BMH MEDICAL JOURNAL

BMH Med. J. 2020;7(4):17-22. **Case Report**

Surgical Management of a Challenging Dermatofibrosarcoma Protuberans of the Abdominal Wall

Mohamed Raboune, Abdelmoughit Echchaoui, Jawad Hafidi, Nour-eddine Gharib, Samir El Mazouz

Department of Plastic Surgery and Burns, Mohamed V University, Avicenna teaching Hospital, Rabat, Morocco

Address for Correspondence: Dr. Abdelmoughit Echchaoui, Department of Plastic Surgery and Burns, Mohamed V University, Avicenna teaching Hospital, Rabat, Morocco. Email: e.moughit@hotmail.fr

Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue neoplasm which arises from the all part of body especially trunks and limbs, occurs more often in young people; the definite diagnosis depends mainly on the pathological examination; early detection and complete excision remain the foundation of treatment. The challenge of management for large sized tumors is presented as surgery may be associated with poor cosmetic and functional outcomes or inadequate surgical margin, hence the interest of a careful clinical long-term monitoring. We report a case of a young man, who presented with DFSP in his left abdominal wall, which was successfully removed by two-stage surgery, without recurrence at fifteen months of follow-up.

Keywords: Dermatofibrosarcoma protuberans; management challenge; two-stage surgery

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade malignant mesenchymal tumor of the soft tissue representing about 1% of all tumors [1], characterized by its aggressive local growth and common local recurrence after surgical excision, with rare distant metastases [2]. The greatest challenge in the management of DFSP is performing a good surgery with high oncological, cosmetic and functional outcomes.

Through a case of DFSP of abdominal wall, we identify the challenges encountered in managing this condition, and we emphasize the importance of early recognition and clinical long-term monitoring.

Case report

We report the case of a young male, who presented with an eight-month history of a gradually increasing asymptomatic movable firm red mass of 7x5cm of diameter on his left

abdominal wall (**Figure 1**). The patient was a smoker, biopsy of the lesion revealed a Darier and Ferrand dermatofibrosarcoma. Surgical management was planned as a two-stage intervention: the first stage was microsurgical transfer of the free latissimus dorsi musculo-cutaneous flap (**Figure 2**) being folded and sutured on itself with an interposed silicone sheet of 2 mm thick in its internal part (**Figure 3**). The donor site of the flap was closed with a thin skin graft. The recipient vessels were the left superficial femoral artery and vein (**Figure 4**), the second stage was planned after two weeks, by direct reconstruction after resection of the tumor with 5 cm surgical margins and deep resection to a disease-free anatomical structure (in this patient it was the rectus abdominis muscle) (**Figure 5**). Given that the peritoneal plane was intact; the reconstruction of the musculoaponeurotic plane was performed with a polypropylene mesh. The superficial layer was then covered with the free flap unfolded from its proximal base (pedicle) to cover the whole defect (**Figure 6**). Histological examination confirmed the diagnosis, The postoperative courses were uneventful and the length of hospital stay was four weeks. There was no recurrence at 15 months of followup.

Figure 1: A protuberant mass on the abdominal wall measuring 7x5cm in size, with surgical margins of 5cm

Figure 2: Microsurgical transfer of the free latissimus dorsi musculo-cutaneous flap

Figure 3: A silicone sheet of 2 mm thick sutured in the internal part of the flap after microsurgical transfer

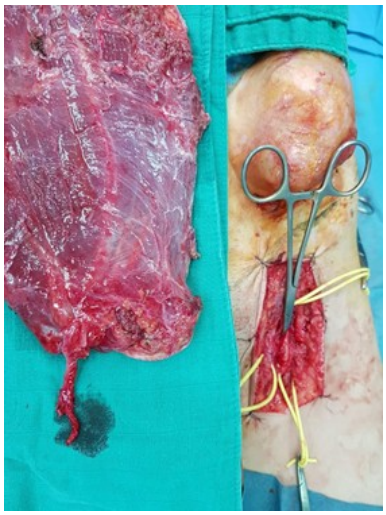

Figure 4: The recipient vessels (femoral artery and vein) of the free flap

Figure 5: A large and deep defect post wide local excision; A=Rectus Abdominis, B= Iliac Bone, C= the flap folded and sutured on itself with an interposed silicone.

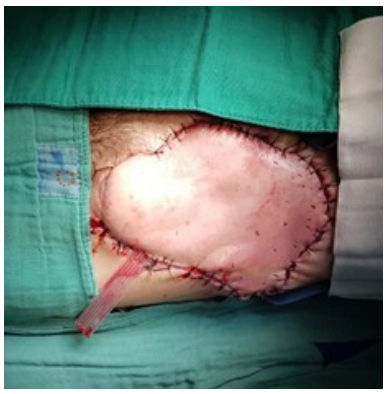

Figure 6: Coverage of the defect with the free flap unfolded from its proximal base (pedicle) and silicone sheet removal.

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing tumor that constitutes less than 0.1% of all malignancies [3]. DFSP has an intermediate to low malignant tendency. The potential for distant metastasis is low, but DFSP frequently recurs locally after incomplete excision [4]. Confirmed by biopsy and subsequent histopathology, its clinical diagnosis is difficult and often delayed by the variable presentation of the tumor.

Surgery plays a major role in the curative treatment of DFSP; it should consider the oncologic outcomes, functional and cosmetic issues [5]. Wide local excision (surgical margins of 5cm) has been the gold standard of treatment [6], although Mohs Micrographic Surgery (with surgical margins of 1 to 2 cm) is being advocated as preferred choice [7]; it offers histopathologic control while providing maximum tissue conservation with much lower risk of recurrence [8]. In our context, Mohs surgery is not available in our country, so conventional wide local excision remains the only choice.

Most abdominal wall defects post excision are amenable to local soft tissue closure, Due to the abundant and lax tissue of the abdominal wall [9], but sometimes may represent a challenge, when large excisions cause a large skin defect which is difficult to cover and requires the use of free flaps [10].

Thin skin graft is indicated for all superficial defects; however, the use of flaps remains crucial when noble elements are exposed (fascia, bone, nerves and vessels…) [11]. Several flaps are indicated for deep and large abdominal defects such as Mac Gregor flap, deep inferior artery perforator flap, latissimus dorsi flap, tensor fascial lata flap etc [9]. The choice will depend on the site and the size of the defect as well as the exposure or not of the underlying noble elements. The use of free flaps, in particular the free Latissimus Dorsi flap provides soft tissue coverage for defects that are not amenable to either local or regional flap coverage, it may be used like our case as a two-stage intervention for minimizing the time of surgery, thus the risk of failure [10].

The role of adjuvant radiotherapy is limited, it may be used efficiently for unresectable lesion or resected lesion with a positive margin and may decrease the rate of local recurrence [12]. Targeted therapies such as imatinib mesylate (Glivec®), has been currently FDA-approved its efficiency for adult with unresectable, recurrent, and/or metastatic cases [13]. Long-term follow-up is crucial as majority of the local recurrences occur within 3 years after the surgery [14].

Conclusion

DFSP is a rare disease with local aggressive behavior and high capacity of recurrence. Mosh

surgery technique is being advocated as best choice; it may improve the oncologic outcomes, functional and cosmetic issues. The wide variability of clinical expressions and unpredictable evolutionary nature require regular long-term monitoring in detecting local recurrence.

Conflict of Interest: None declared

Funding Sources: None declared

References

1. Wiesmueller F, Agaimy A, Perrakis A, et al. Dermatofibrosarcoma protuberans: surgical management of a challenging mesenchymal tumor. World J Surg Oncol. 2019; 17(1):90.

2. Kashyap L, Noronha V, Patil V, Joshi A, Mahajan A, Mittal N, Prabhash K. Dermatofibrosarcoma protuberans of head and neck: Clinical outcome of nine cases treated with imatinib. Cancer Res Stat Treat 2019; 2: 112-8.

3. S. El Mazouz, J. Hafidi, A. Echchaoui, M. Benyachou, N. Gharib, A. Abbassi. Le dermatofibrosarcome de Darrier et Ferrand. Maroc Médical. 2013; 35(2): 111-118.

4. A Echchaoui, M Benyachou, J Hafidi, H Zaouri, J Lamine, S El Mazouz, N-E Gharib, A Abbassi. Dermatofibrosarcoma protuberans and corticosteroids: a possible etiologic link. Research 2014; 1: 1233.

5. Bouhani M, Fertani Y, Zemni I, et al. Dermatofibrosarcoma Protuberans of the Breast in Man: An Extremely Rare Entity With a Review of the Literature. J Investig Med High Impact Case Rep. 2019;7: 2324709619875634.

6. Malumani Malan et al. The efficacy of Mohs micrographic surgery over the traditional wide local excision surgery in the cure of dermatofibrosarcoma protuberans. Pan African Medical Journal. 2019;33:297.

7. Foroozan M, Sei JF, Amini M et al. Efficacy of Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans: systematic review. Arch Dermatol 2012; 148: 1055–63

8. Hafner HM, Moehrle M, Eder S et al. 3D-Histological evaluation of surgery in dermatofibrosarcoma protuberans and malignant fibrous histiocytoma: differences in growth patterns and outcome. Eur J Surg Oncol 2008; 34: 680–6.

9. Roubaud MS, Baumann DP. Flap Reconstruction of the Abdominal Wall. Semin Plast Surg. 2018;32(3):133-140.

10. Leila Essid et al. The surgical management of a hypogastric dermatofibrosarcoma protuberans. Pan African Medical Journal. 2019;34:116.

11. Arnaud E, Couturaud B, Revol M, Servant JM, Banzet P. Surgical repair of abdominal wall. Ann Chir Plast Esthet. 1999;44(4):357-372.

12. Prakash G, Midya M, Sharma P, Jain RK. Neglected giant dermatofibrosarcoma protuberans. Formos J Surg 2019;52:24-7.

13. 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):361-369.

14. Xiao W, Que Y, Peng R, Ding Y, Zhao J, Wen X, Weng D, Zhang X, Guan Y, Zhang X. A favorable outcome of advanced dermatofibrosarcoma protuberans under treatment with sunitinib after imatinib failure. Onco Targets Ther. 2018;11:2439-2443.