Giant dermatofibrosarcoma protuberans on the abdominal wall
Dermatofibrosarcoma protuberans gigante de parede abdominal

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ABSTRACT
Dermatofibrosarcoma protuberans is probably best considered as a low-grade dermal sarcoma. It is an uncommon type of sarcoma that may occur anywhere in the body. It rarely arises on the abdominal wall. It often affects young patients and appears as a nodular mass. The growth pattern is usually slow and persistent. These lesions enlarge over many years, becoming protuberant. These tumors have a locally aggressive nature and may recur frequently. They rarely metastasize. A wide resection with 2-3 cm free margins is the best treatment. This paper reports a rare type of abdominal wall sarcoma. A brief literature review is presented showing the surgical principles for treating abdominal wall tumors and their outcome. Radical surgery and good locoregional control are shown to be significantly related.

Keywords: Dermatofibrosarcoma; Abdominal wall/pathology; Soft tissue neoplasms

INTRODUCTION
Soft tissue sarcomas are mesenchymal neoplasms comprising 1% of adult malignant tumors. Among soft tissue tumors, abdominal wall tumors are uncommon, accounting for less than 5% of these neoplasms(1,2). Although clinically similar, these tumors have many distinct histological subtypes with significantly variable biological behaviors. It is advisable to know the type of primary tumor for optimal treatment planning and outcome. Since these tumors are rare, their behavior is sometimes poorly understood. Their treatment can be challenging regarding resection and reconstruction(3).

The most common soft tissue tumor of the abdominal wall is the desmoid tumor while the least frequent is dermatofibrosarcoma protuberans(3). Generally speaking, dermatofibrosarcoma protuberans (DFSP) is a rare tumor, accounting for less than 5% of adult soft tissue sarcomas. Historically, this neoplasm was first described by Taylor, in 1890, but Hoffman coined the currently accepted term when he reported three cases in 1925(2).

DFSP is a low-grade mesenchymal tumor that originates in the dermis. These tumors occur as pink or violet-red plaques. It was also described as a nodular cutaneous mass and its surrounding skin may be telangiectatic. They are characterized by progressive, locally infiltrative behavior. If left untreated, these tumors continue to grow slowly, invading the surrounding tissue, including neurovascular bundles. They usually present on the trunk and extremities, between the second and fifth decades of life. Lung and regional lymph node metastases are rare and usually preceded by multiple local recurrences. The...
The treatment of choice is surgical resection with an adequate margin of uninvolved tissue\(^{(4,6)}\).

The authors describe one patient with a giant dermatofibrosarcoma protuberans on the abdominal wall treated with a wide resection. Reconstruction was performed by primary wound closure with a prosthetic mesh.

**CASE REPORT**

Patient ABS, a black, 20-year-old man presented with a painless skin mass in the abdominal wall. The patient had noticed a slow growth of this lesion over the previous 10 years. He had previously been to many medical services to treat this disease.

The clinical examination showed a 20 x 15 cm size tumor, violet-red, nodular and firm that was attached to the dermis but moved freely over deeper-lying tissue. The lesion was located in the left flank and mid-abdomen (figure 1). The computed tomography scans showed a solid skin-subcutaneous lesion not invading the underlying fascia or musculature.

The main diagnosis was soft tissue tumor, probably a DFSP. The clinical differential diagnosis included desmoid tumor, dermatofibroma, metastatic tumor or a benign/malignant fibrous histiocytoma. Subsequently, an incisional biopsy was performed for an etiologic diagnosis.

The histopathological findings revealed monomorphous spindle cells in a storiform pattern, embedded in a scarcely to moderately dense fibrous stroma compatible with dermatofibrosarcoma protuberans. Immunohistochemistry showed that tumor cells were positive for CD34 and XIIIa.

As definitive treatment, this patient underwent a partial-thickness abdominal wall resection with a 2.5 cm margin of normal tissue (figure 2). The previous scar with surrounding skin, subcutaneous tissue and anterior abdominal aponeurosis were excised. The reconstruction was performed with primary wound closure and propylene mesh. A closed-suction catheter was used (figure 3).

The patient was discharged from hospital within five days. There were no surgical complications. Histological examination revealed free margins. No adjuvant treatment was administered. To date, two years after surgery, the patient remains fit. No evidence of local or distant recurrence has been found.

**DISCUSSION**

Sarcomas of the abdominal wall are difficult-to-treat neoplasms. Due to their variable histological type and grade, there are many different surgical approaches. The general recommendation is to perform a wide excision with free margins. Narrow margins were related to a dismal prognosis. A wide resection with a 2-3 cm margin in the treatment of abdominal wall sarcomas is associated with a good regional and distant control. To date, only one series has been described on treatment of abdominal wall DFSP\(^{(3)}\).
DFSP usually presents as a nodular, violet-red skin mass on the trunk and proximal extremities. It tends to present a slow growth pattern and, in many cases, its symptoms are long lasting\(^{(2,4,5)}\). Most lesions are smaller than 5 cm. In a large series of 159 patients treated at the Memorial Sloan-Kettering Cancer Center (New York, NY), between 1950 and 1998, Bowne et al. reported only 4 patients (3\%) with large tumors (≥ 10 cm)\(^{(7)}\). In the present study, the authors report a DFSP that arose in an unusual site, in addition to showing an exceptional size\(^{(2-6,8)}\).

Although specimen collection could be performed by core biopsy, the classical approach has been incisional biopsy. According to experts, an incisional biopsy would be adequate for large lesions. The amount of tissue resected should be suitable for a good histopathological assessment\(^{(1-8)}\). An incisional biopsy should be performed planning for the definitive surgical incision at Langer’s lines. The definitive incision must include the previous scar and catheter incision\(^{(3)}\).

DFSP spreads locally, with regional infiltration of surrounding structures as fascia, aponeurosis, muscles, peritoneum and bone\(^{(3-8)}\). Computed tomography (CT) or magnetic resonance imaging (MRI) has been indicated to stage these tumors. These imaging techniques are important for resection planning\(^{(2-6,8)}\).

Essentially, the optimal treatment for DFSP has been a wide resection. Since these tumors are locally infiltrative, the general surgical principles used for sarcomas should be used to properly treat them. A three-dimensional wide resection of skin and surrounding structures must be performed. Most authors recommended a 2-3 cm local margin including the underlying deep fascia and overlying skin\(^{(2,6,8)}\). When resections are performed with inadequate margins, the reported local recurrence rate can be as high as 60\%\(^{(5)}\).

Adjuvant treatment with radiotherapy seems controversial. There are specialized services that have recommended adjuvant radiotherapy for large low-grade (> 5 cm) or high-grade sarcomas\(^{(2,8)}\). Other authors believe radiotherapy should be reserved for close or positive surgical margins\(^{(4,6)}\). In the present study, the authors believe that radiotherapy is not necessary for most cases since it has risks and may be detrimental to clinical follow-up. Most experts reserve it for microscopic margins or irresectable tumors.

**CONCLUSION**

In sum, when adequate principles are followed, DFSP has a good prognosis. However, close surveillance is required since local recurrence rates are high\(^{(2,5)}\). The present study showed that safe diagnosis can lead to excellent surgical results in the treatment of DFSP.

**REFERENCES**