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DERMATOFIBROSARCOMA OF DARIER-FERRAND: A CASE REPORT OF A BREAST LOCATION

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ABSTARCT

Dermatofibrosarcoma protuberans (DFSP) or Darier et Ferrand tumor is an uncommon mesenchymal skin tumor. Mammary localization remains rare. Its diagnosis is essentially histological. It is characterized by its locally aggressive potential which requires a large surgical resection and adjuvant treatment is reserved for metastatic and recurrent forms**.**

KEYWORDS: Mammary localization remains rare.

INTRODUCTION

Darier-Ferrand dermatofibrosarcoma is a relatively rare neoplasia, described in 1924 by those who gave it its name.[1] It is a variety of fibrosarcoma of the skin with low growth rate and attenuated malignancy. It occurs mainly in children and young adults, and its cause remains unknown for the moment. However, genetic abnormalities and repeated trauma, as well as development on scars (burns), appear to be the cause of this variety of sarcoma.[2]

We report the case of a dermatofibrosarcoma of Darier Ferrand with mammary localization simulating a mammary myofibroblastoma and we will discuss through a review of the literature, the diagnostic and therapeutic difficulties imposed by this entity.

CASE REPORT

A 16-year-old female patient, without any particular pathological history, consulted for a left breast erythematous lesion evolving for 5 years.

The clinical examination revealed a patient in good general condition, apyretic, with a macular skin lesion in the upper inner quadrant of the left breast with more or less clear boundaries measuring 4 cm in length, painless and without inflammatory signs or locoregional adenopathies.

The breast ultrasound was unremarkable, apart from a subcutaneous swelling.

Anatomopathological examination after biopsy shows in the dermis and hypodermis, a massive fuso-cellular tumor infiltration made of short intertwined bundles and shows by place a storiform architecture; the cells present an oval nucleus with fine chromatin, inconspicuous nucleoli and eosinophilic cytoplasm with invisible cytoplasmic boundaries; rare mitosis figures are found; consistent with Darier-Ferrand dermato fibrosarcoma.

The extension workup does not detect any metastases.

We proceeded with surgical removal of the tumor with a safety margin of 5 cm. Histological examination of the excision specimen confirmed the diagnosis and showed that the excision margins were not affected. The patient was referred for radiotherapy.

DISCUSSION

Dermatofibrosarcoma of Darrier and Ferrand or dermatofibrosarcoma protuberans (DFSP) is a rare mesenchymal tumor, developed at the expense of the dermis and representing less than 2% of all soft tissue sarcomas, with an estimated incidence of about four cases per million.[3]

Node invasion is absent, and visceral metastases are exceptional.

The most frequent locations are the trunk and the extremities.[4] Mammary localization remains exceptional.[5]

Clinically, most patients present with a solitary, well-circumscribed, slow-growing nodule in the breast, with lesions occurring most often in postmenopausal women.[6] According to Wargotz et al.

Histologically, it is a tumor composed of a proliferation of spindle cells arranged in short irregular bundles dissociated by clusters of hyalinized collagen.

It is a cancer with a good prognosis, because despite local recurrences, the prognosis is only exceptionally vital.[2]

The treatment of cutaneous sarcoma is based on the evaluation of prognostic factors, namely local extension, histological type and grade, and general extension. The therapeutic means are surgery, which is the reference treatment, radiotherapy and targeted therapy.

For localized forms, the reference treatment is surgical with wide excision and strict histological control of the margins (Mohs' technique or modified vertical technique). In the most recent series, recurrence rates after surgery are reported to be less than 10%.[3-5]

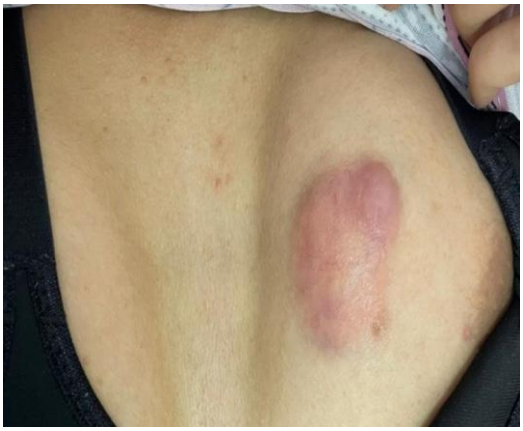
Radiotherapy remains a therapeutic modality and is indicated in case of unhealthy margins after re-excision or in case of recurrence.

Dermatofibrosarcoma protuberans is a radiosensitive disease with excellent local control after conservative surgery and radiotherapy.

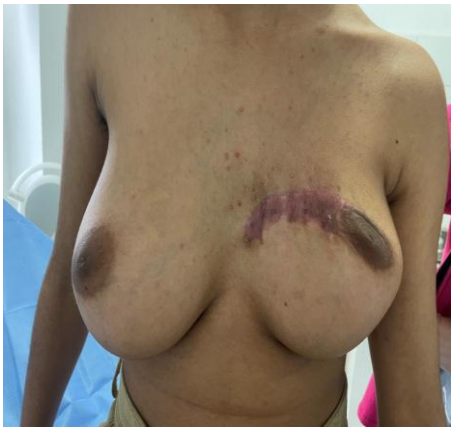
Adjuvant radiotherapy should be considered for patients with large or recurrent tumors.

Chemotherapy is reserved for metastatic stages.

Immunotherapy is based on a tyrosine kinase inhibitor: imatinib mesylate, which primarily targets the PDGFB receptor, is indicated for unresectable tumors, recurrences and metastases.[8]



Nodule sein gauche pré opératoire



Tumorectomie du nodule (post opératoire)

CONCLUSION

Dermatofibrosarcoma of Darier Ferrand of the breast type is rare.

The diagnosis can be suspected by imaging and confirmed by histology.

Treatment consists of wide surgery of the lesions with excision margins of more than 2 cm, on which the prognosis depends mainly. Breast oncoplastic surgery could be of major interest in this localization.

REFERENCES

1. Dawes KW, Hanke CW. Dermatofibrosarcoma protuberans treated with Mohs micrographic surgery: cure rates and surgical margins. Dermatol Surg, 1996; 22: 530-4.
2. Gloster HM Jr, Harris KR, Roenig RK. A comparison between Mohs micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans. J Am Acad Dermatol, 1996; 35: 82-7.
3. Flugstad DL, Wilke CP, Mc Nutt MA, Welk RA. Importance of surgical resection in the successful management of soft tissue sarcoma. Arch Surg, 1999; 134: 856-62.
4. Joucdar S, Kismoune H, Boudjemia F, Bacha D, Abed L. Darier and Ferrand dermatofibrosarcomas-retrospective analysis of 81 cases over ten years (1983-1994). Ann Chir Plast Esthet, 2001; 46: 134-40.
5. Bilger, G., Nicolaï, M., Perez, M., et al. (2021) Cas clinique: Reconstruction par abdominoplastie inversée et transposition ombilicale d'un dermatofibrosarcome épigastrique. Annales de Chirurgie Plastique Esthétique, 66: 261-267. https://doi.org/10.1016/j.anplas.2021.02.001
6. Brooke, H., Christopher, D.M., Fletcher, C.D., et al. (2016) Mammary Type Myofibroblastoma Clinicopathologic Characterization in a Series of 143 Cases. The American Journal of Surgical

Pathology, 40: 361-367. https://doi.org/10.1097/PAS.0000000000000540

7. Labhali, O., Azami, A., Tbouda, M., et al. (2017) Un myofibroblastome de type mammaire pas comme les autres: À propos d'un cas. Pan African Medical Journal, 26, Article No. 69. https://doi.org/10.11604/pamj.2017.26.69.11339
8. Castle, K.O., Guadagnolo, B.A., Tsai, C.J., et al. (2013) Dermatofibrosarcoma Protuberans: Long-Term Outcomes of 53 Patients Treated with Conservative Surgery and Radiation Therapy. International Journal of Radiation Oncology, Biology, Physics, 86: 585-590. http://www.ncbi.nlm.nih.gov/pubmed/23628134 https://doi.org/10.1016/j.ijrobp.2013.02.024
9. McArthur, G.A. (2007) Molecular Targeting of Dermatofibrosarcoma Protuberans: A New Approach to a Surgical Disease. Journal of the National Comprehensive Cancer Network, 5: 557-562. http://www.ncbi.nlm.nih.gov/pubmed/17509258 https://doi.org/10.6004/jnccn.2007.0049.