AUCTORES
Globalize your Research

Journal of Dermatology and Dermatitis
Najoua Ammar *

Open Access

Case Report

Dermatofibrosarcoma of Darier-Ferrand with an Atypical Location

Najoua Ammar *, Mariame Meziane, Laila Benzekri, Nadia Ismaili, Karima Senouci

Department of Dermatology and Venereology, CHU Ibn Sina, Mohammed V University, Rabat

***Corresponding Author:** Najoua Ammar, Department of Dermatology and Venereology, CHU Ibn Sina, Mohammed V University, Rabat

Received date: November 29, 2022; **Accepted date:** December 17, 2022; **Published date:** January 20, 2023

Citation: Najoua Ammar, Mariame Meziane, Laila Benzekri, Nadia Ismaili, Karima Senouci (2023), Dermatofibrosarcoa of Darier-Ferrand with an Atypical Location. *Dermatology and Dermatitis*. 8(1); DOI:10.31579/2578-8949/108

Copyright: **©2023** Najoua Ammar, This is an open-access article distributed under the terms of The Creative Commons. Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Dermato fibrosarcome protuberans (DFSP) is a rare malignant soft tissue tumor characterized by aggressive local béhavioral, high frequency of local recurrence with low metastatic potential. It occurs most often in patients between 20 and 50 years of age, and is preferentially located on the trunk. The standard treatment remains complete surgical excision with wide margins (>3 cm) or Mohs micrographic surgery. We report here a case of DFS unusual by its location on the leg

Keywords: darier–ferrand dermatofibrosarcoma; dermatofibrosarcoma protuberans

Introduction

Darier-Ferrand dermatofibrosarcoma or dermato fibrosarcome protuberans (DFSP) is a rare cutaneous fibrous tumor, characterized by aggressive local behavior and low metastatic potential.

It was first described in 1924 by Darier and Ferrand. It represents 0.1% of all cancers and 1.8% of all soft tissue sarcomas [1].

The most frequent tumor location is the trunk but rarely the extremities. Herein we are reporting a case of a Darier-Ferrand dermatofibrosarcoma of the leg

Observation

We present the observation of a 45-year-old patient, with no previous medical history, who was referred to us for the management of a protruding mass in the leg that had been evolving for at least 8 years and that had rapidly increased in size and become painful over the past year. No other systemic or local symptoms were reported. Physical examination revealed a 10-cm, firm, tender mass on the left leg (Figure. 1). There was no evidence of ulceration or bleeding. No adenopathy was found. The lesion was fixed to the overlying skin but mobile in the deeper tissues.

Standard radiographs showed a softs tissus mass without bony involvement.

A punch biopsy was performed, revealing a malignant mesenchymal proliferation, composed of spindle cells with little atypical and low mitotic activity arranged in storiform fascicles. Immuno histo chemistry showed a high expression of CD34 concluding to a dermatofibrosarcoma.

The patient underwent a wide local excision of the mass.

Discussion

Dermatofibrosarcoma protuberans (DFSP) was first described by Taylor en 1890, followed by Darier and Ferrand en 1924 as dermatofibroma or recurrent progressive fibrosarcoma of the skin, and finally by Hoffmann en 1925 as dermatofibrosarcoma protuberans [2].

It is a rare mesenchymal tumor that belongs to the sarcomas of low grade of malignancy with an aggressive local behavior and has a low metastatic potential. It represents 1.8% of all soft tissue sarcomas and only 0.1% of all cancers with a higher incidence in women and in the black population. It is most often seen in patients between 20 and 50 years of age and localizes preferentially on the trunk and rarely on the extremities [3].

Clinically, it begins as a simple brownish or purplish infiltrated plaque or as a small nipple nodule, usually asymptomatic. It evolves slowly over several years to a large multinodular mass with progressive evolution. There are no known predisposing risk factors for the development of DFSP, but a history of trauma has been noted [4,5].

Histologically, the cells are monomorphic, spindle-shaped, cartwheel-shaped, with few atypia and low mitotic activity. The neoplastic cells often infiltrate the surrounding adipose tissue in a honeycomb pattern. Immunohistochemistry shows diffuse and strong expression of CD34, and loss of expression of other biomarkers such as S100 protein, factor XIIIa, smooth muscle alpha actin and melanin [6].

Differential diagnoses are mainly neurofibroma, dermofibroma, hemangioma, giant cell fibroblastoma, pilomatrixoma and malignant melanoma.

The gold standard treatment for DFSP is wide local excision (WLE), with safety margins ranging from 3 to 5 cm Re-excision is recommended if the surgical margin is positive. Mohs micrographic surgery (MMS) is an alternative to wide local excision [6].

Conclusion

Darier-Ferrand dermatofibrosarcoma is an uncommon recurrent tumor that can occur in sites other than the trunk.

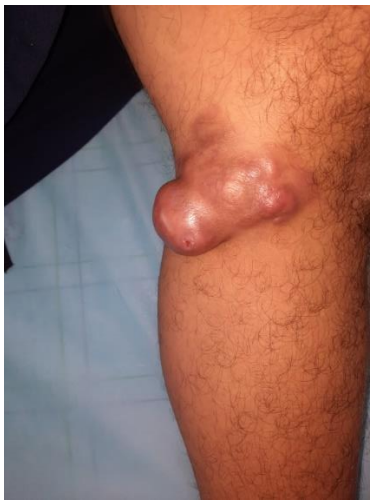

Figure 1: Dermatofibrosarcoma of Darier and Ferrand presented as a mass on the left leg

Conflict of interest

None of the authors has a conflict of interest to declare.

References

1. Darier J and Ferrand M. (1924). Drmatofibromes progressifs et recidivants ou fibrosarcomes de la peau. Ann Dermatol Syphiligr (Paris). 542-562.
2. C.A. Liang, A. Jambusaria-Pahlajani, P.S. Karia, R. Elenitsas, P.D. Zhang, C. D. Schmults, A systematic review of outcome data for dermatofibrosarcoma protuberans with and without fibrosarcomatous change, J. Am. Acad. Dermatol. 71:(4)781–786
3. Hoffman E (1924). Ueber das knollentribende fibrosarkom der haut (dermatofibrosarcoma protuberans). Dermatol Z. 43:1-28.
4. Taylor HB and Helwig EB (1962). Dermatofibrosarcoma protuberans. A study of 115 cases. Cancer 15 : 717-725.
5. C.J. McPeak, T. Cruz, A.D. (1967). Nicastri, Dermatofibrosarcoma protuberans : an analysis of 86 cases–five with metastasis, Ann. Surg. 166:(5)803–816.
6. B. Mujtaba, F. Wang, A. Taher, R. Aslam, J.E. Madewell, R. Spear, et al. (2021). Dermatofibrosarcoma protuberans pathological and imaging review, Curr. Probl. Diagn. Radiol. 50:(2)236–240.

This work is licensed under Creative Commons Attribution 4.0 License

To Submit Your Article Click Here: **Submit Manuscript**

DOI:10.31579/2578-8949/108

Ready to submit your research? Choose Auctores and benefit from:

- fast, convenient online submission
- rigorous peer review by experienced research in your field
- rapid publication on acceptance
- authors retain copyrights
- unique DOI for all articles
- immediate, unrestricted online access

At Auctores, research is always in progress.

Learn more https://auctoresonline.org/journals/dermatology-and-dermatitis