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# Darier-Ferrand Dermatofibrosarcoma of the Chin: A Case and Review of the Literature

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**Abstract:** Darier-Ferrand Dermatofibrosarcoma is a rare but not exceptional malignant cutaneous mesenchymal tumor, representing 0.1% of malignant skin tumors and less than 5% of adult soft tissue sarcomas. We report a case of unusual localization of a Darier-Ferrand dermatofibrosarcoma of the chin that's been developing for 2 years in 45-year-old patient. The clinical examination showed a rounded mass of firm consistency. The surgical resection was complete. Histological examination of the operative specimen confirmed the diagnosis of Darier-Ferrand dermatofibrosarcoma with healthy exeresis limits. The coverage of the loss of substance has been achieved.
**Keywords:** Dermatofibrosarcoma, Exerese margins, Reconstruction.

## INTRODUCTION

The dermatofibrosarcoma of Darier-Ferrand is a fibrous tumor of the skin with high local malignancy, progressive evolution and high potential for recidivism [1]. Described for the first time by Taylor in 1890 and then in 1924 by Darier under the name of progressive and recurrent dermatofibromas and later in 1925 Hoffman gave it the name of dermatofibrosarcoma protuberans (DFSP)[2-4].

All age groups may be concerned by this tumor, but it is more common in adults and the elderly, with a small male predominance [5]. Its sites of predilection are the trunk in 50 to 60% of cases followed by the limbs which represent 20 to 30% of the localizations, then the head and the neck which in 15 to 20% of the cases [6].

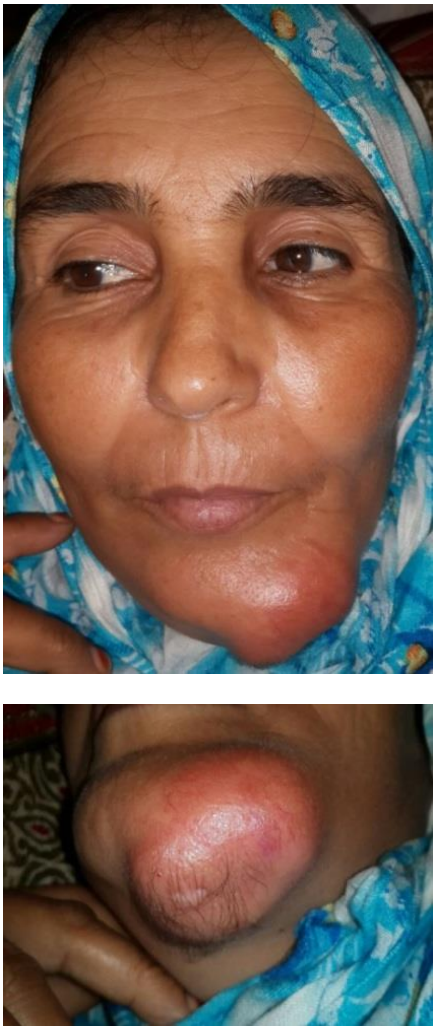
The diagnosis is often clinical and confirmed by the histological study. The use of immunohistochemical techniques may be necessary in some cases to eliminate some differential diagnoses, mainly histiocytofibroma. The treatment is surgical based on extensive and deep lesional excision. We report the case of a Darier-Ferrand dermatofibrosarcoma treated by surgery.

## CASE REPORT

The patient is a 45-year-old married woman and mother of 4 children with no particular pathological history. The beginning of the symptomatology goes back to 2 years by the installation of a chin mass gradually increasing in size, painless and non pruriginous with a good general state.

Clinical examination revealed a budding lesion on the left hemi-chin, firm, painless, with a smooth, even surface with fluctuating areas, not integral with the bone and measuring 6 x 5 cm without filling of the vestibule or ulceration of the mucosa.

The floor of the mouth was normal, there was no tooth mobility and the labio-chin sensitivity was preserved. The rest of the clinicalexamination was normal (Fig: 1).



**Fi-1: clinical aspect of the tumor**

A cervicofacial CT showed a tissue mass in the region under the chin without local or vascular infiltration. A biopsy of the mass had objectified a morphological and immunohistochemical aspect in favor of a Darier-Ferrand fibrosarcoma.

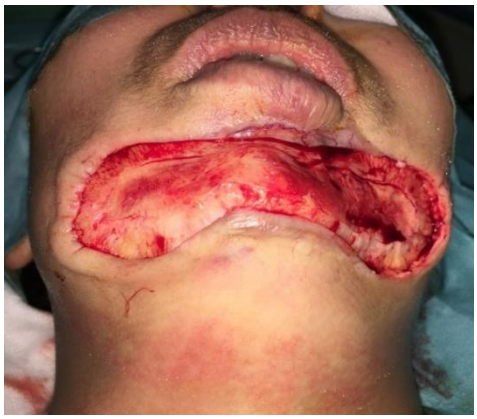


**Fig-2: appearance a few days after the tumor exeresis**

Surgical resection was complete, reaching 3cm in healthy tissue laterally preserving the lower lip and the mandibular bone (Fig: 2).

The anatomopathological study of the operative specimen showed a fusiform cell proliferation whose morphology was compatible with Darrier Ferrand dermatofibrosarcoma with healthy exeresis limits.

The follow-up was normal without any hemorrhagic or infectious complications. From the aesthetic point of view, we obtained a good directed healing of the excision site with a skin graft (Fig3).

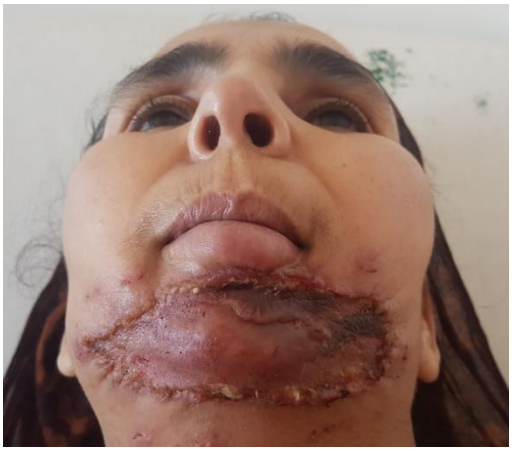


**Fig-3: Appearance 15 days after skin grafting**

## DISCUSSION

Dermatofibrosarcoma of Darier-Ferrand is a rare tumor, representing 0.1 to 1% of malignant skin tumors, but remains the most common cutaneous sarcoma[8,9].

It is characterized by its slow development, low metastatic potential and high local recurrence rate. This tumor occurs at any age with an average age of 20 to 50 years-old [5], affecting both sexes with a slight male predominance. It is more common among black people [10].

Our dermatofibrosarcoma case is unusual in its clinical appearance and chin location, it has never been published. In fact, the localizations are usually the trunk, the proximal extremities of the limbs and rarely the cephalic extremity. Clinically, DFS can take many aspects, making its clinical diagnosis difficult.

In the infiltrative stage, the lesion is an indurated plaque. At a more advanced stage (nodular stage), lesion expands after a few months to a few years to a multi-nodular mass, painless showing the appearance of a dermohypodermal plaque, dotted with nodules, white polychrome yellowish, brown or pink, and sometimes telangiectatic, variable in size, attached to the skin, mobile compared to the deep levels [11].

This two stage evolution is not always present because some forms are uni-nodular or multinodular. In the majority of cases, the lesion evolves slowly and gradually, without any functional signs or general disorders. Untreated, these lesions can become very large, or ulcerate to become painful and hemorrhagic. However, the general state of the patients remains long preserved, explaining the delayed consultation. Our observation is a good illustration of this problem.

Histologically, the microscopic appearance of Darier-Ferrand dermatofibrosarcoma was described for the first time by Taylor and Helwing in 1962. This histological description facilitated the differentiation between DFS and other spindle-shaped skin tumors.

It is a dermal proliferation infiltrating the hypodermis, organized into interlaced short bundles producing a “storiform” aspect, very suggestive of dermatofibrosarcoma in the most cellular zones. In the less dense areas, the cells are arranged in parallel or form wavy, flexuous sheets whose appearance is reminiscent of a neurofibroma. The invasion of the hypodermis is in the form of small tumor flows dispersing in “honeycomb pattern” between adipocytes whose appearance is not modified or massively dissociating to the adipocyte lobules.

Tumor proliferation is separated from the epidermis by a thin border. The tumor cells have a scanty cytoplasm and an elongated nucleus. There are no nucleo-cytoplasmic atypias. Mitoses are rare (1 to 2 mitoses per 10 fields at high magnification).

DFS is a spindle cell tumor that is sometimes difficult to differentiate from other fibrohistiocytic tumors. The use of immunohistochemistry techniques makes it possible to identify the antigens characteristic of tumor cells and thus contributes to the elimination of differential diagnoses[13].

Surgical excision is the standard treatment. It must meet certain specific rules to reduce the risk of relapses such as monobloc resection with margins of safety 3 to 5 cm wide healthy tissue, carrying a healthy barrier in depth.

However, the advent of micrographic surgery according to the Mohs technique allowed the reduction of the margin of exeresis it requires the presence of a

pathologist in the operating room in order to perform a thorough examination. This technique allows tissue saving with extensive analysis of the excisional banks, hypodermic infiltration at a distance can thus be detected and removed.

For technical reasons, we could not do it. Teams using this technique have shown that margins of lateral excision of 3 cm or even 2.5 cm are sufficient [14,15]. The frequency of recurrence is based on margins of excision: 70% for margins of 1 cm, 40% for margins of 2 cm, 10 to 20% for margins of 3cm, 5% for margins of 4cm and 1.75% of cases for margins of 5 cm [16].

The margin of excision of 5 cm remains a margin of safety for certain locations this margin cannot be respected (periorificial regions, breasts and extremities) where anatomical conditions impose a reduction of margins to 3 cm or even less. Our case illustrates this situation well.

The coverage of the loss of substance caused by the excision involves the different reconstruction techniques ranging from the direct skin grafts to the transfer of musculocutaneous flaps. In addition to surgery, radiotherapy and chemotherapy may be indicated in some cases.

The discovery of the COL1A1-PDGFB fusion gene, which results in abnormal activation of the PDGFB receptor with tyrosine kinase activity, has paved the way for clinical trials using imatinib mesylate [17]. The development of dermatofibrosarcoma of Darier-Ferrand is marked by the risk of local recurrence, frequent after excision, and by the possibility of metastases. Its surveillance is therefore particularly important.

The prognosis of DFS is characterized by its high potential for recurrence. The percentage of recidivism, meanwhile, varies according to the margins of excision. Radical initial surgical excision is therefore the essential prognostic factor, conditioning the risk of local relapse. In case of metastasis, the prognosis becomes particularly bleak, most deaths occur in the first year following the discovery of metastasis.

## CONCLUSION

The tumor of Darier-Ferrand is a particular variety of dermatofibrosarcoma whose positive diagnosis is histological. It appears at any age but especially in adulthood with a slight male predominance. Its etiopathogenesis remains unclear. DFS is characterized by its slow evolution, its rarity of metastasis and especially its strong tendency to recidivism.

**Contributions of the authors**

All the authors contributed to the medical care of the patient, as well as the writing this article they approved.

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