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Recurrent Dermatofibrosarcoma Protuberant of the Hand: A Case Report

Toky Rakotoarivo[1], Malala Razakanaivo[2], Norosoa Randriamaroson[2], Clairette Raharisolo[3], Jean Claude Razafimahandry[4], Florine Rafaramino[2]

[1]Department of Oncology, University Hospital Tanambao, Antsiranana, Madagascar
[2]Department of Radiotherapy, University Hospital Joseph Ravoahangy Andrianavalona, Antananarivo, Madagascar
[3]Department of Anatomy Pathology, Institut Pasteur of Madagascar, Antananarivo, Madagascar
[4]Department of Traumatology and Orthopedics, University Hospital Joseph Ravoahangy Andrianavalona, Antananarivo, Madagascar
Email: rakotoarivotoky@gmail.com

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Abstract

Dermatofibrosarcoma is a rare, slow growing tumor with a tendency to local recurrence. The treatment is mainly surgical. In Madagascar there is no specialized center for the management of soft tissue sarcomas. It is in this context that we report a case of a 45-year-old man, driver, presenting with recurrent dermatofibrosarcoma of the left hand. The diagnosis was confirmed by the positivity of the CD34 marker. Extensive local excision surgery was performed with the patient's consent. After free years of treatment, there is no tumor recurrence.

Keywords

Dermatofibrosarcoma, Hand, Wide Surgery, Recurrence

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a low to intermediate grade soft tissue mesenchymal tumor arising from the dermis of the skin. It is a rare tumor representing 0.1% of malignant skin tumors and less than 5% of all soft tissue sarcomas [1]. The DFSP is of very slow evolution; metastases are rarely seen in 3% to 5% of cases. Recurrence is essentially local and can reach up to 50% of cases [1]. We report a case of a 42-year-old man with recurrent dermatofibrosarcoma of the left hand requiring wide surgical resection.

2. Case Report

A 42-year-old man, driver consulted for a pink lesion of the palm of the hand in

June 2017. The patient has no history of skin tumor. He described a painless, non-pruritic lesion that had been evolving for 3 months. On physical examination, there was an ulcero-budding swelling, pink, indurated, circumscribed 6 cm × 5 cm occupying half of the palm of the left hand (Figure 1). This lesion was mobile relative to the deep plane, painless on palpation. The rest of the exam was unremarkable. The X-ray of the hand did not show any bone lesions. The extension assessment including a thoraco-abdomino-pelvic CT scan did not show any secondary localization. Excisional surgery was performed. The anatomopathological result of the surgical specimen was in favor of a myxoid-type DFSP. Immunohistochemical examination confirmed the diagnosis by the positivity of the CD34 marker (Figure 2). At 1 year after surgery, there was a reappearance of swelling at the level of the interphalangeal space of the fourth and fifth fingers of the left hand (Figure 3). Extensive local excision surgery was performed with the patient's consent. At free years after surgery, there was no sign of tumor recurrence.

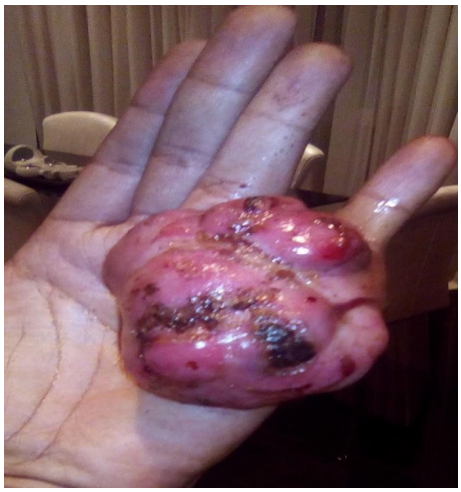

Figure 1. Nodular and ulcerated lesion on palm of the hand, 6 × 5 cm.

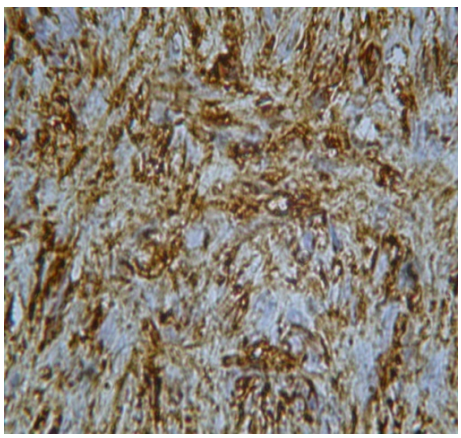

Figure 2. CD34 stain showing strong and diffuse staining in tumor cells.

Figure 3. Tumor recurrence 1 year after surgery.

3. Discussion

DFSP is a locally aggressive tumor. It most frequently occurs in young and middle-aged patients [2]. Most DFSPs are localized on the trunk (40% - 50%), followed by proximal extremities (30% - 40%), and then the head and neck (10% - 15%) [3]. Infrequently documented cases have been reported on the hand [4]. The reported tumor sizes vary in a wide range from 0.5 to 10 cm in diameter, with a mean of 2 - 3.5 cm [5]. DFSP usually develops as purple, pink, and reddish-brown plaques on the surface of the skin and gradually becomes multiple protuberant nodules [6]. The tumor is mobile relative to the deep plane and painless [1]. For our patient, the tumefaction was ulcero-budding, mobile pink in the deep plane.

Histologically, DFSP is characterized by a dense collection of bland cells with spindle nuclei arranged in irregular, interwoven fascicles in the dermis that often have a storiform appearance [1]. Immunohistochemistry was diagnostic of DFSP-positive for CD 34 and vimentin and negative for S-100 ans smooth muscle actin [7]. In our case, tumor cells were positive for CD34.

The pathogenesis of DFSP is commonly thought to be related to chromosomal fusions secondary to translocations on chromosomes 17 and 22 [8]. More than 90% of patients diagnosed with DFSP carry t(17; 22) (q22; q13) translocation that results in upregulation of platelet-derived growth factor (PDGF) expression, leading to tumor formation. This translocation manifests as a fusion of the type 1 collagen alpha 1 gene (COL1A1) and the platelet-derived growth factor-β polypeptide gene (PDGFB) on chromosome 17, which ultimately leads to the overproduction of PDGF [9].

The prognosis of dermatofibrosarcoma depends on the quality of the first surgery, is characterized by its high potential for local recurrence. The standard treatment is complete surgical excision [10]. Wide local excision, Mohs micrographic technique and amputation have been established as the treatment modalities. Wide local excision requires wide and deep excision of the tumor with at

least 2 to 3 cm free margins from the periphery of the tumor [11]. For the Mohs micrographic technique, the surgery was done in horizontal and serial sections which makes it possible to visualize the entire resection margin and precisely locate all residual tumor areas, then excise these regions until healthy tissue is obtained [11].

For difficult anatomical locations such as the hand as in our case, the Mohs technique is the most indicated. In Madagascar the current conditions do not allow the realization by the lack of technical platforms and the unavailability of anatomo-pathologist dedicated to this technique. Our patient had undergone radical surgery with macroscopically tumor-free resection margins. At free years after surgery, there is no local recurrence. Radiation therapy can be used as adjuvant therapy in cases with positive margins or with recurrent disease, and as palliative radiation therapy for incurable disease [12]. The targeted drug imatinib can be used in patients with inoperable primary tumors, locally recurrent and metastatic DFSP patients, and as a new neoadjuvant drug for preoperative reduction of tumor volume [13]. In addition, studies have shown that PD-1 monoclonal antibodies and CDK4/6 inhibitors may be used as new targeted drugs [14].

4. Conclusion

The prognosis of a DFSP is linked to the quality of the first excision. Surgery should be extensive to decrease the risk of recurrence. The establishment of a specialized medical and surgical center could improve the quality of care and the realization of one of the Mohs techniques.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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