

Case Report

Large anterior abdominal wall fibrosarcoma in a known case of recurrent dermatofibrosarcoma protuberans

Snigdha Kamini*, Sunil Kumar Jain, Jaspreet Bajwa, M. Nafees Ahamad,
Sumit Kumar Prasad, Shivam Sharma, Pradeep Mekha Srinivas

Department of General Surgery, VMMC and Safdarjung Hospital, Delhi, India

Received: 27 May 2020

Revised: 12 July 2020

Accepted: 14 July 2020

***Correspondence:**

Dr. Snigdha Kamini,

E-mail: snigdhakamini5@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Dermatofibrosarcoma protuberans is a rare soft tissue sarcoma of low to intermediate malignant potential. Incidence is estimated to be 0.8 to 4.5 cases per million persons per year. The tumor is highly aggressive locally and is known to recur. Punch biopsy or excisional biopsy (in case of small tumors) are diagnostic. Computer tomography (CT) and magnetic resonance imaging (MRI) are useful in determining the size and extent of the tumor's treatment is primarily surgical, with a wide local excision with at 2 cm margin. However, local recurrence after apparently adequate surgical excision is common. Mohs micrographic surgery has been recommended as it enables maximum preservation of tissue. When surgery is insufficient, imatinib mesylate is shown as a safe and effective treatment in dermatofibrosarcoma protuberans (DFSP), especially in cases of locally advanced or metastatic disease. Here, we report a case of a giant fibrosarcoma on the anterior abdominal wall, measuring 27×18×9 cm, which occurred in a recurrent dermatofibrosarcoma protuberans. Diagnosis was done by histopathological examination (HPE) of the previously excised tumor and CT was done. The tumor was excised with a 3 cm margin and extensive reconstruction of the anterior abdominal wall defect was done using synthetic mesh, myocutaneous flaps and split skin grafting. Keeping in view the recurrent nature of our case, large tumor size, DFSP-fibrosarcomatous (FS) transformation and close negative margins in the HPE of the postoperative specimen, the patient was planned for adjuvant radiotherapy.

Keywords: Dermatofibrosarcoma protuberans, Fibrosarcoma, Reconstruction

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma of low to intermediate malignant potential. Incidence is estimated to be 0.8 to 4.5 cases per million persons per year.¹ It most commonly arises in adults in their late twenties to thirties, but it has been described in all age groups. Although this tumor has a low propensity to metastasize, it is highly aggressive locally and is known to recur.

These tumors most commonly occur on the trunk, as is our case and to a lesser extent on the extremities. These are

morphologically characterized by single, red to blue, bleachable, firm, cutaneous nodule.² During the late stage, rate of growth accelerates, producing the characteristic protrusion from the skin. The growth rate is variable, and lesions may remain stable for many years or they may grow slowly with periods of accelerated growth. Several histological subtypes of the tumor have been described, which include pigmented (Bednar tumor), myxoid, myoid, granular cell, sclerotic, atrophic DFSP, giant cell fibroblastoma, and DFSP with fibrosarcomatous areas. Of all these variants, only the DFSP with fibrosarcomatous areas is high grade, with a higher rate of local recurrence and distant metastasis.³

DFSP is genetically characterized by the t (17;22) (q22;q13), resulting in the fusion of alpha chain type 1 of collagen gene and platelet-derived growth factor beta gene.⁴ This translocation is present in 90% of DFSP and is useful in the differential diagnosis of DFSP with other tumors with similar histology.

Punch biopsy or excisional biopsy (in case of small tumors) are diagnostic. Computer tomography (CT) and magnetic resonance imaging (MRI) are useful in determining the size and extent of the tumor.

The treatment is primarily surgical, with a wide local excision with at 2 cm margin.⁵ However, local recurrence after apparently adequate surgical excision is common. Mohs micrographic surgery has been recommended as it enables maximum preservation of tissue.⁶ When surgery is insufficient, imatinib mesylate is shown a safe and effective treatment in DFSP, especially in cases of locally advanced or metastatic disease.^{7,8} Transformation to fibrosarcoma, as had occurred in our case occurs very rarely.^{9,10} Here we present a case report of a patient with a recurrent, large, anterior abdominal wall fibrosarcoma arising from a recurring DFSP, which we managed with wide local excision and extensive abdominal wall reconstruction.

CASE REPORT

A 26-year-old male patient had presented to the surgical outpatient department with complaint of a large growth on the anterior abdominal wall. Patient was a chronic smoker, with no significant family history. He had previously undergone surgery for the same complaint thrice, the last being done 4 months ago. There were no records of any of the previous surgeries but the histopathological examination (HPE) report of the mass excised from the last surgery suggested Dermatofibrosarcoma Protuberans.

On examination, there was a large 27 * 18 * 9 cm swelling over the left anterior abdominal wall extending from above the left costal margin to 4 cm below the umbilicus. It occupied the entire left hypochondrium, left anterior lumbar region. The skin over the swelling was stretched out, with hyper and hypopigmented areas and had dilated veins. The swelling was well defined, had irregular borders, bosselated surface and was firm in consistency.

A contrast enhanced computer tomography (CECT) of the abdomen was done which showed a large well defined heterogeneously enhancing soft tissue density lobulated mass lesion measuring 13.6×15.7×8.4 cm [Transverse (TR)*Cranicaudal (CC)*Anterior-posterior (AP)] seen arising from the upper antero-lateral abdominal wall on the left side with multiple areas of non-enhancement. The lesion was seen to reach up to the overlying skin surface with involvement of subcutaneous tissue.

Posteriorly it was seen to infiltrate the external intercostal, rectus abdominus and external oblique muscle. There was

no involvement of the ribs or any intraperitoneal extension. There were no features suggestive of distant metastasis.

The patient was duly taken up for elective surgery. A wide local excision of the tumor with a 3 cm margin was done. The underlying left external intercostal, left rectus abdominus and left external oblique muscles were removed at the site of infiltration.

The peritoneum was not breached. The excision of the tumor resulted in a large defect in the left anterior abdominal wall with the peritoneum exposed. A 30” *30” inch Ultrapro mesh was used over the peritoneum and the defect was reconstructed using a right pedicled anterolateral thigh flap, right FC flap and split skin graft (SSG) harvested from the left thigh.

Post op recovery and HPE

Patient was monitored intensively in the ICU and shifted to the post-operative ward after 24 hours. Post-operative recovery was good. Flaps were examined every day. On post op day 6 the superior margin of the right FC flap showed necrosis and was debrided under local anesthesia and was left to heal by secondary intention. Flap drains were sequentially removed. The SSG donor site dressing was changed and patient was discharged on post op day 20.

The HPE report showed a cellular infiltrative tumor composed of spindle cells arranged in form of diffuse sheets and intersecting fascicles. The individual tumor cells had elongated spindle shaped hyperchromatic nuclei. The mitotic count was >20/hpf. All the peripheral resected margins were free of tumor, but the tumor was reaching close to the deep resected margin falling short of 2 mm. The tumor cells were positive for vimentin and focally for cluster of differentiation 34 (CD 34). The picture was that of a fibrosarcoma arising in a pre-existing DFSP. The patient was followed up regularly and after healing of the surgical wound was referred to the oncology team for adjuvant radiotherapy.



Figure 1: 30×18×9 cm tumour overlying the left anterior abdominal and chest wall.

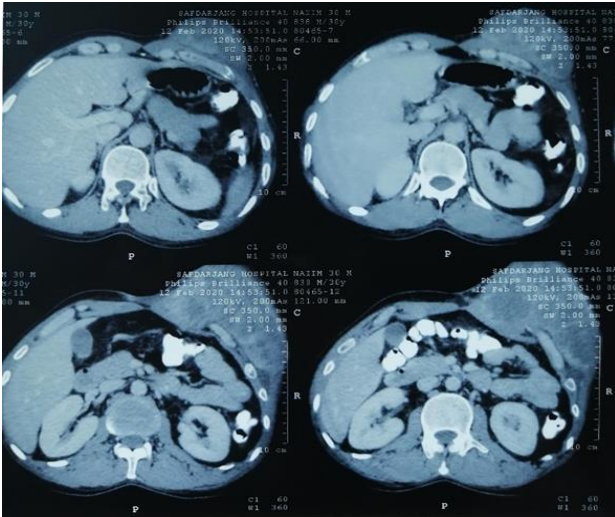


Figure 2: CECT image showing tumour.



Figure 3: Post-operative specimen and subsequent abdominal wall defects.



Figure 4: Abdominal wall after reconstruction.

DISCUSSION

Sarcomas of the anterior abdominal wall are rare and difficult to treat tumors. The recognition of the lesion as an entity is generally attributed to Darier and to Hoffmann, first employed the term “dermato- fibrosarcoma protuberans”.¹¹ DFSP usually presents as a nodular, violet-red skin mass on the trunk and proximal extremities. 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).⁹ Li Min Sun et al, in their study of 35 cases reported a tumor with maximum dimension 25 cm.¹²

In this study, we report a large, recurrent DFSP that grew to an exceptional size of 27×18×9 cm and showed degeneration to fibrosarcoma. Core biopsy or specimen biopsy maybe used in the histopathological diagnosis. In our case, the patient had the excisional biopsy record of the tumor excised during the previous surgery which gave us the preoperative histopathological diagnosis of DFSP.

The optimal treatment option for DFSPs and DFSP-FSs is resection with wide margins. Gloster et al., in their review of 84 cases, suggested that the optimal treatment option for DFSPs and DFSP-FSs is resection with wide margins; the likelihood of local recurrence after this procedure is performed is less than 10%.¹³ A three-dimensional wide resection of skin and surrounding structures must be performed. Rutgers et al., and most authors recommended a 2-3 cm local margin including the underlying deep fascia and overlying skin although the ideal margin to be taken remains controversial.¹⁴

In our study we had taken a margin of 3 cm. Limited experience with Mohs surgery indicates that this procedure is associated with a high probability of cure provided that the final margins are negative.^{6,15} Adjuvant radiotherapy (RT), administered either before or after surgery, significantly reduces the risk of local recurrence in patients who have positive margins or unsatisfactory surgery.^{16,17} Radiotherapy may also be used in cases of recurrence, metastasis and select inoperable cases. Maki et al, first showed that cells transformed with the collagen type 1 alpha 1–platelet derived growth factor beta chain (COL1A1-PDGFB) are inhibited by the tyrosine kinase inhibitor imatinib mesylate. Since then, several published reports indicate that imatinib has significant activity against DFSP.^{8,18}

Transformation of DFSP to Fibrosarcoma is a rare occurrence. In our patient, the transformation to fibrosarcoma was diagnosed by the Histopathological examination of the post-operative specimen. These tumors are known to have higher recurrence and metastatic rates and a worse prognosis.^{19,12} Keeping in view the recurrent nature of our case, large tumor size, DFSP – FS transformation and close negative margins, the patient was planned for adjuvant radiotherapy after complete healing of the wound, after consultation with a multidisciplinary tumor board. 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. We had a large defect, and extensive reconstruction was done using synthetic mesh, flaps and skin grafting.

CONCLUSION

Here we report a rare case of a large fibrosarcoma of the anterior abdominal wall measuring 27×18×9 cm. This tumor arose in a known case of recurrent Dermatofibrosarcoma Protuberans. The tumor was removed by wide local excision with a 3 cm margin and meticulous reconstruction of the anterior abdominal wall was done using synthetic mesh and tissue transfer techniques. The diagnosis was confirmed through histopathology. Post-operative recovery was good, and the patient was referred for adjuvant radiotherapy. This tumor was aggressive as it had a history of two recurrences and showed a high mitotic index. There were no records of the previous surgery, but we hypothesize that the surgical excision might have been inadequate. Keeping in view the aggressiveness of the tumor, and the positive margins in our specimen, patient was referred for adjuvant radiotherapy.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

- Criscito MC, Martires KJ, Stein JA. Prognostic factors, treatment, and survival in dermatofibrosarcoma protuberans. *JAMA Dermatol* 2016;152(12):1365-71.
- Weinstein JM, Drolet BA, Esterly NB, Rogers M, Bauer BS, Wagner AM et al. Congenital dermatofibrosarcoma protuberans: variability in presentation. *Arch Dermatology*. 2003;139(2):207-11.
- Llobart B, Serra-Guillén C, Monteagudo C, Guerrero JA, Sanmartín O. Dermatofibrosarcoma protuberans: a comprehensive review and update on diagnosis and management. In *Seminars in diagnostic pathology*. WB Saunders; 2013:30:13-28.
- Rutkowski P, Przybył J, Świtaj T. Genetics of rare mesenchymal tumors: implications for targeted treatment in DFSP, ASPS, CCS, GCTB and PEComa. *Int J Biochem Cell Biol*. 2014;53:466-74.
- Farma JM, Ammori JB, Zager JS, Marzban SS, Bui MM, Bichakjian CK et al. Dermatofibrosarcoma protuberans: how wide should we resect? *Ann Surg Oncol*. 2010;17(8):2112-8.
- Roh MR, Bae B, Chung KY. Mohs' micrographic surgery for dermatofibrosarcoma protuberans. *Clin Exp Dermatol*. 2010;35(8):849-52.
- Rutkowski P, Dębiec-Rychter M, Nowecki ZI, Michej W, Symonides M, Ptaszynski K et al. Treatment of advanced dermatofibrosarcoma protuberans with imatinib mesylate with or without surgical resection. *J European Academy Dermatol Venereol*. 2011;25(3):264-70.
- Malhotra B, Schuetze SM. Dermatofibrosarcoma protuberans treatment with platelet-derived growth factor receptor inhibitor: a review of clinical trial results. *Curr Opin Oncol*. 2012;24(4):419-24.
- 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.
- Stacchiotti S, Pedetour F, Negri T, Conca E, Marrari A, Palassini E et al. Dermatofibrosarcoma protuberans-derived fibrosarcoma: Clinical history, biological profile and sensitivity to imatinib. *International J Cancer*. 2011;129(7):1761-72.
- Pack GT, Tabah EJ. Dermatofibrosarcoma protuberans: a report of thirty-nine cases. *Arch Surg*. 1951;62(3):391-411.
- Sun LM, Wang CJ, Huang CC, Leung SW, Chen HC, Fang FM et al. Dermatofibrosarcoma protuberans: treatment results of 35 cases. *Radiotherapy and oncology*. 2000;57(2):175-81.
- Gloster JH, Harris KR, Roenigk RK. A comparison between Mohs micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans. *Journal of the American Academy Dermatol*. 1996;35(1):82-7.
- Rutgers EJ, Kroon BB, Albus-Lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. *European J Surgical Oncol*. 1992;18(3):241-8.
- Loghdey MS, Varma S, Rajpara SM, Al-Rawi H, Perks G, Perkins W. Mohs micrographic surgery for dermatofibrosarcoma protuberans (DFSP): a single-centre series of 76 patients treated by frozen-section Mohs micrographic surgery with a review of the literature. *J Plastic, Reconstructive Aesthetic Surg*. 2014;67(10):1315-21.
- Hamid R, Hafeez A, Darzi MA, Zaroo I, Rasool A, Rashid H. Outcome of wide local excision in dermatofibrosarcoma protuberans and use of radiotherapy for margin-positive disease. *Indian Dermatol Online J*. 2013;4(2):93.
- Uysal B, Sager O, Gamsiz H, Cicek A, Demiral S, Dincoglan F et al. Evaluation of the role of radiotherapy in the management of dermatofibrosarcoma protuberans. *J BUON*. 2013;18(1):268-73.
- Navarrete-Dechent C, Mori S, Barker CA, Dickson MA, Nehal KS. Imatinib treatment for locally advanced or metastatic dermatofibrosarcoma protuberans: a systematic review. *JAMA Dermatol*. 2019;155(3):361-9.
- Cai H, Wang Y, Wu J, Shi Y. Dermatofibrosarcoma protuberans: clinical diagnoses and treatment results of 260 cases in China. *Journal Surgical Oncol*. 2012;105(2):142-8.

Cite this article as: Kamini S, Jain SK, Bajwa J, Ahamad MN, Prasad SK, Sharma S, et al. Large anterior abdominal wall fibrosarcoma in a known case of recurrent dermatofibrosarcoma protuberans. *Int Surg J* 2020;6:3120-3.