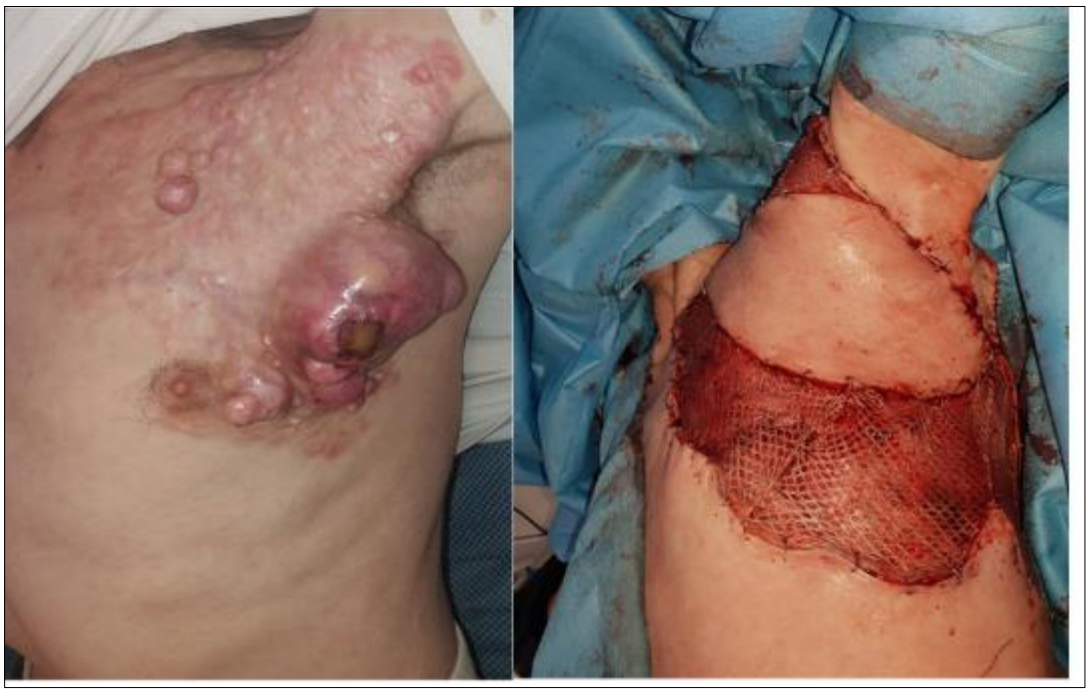

Figure 1 Clinical presentation of dermatofibrosarcoma protuberans, showing multiple raised, erythematous, and confluent nodules on the thorax. Post-excision, the defect was reconstructed using a latissimus dorsi flap and skin graft

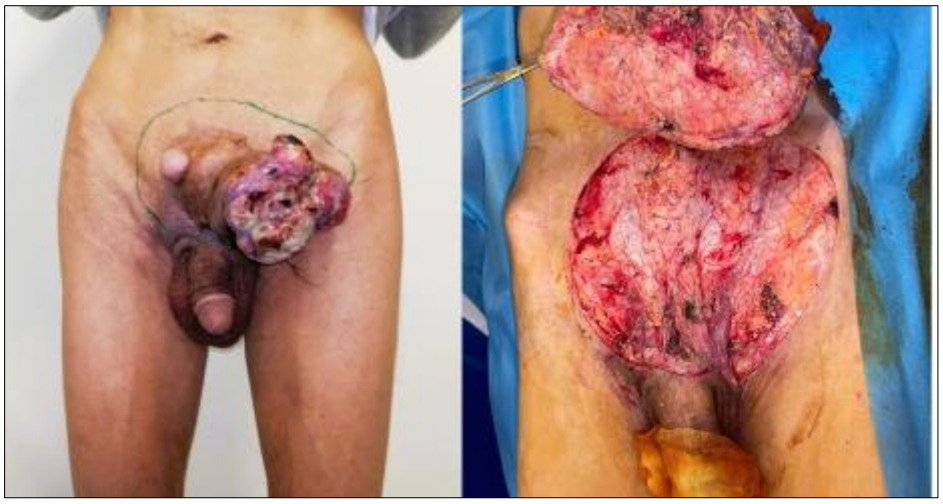

Figure 2 Dermatofibrosarcoma protuberans (DFSP) in the suprapubic region, with post-excision showing a 3–5 cm margin

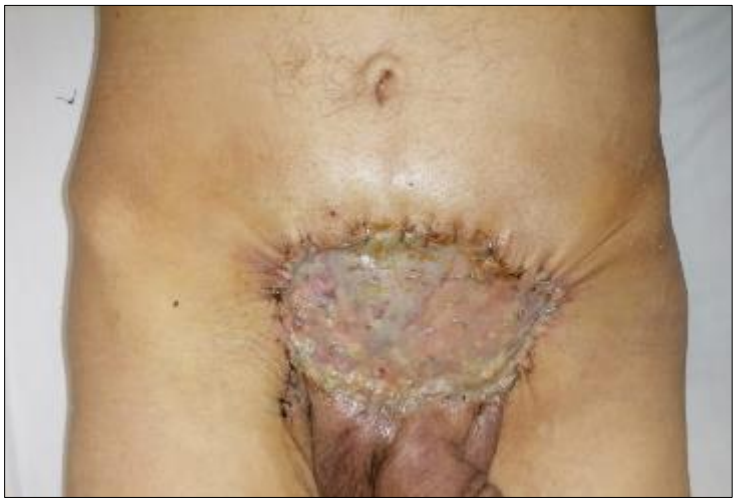

Figure 3 Reconstruction of the defect using a skin graft

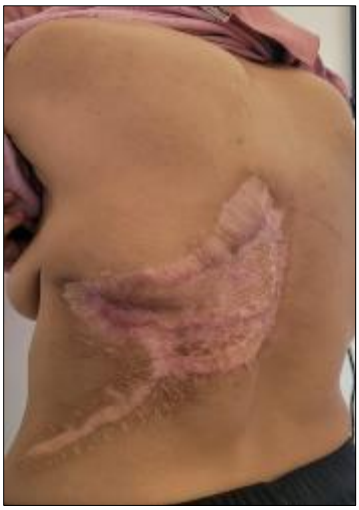

Figure 4 Recurrence of dermatofibrosarcoma protuberans (DFSP) in the back region after 4 months

4. Discussion

The reported incidence of DFSP ranges from 0.8 to 4.5 cases per million inhabitants per year [2]. Unlike most skin neoplasms, DFSP is more commonly seen in younger individuals, typically presenting in the third or fourth decade of life [1],[2]. In our series, the mean age at diagnosis was 43.5 years. Some studies report a higher incidence in males[5], [6], whereas others find it more frequent in females [7]. In our study, DFSP was more common in females, with an incidence of 60% in women compared to 40% in men.

DFSP initially presents as an asymptomatic, indurated plaque that gradually enlarges over months or even years. The lesion may be covered by skin that appears normal or displays yellowish-brown, erythematous, sclerodermiform, or atrophic changes, sometimes with telangiectasia[2]. As DFSP progresses, it becomes raised, firm, and nodular. Being a slow-growing tumor with minimal discomfort, its diagnosis is often delayed[2]. In our patient group, the median time from onset to diagnosis was 28 months.

DFSP most commonly occurs on the trunk and the proximal regions of the extremities[1], [2]. In our patient group, DFSP was predominantly located on the trunk (9 cases, 90%), with the back being the most affected area (6 cases).

DFSP is considered a low-grade sarcoma[2]. Local recurrence of DFSP has been reported to vary from 5.5% up to 60% of cases[1], [8]. In the present study, 3 recurred (30 %), most of them locally in adjacent skin.

The classical therapy of DFSP is wide surgical excision trying to achieve 2–5 cm tumour-free margins. However, several recent studies have demonstrated that Mohs micrographic surgery is associated with a much lower recurrence rate than wide local excision[9], [10], [11]. In our series; 9 patients were treated by wide surgical excision, aiming to achieve 3–5 cm tumor-free margins. Defects were treated with skin grafts in 8 cases and with a latissimus dorsi flap in 1 case.

5. Conclusion

In conclusion, in our population, DFSP is most commonly located on the trunk, particularly on the back. Wide local excision with safe margins is the preferred treatment for most cases of DFSP. Adjuvant radiotherapy is recommended for patients who are not candidates for surgical excision. Regular follow-up is advised, with surveillance every 6 months during the first five years, followed by annual follow-up to monitor for potential 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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