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Dermatofibrosarcoma Protuberans (DFSP) of the Vulva: Mohs Micrographic Surgery or Radical Vulvectomy? – A Case Report

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Abstract

Background: Dermatofibrosarcoma Protuberans (DFSP) of the vulva is rare. Primary management is surgery. Positive margins are associated with higher recurrence rates. The disease characteristically requires large resections to achieve negative margins, which given the location of vulva is a challenging to devise the least morbid surgery. The management typically requires a multidisciplinary approach for resection or reconstruction.

Case Report: A 25-year-old patient with an incisional biopsy in the left vulvar region diagnosed as DFSP underwent Mohs Micrographic Surgery (MMS) and radical partial vulvectomy with the preservation of all genitals. A 1cm margin resection followed by Complete Circumferential and Peripheral Deep Margin Assessment (CCPDMA) was performed. During MMS, the margins were positive with each resection as it approached the vaginal wall and definitive MMS was aborted. She was then referred back to gynecologic oncology for radical surgical treatment. Subsequent anatomy-sparing radical partial vulvectomy was done and successfully cleared all the positive margins near the vagina and clitoris.

Conclusion: Young patient with DFSP of the vulva is challenging to decide surgical treatment between dermatology and gynecologic oncology. Recommend MMS approach if feasible and in the event of the need of more radical surgery by gynecologic oncology, consider a step-wise approach to preserve as much normal anatomy as possible.

Keywords: DFSP; Vulva; CCPDMA; Mohs; Surgical oncology; Gynaecological cancer

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon slow-growing sarcoma tumor of the skin and subcutaneous tissue that rarely metastasizes (fewer than 5% of the cases) but recurs frequently. The incidence rate of DFSP is estimated to be 0.8 to 5 cases per million per year, which usually occurs over the trunk, proximal extremities and scalp locations [1,2]. However, vulvar DFSP is extremely rare with less than 40 cases previously reported in the literature [3].

Surgical removal has been reported as the main therapeutic plan for DFSP treatment. Wide local excision, vulvectomy and Mohs Micrographic Surgery (MMS) are reported most in the literature for vulvar DFSP cases [3]. The main difference between these surgical options is the recurrence rate, functional damage and anesthesia method. MMS reported having a 98% cure rate of DFSP, while traditional gynecologic surgery may have a 20-49% recurrence rate [4]. Tissue conservation is fundamental to preserve function, especially for the clitoris [5]. MMS uses systematic sectioning with 100% of the tumor margin being microscopically assessed which can preserve all the healthy tissues while minimizing recurrence, maintaining aesthetic and functional outcomes. Traditional vulvectomy or wide local excision (WLE) surgery will perform recommended 3cm 3-dimensional specimen to obtain negative margins [6], thereby threatening the functionality, sexuality and aesthetical issue of the vulva. MMS performs with local anesthesia (LA) while traditional surgeries undergo general anesthesia (GA). Having MMS over the labium major with LA is much more challenging than other DFSP over extremities, shoulder, or scalp since the nerves are more sensitive and patient is fully conscious for the procedure. However, there is no consensus on the treatment and margin assessment for vulva DFSP currently.

We present a case where a new case of vulvar DFSP initial MMS was unsuccessful and was followed by serial partial radical vulvectomies with a priority of preserving normal anatomy in a young patient.

Case Report

A 25-year old otherwise healthy female patient without trauma history over the vulva area, presented with an asymptomatic left subcutaneous labium major mass for two years. She attended several gynecology consultations before, but regrettably, no invasive diagnostic were performed due to the absence of pain or irritating symptoms and low potential for developing malignant tumors over the vulva at such young age. The patient advocated for excision. The simple vulvectomy resection yielded a 3-4cm lesion that was removed piecemeal. Histopathology diagnosed as Dermatofibrosarcoma Protuberans (DFSP) and CD 34 positive. She was referred to gynecologic oncology. Whole-body PET-CT was performed and was negative for regional lymph node or systemic metastasis. She was presented at gynecologic oncology multidisciplinary tumor board recommend to resection with either radical vulvectomy or Mohs Micrographic Surgery (MMS). After discussing and counselling with the patient, MMS was performed. The procedure was stopped as the margin continued to be positive as it approached the clitoris and vaginal wall, necessitating a potentially more extensive surgery and reconstruction. Dermatologist surgeon estimated over 70% of the positive margins located closely to the wall of the vagina and therefore referred her back to gynecology oncology for radical surgery. Gynecologic oncology standard recommendation for an invasive lesion involves a wide-margin definitive resection, and for this case, would include the clitoris, portion of the vaginal wall and possible skin flap as part of reconstruction to achieve negative margin. This was appropriately upsetting and the patient considered not undergoing surgery. In line with the patients informed decision, a modified plan was made to start with an initial anatomy sparing approach to preserve the clitoris and vaginal wall.

Anatomy sparing partial radical vulvectomy was initially performed. Since the mass was subcutaneous, the approach involved resecting very little skin, so much so that her entire labia minora was preserved and the skin island was just a few cm in width. The deep subcutaneous tissue was completely removed down to the fascia and off the left clitoral horn and vaginal wall. The pathology report revealed negative margins at the vaginal wall and clitoris. There was a small focus of positive margin at the inferior aspect. Another partial radical vulvectomy was performed to clear the remaining positive margins. All genitals were preserved. Patient reported with good quality of life and sexual function is not affected much after repeated surgeries. After following up for eight months, the patient does not have any symptoms of recurrence.

Discussion

DFSP over vulva on young women under 30 years old is extremely rare. Limited literature can be found to support treatment decisions. There are only two reported cases. The first one is a 28-year-old female patient presented with a 5.0 cm lesion over the mons pubis, major vulvar reconstruction using skin expansion followed by skin and fat tissue flap was

performed [3]. The second case is a 23-year old female patient presented with a 4.0 cm mass over the right labium major and performed WLE and excisional biopsy [1]. There is just one MMS surgery performed for DFSP in the labium major is documented in 2009. However, this patient is 38 years old and the lesion is much smaller [5].

Apart from the MMS over the DFSP vulva case is limited, there is no failure of MMS reported simultaneously. The patient and our team found challenging to decide surgical treatments due to the limited evidence can be found to justify an option.

There is scarce literature presenting MMS and vulvectomy surgeries together over vulvar DFSP. The absence of choices for recommendation forced our patient to struggle between partial or complete radical vulvectomy for a while. Due to the high tendency of DFSP to infiltrate the surrounding tissue via microscopic projections, a very wide excision was highly recommended to get a higher cure rate. From previously documented cases and according to the margin assessment protocol, radical vulvectomy is suggested for any positive margins found close to the vagina. However, a traditional wide margin radical vulvectomy is a morbid procedure with significant impact on young female patient psychological and sexual quality of life.

Moreover, the limitation of treatment options may be ascribed to the delayed treatment among young patients. From previous literature, 43% of patients reported a long interval to diagnosis time (12.5 vs. 8.1 years) [N=143] [7]. Their sample was drawn from the Disease-specific Facebook support groups (FBSGs) which is a social media website where members with rare diseases such as dermatofibrosarcoma protuberans (DFSP) can share their stories internationally without time and location constraints. However, specific to vulvar DFSP on young women normally takes a few years from the onset of symptoms to diagnosis. From our presented case, the patient might be able to undergo MMS successfully if an earlier diagnosis can be made.

This case illustrates an example of a vulvar DFSP case, performed MMS and followed by radical partial vulvectomy to achieve the prognostically important negative margin while preserving the clitoris and in a young patient. Although the MMS failed to remove all the positive margins in this case, anatomy-sparing partial radical vulvectomy was successful in clearing the positive margins. This did require more than one surgery, however, preserving the functions and aesthetic issues was of high importance to this and many young patients. A step-wise progression of surgery is recommended for surgical treatment in lesions that present in areas that may require extensive surgery or reconstruction as DFSP is a slow-growing sarcoma, that rarely metastasizes, with overall good oncologic outcome if negative margins are achievable.

Conclusion

Vulvar DFSP is a very challenging diagnosis to the young patient due the disease characteristic of expansive microscopic margins that typically necessitate extensive surgical resection.

Any lesion or nodule developed over the vulva should be closely monitored and biopsy procedure should be performed. Smaller lesions may be safely resected with MMS. Multidisciplinary approach should be done in all cases for DFSP and with consideration of primarily utilizing MMS for functional and cosmetic rationale, though if not feasible consider a step-wise anatomy-sparing approach for patients where a more radical excision may significantly impact their quality of life for a highly curable entity.

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Ethical Statement

The patient has given her written informed consent to publish this case. Permission has been obtained from all named persons in acknowledgment.

Conflict of Interest

The authors have no conflicts of interest to declare.

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Author Contributions

Vivian Hui: Conceptualization, Writing-Original draft preparation, Editing.

Yen-Jen Wang: Writing- Reviewing and Editing.

Brian Orr: Supervision and Writing-Reviewing and Editing.

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