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Darier and Ferrand dermatofibrosarcoma: A case report of an unusual presentation and literature review

Prosper Nsengiyumva, Taha Kabbaj, Mohamed Anass Majbar, Amine Benkabbou, Amine Souadka, Raouf Mohsine

ABSTRACT

Dermatofibrosarcoma is a rare fibrous tumor of the skin. Its evolution is slow and indolent with a high potential for recurrence. We report a case of Darier and Ferrand dermatofibrosarcoma that had progressed in a 56-year-old female patient from childhood. Clinical examination revealed an indurated, round, budding, ulcerative mass located to the left hypochondrium. This mass was painful, hard, and mobile at deep levels. Computed tomography of the thorax, abdomen, and pelvis showed no lesions other than a mass in the left hypochondrium. Immunohistochemical studies of previously performed biopsies were unremarkable. A histologic examination of the surgical specimen confirmed the diagnosis. The further course after 12 months was uneventful.

Keywords: Dermatofibrosarcoma, Difficulties in diagnosis and treatment, Slow and indolent course

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Prosper Nsengiyumva[1], Taha Kabbaj[1], Mohamed Anass Majbar[1], Amine Benkabbou[1], Amine Souadka[1], Raouf Mohsine[1]

Affiliation: [1]Department of Digestive Oncology Surgery, Mohammed V University of Rabat, Ibn Sina University Hospital Centre, National Institute of Oncology, Rabat, Morocco.

Corresponding Author: Prosper Nsengiyumva, Department of Digestive Oncology Surgery, Mohamed V University of Rabat, Ibn Sina University Hospital Centre, National Institute of Oncology, Rabat, Morocco; Email: nspmedecine@gmail.com

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INTRODUCTION

Dermatofibrosarcoma (DFS) is a rare fibrous skin tumor that accounts for 0.1% of malignant skin tumors and 2–6% of all soft tissue sarcomas [1]. Histologically confirmed diagnosis is difficult and often delayed by etiologic, evolutionary, clinical, and topographic polymorphism. The specificity and severity of the disease are related to its local aggressiveness and a high potential for recurrence. Initial treatment must follow the strict rules that apply to any treatment of this pathology. Before any surgical treatment, it would be advisable to undertake an approach to prevent recurrence. Thus, the realization of biopsies or the presence of a pathologist in the operating room to conduct an extemporaneous analysis of the specimen allows a wide excision of the lesion and therefore avoids recurrence [2]. This lesion continues to pose several problems related to its evolution. It is a lesion that can evolve over a long period without causing any health concern to the patient. However, at a certain stage the tumor becomes painful; hence, patients consult late. The characteristics of DFS are not recognized by the vast majority of hospital staff and the misleading clinical appearance in combination with other challenges mentioned above are often responsible for a delay in diagnosis. To try to answer these questions, this paper proposes an analysis of a case in conjunction with the literature, in which the authors share their experience and report the various epidemiological, clinical, therapeutic, and prognostic features of this rare tumor.

CASE REPORT

A 56-year-old woman, married, mother of three children, hypertensive, consulted us for ulcerative

swelling located to the left hypochondrium that had appeared from childhood. The patient had a body mass index (BMI) of 22.86 kg/m^2. Clinical examination revealed a budding lesion of the left hypochondrium that was roundish, indurated, necrotic-ulcerative, painful, mobile concerning the deep levels, and hard (Figure 1).

Immunohistochemical studies of the original biopsies taken were inconclusive. Thoraco-abdominopelvic CT scan revealed a budding mass of 7.2×5.6 cm in the left hypochondrium with no secondary location (Figure 2).

In the tumor board meeting (TBM), surgical excision of the lesion was decided and then performed. Macroscopically, the surgical specimen was weighted at 250 g and measured at 12×7×7 cm (Figure 3).

From a microscopic point of view, the various sections of the tumor showed dermal and subcutaneous tumor proliferation with dissociated adipocytes of moderate cell density and storiform architecture. They consisted of spindle-shaped, monomorphic cells with abundant eosinophilic cytoplasm, and ovoid, elongated nuclei that were slightly atypical. Mitotic divisions were rare, and the stroma was fibrous. Resection margins were healthy, increasing laterally to 4 cm and in-depth to 1.1 cm. Immunohistochemical studies revealed positive staining of tumor cells with anti-CD34 antibodies and a lack of labeling of tumor cells with anti-AE1/AE3, anti-EMA, anti-smooth muscle actin, anti-desmin, and anti-PS100 antibodies. Thus, it was a morphological aspect and immunohistochemical profile of dermatofibrosarcoma according to Darier and Ferrand. A follow-up period of 12 months was uneventful (Figure 4).

Figure 1: Lesion image before surgery.

Figure 2: CT scan image showing budding cutaneous–subcutaneous mass of the left hypochondrium.

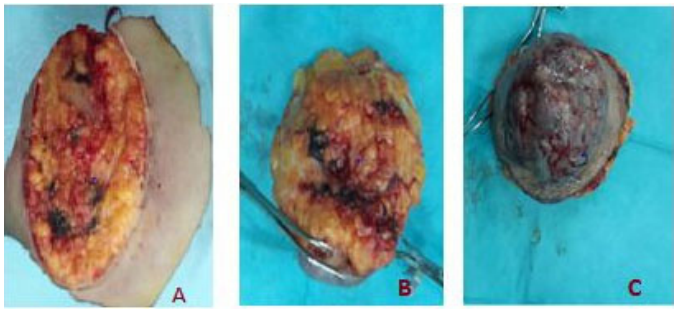

Figure 3: Per operative image after removal the mass (A) and surgical specimen image (B and C).

Figure 4: Scar image 12 months after surgery.

DISCUSSION

Dermatofibrosarcoma occurs preferentially in young adults between 20 and 50 years. There is no clear gender predominance. Regarding the origin of patients, the data are contradictory: Taylor counts a majority of Caucasian patients in a population of American military personnel, whereas in a retrospective American multicenter study, patients of black ethnicity are almost twice as likely to be affected as whites. This tumor may exceptionally occur in children (1.5%) or even rarely be congenital (3%) [3]. The tumor is commonly located on the legs (47% of cases), extremities (38% of cases), and head and neck (14% of cases). This is a single, slowly developing tumor, over several years, and causes little concern to the patient. These features often lead to diagnostic and therapeutic delays that can last for years. The conventionally described protuberant form corresponds to an advanced stage of the tumor. It is a firm, multinodular mass that is superficially fixed to the skin but mobile concerning the underlying planes. The tumor is not painful unless ulcerated, and its coloration is variable. Patients often seek medical attention when the tumor has suddenly enlarged. Even in these cases, the patient's general health is well preserved. The size of the tumor is variable, ranging from 0.5 to 12 cm. In a series of 134 cases, the following distribution was observed: Size of 15 cm: 84%; from 5 to 10 cm: 13%; 10 cm: 3%. Cases of "monstrous tumors" weighing up to 6.5 or even 7 kg or up to 25 cm in diameter have been described [4]. In 43% of cases, the so-called multinodular form may develop after a latency period of several months to several years (on average 7.6 years) and from a flat lesion designated as non-protrusive [5].

The diversity of clinical forms is another cause of delay in diagnosis. It should also be noted that the protruding and nonprotruding forms share the same histologic, immunohistochemical, and cytogenetic features.

According to some authors, several exogenous factors favor the occurrence of this disease, such as burn scars, vaccinations, radiotherapy, traumatized nevi, syphilitic lesions, microtraumas on healthy skin, and lesions of iatrogenic or occupational keratoses.

While others have described molecular pathogenesis, two types of karyotype abnormalities are found in 95% of DFS [2]. In our case, we found no precipitating factor (trauma, scar, etc.) as described in the literature. Therefore, oncogenetic monitoring should be performed in search of molecular etiology.

Darier and Ferrand dermatofibrosarcoma is a dermal connective tissue tumor with spindle cells. Unlike tumors in subcutaneous tissues, DFS lies close to the overlying skin and may fuse with it. Normally, DFSs do not adhere to the underlying structures of the epidermis, and most DFSs are superficial and less than 5 cm in size at the time of diagnosis. Tumor progression ranges from months to years and in some cases may extend over decades [6]. Some differential diagnoses should be considered such as lipomas, deep epidermal cysts, scarring or hypertrophic keloids, dermatofibromas, nodular fasciitis, and insect bites. Dermatofibrosarcoma is a malignant tumor that metastasizes in only 1–4% of cases. In exceptional cases and at a very late stage, it is an overtly metastatic sarcomatous transformation [7]. A positive diagnosis of DFS is confirmed by histologic and immunohistochemical studies of the needle or incisional biopsies. The role of fine needle aspiration has been demonstrated in cases of disease recurrence. Initial biopsies require the collection of larger samples that can show the histologic architecture of the tumor. In our case, it was difficult for us to make a definitive histologic diagnosis, probably because the sample was not large enough. Therefore, we would like to suggest that when the diagnosis of a skin tumor is uncertain, it is essential to confirm the histologic nature, as this will influence surgical therapy. Although routine imaging is not required, magnetic resonance imaging (MRI) can help assess the local extent of the tumor and may prove to be an important step in surgical planning for larger tumors. As with many other soft tissue tumors, T1-weighted images show a weak signal, whereas T2-weighted images show a higher signal [8].

Surgery is the only therapeutic method that is effective in eliminating tumor and preventing recurrence. The excision must comply with certain precise rules: one-piece excision with wide safety margins of 3–5 cm of healthy tissue, and the wearing of a healthy barrier at depth. If the resection is not one-piece, there is a risk of seeding into the surgical site. The Mohs technique is most appropriate and requires the presence of a pathologist in the operating room to carefully examine the fresh surgical specimen. The resection is performed until sections without tumor cells are obtained. This contributes to a significant prolongation of the surgical time. In our case, an immunohistochemical examination of the original biopsies taken was not helpful. For this reason, we could not perform Mohs surgery, which would have been the best surgical treatment method. Several teams who used the Mohs technique with extemporaneous examination found that margins of 3 cm were sufficient to remove all tumor cells. Other authors even reported excision margins of 2.5 cm without recurrence. The frequency of recurrence depends on the resection margins: 70% for margins of 1 cm, 40% for margins of 2 cm, 10–20% for margins of 3 cm, 5% for margins of 4 cm, and 1.75% for cases for margins of 5 cm. The deep margin remains controversial, as it is defined differently both anatomically and surgically [9].

Radiotherapy is part of the therapeutic armamentarium used for the local treatment of DFS. It is recommended for multiple recurrences, inadequate or invasive excision margins, very large tumors, and sites that oppose major surgery [10]. Drug treatment can be considered, for example, taking imatinib mesylate. It is already used to treat chronic myeloid leukemia and gastrointestinal stromal tumors. This molecule has been approved (MA) for the treatment of non-resectable forms. A French

study, the results of which were presented at the American Society of Clinical Oncology (ASCO), evaluated the use of this drug as neoadjuvant therapy over two months to reduce the size of the tumor before its removal. In this study, a preoperative clinical response was achieved in 9/25 cases (36%) [3].

CONCLUSION

Darier and Ferrand tumor is characterized by its difficult diagnosis, its tendency to recur, and its possibility of degeneration into fibrosarcoma after several recurrences. Our observation highlights the possibility of a slow evolution of this rare tumor, and the difficulty to make the histological diagnosis early and performing a major surgical resection since the whole prognosis depends on it.

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Author Contributions

Prosper Nsengiyumva – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Taha Kabbaj – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mohamed Anass Majbar – Conception of the work, Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Amine Benkabbou – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Amine Souadka – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Raouf Mohsine – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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ABOUT THE AUTHORS

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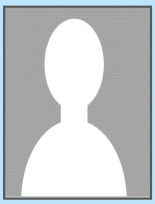
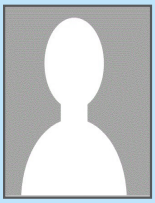
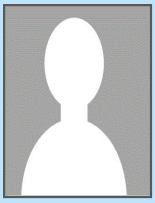
Prosper Nsengiyumva, Oncological Digestive Surgery Department, National Institute of Oncology, University Hospital Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco, Resident in General Surgery. He has published 4 articles in international academic journals. Email: nspmedecine@gmail.com



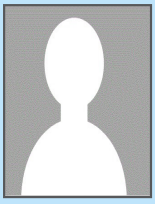
Taha Kabbaj, Oncological Digestive Surgery Department, National Institute of Oncology, University Hospital Centre Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco Resident in Oncological Surgery.
Email: Taha.kabbaj.chuis@gmail.com



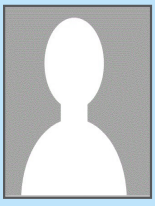
Mohamed Anass Majbar, Professor of Visceral and Digestive Surgery, Oncological Digestive Surgery Department, National Institute of Oncology, University Hospital Centre Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco. His research interest is in colorectal surgery.



Amine Benkabbou, Professor of Visceral and Digestive Surgery, Oncological Digestive Surgery Department, National Institute of Oncology, University Hospital Centre Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco. His research interests include hepatobiliary surgery and liver transplantation.



Amine Souadka, Professor of Visceral and Digestive Surgery, Oncological Digestive Surgery Department, National Institute of Oncology, University Hospital Cente Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco. His research interests include colorectal surgery and public health.



Raouf Mohsine, Professor of Visceral and Digestive Surgery, Head of Oncological Digestive Surgery Department, National Institute of Oncology, Head of University Hospital Centre Ibn Sina, Mohamed V University, Faculty of Medicine and Pharmacy, Rabat, Morocco.

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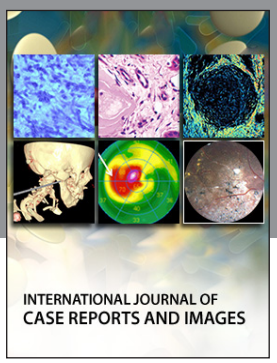

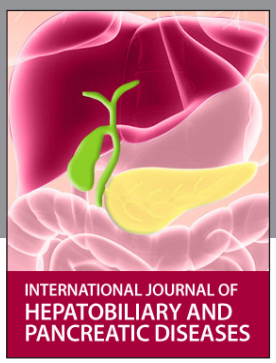

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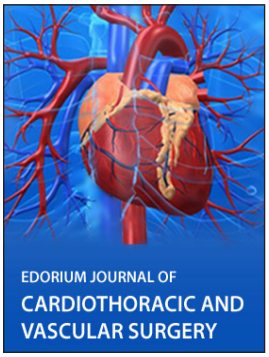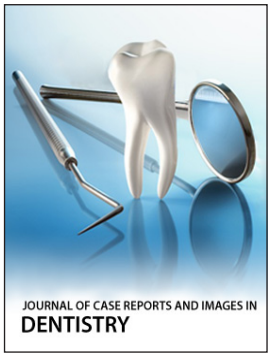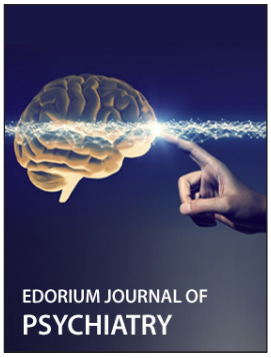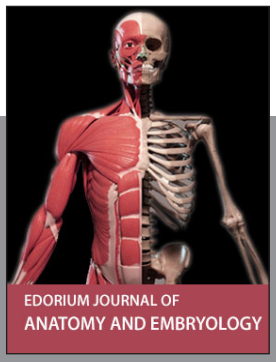
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