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Case Study

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METASTATIC PULMONARY DARIER AND FERRAND'S DERMATOFIBROSARCOMA: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Dermatofibrosarcome de Darier et Ferrand (DFS) is a skin tumor with intermediate malignancy. Although rare, it represents about 0.1% of malignant skin tumors. The authors report the case of a 48-year-old patient who was admitted to the emergency department due to diffuse chest pain persisting for 3 months, without any other associated symptoms. The patient's medical history includes the excision of nodules located at the root of the left thigh, but there are no available records regarding this prior procedure. The diagnosis of dermatofibrosarcome de Darier et Ferrand was suspected during the standard histological examination and confirmed by the observation of intense and diffuse expression of the CD34 protein. The treatment involved surgery, in combination with chemotherapy. It is essential to perform a biopsy and histological examination to establish an accurate diagnosis. The prognosis of this tumor is primarily influenced by its potential for local malignancy and the high risk of recurrence. Transformation into a markedly malignant, metastasizing sarcomatous form is rare.

KEYWORDS: Skin tumor, dermatofibrosarcoma of Darier and Ferrand, histopathology, CD34, recurrence.

INTRODUCTION

Dermatofibrosarcoma of Darier and Ferrand (DFS) is a rare fibrous cutaneous tumor originating from the dermis of the skin, classified as a low to intermediate-grade soft tissue sarcoma. It represents a small proportion of malignant skin tumors, approximately 0.1%, and accounts for 2 to 6% of all soft tissue sarcomas. This condition predominantly affects young adults aged 20 to 50 years, with no significant gender predominance.

In general, DFS presents as a nodular cutaneous mass, often localized on the trunk and extremities. Its primary characteristic is its local aggressiveness, with no tendency to spread through the lymphatic system, and the rate of distant metastases is less than 5%. The primary treatment involves surgical intervention to remove the tumor with substantial margins, typically 3 to 5 cm, to reduce the risk of recurrence. Inadequate excision is associated with a high rate of local recurrences, reaching approximately 40%. The prognosis of the disease is strongly influenced by the quality of the surgical excision.[1,2]

PATIENT AND MEDICAL OBSERVATION

We report the case of a 48-year-old patient who was admitted to the emergency department of the university hospital (CHU) with diffuse chest pain that had been present for three months, without any other associated symptoms. In the patient's medical history, there is a mention of the excision of nodules at the root of the left thigh without any available documentation.

Upon clinical examination, the patient presented with two nodular lesions at the root of the left thigh, which were firm in consistency, mobile in relation to the deeper tissues, and had a smooth, flesh-colored surface. The rest of the clinical examination did not reveal any significant findings.

A chest X-ray showed an opacity in the lower third of the left lung (radiological images are not available). A scan-guided biopsy of the right basal pyramid indicated a proliferation of spindle-shaped cells suggestive of a low-grade fusocellular sarcoma with a positive anti-CD34 immunostaining. The histological and immunostaining findings were consistent with those of a biopsy-excision of the skin tumor.

The diagnosis of a pulmonary metastasis of Darier and Ferrand was confirmed, and the treatment involved wide excision of the skin tumor and chemotherapy consisting of doxorubicin and ifosfamide.

DISCUSSION

Dermatofibrosarcoma of Darier and Ferrand is a rare tumor, accounting for only 0.1% of skin cancers. Its global incidence is estimated to be approximately 0.8 to 4.2 cases per million per year.[3] It tends to develop in adults aged 20 to 50. However, it is extremely rare in children and infants.[4,5]

Le DFS affecte both genders with a slight male predominance.[6]

The microscopic appearance of the characteristic typical form was first described by Taylor and Helwing in 1962.[7] Immunohistochemistry is essential for diagnosis, as DFS expresses the CD34 antigen and vimentin.[6,8]

Cytogenetic techniques reveal two types of karyotype abnormalities in the form of a supernumerary ring chromosome r(17,22) or a translocation t(17,22).[9] The clinically described classical form, which appears as a protuberant growth, corresponds to an advanced stage of the tumor. It presents as a firm, multinodular mass, attached to the skin but movable in relation to underlying tissues. The tumor is typically non-painful, except in cases of ulceration.[10] The diversity of clinical presentations can lead to diagnostic delays. DFS can occur on any part of the body, with a preference for the trunk and extremities.[6]

Surgery plays a pivotal role in the curative treatment of DFS. Two excision techniques achieve tumor control in over 90% of cases: the conventional wide excision and the Mohs micrographic surgery, which allows tumor removal with histological control to ensure the absence of tumor cells at the excision margins.[11–13] Conventional excision involves the necessity of wide and deep excisions, removing a peripheral margin of healthy skin of 3 to 5 cm, along with a healthy anatomical barrier in depth.[5,6,14–19]

Surgical excision is then tailored based on considerations of anatomical territories, functional aspects, and aesthetic units.[20]

Systematic lymph node dissection has no utility.[7,15,21] Most authors describe DFS as a radioresistant tumor.[7,5,22,23] However, others have asserted that radiotherapy reduces the rate of local recurrences and enables more conservative surgery.[24–26] According to HAAS et al., local tumor control was 82% with adjuvant radiotherapy for insufficient or involved excision margins, with a follow-up ranging from 1 to 22 years.[25] Radiotherapy is recommended for cases of multiple recurrences, inadequate or involved excision margins, very large tumors, and locations that preclude extensive surgery.[1]

The combination of surgery and radiotherapy appears to be effective in preventing recurrences.[25–27] The recommended dose is 50 Gray in cases of R0 surgery and 60 Gray in cases of R1 surgery, administered in 2 Gray fractions, 5 days a week.[28] As for exclusive radiotherapy, it can be attempted in unresectable tumors, in cases of inoperable patients, or those refusing surgical treatment, with doses potentially reaching up to 66 Gray.[1,29]

Several chemotherapy protocols, primarily based on doxorubicin, ifosfamide, methotrexate, and dacarbazine, have shown no significant improvement in terms of survival.[14,30]

Targeted therapy with Imatinib Mesylate has been tested in vivo for unresectable tumors and cases of metastatic DFS. Its effectiveness is not complete and may be absent in the absence of the t(17,22) translocation.[31,32] The prognosis of DFS is characterized by its high recurrence potential. The percentage of recurrences varies depending on the extent of excision. Therefore, the essential prognostic factor is the completeness of the initial surgical excision, determining the risk of local recurrence. Several poor prognostic factors have been identified in different published series, including incomplete excision, location on the cephalic extremity where wide excision principles are more challenging to apply, the presence of fibrosarcoma areas within the tumor, and the depth of the tumor.[33,34] DFS rarely metastasizes, with various series reporting a metastasis rate of 3 to 5%, and the prognosis for these cases is bleak. The 5-year survival is estimated at 20%.[35]

CONCLUSION

Darier and Ferrand's dermatofibrosarcoma (DFS) is a rare skin tumor characterized by a slow, gradual progression over several years. It is marked by diagnostic challenges, a tendency for recurrence, and the rarity of metastases, primarily to the lungs. Diagnosis is often suspected clinically and confirmed through histological examination.

The primary treatment for DFS is surgical, with the conventional excision approach. An alternative to conventional surgery is the Mohs micrographic surgery technique, which allows for more sparing of healthy tissue. However, this technique is not yet practiced in Morocco due to its high cost and the lack of trained teams. Overall, DFS has a good prognosis when well-managed, but it still requires lifelong clinical monitoring, as some cases of late recurrences have been documented.

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