Case Report

Recurrent Dermatofibrosarcoma Protruberans: For all Surgeons or the Right Surgeons – Challenges and Management Approach

CM Isiguzo, C Opara, C Onyeokpere, A Uzoho, IO Ifeakandu, N Ilokanuno[1]

Department of Surgery, Plastic and Reconstructive Surgery Unit, Federal Teaching Hospital, Owerri, Imo State, [1]Department of Surgery, Plastic and Reconstructive Surgery Unit, Nnamdi Azikiwe University Teaching Hospital, Nnewi, Anambra State, Nigeria

Received: 09-Apr-2024;
Revision: 27-Jul-2024;
Accepted: 16-Oct-2024;
Published: 04-Dec-2024

Abstract

Dermatofibrosarcoma protuberan is a locally aggressive tumor that can be challenging to manage if it becomes recurrent. The best attempt at achieving cure is the first surgery. The outcome of the first surgery is dependent on who the initial surgeon is – right surgeon or not. Multidisciplinary care offers good outcomes when reconstructive needs are catered for. In our resource-poor setting, it will be beneficial to have an adequate (more than 5 cm margin) excision margin followed by reconstruction of defects by surgeons with adequate training in management of the pathology and tissue reconstruction.

Keywords: *Dermatofibrosarcoma protuberans, multidisciplinary, surgeon, wide local excision*

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare local aggressive tumor arising from the dermal fibroblast or subcutis. In some cases, it may involve muscles and connective tissues near the skin.[1] It is a low-grade sarcoma that is marked by high recurrence rate, which reflects directly on the extent of initial resection. It is characterized by intermediate malignant potential between common fibrous histiocytoma and malignant fibrous histiocytoma. Rarely does it metastasize, but it is locally invasive and aggressive. Ten percent (10%) of cases can undergo fibrosarcomatous transformation and can metastasize. Cytogenic studies implicate translocation characteristic morphology and chromosomal translocation (17;22)(q21;q13) with gene fusion COL1A1-PDGFB.[2] It is characterized by a uniform spindle cell arrangement, classically with a storiform pattern and CD34 immunoreactivity.[3] Wide local excision with 5 cm margin of normal skin and deep to the fascial plane has a low incidence of recurrence which can be equated to results from a more technically demanding Mohs micrographic surgery. Recurrence can occur after more than 20 years.

It usually appears as a slow-growing papule or nodule. It may progress to be lumpy, sclerotic, or atrophic plaque.[4] It can affect any part of the body and any age. However, it is usually more common between the ages of 20 and 50 years. It affects more males than females.[1]

Cure can only be achieved by surgery if the resection margin is clear of the tumor. Achieving a clear margin is the biggest challenge of this surgery. Wide local excision (WLE) with 3–5 cm margin is the most preferred technique and reconstruction of the defect with skin graft or flaps. Tissues excised are the skin, subcut, and superficial fascia. In order to achieve a tumor-free

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/njcp.njcp_264_24

Address for correspondence: Dr. CM Isiguzo, Department of Surgery, Plastic and Reconstructive Surgery Unit, Federal Teaching Hospital, Owerri, Imo State, Nigeria.
E-mail: mcisiguzo@gmail.com

How to cite this article: Isiguzo CM, Opara C, Onyeokpere C, Uzoho A, Ifeakandu IO, Ilokanuno N. Recurrent dermatofibrosarcoma protruberans: For all surgeons or the right surgeons – challenges and management approach. Niger J Clin Pract 2024;27:1340-3.

margin, Mohs micrographic surgery can be used to detect microscopic clearance of tumor cells.[5]

Tumors can be locally invasive spreading radially along collagen bundles in the skin and deeply along connective tissue septae. The risk of recurrence is high if there are residual tumors in the margin. The reoperation will involve excision of the previous scar.[5] Therefore, the best time to achieve cure is the first WLE. This will require a delicate balance between undertreatment and overtreatment. It is generally resistant to conventional chemotherapy, even though targeted immunotherapy gives some encouraging report.[6]

Aim

This report will demonstrate the need for adequate excision at the initial presentation by the right surgeon with appropriate training and experience to reduce the incidence of recurrence. It will also emphasize the need for long follow-up of patients after resection of DFSP.

Patients and Methods

This study involves a case report of 49-year-old female with recurrent DFSP of the left groin, thigh, and vulva of 28 years duration and who has had nine inadequate resections at peripheral centers. Wide local excision with 5 cm skin margin and down to the fascial plane was carried out to achieve cure.

Case Report

A 49-year-old female presented to the clinic with a recurrent mass on the left groin, thigh, and vulva of 28 years duration [Figures 1 and 2]. There were no associated local symptoms except for occassional nonradiating pain, with no known aggravating or relieving factor. The esthetic disfigurement was undesired. Weight loss or features of metastases to other regions were not noted. She has had nine excisions with serial recurrence before presentation. The current histology report showed DFSP. The patient was a known hypertensive for about 1 year before presentation with poor control.

Examination revealed a massively disfigured left groin with a bosselated appearance. There are predominantly two masses located on the femoral triangle with the superior extending to the mons pubis and deforming the left labium majora. Superior mass extends above the inguinal ligament, irregularly shaped, 16 × 12 cm in widest limits across two planes. The mass was firm in consistency and tender, attached to the skin but not to the underlying structures. The skin was hyperpigmnted. The surface had multiple incisional scars. Differential warmth was absent; lymph nodes were not palpable on the ipsilateral or contralateral side. The inferior mass is

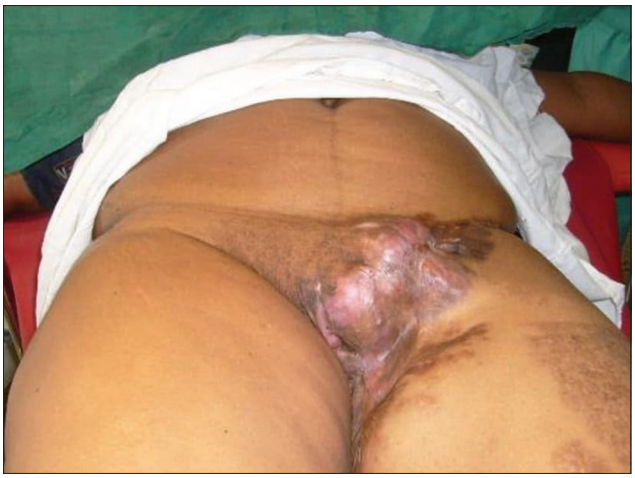

Figure 1: Recurrent lesion involving the left groin and thigh

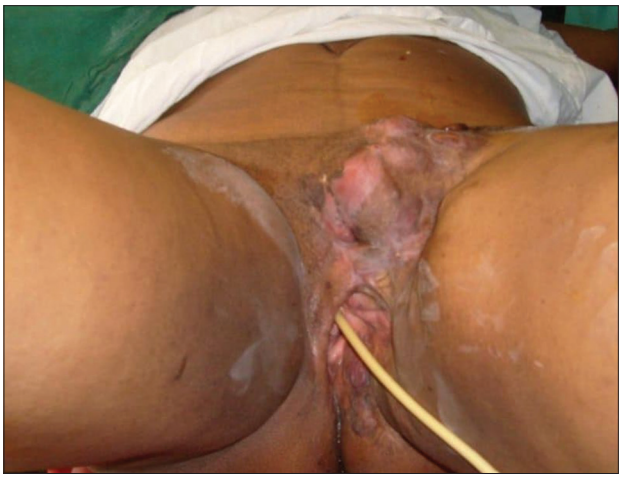

Figure 2: Lesion extending and affecting the vulva

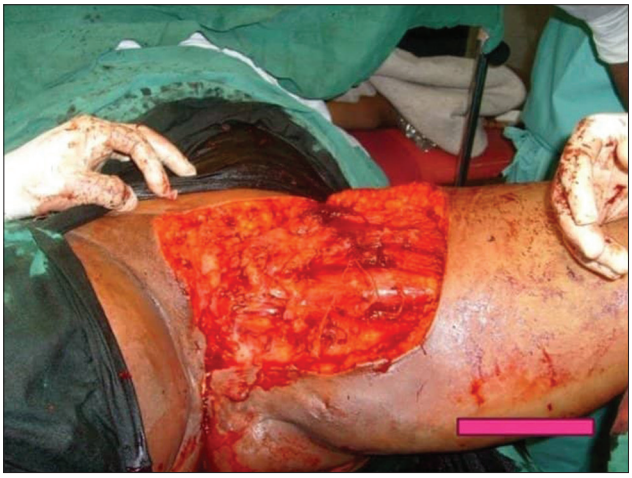

Figure 3: Wide Local Excision defect intraop

5 cm separated from the superior mass and below the inguinal ligament with a size of 6 × 8 cm, bosellated, irregular, and firm in consistency. Distal pulses were intact with no varicosities.

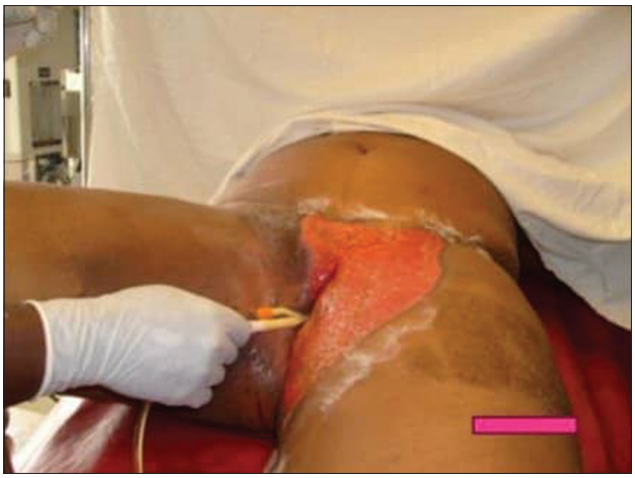

Figure 4: Post Excision granulation tissue after 14 days and ready for skin grafting

The blood pressure was persistently high (290/130 mmHg). Cardiology review was requested for and 24 h vanyl mendelic acid (VMA) assesment was done, and the report came back normal (2.3 mg/24 h (normal value: 0.4-15 mg)) to rule out phaechromocytoma. The blood pressure was stabilized with antihypertensives.

WLE down to the deep fascia involving the left labium majora under spinal anaesthesia after 5 cm margin was marked out [Figure 3]. The excision removed the roof of the femoral triangle exposing the femoral vessels. The middle 2/5 of the satorius muscle was excised because of tumor infiltration. The vessels were covered with the remaining upper 1/5 of satorius by suturing it to pectineus to achieve a graftable bed and prevent vesel desication. Since Mohs surgery was not available, we decided to prepare the bed for grafting. After 14 days, the granulation tissue was good for grafting [Figure 4]. Graft take was 100%. The patient was discharged and was followed up for 2 years, after which she was lost.

Discussion

Evidence has shown that high recurrence rate in DFSP reflects directly on the extent of resection.[7] The index patient had nine incomplete resections that resulted in multiple surgeries. Therefore, in the light of the aforementioned, undertreatment causes more morbidity, and hence, the surgery should not be undertaken by a nonspecialist who does not understand the pathology very well. It is imperative that a histological diagnosis be established before any intervention with an incisional biopsy or trucut if there are clinical features suggestive of this locally aggresssive tumor. This early diagnosis aids careful planning of resection and the consequent reconstructive needs.

It is evident that the most optimal treatment is Mohs micrographic surgery (MMS), in which under local aneasthesis, serial excisions of lesion are made until a tumor-free margin is achieved with simultaneous histological assesment.[8] The rate of recurrence for MMS is reduced markedly following this mode of treatment. In our low-resource setting where availability of MMS is limited or nonexistent, it may be wise to have a treatment protocol that will almost always achieve cure with the initial surgery. The pathological behavior of invading sorrounding tissues through tentacle-like projections into septae and fat tissues makes local agressiveness and destruction of tumor to be worrisome.[9] The projections usually look like normal tissues grossly making it hard to establish tumor-free margins without MMS.

Even though WLE is considered an overtreatment, it still shows a recurrence risk as high as 9.1% compared to that of MMS of 2.72%.[10] In a resource-poor setting, WLE is the most available and accesssible method of treatment because of the scarcity of equipment needed for the serial-frozen section. Ratner *et al*.[11] analyzed records of 58 patients with primary and recurrent DFSP treated with Mohs surgery and reported that 70% had positive margins with a 1 cm margin, 39.7% with 2 cm, 15.5% with 3 cm, and 5.2% with 5 cm margins. Therefore, excisions may have to go beyond 5 cm in WLE to come close to cure in the low-resource setting without MMS. The wide resection should be balanced by reconstructive needs. The recommended excision margin is 3 cm (range 1–5 cm).[12]

Therefore, before prescribing surgery, the surgeon should thoroughly explain to the patient about the risk of recurrence and hence possible reoperation. Surgery should not be carried out by any surgeon who does not understand the pathology of the tumor and has no structured training on the surgical care of tumor. The reoperation has severe implications for our patients because of cost and risk of losing them to follow-up. Many of the indigent patients will likely stop presenting and will later show up with severe disease. The surgeon factor that contributes to recurrence even when diagnosis is known is if the surgeon lacks reconstructive techniques. The surgeon should ascertain his skill and balance the excision with availability of reconstructive techniques in a ‘surgeon-poor setting’ now emanating because of migration of highly skilled practitioners to greener pastures.

The role of multidisciplinary care for tumor patients administered through a Multidisciplinary Tumor Board (MTB) offers the best of outcomes. MTB is a treatment planning approach in which a group of health-care professionals, who are experts in different

specialties, review and discuss the medical condition and treatment options of patients.[13]

If a surgeon can excise tumor but needs reconstructive support, proper and early consults to reconstructive surgeons are necessary in order to develop a useful surgical plan that will achieve cure were possible. The consults will depend on the site and location of tumor. A situation whereby the requisite support subspecialities are not available, where the patient presents appropriate and early referral to a center, and where all the necessary subspecialities are found should be considered.

The follow-up of 2 years for our patient was exceptional in the case. Obviously, the motivation for her compliance was because of frequency of recurrence prior to presentation. In our environment, follow-up is usually a challenge because of poverty and ignorance. The later is a big limitation to follow-up in our environment, and as such, every surgeon should take time to appropriately educate all patients before discharge on the need for compliance with such long follow-up in order to identify and treat early recurrences.

Conclusion

Adequate wide local excision is imperative in the low-resource setting to reduce the incidence of recurrence in DFSP. The surgery should be carried out by surgeons who are well trained on managing the tumor and defect. An MTB is most preferred for tumor management for optimal outcomes. Also, follow-up of such patients should be for life.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Dermatofibrosarcoma protuberans (DFSP). Available from: https://sarcoma.org.uk/about-sarcoma/what-is-sarcoma/types-of-sarcoma/dermatofibrosarcoma-protuberans-dfsp/. [Last accessed on 2023 Nov 22].
2. Hrudka J, Charvát M, Grossmann P, Kinkor Z. Dermatofibrosarcoma protuberans with fibrosarcomatous transformation: A case report. Cesk Patol 2020;56:89-93.
3. Hao X, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, *et al.* Dermatofibrosarcoma protuberans: Update on the diagnosis and treatment. J Clin Med 2020;9:1752.
4. Dermatofibrosarcoma Protuberans Clinical Presentation. Available from: https://emedicine.medscape.com/article/1100203-clinical?form=fpf#b2. [Last accessed on 2023 Nov 23].
5. Wiesmueller F, Agaimy A, Perrakis A, Arkudas A, Horch RE, Grützmann R, *et al.* Dermatofibrosarcoma protuberans: Surgical management of a challenging mesenchymal tumor. World J Surg Oncol 2019;17:90.
6. Thway K, Noujaim J, Jones RL, Fisher C. Dermatofibrosarcoma protuberans: Pathology, genetics, and potential therapeutic strategies. Anna Diagn Pathol 2016;25:64-71.
7. Efterpi CD, Dimitrios DD, John MK, Constantina P, Dimitrios KP. Multiple recurrent dermatofibroma protuberans of the hand. J Plast Reconstr Aesthet Surg 2008;61:842-5.
8. Menon G, Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2023. Available from: http://www.ncbi.nlm.nih.gov/books/NBK513305/.
9. Dermatofibrosarcoma Protuberans: Treatment-UpToDate. Available from: https://www.uptodate.com/contents/dermatofibrosarcoma-protuberans-treatment. [Last accessed on 2023 Dec 31].
10. Xingpei H, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, *et al.* Dermatofibrosarcoma Protuberans: Update on the Diagnosis and Treatment. J Clin Med 2020;9:1752.
11. Ratner D, Thomas CO, Johnson TM, Sondak VK, Hamilton TA, Nelson BR, *et al.* Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans. results of a multiinstitutional series with an analysis of the extent of microscopic spread. J Am Acad Dermatol 1997;37:600–13.
12. Kim BJ, Kim H, Jin US, Minn KW, Chang H. Wide Local excision for dermatofibrosarcoma protuberans: A single-center series of 90 patients. BioMed Res Int 2015;2015:642549.
13. Joan O, Bernstein C. Multidisciplinary tumor boards and guiding patient care: The AP role. J Adv Pract Oncol 2022;13:227–30.