International Journal of Research in Medical Sciences
Winarta GK et al. Int J Res Med Sci. 2019 Sep;7(9):3586-3590
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20193954

Case Report

Pitfall management of dermatofibrosarcoma protuberans on right leg

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Received: 21 July 2019
Revised: 09 August 2019
Accepted: 13 August 2019

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally invasive soft tissue sarcoma. The local recurrence rate is high, in some studies upwards of 60%, likely reflecting a failure to remove occult extensions of tumor. Surgical excision has been the treatment of choice for the resection of DFSP. Any pitfall on management therapy of DFSP not only increase the recurrence rate but also add new problems to patients with DFSP. 58-year-old male patient, came with a local recurrent of DFSP on his right leg. Then performed excision with margin one centimetre, and closing defect using pedicle sural flap, durante surgery turned pedicle on the flap is too short and because of poor soft tissue handling pedicle was injured. It consulted into the vascular division for evaluated and treatment pedicle. Postoperative evaluation, the flap experiences bluish due to vascularization compromised. It was decided to expose the flap and the defect was covered with skin graft. 2 months postoperative evaluation found local recurrent on skin graft area. Excision margins between 2 to 5 cm can reduce the recurrence rate. Proper planning in designing flaps to cover defects and the ability of good soft tissue handling is required to avoid new problems on management DFSP.

Keywords: Dermatofibrosarcoma protuberans, Local recurrent, Sural flap, Wide excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally invasive soft tissue sarcoma with a propensity for extensive subclinical involvement. Originating in the dermal layer of skin, microscopic tendrils of tumor may extend far beyond the margin of clinically evident tumor.

While painless and often indolent appearing, the tumor can grow quite large before receiving clinical attention. Local invasion can include subcutaneous tissue, muscle, fascia and bone.

The local recurrence rate is high, in some studies upwards of 60%, likely reflecting a failure to remove occult extensions of tumor.[1] Fortunately, distant metastasis is rare (between 1 and 4%) and complete removal is considered curative.[2]

CASE REPORT

By case report, 58-year-old male patient came with complaints of multiple lumps and ulcer appearing in the cruris anterior dextra area. History of lump excision on this similar area three months ago, from the results of examination pathology anatomy diagnosed with dermatofibrosarcoma protuberans.

Performed therapeutic management by performing re-wide excision. A wide-circle excision is performed with a one-centimeter excision margin of the outer lesion. and with the depth of excision to fascia layer.

To close the defect, it is taken from the sural flap. During durante operation, good soft tissue handling is very big role in the identification and keep the vascularization through pedicle flap remain good. On this case poor

operator soft tissue handling make the pedicle edema. but still try to close the defect by the sural flap that has been taken.

Figure 1: Local residif of DFSP on regio cruris anterior dextra.

Figure 2: Post Excision.

Figure 3: Edema Pedicle Sural Flap.

Evaluation of the third day post-operative, the flap starts to look bluish but still felt warm. The ten days there was a reddish flap as a sign of venous compensation failure. Then it was decided to expose the flap and close the defect with the skin graft.

Figure 4. Third Days Flap.

Figure 5. Ten Days Flap.

Figure 6: Local Residif DFSP On Skin Graft.

Follow up post-surgery, skin graft used to cover defects is growing well. But in second months follow up post-operative recovered local residif on skin graft area.

DISCUSSION

The cell of origin for DFSP is controversial. Several authors theorize that DFSP arises from fibroblasts, as tumor cell features that are consistent with modified fibroblasts have been observed on electron microscopy.[3] In addition, like fibroblasts, DFSP cells stain with vimentin and contain active endoplasmic reticulum that readily synthesize collagen.[4] However, several studies in tissue culture indicate that tumor cells may be histiocytes that have acquired fibroblastic elements.

The growth pattern of DFSP resembles that of fibroblasts in the body, which serve to support tissue through formation of a lattice network around cells. DFSP cells mimic the fibroblast infiltrative growth pattern with pseudopod like extensions from a central mass that penetrate fat and adjacent tissue over time.[3] It may be that this similarity to the fibroblast growth pattern explains the low rate of blood borne metastasis, as fibroblasts tend to remain enmeshed in the area they stabilize.[5] Histologically, DFSP is characterized by a fibroblastic proliferation of tumor cells arranged about a central hub in a storiform pattern.[2]

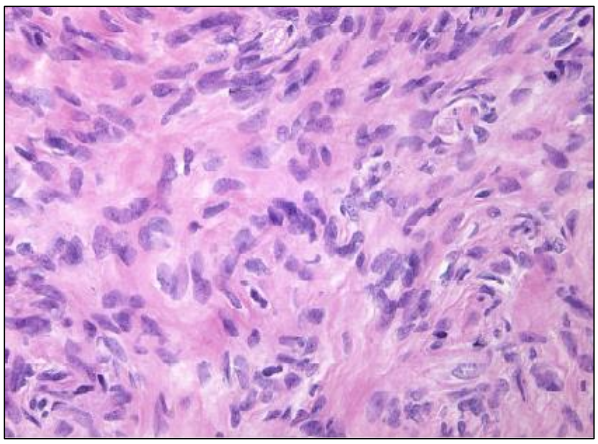

Figure 7: Microscopic Appearance of DFSP.[6]

Ninety percent of DFSP tumor cells exhibit a chromosomal translocation of genes COL1A9 and PDGFβ (t17;22,) which encode the alpha chain of type I collagen and the beta chain of platelet-derived growth factors, respectively.[6] The fusion protein produced by this translocation causes continuous stimulation of the PDGF protein tyrosine kinase, resulting in increased production of PDGF and abnormal cellular proliferation.[7] Fibrosarcomatous-DFSP is a more aggressive variant of DFSP and likely represents dedifferation. The increased cellularity and mitotic activity observed on histology for these tumors are indistinguishable from the cytologic and architectural pattern of a high grade fibrosarcoma, and this variant is associated with a higher rate of recurrence and metastasis.[6,8,9]

DFSP is a locally aggressive tumor characterized by low rate of metastasis and high capacity for local invasion. Consequently, the treatment of choice is resection with wide margins. In the early 1990s the gold standard of treatment for DFSP was surgical excision, with 3-5cm margins of healthy skin, while the underlying subcutaneous tissue, including fascia, was removed en block.[10]

Historically, surgical excision has been the treatment of choice for the resection of DFSP. However, the ideal width of margins remains undefined. Several authors have suggested that traditional surgical excision, which removes tissue in a concentric ring based on the macroscopic extent of the tumor, is not well suited for removal of DFSP.[10] This surgical approach is predisposed to remove too much healthy tissue without eradicating the extensive, asymmetric projections of tumor cells.[11] Most authors currently suggest a margin of 2 to 3 cm with a three dimensional resection including skin, subcutaneous tissue, and the underlying investing fascia.[8]

Excision with wide margins leads to improvement of the recurrence rate. When the surgical margins are at least 3cm and there is a three-dimensional resection that includes skin, subcutaneous tissue and the underlying fascia, the rate of recurrence is 20%.[12] It is necessary to be mentioned that in cases where the underlying bone structures are too close to the lesion, the periosteum and the portion of the bone may also need to be removed in order to achieve negative deep surgical margins. In cases where the margins were more than 5cm, the rate of recurrence was less than 5%.[10]

In the last several decades, Mohs surgery has emerged as a promising treatment option that may achieve superior results to surgical excision.[13] Mohs surgery provides a method of eradicating tumor that rests on intraoperative evaluation of tumor margins.[14] The tumor is resected in a stepwise fashion with tissue removal that is based on the presence of tumor cells. In addition to conserving tissue, the pathologic techniques used in Mohs surgery have been shown to provide an excellent rate of cure with very few documented recurrences.[11]

In this case the selection of excision margins that are only one centimeter from the outer border of the tumor may be due to consideration for easier closure of the defect. But for the case DFSP corresponds to some literature saying that wide excision margins are usually two to four centimeters can reduce the local number of recurent on DFSP. In this case it is likely that the small excision margin still leaves the tumor cell tissue causing local residif.

Mohs surgery seems to be very useful in cases where wide excision is not feasible. DFSP is considered as

radiosensitive tumor.[15] However, adjuvant radiotherapy is not extensively studied, even though it was found that has successfully contributed to local tumor control.[16] In the study of MD Anderson Cancer Center in 1998 the estimated rate of local control in 19 patients who received radiotherapy as adjuvant to surgical resection was 95% at 10 years.[17] It may be preferable in the latter case, patients with DFSP performed wide excision with a wide margin of about four centimeters. And, if possible, cooperate with pathologicanathomist during surgery by using mohs surgery methode. To ensure that excision restrictions are made free of tumor cell tissue, so there is no residual tumor.

After resection of DFSP, the National Comprehensive Cancer Network (NCCN) guidelines recommend immediate reconstruction in most cases, but state that it is preferable to delay deep undermining or flap reconstruction until negative surgical margins are assessed.[18] These recommendations for treatment of DFSP are potentially problematic because of the emphasis on clinical judgment for determining whether to perform immediate or delayed reconstruction. It is often difficult to predict the extension of the tumor, because of the eccentric pattern of invasion characteristic of DFSP that mimics normal tissue.[19] Immediate reconstruction can compromise options for subsequent surgery if positive margins found on permanent pathology necessitate further excision. In addition, immediate reconstruction in the context of residual tumor may pose a risk for the spread of microscopic disease.

In this case the selection of defect closure using a sural flap is due to the location of the adjacent defect. And vascularization of the sural flap is expected to help close the defect well. But the correct operation technique, and the skill of the operator in good soft tissue handling is very influential to pedicle which will guarantee vascularization of the flap to be used.

Surgical technique of sural flap are start from attention is directed to the posterior central third of the proximal leg where a skin island is created based on the dimensions and distance of the recipient site.

The flap should be created slightly larger than the defect needing to be covered and when establishing the margins of the flap, it is vital to maintain enough overall length to prevent tension or kinking during rotation of the flap. Finally, the peroneal perforators and lesser saphenous vein need to be identified.

Located superior to the lateral malleolus, the course of these vessels will influence the overall path of the pedicle and it is imperative to establish these vessels beforehand in order to preserve the major circulation and subsequent survival of the island flap. Dissection is begun proximally and carried down through to the deep fascial level until the sural nerve, artery, and lesser saphenous vein are identified.

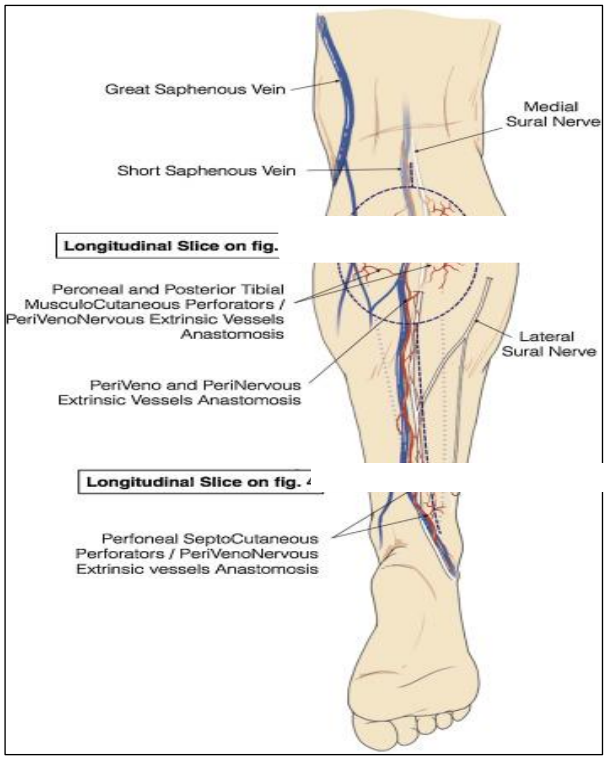

Figure 8: Anatomical of Sural Nerve.[20]

At this point, the island flap including the skin, subcutaneous tissue, and the deep fascial tissues containing the neurovascular structures, are elevated off the surface of the gastrocnemius muscle bellies. The pedicle is created as dissection continues distally, elevating these tissue layers as a single unit until the pivot point of the flap is reached. The pivot point is dependent upon the location of the recipient site. The pedicle is approximately 2 to 3 cm in width once dissected freely from the surrounding tissues. Once the flap is mobile, it is repositioned distally and sutured to the recipient site with care to not kink the flap as it is folded back upon itself.[20]

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally invasive soft tissue sarcoma with a propensity for extensive subclinical involvement. Surgical therapy is the modality therapy of DFSP. but there are some pitfalls to consider in order to avoid complications and local residual postoperative.

The determination of the margin of wide excision margin according to say some literatures about four centimeters can reduce the local residual number. At a central hospital with anatomy pathology specialists, the mohn surgery method can be used to ensure that excisional tissue boundaries are free of tumor cells. to ensure no residual tumor cells are at risk of inducing local residif.

Before carrying out extensive excision actions, it should also be noted about the action of reconstruction of the closure of the defect to be performed. The correct operation technique and soft tishue handling capability of

the operator when the flap action is very important. For pedicle and vascularization of the flap to be closed to close the defect as expected.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not Required

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Cite this article as: Winarta GK, Suryawisesa IBM. Pitfall management of dermatofibrosarcoma protuberans on right leg. Int J Res Med Sci 2019;7:3586-90.