Dermatology Research and Reports
2024 Volume 3, Issue 1
DOI: 10.59657/2993-1118.brs.24.018

BioRes Scientia
Value your Research

Case Report

Open Access

Darier-Ferrand Dermatofibrosarcoma of the Beard: Diagnostic Wandering and Therapeutic Challenge in a Sub-Saharan African Patient

Mame Téné Ndiaye Diop[1,2*], Khadim Diop[3], Birame Seck[4], Assane Diop[1], Souleymane Maiga[5], Fatou Diasse Fall[2], Maodo Ndiaye[1], Moussa Diallo[1], Suzanne Oumou Niang[1], Fatimata LY[1]

[1]*Dermatology Department, Cheikh Anta Diop University, Dakar, Senegal.*
[2]*Pediatric Derm, Atology Department, Albert Royer Children Hospital.*
[3]*dermatopathology Department, Cheikh Anta Diop University, Dakar, Senegal.*
[4]*Dermatology Department, Gaston Berger of Saint Louis University, Saint-Louis, Senegal.*
[5]*Orl Department Cheikh Anta Diop University, Dakar, Senegal.*
**Corresponding author: Mame Tene Ndiaye Diop.*

Abstract

Darier-Ferrand Dermatofibrosarcoma accounts for 0.1% of malignant cutaneous tumours. This rarity explains why it is not well known and why it is misdiagnosed in our practice. Furthermore, immunohistochemistry with CD34 antigen assay is inaccessible in our region, making it difficult to rule out certain differential diagnoses such as sarcoma. A positive diagnosis is most often made on the basis of a constellation of anamnestic and histopathological evidence. We report a particular case due to its location in the beard area, which is a contributing factor for both occurrence and recurrence due to the repeated microtrauma induced by shaving, especially on black skin. Additionally, this case highlights the therapeutic challenge posed by this type of tumour, necessitating carcinological surgery with wide excision margins exceeding 2.5 cm, or ideally, Mohs micrographic surgery, which is not available in our resource-constrained settings.

Keywords: darier-ferrand dermatofibrosarcoma; beard area; Dakar; recurrent tumour

Introduction

Darier-Ferrand dermatofibrosarcoma is a dermal connective tissue tumour with spindle cells. Its histological structure is more or less similar to sarcomatous tumours, but it differs from true primary fibrosarcoma's in that it always originates in the skin and develops very slowly [1]. It represents 0.1% of malignant skin tumours [2]. We report a particular case due to its location in the beard area, which is a contributing factor of both occurrence and recurrence due to the repeated microtrauma induced by shaving, especially on black skin. In this patient, herbal therapy was used as the first therapeutic resort leading to a diagnosis wandering. Additionally, this case highlights the diagnostic challenges along with the absence of immunohistochemistry [3,4] of this type of tumour and its therapeutic approach in our practice. Indeed, managing Darier-Ferrand dermatofibrosarcoma requires carcinological surgery with wide excision margins of 2 to 3 cm. Though, Mohs micrographic surgery would be the best approach, however, it is not available in our resource-constrained settings.

Observation

We present the case of a 32-year-old male patient with a surgical history of a submental tumour 6 years previously. The surgical specimen didn't undergo a histopathological examination. 8 months after the initial surgery, the patient sought dermatology consultation due to a recurrence of the tumour. He was taking an unspecified oral and local herbal medicine to treat the tumour without experiencing relief. There were no functional signs and the patient's general condition remained unchanged. Dermatological examination revealed a large, multi-nodular mass, pink to reddish, hanging, firm, and mobile regarding the deep plane. The largest nodule of which measured 10 cm in diameter at its greatest axis (Figure 1). Remarkably, this mass occupied the entire submandibular region (Figure 2) and had been evolving for at least 5 years after the initial removal surgery. There was no evidence of adenopathy, and the physical examination of the other systems was unremarkable. However, the cutaneous histopathology revealed a malignant connective tissue

tumour proliferation composed of spindle-shaped cells with discrete atypia. These cells formed short intersecting bundles giving a storiform appearance (Figure 3). Immunohistochemistry with CD34 antigen assay was not performed due to financial constraints (the patient was of low socio-economic status) and its unavailability in our public healthcare facilities. Consequently, the diagnosis of Darier-Ferrand Dermatofibrosarcoma was accepted. The cervico-thoracic CT scan, performed to assess local and secondary tumour extension, was normal. However, cervical MRI, which was indicated to detect submental infiltration, could not be performed due to financial constraints. The management approach consisted of complete tumour removal, with 3 cm margins, followed by a flap procedure. It was performed in an ENT department.

Figure 1: sub-mandibular pinked multinodular hanging tumour

Figure 2: mass of the submandibular region located at the beard area

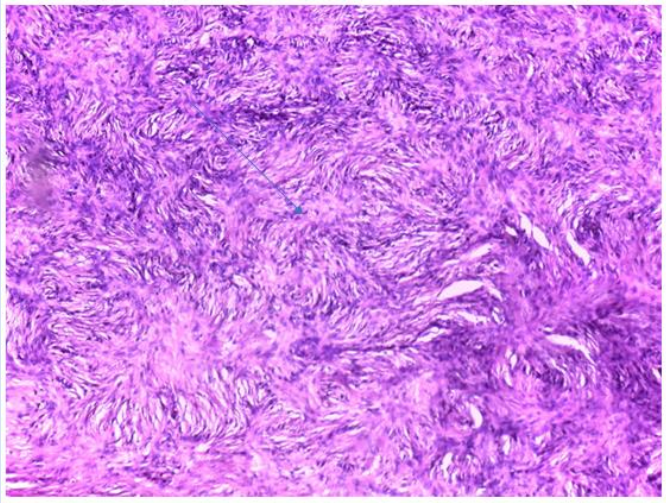

Figure 3: dermal proliferation of spindle-shaped cells with a storiform appearance

Discussion

We report a case of Darier-Ferrand dermatofibrosarcoma, which is unusual due to its location in the beard. This malignant tumour is rare, leading to diagnostic challenges, particularly in our resource-constrained settings. It can be mistaken for other conditions such as keloid, epidermised botriomycoma, or sarcoma. However, the recurrence of the tumour serves a significant suggestive pattern [5,6] and cutaneous histopathology helps to confirm the diagnosis. Unhopefully, more specific immunohistochemistry using CD34 antigen assay was not performed due to both its unaffordability for our patient, who is of low socio-economic status, and unavailability in our healthcare facilities. Darier-Ferrand dermatofibrosarcoma tumor cells strongly and uniformly express the CD34 antigen, and this could help to eliminate certain differential diagnoses, in particular sarcoma [7]. As per some authors, the mechanism of occurrence of this tumour involves exogenous factors, such as microtrauma to healthy skin [8,9]. In our patient, the tumour was located on the beard, so we can assume that microtrauma induced by shaving would be the exogenous triggering factor. This hypothesis holds even more validity considering black skin, where the hair is physiologically curved. Thus, after each shave, individuals may develop pseudo folliculitis of the beard, which is a specific pathology in black skin and may be considered a contributing factor. Since its initial description in 1924 by the author whose name it bears, it has been accepted that Darier-Ferrand dermatofibrosarcoma is a very slowly progressing cutaneous tumour, exceptionally metastatic, yet primarily exhibits local malignancy, with a strong tendency for local recurrence [3]. However, cases of metastatic dermatofibrosarcoma have been reported in the literature, with an estimated frequency of 3% according to Berbis et al [10]. The risk factors for metastasis include the tumour's prolonged evolution (over 20 years) and the frequency of recurrence after excision. Notably, the lung is the most common site for metastasis [11,12], emphasizing the importance of assessing lung involvement. In our patient, misdiagnosis was mainly due to the use of traditional medicine as a first therapeutic resort. Indeed, the efficacy of herbal therapy in the treatment of all dermatoses, commonly known as "ndoxum siti", is culturally accepted by patients in our country, Senegal. Fortunately for our patient, despite 6 years of misdiagnosis, there was no pulmonary metastasis, as confirmed by his normal chest CT scan. The therapeutic approach is surgical and consists of a monobloc excision with margins exceeding 2.5cm to avoid recurrence [13], as we did in our patient. However, some authors advocate Mohs' micrographic surgery, which is not available in our setting, as the optimal method for reducing the risk of recurrence. It consists of performing excision surgery with a simultaneous real-time anatomopathological examination during which an anatomopathologist provides proof of complete excision of the tumour directly in the operating theatre. Surgical excision continues until the surgeon reaches the healthy tissue zone [14].

References

1. Nora Naqos. (2018). Darier and Ferrand dermatofibrosarcoma of the breast. *Pan African Medical Journal*, 31:179.

2. Ahmed AF, Barzideh S, Angustiri S. (1986). Dermatofibrosarcoma protuberans: an unusual presentation. *N Y State J Med*, 86:543.
3. Shayan Khalid Ghaloo, Rahim Dhanani, Hamdan Ahmed Pasha, Mohammad Sohail Awan, Aisha Memon. (2019). Recurrent Dermatofibrosarcoma Protuberans of the Parotid: A case report and review of literature. *J Pak Med Assoc*, 69(1):113-115.
4. Kaya G. (2009). Nouvelles approches moléculaires dans le dermatofibrosarcome protuberans. *RevMed Suisse*, 211:1519-1524.
5. A. Triki, M. Aloui, M. Ghalleb, I. Jbir, Ben Naceur J. (2021). Ben Hassouna, Giant Darier–Ferrand dermatofibrosarcoma protuberans of the abdomen and pelvis: a case report. *J Med Case Rep*, 15:120.
6. Traoré SS, Zida M, Baro FT, et al. (2007). Le dermatofibrosarcome de Darier et Ferrand (DFDF). À propos de 7 cas au CHU de Ouagadougou. *Bull Soc Pathol Exot*, 100:105-106.
7. Abenoza P, Lillemoe T. (1993). CD34 and factor XIIIa in the differential diagnosis of dermatofibroma and dermatofibrosarcoma protuberans. *Am J Dermatopathol*, 15:429-434.
8. Burkhard BR, Soule EH, Chahbra H, Postel A. (1966). Dermatofibrosarcoma protuberans. Study of fifty-six cases. *Am J Surg*, 111:638-644.
9. Costa OG. (1924). Progressive reccurent dermatofibroma (Darier Ferrand). Anatomical study. *Arch Derm Syphilol (Paris)*, 5:432-454.
10. Berbis P1, Devant O, Echinard C, Le Treut YP, Dor AM, Privat Y. (1987). Metastatic Darier-Ferrand dermatofibrosarcoma. Review of the literature a propos of a case. *Ann Dermatol Venereol*, 114(10):1217-1227.
11. Grosdidier G1, Sibille P, Borrelly J, Thomas C. (1991). Pulmonary metastasis of a dermatofibrosarcoma. *Ann Chir*, 45(8):724-725.
12. Ouédraogo Nde NA, Ouédraogo NA, Bambara HA, Tiemtoré-Kambou BM, Traoré F, Zongo N. (2017). Breast and pleuropulmonary metastasis of multi recurrent scalp dermatofibrosarcoma protuberans: a case report. *J Med Case Rep*, 11(1):96.
13. Doreen Lemm, L-O Mügge, T Mentzel, K Höffken. (2009). Current treatment options in dermatofibrosarcoma protuberans J Cancer Res Clin Oncol, 35(5):653-665.
14. Hajar Ouahbi, Lamiae Amadour, Fatima zahra Elmernissi, Layla Tahiri, Taoufik Harmouch, Zineb Benbrahim. (2019). Une réponse complète d'un dermatofibrosarcome de Darrier et Ferrand localement avancé : à propos d'un cas et revue de la littérature. *Pan Afr Med J*, 32:97.

Cite this article: M.T.N. Diop, Diop K, Seck B, Diop A, Maiga S, et al. (2024). Darier-Ferrand Dermatofibrosarcoma of the Beard; Diagnostic Wandering and Therapeutic Challenge in a Sub-Saharan African Patient. *Dermatology Research and Reports*, BioRes Scientia Publishers. 3(1):1-4. DOI: 10.59657/2993-1118.brs.24.018

Article History: **Received:** May 06, 2024 | **Accepted:** May 20, 2024 | **Published:** June 05, 2024