

Localization on the palmar face of the hand of a Darier-Ferrand dermatofibrosarcoma. About a case

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Abstract

Darier-Ferrand dermatofibrosarcoma is a rare but not exceptional malignant mesenchymal skin tumor, representing 0.1 % of malignant skin tumors. It is characterized by high recurrence, slow growth, and low metastatic potential. Although several clinical cases of an unusual variant of Darier-Ferrand dermatofibrosarcoma have been reported in the literature, localization on the palmar face of the hand is not common. We report a case of Darier and Ferrand dermatofibrosarcoma at the level of the fourth commissure of the palm of the left hand in a 43-year-old young Malagasy adult treated by a large surgical excision with a healthy margin of two centimeters associated with adjuvant chemotherapy with Imatinib.

Key words: Darier-Ferrand Dermatofibrosarcoma, Palmar Face & Hand

Introduction

Darier-Ferrand dermatofibrosarcoma (DFS) is a rare but not exceptional malignant mesenchymal skin tumor, representing 0.1% of malignant skin tumors (Triki et al., 2021). It is characterized by high recurrence, slow growth, and low metastatic potential and occurs mainly in adults at any age, but mostly between their second and fifth decades of life (Al Barwani et al., 2016). The diagnosis is confirmed by histopathological examination and treatment involves surgery with wide margins of three to five centimeters (cm) to reduce the rate of recurrence (El Kacemi et al., 2014). Although several clinical cases of an unusual variant of DFS have been reported in the literature, localization on the palmar face of the hand is not common. We report a case of Darier-Ferrand dermatofibrosarcoma of the palm of the left hand in a young Malagasy adult.

Case Report

It was a 43-year-old man, driver, without medical or surgical history, seen in consultation at the Joseph Ravoahangy Andrianavalona University Hospital Center (CHU-JRA) Antananarivo Madagascar for a tumor of the palmar face of the left hand evolving since 18 months. On examination, the patient was in good general condition, presented a multinodular tumor, mobile and hard on palpation, not adhering to the deep level, protruding, itchy, non-painful, covered with thinned more or less ulcerated skin, measuring 5, 8 cm in diameter, located at the level of the fourth commissure of the palm of the left hand (Figure 1), the lymph node areas were free and the rest of the clinical examination normal. The complete blood count, liver tests, and kidney tests were normal, the x-ray of the left hand did not show any bone damage.

Histopathological examination of a first biopsy performed confirmed the diagnosis of Darier-Ferrand dermatofibrosarcoma.



Figure 1. Clinical Appearance of the Tumor

According to a multidisciplinary consultation meeting, medical treatment with Imatinib (GLIVEC®) at a dose of 800 mg per day in two divided doses was initiated. The response to medical treatment at the two-month follow-up was satisfactory with a reduction in tumor volume from 5.8 cm to 4.3 cm in diameter. Total tumor excision was performed with a healthy margin of two centimeters; total skin autografting was performed from an abdominal skin graft. The surgical specimen was sent for histopathological examination (Figure 2).



Figure 2. a) Surgical excision, b) The surgical specimen, c) Appearance after tumor excision d) Total skin autograft.

The postoperative follow-up was simple, adjuvant chemotherapy with Imatinib was continued until the sixth postoperative month at a dose of 600 mg per day in two divided doses. A periodic checkup every six months up to three years was done after chemotherapy. At three years postoperative follow-up, the patient was in complete remission but he presented a retractile sequela of the fifth finger of the left hand (Figure 3).



Figure 3. Evolution at three years postoperative.

Results and Discussion

Darier-Ferrand dermatofibrosarcoma was first described by Darier and Ferrand in 1924, it appears at any age and generally affects young adults between the ages of 20 and 50. It is rarely metastatic but at high risk of local recurrence after tumor excision (El Kacemi et al., 2014). It is a rare tumor of the dermis of mesenchymal origin which accounts for 1 % of malignant soft tissue tumors and 0,1 % of all other malignant tumors (Sevim et al, 2019). The cause remains unknown, but some author reports that genetic abnormalities, repeated trauma as well as a burn seem to be at the origin of DFSP (Flugstad et al., 1999). The most frequently affected site is the trunk, followed by the proximal extremities and scalp. The head, neck, and genitals are rarely involved (Malkud et al., 2017). Its localization in the palmar face of the hand is rarely published in the literature.

Clinically, the appearance of the tumor depends on the stage of the disease. Initially, DFS appears as a painless, pink to bluish red trophic and/or sclera skin mass that develops into a lumpy tumor, over time to a protruding, ulcerative hemorrhagic tumor. It is a mobile tumor on palpation and does not adhere to underlying tissues, attachment to deeper structures such as fascia and muscle can be found in advanced cases (Elamrani et al., 2014).

Regarding imaging, due to its rarity, the literature has only shown a few studies that discussed imaging results. Al Barwani AS et al. in 2016 did a study by examining the literature published over the past 40 years, they did not find that two studies that examined the imaging appearance of this lesion, another factor explaining the limited information obtained on DFS imaging results is its superficial location in most cases (Al Barwani et al, 2016). A very recent publication by Kim on breast DFS reported that ultrasound revealed an oval-shaped hypoechoic mass with a hyperechoic edge in the subcutaneous layer with slightly increased vascularity and several microcalcifications and imaging. A magnetic resonance (MRI) scan showed a subcutaneous mass with well-defined margins and irregular lobulations (Kim, 2021). Ouahbi H et al. had done a cranial CT scan on the localization of DFS in the scalp, which revealed a large tumor process developing in the soft parts in the subcutaneous area without bone lysis (Ouahbi et al., 2019).

On histopathological examination, the tumor is made up of a dense, poorly defined, non-encapsulated cell proliferation occupying the dermis. It sends fine extensions sometimes very deep into the hypodermis, which would explain the occurrence of recurrence even with wide resection margins, the epidermis is respected, the cells are elongated, spindle-shaped, with more or less abundant cytoplasm, the oval nucleus, regular, mitoses are variable with rare

atypia, the stroma is variable from one area to another (Hammas et al., 2014). In doubtful cases, Muhamed et al. had published that immunohistochemistry makes it possible to distinguish DFSP from other spindle cell tumors, it shows an intense and diffuse positivity of CD34 in 90% of cases and negativity of actin, of desmin, and the S-100 protein (Muhammed et al., 2018).

The differential diagnosis are recurrent dermatofibroma, hypertrophic scars, keloid, skin manifestations of myofibroblastoma, metaplastic carcinoma (Chan et al., 2014),

The treatment of choice is a large surgical resection ensuring healthy margins of three to five centimeters (Chan et al., 2014), the local recurrence rate tends to decrease with increasing surgical margins but a recurrence rate of 20 to 50% in cases of incomplete resection (Al Tarakji et al., 2015). Some guidelines recommend radiotherapy in the event of incomplete and unresectable tumor resection or multiple recurrences (Huis in 't Veld et al., 2020). Adjuvant radiotherapy can effectively reduce the rate of local recurrence (Suna et al, 2000). For adjuvant chemotherapy, Imatinib has been shown to successfully inhibit the growth and survival of DFSP cells in vitro (Huis in 't Veld et al., 2020). Imatinib is one of the major targeted therapies of DFSP, its place in the management of this pathology derives directly from its mechanism of action (Pagès et al., 2013). Hajar O et al. reported that the use of Imatinib in a patient with inoperable DFSP was dramatic (Ouahbi et al., 2019). The National Comprehensive Cancer Network (NCCN) guideline recommends considering Imatinib in unresectable PSFPs due to multiple resections or unacceptable functional or aesthetic results when a new resection is performed (Huis in 't Veld et al., 2020). In our case, adjuvant chemotherapy was performed until the sixth postoperative month because the patient had already started chemotherapy two months before the surgical excision and we were also limited to a margin of two centimeters for the functional result and aesthetics of the hand afterward.

Concerning the evolution, and prognosis, even if DFS metastases are rare, it is locally aggressive and local recurrence after resection is observed in 50 to 70% of patients. For this reason, patients should be observed periodically after surgery for a long time. Patients with complete resection have an excellent prognosis with a five-year survival rate of 99% (Kim, 2021). In our case, even if the tumor excision margin was limited, with the continuation of adjuvant chemotherapy with Imatinib until the sixth post-operative month, no recurrence was detected during the monitoring. At the three-year follow-up, the patient was in complete remission with the retractile complication of the fifth finger.

Conclusion

Darier-Ferrand dermatofibrosarcoma is a rare but not exceptional malignant skin mesenchymal tumor, localization in the palm is not common in the literature. Its diagnosis is made after carrying out a histopathological examination, immunohistochemistry is essential to rule out differential diagnoses. A large surgical excision ensuring healthy margins of three to five centimeters is the treatment of choice. Due to the frequent recurrence of this tumor, periodic follow-up after surgery is necessary .

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