

Case Report

A rare large anterior abdominal wall dermatofibrosarcoma protuberans treated by wide local excision and abdominoplasty: a case report and review of the literature

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare soft tissue neoplasm characterized by uniform, slender, spindle shaped, fibroblast like cells, arranged in a typical storiform or cartwheel pattern. Diagnosis is established after excisional biopsy or punch biopsy, CT scan and MRI are useful in both diagnosis and planning the magnitude of wide, free margins surgical excision. Large anterior abdominal wall DFSP represents a challenging modality in reconstructive options. In our paper, we introduce the technique of spindle shaped wide local surgical excision followed by abdominoplasty for a rare large 15×7×4.5 cm, anterior, middle abdominal wall dermatofibrosarcoma protuberans (in the umbilical region). In spite of the large tumor size, and the rare location, the technique is simple, easy to plan and perform, safe for free surgical margins resection, with good cosmetic results, and excellent post operative recovery.

Keywords: Dermatofibrosarcoma protuberans, Spindle cell neoplasm, Abdominoplasty

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a soft tissue neoplasm of intermediate to low-grade malignancy. Although metastasis rarely occurs, DFSP has a locally aggressive behavior with a high recurrence rate.¹ DFSP is a rare tumour that represents 1% of all soft tissue sarcomas. It accounts for less than 5% of soft tissue tumours and 0.1% of all malignancies with an annual worldwide incidence of 0.8 to 4.5 per million.^{2,3}

Historically, Darrier and Fernand first recognized DFSP as a clinicopathological entity and Hoffman introduced the term "DFSP".^{4,5} It is a slow growing, nodular,

polypoid neoplasm that occur almost exclusively in the dermis, but it can also occur in the deeper soft tissues, most commonly found in the trunk and proximal extremities.⁶ It may occur at sites of previous trauma, affecting more commonly men than women and has a peak incidence during the third decade of life, and most of the cases occur in the adults, but they have also been seen in infancy and childhood.⁷

DFSP is a locally aggressive tumor with a high rate of recurrence that can occur in 20-55% of cases and with an extremely low rate of distant metastasis.⁸ The tumor first appears as a single, red to blue, bleachable, firm, cutaneous nodule. During the late stage, rate of growth

accelerates, producing the characteristic protrusion from the skin.⁹ The growth rate is variable and lesions may remain stable for many years or they may grow slowly with periods of accelerated growth.¹⁰

Clinically, it presents as an asymptomatic raised, firm nodular lesion fixed to the skin but mobile over the deep fascia, with a pattern of slow, progressive growth.^{11,12}

There are conditions that can mimic DFSP, which should be considered in the differential diagnosis. They are: dermatofibroma, epidermal inclusion cyst, keloid and hypertrophic scar, malignant melanoma and metastatic carcinoma of the skin.¹³

Diagnosis is established after excisional biopsy or punch biopsy.¹⁴ CT-scan is useful to determine the tumor extent of penetration.¹⁵ The CT appearance of DFSP is well defined, as unmineralized, nodular soft tissue mass involving the skin and subcutaneous adipose tissue. CT scans or MR images are well suited to show this location, the relation of lesion to underlying structures and the distinct lobular or nodular architecture.⁹

The disease is thought to be of histiocytic origin and characterized by uniform, slender, spindle shaped (Figure 1), fibroblast like cells, arranged in a typical storiform or cartwheel pattern. Other features of diagnostic importance are the high cellularity, monomorphic appearance, moderate to high mitotic activity, lack or inconspicuousness of foamy or hemosiderin laden macrophages and or multinucleated giant cells and entrapment of fat cells when there is an invasion of subcutaneous tissue.¹⁶

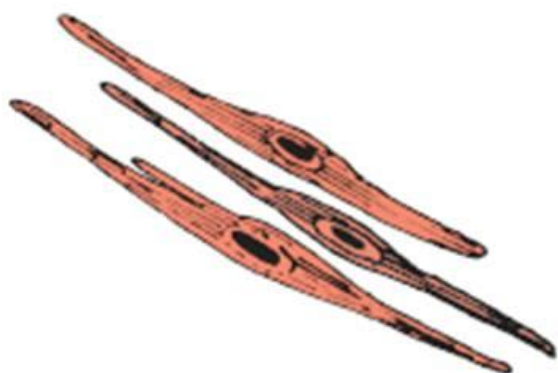


Figure 1: Spindle cell.

Immunohistochemical staining was strongly positive for vimentin and CD34, and cytogenetically DFSP commonly has translocation involving PDGF-beta & COL1A1.¹⁷ Chromosomes 17 and 22 have been incriminated in the aetiopathogenesis of DFSP. There is translocation t(17;22) involving COL1A1 (collagen type 1α1 gene) and PDGF β (platelet-derived growth factor) genes respectively.¹⁸ DFSP has two histologic variants:

the more typical low-grade tumor, and a high-grade rare fibrosarcomatous variant demonstrating necrosis, high mitotic rate (> 10 mitoses/high-power fields) and the presence of pleomorphic areas.¹⁵ This last variant is associated with a poor clinical outcome.

Metastasis is rare. Lung metastasis is most common, while lymph node metastasis is exceedingly rare.¹⁵

Wide surgical excision using a margin of three centimeters with inclusion of superficial fascia is currently the standard therapy in children.¹⁹ Mohs micrographic surgical excision has wide acceptance among adult cases.²⁰ Imatinib has shown clinical activity against both localized and metastatic DFSP with t(17;22). However, fibrosarcomatous variants of DFSP lacking t(17;22) may not respond to imatinib.²¹ 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.²²

CASE REPORT

A 32-year-old male came to the hospital with chief complaints of painless swelling in the umbilical region for one year. The swelling had been slowly growing in size over the last year. The swelling, which was earlier painless, had now become painful after it ruptured with a bloody discharge. The swelling was initially small in size, nodular, gradually increasing in size and had currently achieved the dimensions of about 15×7 cm, composed of 3 large masses attached together, with dark skin pigmentation surrounding the entire swelling. Additionally, the right lateral nodule showed a ruptured surface of 3×2 cm (Figure 2-4).



Figure 2: Anterior view of the swelling showing 3 nodules with ruptured right lateral one.



Figure 3: Lateral view showing the swelling and skin dark pigmentation surrounding it.



Figure 4: Anterior view of the swelling showing 3 nodules with ruptured right lateral one.

Clinical parameters were within normal limits. Blood and Chemistry laboratory investigations were within normal ranges. Chest X-ray was normal.

Computed tomography scan (CT) showed an irregular, nodular, soft tissue density mass, occupying the umbilical and paraumbilical regions of the anterior abdominal wall, protruding anteriorly, measuring collectively 15×7 cm, (pre contrast 21, post contrast 23 H.U) not enhancing, no signs of metastasis detected (Figure 5-10).

A core biopsy was taken under local anesthesia to establish the diagnosis, it showed oval to spindle tumor

cells, arranged in storiform and parallel pattern exhibiting mild to moderate pleomorphism, at places with scant to abundant collagen within tumor cells and few dilated vessels. Mitosis was very minimal (2 mitosis seen in entire biopsy). The tumor is CD34 positive, but negative for Msa, S100, Calponin, Desmin, Beta Ketanin, Pankeeratin.

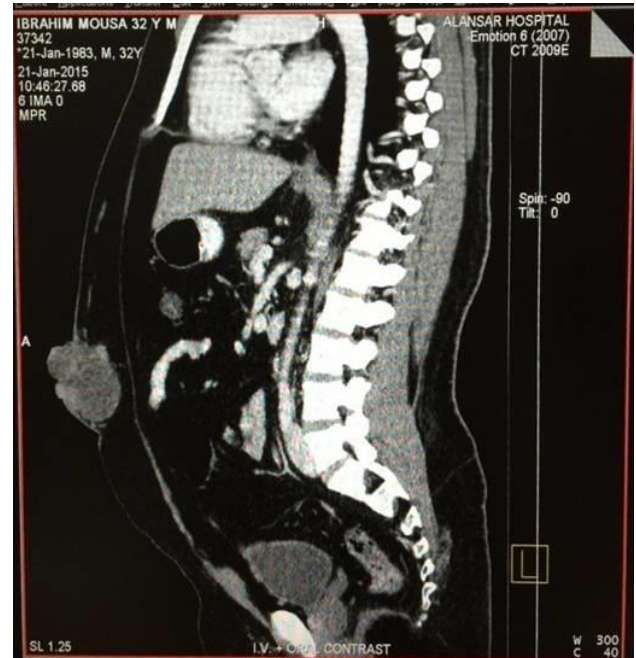


Figure 5: Sagittal view of soft tissue density nodular mass at umbilical region.



Figure 6: Coronal view of soft tissue density nodular mass, only soft tissue area seen.



Figure 7: Sagittal view of soft tissue density nodular mass, only soft tissue area seen.

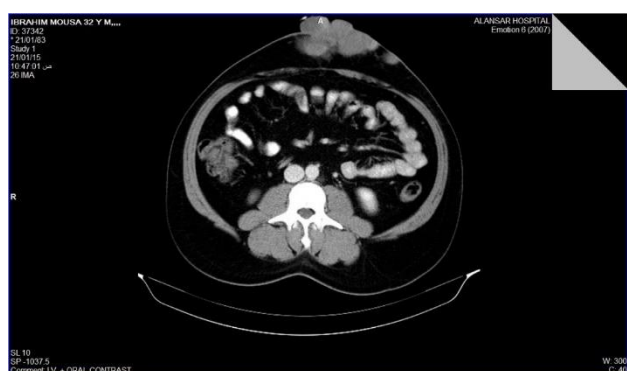


Figure 8: Axial view showing nodular masses connected together, on the right and left side of the umbilical region of the anterior abdominal wall.

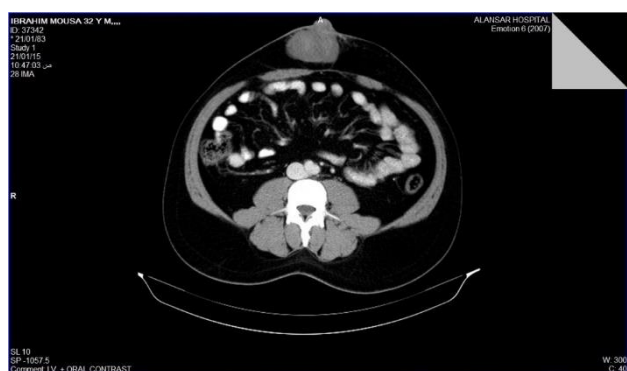


Figure 9: Axial view of soft tissue density nodular mass.

A diagnosis of Spindle cell neoplasm was reached (Figure 11 & 12).

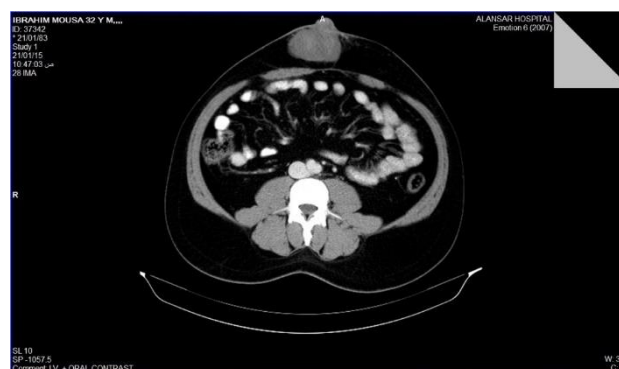


Figure 10: Axial view of soft tissue density nodular mass, showing the deepest part separated from the rectus sheath.

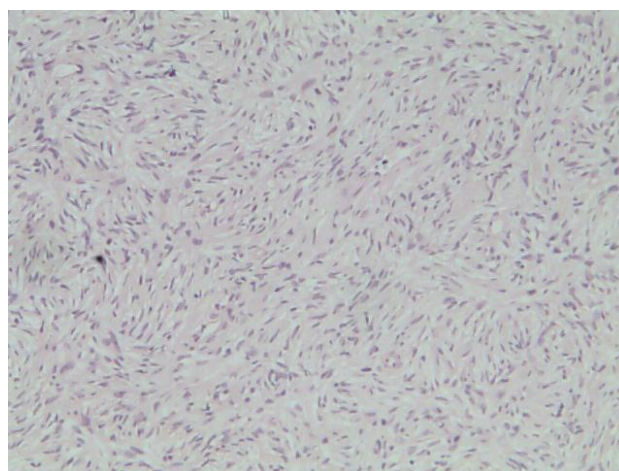


Figure 11: Core biopsy showing spindle cell neoplasm.

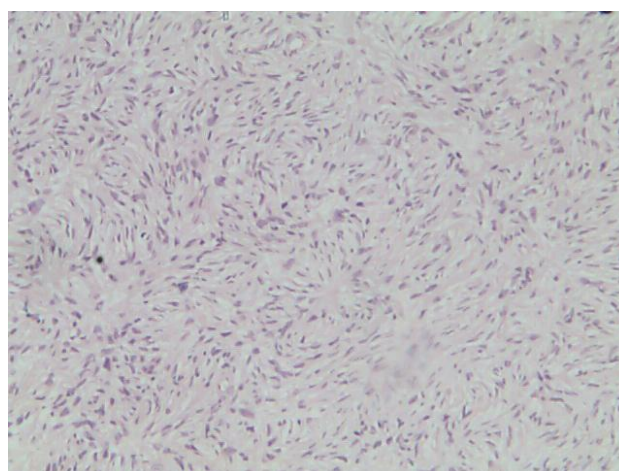


Figure 12: Core biopsy showing spindle cell neoplasm.

The patient underwent a wide local excision of the mass (en block), the skin incision was carefully planned in a transverse spindle shape (from spindle cell) depending on CT scan measurement and outline, in addition to on table examination, dissection down to the rectus sheath was carefully carried out, the rectus sheath was intact with 2.5 cm save margin from the deepest area of the mass, and 3 cm save margins was taken in all directions. The umbilicus was included in the en block resection due to very close proximity to the mass (Figure 13-16).

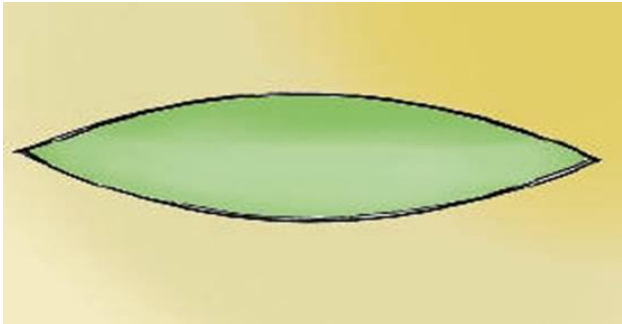


Figure 13: The planned skin incision – spindle in shape.

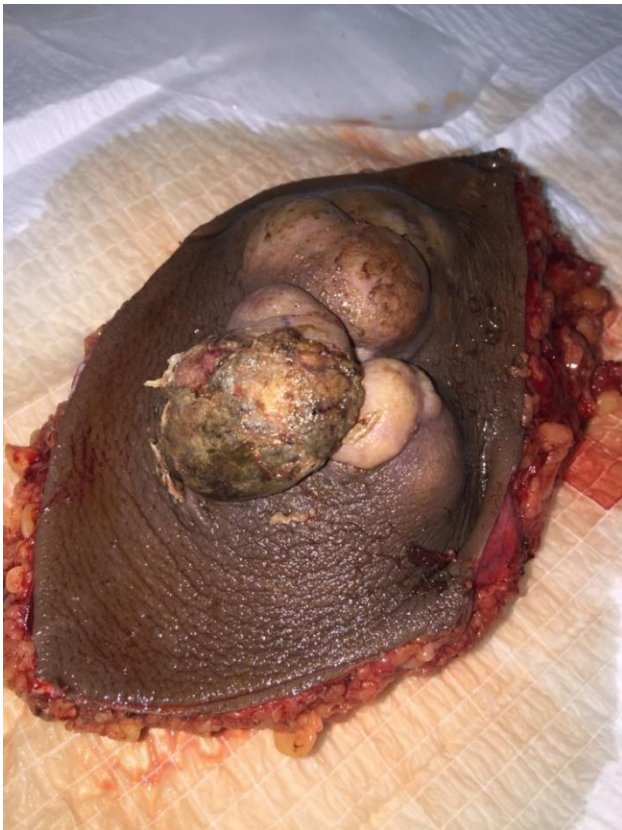


Figure 14: The resected mass (en block) – spindle in shape.



Figure 15: The resected mass showing full thickness.



Figure 16: The resected mass showing all nodules.

A spindle shaped defect of 18×10 cm resulted from the en block excision. Abdominoplasty was performed gaining 10 cm of skin and subcutaneous tissue of the abdominal wall from above and below the defect, two surgical drains were inserted, one in the upper part, one in the lower part, primary closure of the defect was achieved by approximating the two edges tension free by 0-vicryl sutures, and the skin was closed by surgical staples (Figure 17 & 18).



Figure 17: Anterior view of the abdomen post operative showing closed wound.

Histopathology examination of the resected mass grossly revealed an ellipse of skin with underlying tissue measures 18×10×7 cm, with nodular surface, tumor mass measures 15×7×4.5 cm, cut section showed multiple yellow firm nodules.

Microscopically, section showed skin exhibiting dermal tumor extended in to the subcutis entrapping adipose tissue. The tumor consists of sheets of spindle shaped cells displaying mildly pleomorphic nuclei and arranged in storiform, fascicular and cartwheel pattern. Mitotic activity was present. The surgical margins were clear. The tumor cells are positive for CD34 and negative for Desmin (Figure 19-25).



Figure 18: Anterior view of the abdomen post operative showing closed wound.

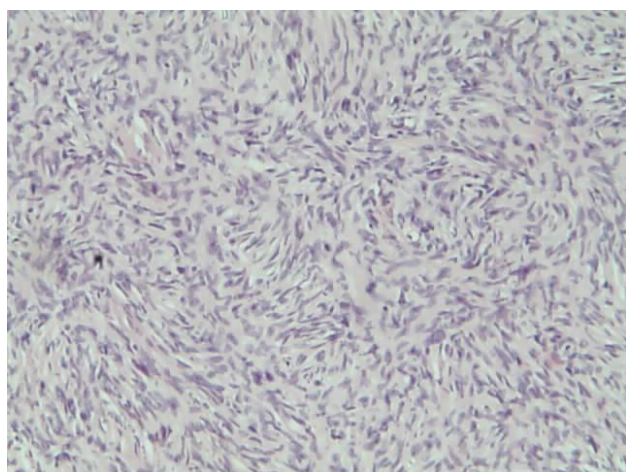


Figure 19: Storiform or cartwheel pattern of dermatofibrosarcoma protuberans.

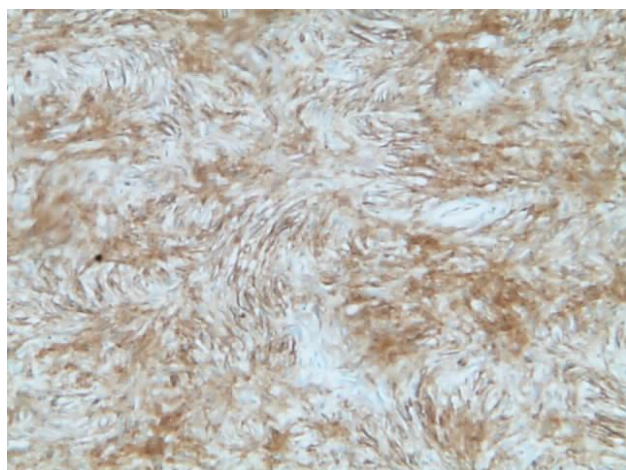


Figure 20: Section showing positivity for CD34 immunohistochemical stain of tumor cells.

Recovery was uneventful and patient was discharged on the fifth post-operative day. Patient was asymptomatic one month after surgery (Figure 26-29), and has been coming for regular follow-up, he was referred to a

specialized cancer center for the option of radiation therapy and imatinib.

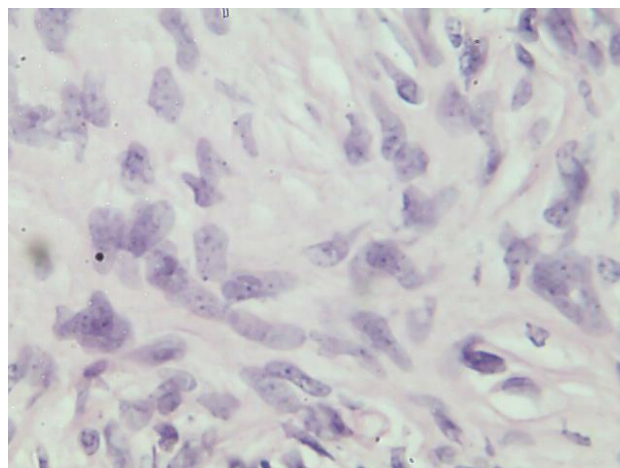


Figure 21: High power field showing spindle shaped cells with atypia.

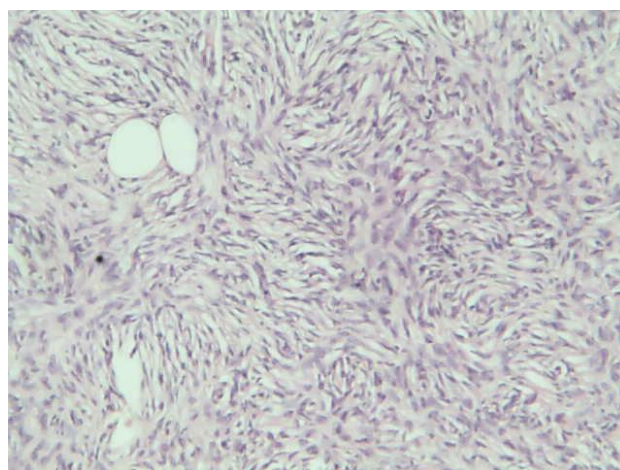


Figure 22: Section showing invasion of subcutaneous fat.

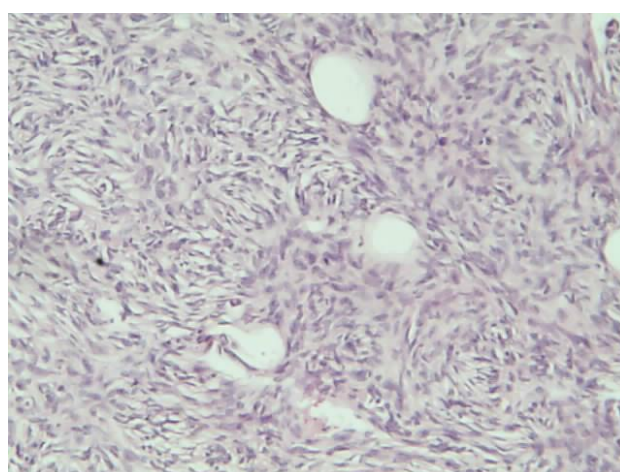


Figure 23: Section showing invasion of subcutaneous fat.

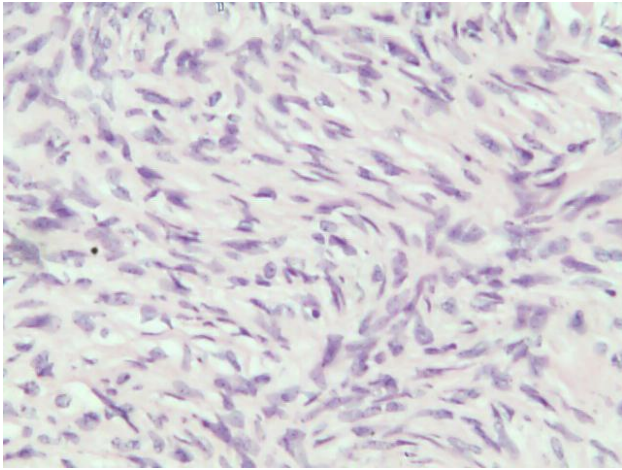


Figure 24: High power field showing mitotic figure.

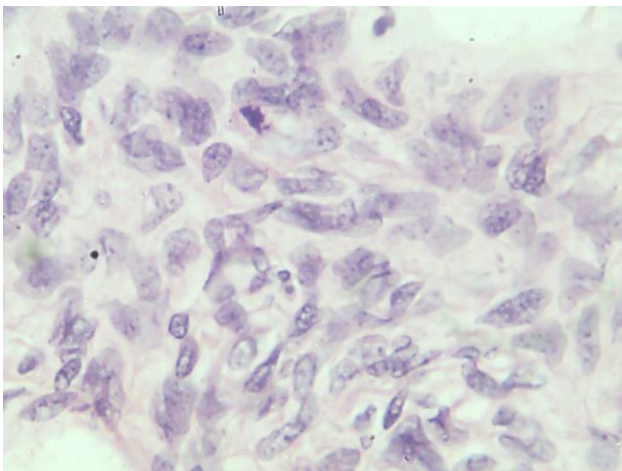


Figure 25: High power field showing mitotic figure.



Figure 26: The wound 7 days post-operative (1st clinic visit).



Figure 27: The wound 14 days post operative (2nd clinic visit).



Figure 28: The wound 21 days post operative (3rd clinic visit).



Figure 29: The wound 28 days post operative (4th clinic visit).

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.^{23,24}

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.^{25,26} 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).¹⁷ In this study, we report a DFSP that arose in an unusual site, in addition to showing an exceptional size.

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.^{27,28} 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.²⁴

DFSP spreads locally, with regional infiltration of surrounding structures as fascia, aponeurosis, muscles, peritoneum and bone.²⁴⁻²⁶ Computed tomography (CT) or magnetic resonance imaging (MRI) has been indicated to stage these tumors. These imaging techniques are important for resection planning.^{8,9,24,25} 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.²⁴⁻²⁶ When resections are performed with inadequate margins, the reported local recurrence rate can be as high as 60%.²⁶ Wide resection of DFSP (whether recurrent or primary) with negative histological margins predicts a superior local recurrence-free survival.²²

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.^{24,25,28} Other authors believe radiotherapy should be reserved for close or positive surgical margins.²⁵⁻²⁸

Several published reports indicate that imatinib has significant activity against DFSP. The first experience was published in 2002 by Maki, Cells transformed with the COL1A1-PDGFB gene as well as cell cultures derived from patients with DFSP are inhibited by the tyrosine kinase inhibitor imatinib mesylate (Glivec or STI-571; Novartis, Basel, Switzerland). In challenging, locally advanced disease and in the rare metastatic cases imatinib mesylate has shown impressive single agent activity with limited toxicity.²⁹

Reconstruction of large and complex abdominal wall defects after wide local excision represent a challenging problem, and the options include primary closure, skin grafts, local and distant flaps, synthetic and biologic mesh reconstruction. Abdominoplasty as a means of reconstruction has been reported after traumatic injuries of abdominal wall.^{30,31}, but rarely has been used post sarcoma excision. Using of abdominoplasty is feasible for lower abdominal defects. Good cosmetic results are achieved without the need for prosthesis or second operative procedure.³²

In conclusion, in our case, we report a rare, large 15×7×4.5 cm, anterior, middle abdominal wall dermatofibrosarcoma protuberans (in the umbilical region), and introduce the technique of the spindle shaped incision, en block tumor wide local excision, and tension free primary closure (modified abdominoplasty procedure). The technique is simple, easy to plan and perform, safe for free surgical margins resection, with good cosmetic results, and excellent post-operative recovery.

Abbreviations

DFSP	: Dermatofibrosarcoma protuberans
CT	: computed tomography scan
MRI	: magnetic resonance imaging
CD34	: hematopoietic progenitor cell antigen
PDGF-beta	: platelet-derived growth factor subunit B
COL1A1	: Collagen, type I, alpha 1
Msa	: muscle-specific actin
S100	: low-molecular-weight proteins
H.U	: Hounsfield unit

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Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Heuvel ST, Suurmeijer A, Pras E, Van Ginkel RJ, Hoekstra HJ. Dermatofibrosarcoma protuberans: recurrence is related to the adequacy of surgical margins. *Eur J Surg Oncol.* 2010;36:89-94.
2. Stojadinovic A, Karpoff HM, Antonescu C, et al. Dermatofibrosarcoma protuberans of the head and neck. *Ann Surg Oncol.* 2000;7(9):696-704.

3. Criscion VD, Weinstock MA. Descriptive epidemiology of Dermatofibrosarcoma protuberans in the United States 1973 to 2002. *J Am Acad Dermatol.* 2007;56:968-73.
4. Darier J, Ferrand M. Dermatofibrosarcoma progressifs et recidivants on fibrosarcomas de la plan. *Ann Dermatol Venereol.* 1924;5:545.
5. Hoffman E. Uber das knollentriebende Fibrokom der Haut. *Dermat Zeitschr.* 1925;43:1-8.
6. Taylor HB, Helwig EB. Dermatofibrosarcoma protuberans - A Study of 115 cases. *Cancer.* 1962;15:717-25.
7. Rockley PF, Robinson JK, Magid M, Goldblatt D. Dermatofibromasarcoma protuberans of the scalp-a series of cases. *J Am Acad Dermatol.* 1989;21:278-82.
8. Rutgers E, Kroon BBR, Albus-lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. *European J Surgical Oncol.* 1992;18:241-8.
9. Miyakawa E, Fujimoto H, Miyakawa K, Nemoto K et al, Dermatofibrosarcoma protuberans - CT findings with pathologic correlation in 6 cases. *Acta Radiol.* 1996;37:362-5.
10. Phelan JT, Juado J. Dermatofibrosarcoma Protuberans. *AM J Surg.* 1963;106:943-8.
11. McKee PH, Fletcher CD. Dermatofibrosarcoma protuberans presenting in infancy and childhood. *J Cutan Pathol.* 1991;18(4):241-6.
12. Keshen TH, Cederna PS, Savell VH Jr, Platz CE, Chang P, Ricciardelli EJ. Clinical and pathological features of pediatric dermatofibrosarcoma protuberans. *Ann Plast Surg.* 1995;35(6):633-7.
13. Kshirsagar AY, Kanetkar SR, Nikam YP, Vasisth GOP. Recurrent Dermatofibrosarcoma Protuberans Over Anterior Abdominal Wall. *J Cutan Aesthet Surg.* 2010;3(3):167-9.
14. Pappo AS, Rao BN, Cain A, Bodner S, Pratt CB. Dermatofibrosarcoma protuberans: the pediatric experience at St. Jude Children's Research Hospital. *Pediatr Hematol Oncol.* 1997;14(6):563-8.
15. Bouyssou-Gauthier ML, Labrousse F, Longis B, Bedane C, Bernard P, Bonnetblanc JM. Dermatofibrosarcoma protuberans in childhood. *Pediatr Dermatol.* 1997;14(6):463-5.
16. Mentzel T, Beham A, Katenkamp D, Dei Tos AP, Fletcher CD. Fibrosarcomatous ("high-grade") dermatofibrosarcoma protuberans: clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance. *Am J Surg Pathol.* 1998;22(5):576-87.
17. Bowne WB, Antonescu CR, Leung DH, Katz SC, Hawkins WG, Woodruff JM, et al. Dermatofibrosarcoma protuberans: A clinicopathologic analysis of patients treated and followed at a single institution. *Cancer.* 2000;88(12):2711-20.
18. Kiuru-Kuhlefelt S, El-Rifai W, Fanburg-Smith J, Kere J, Miettinen M, Knuutila S. Concomitant DNA copy number amplification at 17q and 22q in dermatofibrosarcoma protuberans. *Cytogenet Cell Genet.* 2001;92(3-4):192-5.
19. Marcus JR, Few JW, Senger C, Reynolds M. Dermatofibrosarcoma protuberans and the Bednar tumor: treatment in the pediatric population. *J Pediatr Surg.* 1998;33(12):1811-4.
20. Checketts SR, Hamilton TK, Baughman RD. Congenital and childhood dermatofibrosarcoma protuberans: a case report and review of the literature. *J Am Acad Dermatol.* 2000;42(5):907-13.
21. McArthur GA, Demetri GD, van Oosterom A, Heinrich MC, Debiec-Rychter M, Corless CL, et al. Molecular and clinical analysis of locally advanced dermatofibrosarcoma protuberans treated with imatinib: Imatinib target exploration consortium study B2225. *J Clin Oncol.* 2005;23:866-73.
22. Khatri VP, Galante JM, Bold RJ, Schneider PD, Ramsamooj R, Goodnight JE Jr. Dermatofibrosarcoma protuberans: Reappraisal of wide local excision and impact of inadequate treatment. *Ann Surg Oncol.* 2003;10:1118-22.
23. Costa SRP, Henriques AC. Giant dermatofibrosarcoma protuberans on the abdominal wall. *einstein.* 2007;5(1):63-5.
24. Stojadinovic A, Hoos A, Karpoff HM, Leung DH, Antonescu CR, Brennan MF, et al. Soft tissue tumors of the abdominal wall: analysis of disease patterns and treatment. *Arch Surg.* 2001;136(1):70-9.
25. Chang CK, Jacobs IA, Salti GI. Outcomes of surgery for dermatofibrosarcoma Protuberans. *Eur J Surg Oncol.* 2004;30(3):341-5.
26. Lindner NJ, Scarborough MT, Powell GJ, Sapnier S, Enneking WF. Revision surgery in dermatofibrosarcoma protuberans of the trunk and extremities. *Eur J Surg Oncol.* 1999;25(4):392-7.
27. Brennan MF. Staging of soft tissue sarcomas. *Ann Surg Oncol.* 1999;6(1):8-9.
28. Mendenhall WM, Ziotecki RA, Scarborough MK. Dermatofibrosarcoma protuberans. *Cancer.* 2004;101(11):2503-8.
29. Labropoulos SV, Razis ED. Imatinib in the treatment of dermatofibrosarcoma protuberans. *Biologics.* 2007;1(4):347-53.
30. Brenneman FD, Boulanger BR, Antonyshyn O. Surgical management of abdominal wall disruption after blunt trauma. *J Trauma.* 1995;39:539-44.
31. Okunski WJ, Sonntag BV, Murphy RX Jr. Staged reconstruction of abdominal wall defects after intra-abdominal catastrophes. *Ann Plast Surg.* 1996;36:475.
32. Shenoy R, Beh A, Dhabhar B. *International Journal of Health Information and Medical Research.* 2014; 1(1).

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