

Dermatofibrosarcoma Protuberans of Vulva, A Case Report and Review of Literature

Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma of dermal origin [1-3] that rarely presents in the vulva [1,2]. It typically presents on the trunk of young to middle-aged adults [4]. Even though it has a low potential for distant metastases, it often recurs locally [5]. Histopathologically, DFSP typically consists of spindle cells in a “storiform” pattern and stain positive for CD34, which can aid in establishing a diagnosis [1,6-8]. The traditional treatment of choice has been wide surgical excision, but multiple resections are often required for clearance of the lesion [9]. Mohs micrographic surgery (MMS) has recently been described as an alternative treatment option to help decrease the rate of recurrence.

Objective: To present a case of dermatofibrosarcoma protuberans of vulva in a young patient treated with excisional surgery with a systematic review of the condition.

Case: This patient is a 35-year-old female, G0P0, from The Republic of the Congo (Central Africa), who presented with an eight-year history of a “large left keloid of the mons pubis”. The patient stated the mass was non-tender and the size of a ping-pong ball that changed in size. The patient underwent surgical excision of the “large left keloid of mons pubis”. The specimen consisted of a 6 x 3 x 3 cm elevated portion of smooth tan skin with subcutaneous tissue. It was determined that the specimen was composed of extensive, ill-defined, firm, smooth, homogeneous tan tissue and was sent to pathology for further evaluation. The pathological specimen findings were “Dermatofibrosarcoma protuberans with positive margin”. A second surgery was performed to excise the positive margins of DFSP that remained. Review of an intraoperative frozen specimen found no tumor in the tissue margins. The patient had an uneventful postoperative course and was discharged two days after the surgery was performed.

Conclusion: Dermatofibrosarcoma protuberans (DFSP) infrequently involves the vulva and should be considered in the differential diagnosis of other spindle cell lesions presenting in this unusual site [10,11]. The role of immunohistochemical staining with CD34 is imperative in establishing the diagnosis. The rate of local recurrence is high, but it rarely shows metastasis [12]. Surgical excision is the treatment of choice and close follow-up to detect recurrence is imperative [1].

Keywords: Bednar Tumor; CD34 Positive; Dermatofibrosarcoma; Dermatofibrosarcoma Protuberans (DFSP); Factor XIIIa Negative; Vulvar Carcinoma

Case Report

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Background

Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma of dermal origin [1-3] that rarely presents in the vulva [1,2]. It is an uncommon soft tissue tumor that has a high local recurrence rate. The incidence of DFSP is 0.1% of all cancers and 1% of all soft tissue sarcomas [4]. It most commonly appears on the trunk or proximal extremities of young to middle aged adults. Surgical excision is the treatment of choice and early recognition is extremely important because of the excellent prognosis following adequate excision [13]. We report an unusual case of this tumor involving the vulva [1].

Presentation of the Case

This patient is a 35 years old female, G0P0, from The Republic of the Congo (Central Africa), who presented with an eight year history of a “large left keloid of mons pubis”. The patient stated the mass was non-tender and the size of a ping-pong ball with a changing size. The patient’s past surgical history was significant for two abdominal myomectomies in 2001 and in 2009 for symptomatic uterine leiomyomas. Her past medical history was significant for infertility, large uterine fibroids and oligomenorrhea after a myomectomy in 2009.

The patient underwent surgical excision of the “large left keloid of mons pubis”. The specimen consisted of a 6 x 3 x 3 cm elevated portion of smooth tan skin, with subcutaneous tissue. It was determined that the specimen was composed of extensive, ill-defined, firm, smooth, homogeneous tan tissue and was sent to pathology for further evaluation. The pathological specimen results came back as “Dermatofibrosarcoma protuberans with positive margin” (Figure 1). Subsequent excisional biopsy of the mass was performed.

Histologic evaluation of the specimen showed a spindle cell lesion consisting of fibroblast-like cells arranged in a storiform pattern [1,6-8] (Figure 2). On average, there were 2 to 3 mitotic figures per 10 high power field (HPF). The neoplastic cells showed extension into the surrounding fibro adipose tissue [14,15] (Figure 3). Based on the morphologic staining pattern, a diagnosis of Dermatofibrosarcoma protuberans (DFSP) was rendered [1]. The patient underwent a second resection of positive margin for Dermatofibrosarcoma protuberans of vulva (Figure 4). Wide local excision of tumor positive margin performed (Figure 5). An intraoperative frozen specimen was sent to pathology with negative margins (Figure 6). The patient had an uneventful postoperative course and was discharged on postoperative day two. Patient will be followed up in 6 months and then annually for revelation of possible recurrence.

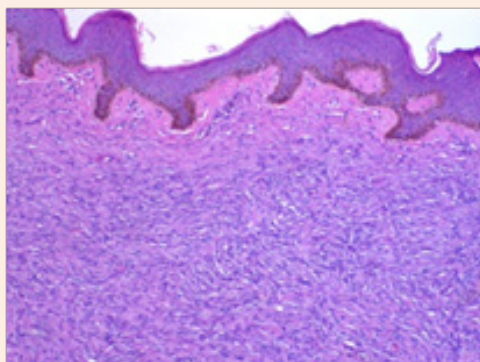


Figure 1: Histopathologic Slide, epidermis, basal layer and Tumor cells, Hematoxylin and Eosin Stain (H&E stain). Low power view demonstrating the tumor, The epidermis appeared thinned [32].

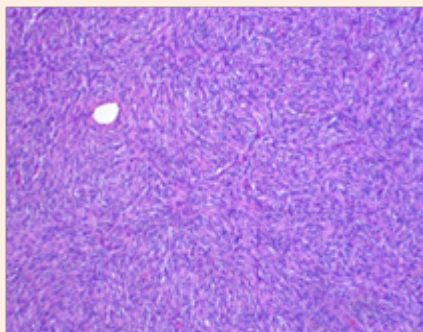


Figure 2: Histopathologic Slide, Tumor cells demonstrating a storiform pattern, Hematoxylin and Eosin Stain (H&E stain).

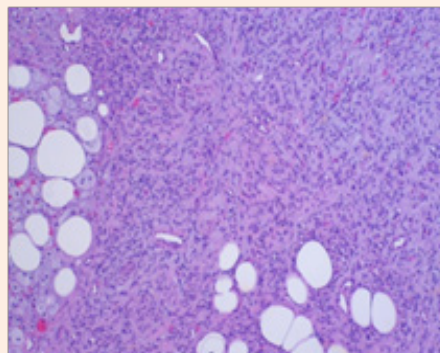


Figure 3: Histopathologic Slide, Tumor cells infiltrating into the subcutaneous fat, Hematoxylin and Eosin Stain (H&E stain).



Figure 4: Intraoperative Pictures: Wide Local Excision performed.



Figure 5: Intraoperative Pictures, wide local re-excision of positive margin performed and skin was approximated.

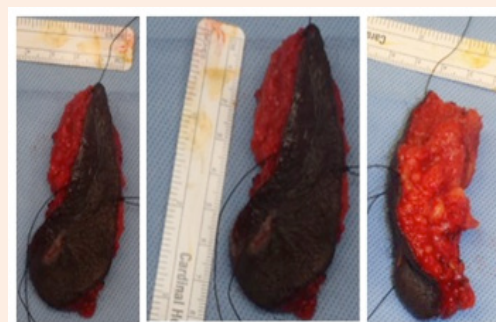


Figure 6: Intraoperative Pictures, specimen from wide local re-excision.

Discussion

DFSP is a rare and slow growing soft tissue tumor of cutaneous origin [4]. It most commonly arises on the trunk and proximal extremities of individuals between the ages of 20 and 50 [4,16]. It can also appear on the head and neck and rarely, it can occur on the vulva or perineum [17]. The cell of origin is believed to be either a dermal stem cell or an undifferentiated mesenchymal cell with fibroblastic features [4]. It rarely presents with distant metastases, but can be locally aggressive. Due to its locally aggressive nature, surgical excision is the treatment of choice but may require multiple excisions to achieve complete resection [18].

The prognosis for DFSP is generally a good with reported survival rates of 91-100% [19]. The deaths that have been reported were mainly due to extensive local spread resulting from inadequate excision. Other rare fatalities are result from metastases. Risk factors for a poor prognosis include an advanced age, high mitotic index and an increased cellularity [17].

DFSP represents only 0.1% of all cancers and 1% of all soft tissue sarcomas [4]. Its annual incidence is estimated to be between 0.8-4.5 cases per million in the general population of the United States [4]. It has been reported in all races, but it has a higher incidence in African Americans [4]. A number of reports have described an association of DFSP with trauma. Several cases to date state an association with sites of surgical scars, burn scars, sites of prior immunization, therapeutic irradiation and even a tattoo site [20].

DFSP usually occurs as a solitary lesion, but can also present with multiple foci [17]. The tumor is often solid and due to its indolent nature, often escapes detection in the early stages [21]. It often begins as a small, firm patch of skin that can be reddish, purplish, or even flesh-colored and grows slowly to become a nodule. It may also present with pain and ulceration if it is in the accelerated growth phase. Lymphatic spread is rare in DFSP. Metastatic DFSP occurs in less than 6% of cases and typically involves the lungs and bones [22].

Histopathologically, DFSP typically consists of spindle cells in a "storiform" pattern [1,6-8]. The fibroblasts are elongated and have small nuclei. The cells also display a uniform appearance with little nuclear pleomorphism and low to moderate mitotic activity [21]. DFSP not only infiltrates the dermis, but also the subcutaneous fat and the fascia [6,23]. Our patient's cells demonstrated this classic storiform pattern and also invaded the subcutaneous fat. Several histopathological variants of DFSP have also been reported, including pigmented DFSP and Fibrosarcomatous DFSP [24]. The pigmented variant of DFSP, also known as Bednar tumor, is identified by the presence of melanin-containing dendritic cells. The incidence of this variant is very low, estimated to be about 5% of all DFSP [24]. The Fibrosarcomatous variant of DFSP is identified when the tumor has areas that are indistinguishable from fibrosarcoma. While extremely rare, this form has a higher incidence of metastasis and therefore a worse prognosis. Another variant that has been described is one that contained a focus of endometriosis [25].

Immunohistochemistry is used to diagnose DFSP, because it can be histologically difficult to distinguish from other fibrohistiocytic

neoplasms, specifically fibrous histiocytoma and benign neural tumors [26]. A panel of immunohistochemical stains including CD34, S-100, melan-A, HMB-45, vimentin and smooth muscle actin (SMA) tested. The neoplastic cells will show diffuse staining with CD34 and vimentin, while the rest were negative. In DFSP, the tumor cells stain positive for CD34 and negative for factor XIIIa [27]. DFSP also has specific cytogenetic features that can be detected using reverse transcriptase polymerase chain reaction (RT-PCR) [28]. It expresses a reciprocal translocation between chromosomes 17 and 22 t(17; 22)(q22; q13) [29]. This translocation results in a fusion of the collagen type I alpha gene (COL1A1) on chromosome 17 with the platelet-derived growth factor B-chain gene (PDGFB) on chromosome 22 [28,30,31]. The resultant overproduction of PDGFB results in autocrine and paracrine stimulation of functional ligand production [32,33].

If an imaging study is warranted to diagnosis DFSP, magnetic resonance imaging (MRI) has been suggested as the best modality because it can clearly show soft tissue, can demonstrate the relationship of the tumor to adjacent structures and show extension and depth of the tumor [17]. It can also show any lymph node involvement.

Traditionally, the treatment of choice was wide local excision (WLE) extending 3 to 5 centimeters beyond the margin [2,34]. The recurrence rates are high after wide local excision, because the tumor often has microscopic projections into the surrounding tissue that extend beyond the central nodule [21]. However, more recent studies have shown that there is a lower recurrence rate with Mohs micrographic surgery (MMS) [35]. MMS allows mapping of the tumor along with microscopic examination of the lateral and deep margins [21]. In one study, DFSP treated with MMS had an overall recurrence rate of 1.6%, versus a 20% recurrence rate with wide excision and 43% after conservative excisions [6]. MMS also aids in tissue conservation. Chemotherapy has been shown to aid in therapy and radiation therapy has been demonstrated to have only a limited role in treatment [13]. Other forms of therapy, such as molecular target therapies like imatinib have been suggested [13]. Imatinib is an option for tumors that overexpress PDGFRB and may be helpful in cases with distant metastases or tumors that are difficult to access [33]. It is also an option for children when mutilating surgery is the only other option. Some authors have suggested that imatinib works by inducing apoptosis whereas others have suggested that it alters the tumor phenotype, decreasing tumor size and proliferation [33].

No matter which treatment option is chosen for DFSP, long-term follow up is necessary. Since recurrence rates are high, some studies have suggested that follow up should include an MRI in order to closely monitor for any sign of recurrence [36]. Some studies also suggest that patients should receive routine chest x-rays, because multiple local recurrences can increase the risk for lung metastases [36].

Conclusion

Dermatofibrosarcoma protuberans (DFSP) infrequently involves the vulva and should be considered in the differential diagnosis of other spindle cell lesions presenting in this unusual site [37]. The role of immunohistochemical staining with CD34

is imperative in establishing the diagnosis [14]. The rate of local recurrence is high, but rarely are metastatic lesions present [38,39]. The infrequent metastasis of fibrosarcoma associated with DFSP may be secondary to its superficial location, low cytologic grade, and small size [7]. Surgical excision is the treatment of choice and close follow-up to detect recurrence is necessary [1].

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