Dermatofibrosarcoma
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Abstract
Dermatofibrosarcoma is a dermic tumor with spindle-shaped cells of intermediate wickedness characterized by a slow evolution with a major risk of recurrence. Its localization in the ankle constitutes a rare variety of this tumor. A 39 years old man who presented a giant egg-shaped tumor on the lateral side of the ankle. A tumoral resection, with 3 cm of margin was realized with a cytopathologic study of the tumor and margins of resection. The loss of cutaneous substance was covered by a f reversal fasciocutaneous sural flap and the bed of the flap put in managed healing. The total healing was obtained at post-operative day 45. The histology of the tumor confirmed the results of the biopsy and the margins of resection were clear. At evaluation, the flap was entirely integrated and the scar of the donor site lightly atrophic without recurrence.

INTRODUCTION
Dermatofibrosarcoma (DFS) is a rare malignant soft tissues tumor. Described at any age, but especially in the adulthood, without distinction of sex nor race it is characterized by exceptional metastases, high inclination to recurrence, and possibility of transformation in a true sarcoma acquiring the capacity of metastasizing 1. It’s usual localizations are in the trunk and in the neck 2, 3. The localizations in the extremities are mainly in the root and rarely in the distal extremities 4, 5, 6. It’s localization in the ankle constitutes a rare variety of this tumor 7. The treatment, eminently surgical, requires an often mutilating large resection which raises a problem of coverage. We report an exceptional localization to the ankle of a voluminous DFS and discuss the modalities of the treatment in this difficult anatomical site.

CASE REPORT
A 39 years old man, without particular pathological antecedent, presented with a spontaneously, occurring tumefaction of the lateral side of the ankle of two years duration. It was at first a cutaneous hardening that gradually increased in volume before beginning to exulcerate. Biopsy of the tumor, performed by dermatologist, revealed DFS of Darier and Ferrand. He then referred us the patient for surgical treatment. At examination we found an egg-shaped tumor on the lateral side of the ankle being 9 cm x 3.5 cm x 4 cm covered with a scab with hemorrhagic tendency to any attempt of ablation (Figure 1). We noted also non-inflammatory inguinal micro adenopathies. X-rays showed the nodular shadow of the tumor without calcification or osseous lesions (Figure 2).

Figure 1
Clinical appearance of the tumor.
Dermatofibrosarcoma

Figure 2
Radiographs of the ankle. Note the nodular shadow of the tumor.

Thoraco-abdominal CT scan did not show metastasis. After dissection of the sural pedicle, the tumoral resection with 3 cm of margin taking the superficial sural aponeurosis and the fibular periost on the opposite page was realized. A cytopathologic study of the tumor and margins of resection was made (Figure 3).

Figure 3
Intraoperative view. Note the satellite nodules along the sural pedicle.

The postoperative course was free of complication in spite of a necrosis of the banks of the flap. The total healing was obtained two months later without grafting of the donor site. The histology of the tumor confirmed the results of the biopsy and the margins of the resection were clear. Three years later, no recurrence was noted. The mobility of the ankle was normal without pain. The scar was atrophic on the donor site.

Figure 4
Clinical outcome. Note the hypochromic scar of the flap bed.

DISCUSSION
DFS is a dermic tumor with spindle-shaped cells of intermediate malignancy characterized by a slow evolution with a major risk of recurrence 8. It does not occur often (0, 1 % of all the tumors). It occurs in both sexes, all the ethnic groups, and arises at any age with two peaks of frequency, at 20 and 40 years of age 1. It can develop on any zone of the body, the face and the hands included, but prevails in more than half of the cases on the trunk and the roots of the extremities (especially shoulders and groin) 1, 2, 3. The localizations in the distal extremities are less usual and are reported in the literature by case reports 7.

The evolution of this tumor is always slow, so that it is often difficult to determine a date of the beginning for the patients. In 20 % of the cases, an etiologic factor like trauma (direct shock, vaccine, burn, clothing friction, previous surgical resection) or a pre-existent congenital cutaneous abnormality is found 7.

The tumor can be in two different presentations8, which never have worrying appearance and explain in our context, the delays in consultation (two years in our patient):
- First as a fibrous plaque pinkish or brownish, slightly raised, sometimes confused with a keloid scar. This plate is gradually expanding and nodules occur;
- Or as a dermic nodule colorized as normal skin, similar to
that of a benign histiocytotifroma. The evolution of the lesion is characterized by satellite nodules in the periphery that converge to make a multinodular closet. This was the form of presentation in our patient with the characteristic of the size of the tumor.

Only histological analysis can confirm the diagnosis. Histological examination revealed a mesenchymatous tumor proliferation by dermal point of departure with fibroblastic spindle elements arranged in wheel spokes. The cast tumors infiltrate the derma and dissociate the fat hypodermic lobules following the vascular axes. This infiltration along the main neurovascular and fascial planes is often much further than is suggested by the clinic or the macroscopic appearance of the pseudo-encapsulated tumor, explaining the presence of satellite nodules away from the tumor focus princeps (Figure 3). The absence of nuclear abnormalities and elevation of the mitotic index differentiates the DFP to the fibrosarcoma 5. The differential diagnosis in tropical areas is mainly done with the botryomycoma and fibrosarcoma 1, 2.

Immunohistochemistry examination often reveals a positive staining for CD34 antigen, but this marker is not specific. The origin of dermatofibrosarcoma protuberans is still mysterious 5. A genetic defect is present in 95% of cases which was discovered in 1990 with the identification of either a translocation of chromosomes 17 and 22, or the formation of a ring.

Only surgical excision of the tumor was so far the indisputable proof of its effectiveness. Because of its low mitotic activity, the tumor is not radiosensitive. Chemotherapy is not yet an effective therapeutic method, however, although there is now some hope with imatinib that may lead to a systematic reduction of margins of excision 8.

Its prognosis is only related to the quality of surgical removal. It is now clear that surgical excision of the surface must pass at least 5 centimeters from the edges of the tumor and should include a deep healthy anatomical barrier. The prognosis depends primarily on the quality of the initial resection 8.

In fact, the optimum margin surgical excision of DFS is not really known. The analysis of the literature shows an overall rate of tumor recurrence, which decreases when the margin of surgical excision increases. We can thus retain the rough orders of magnitude: 20% of recurrences between 2 and 3 cm margin, 15% between 3 and 4 cm, and 8% between 4 to 5 cm.

This rule should be applied wherever it is possible without unbearable mutilation that is to say everywhere outside the periorificals areas (face, perineum), breast, and extremities. Mohs micrographic surgery offers an alternative therapeutic approach. It allows for mapping the extent of the tumor with the microscopic examination of deep and lateral margins. An innovative therapy, Mohs micrographic surgery appears to be a significant step in reducing the recurrence rate. Indeed, lower recurrence rate than wide local excision have been reported in recent preliminary reports 9.

In any case, what is fundamental is not the clinic margin of excision, but the histological margin which should be healthy to minimize the risk of local recurrence 10.

CONCLUSION

The DFSP is a rare tumor with a high tendency to recurrence. Its differential diagnosis in tropical areas, with advanced forms, is mainly with the botryomycoma. The localization to the ankle is exceptional. Its surgical treatment, even though codified, does not always guarantee a cure.

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