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Case Report

A Rare Case of Vulvar Dermatofibrosarcoma Protuberans: A Case Report

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ABSTRACT

Background
Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma of dermal origin with high local recurrence rate that rarely presents in the vulva. It affects adults between the second and fifth decade of life and the involved areas are most frequently the trunk, proximal extremities, head and neck.

Case Presentation
A 45-years-old Para 4 women presented with two raised masses on the right part of vulva for the last 17-month duration which increase in size for the last 6-month. First, incisional biopsy was taken from the mass and diagnosed as "*Suggestive of leiomyoma*". Following to this, excision of the whole mass was done and sent for histopathologic examination. The final histopathologic diagnosis became DFSP with the classic microscopic picture of a storiform and honeycomb pattern of monomorphic bland spindle cells.

Conclusion
Dermatofibrosarcoma protuberans infrequently involves the vulva and should be considered in the differential diagnosis of other spindle cell lesions presenting in this unusual site.

Keywords
Dermatofibrosarcoma protuberance; Leiomyoma; Vulva; Spindle cell lesions.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma of dermal origin that rarely presents in the vulva.[1] It is an uncommon soft tissue tumor that has a high local recurrence rate with an incidence of 0.1% of all cancers and 1% of all soft tissue sarcomas.[2] It affects adults between the second and fifth decade of life and the involved areas are most frequently the trunk, proximal extremities, head and neck.[3]

DFSP is flesh-colored or slightly yellow–brown skin tumour without epidermal invasion but with intracutaneous and subcutaneous spread. Sometimes the tumour presents as a reddish, flat elevated, firm lesion with irregular borders or multinodular appearance.[4]

Histopathology with immunohistochemical findings is used to diagnose DFSP.[3] As metastasis is rare, morbidity due to local recurrence is a more common issue.[5] The standard treatment techniques for resectable DFSP are complete surgical excision with wide local excisions of 2 or 3 cm tumor-free margins whereas; unresectable DFSPs are treated with radiation therapy, which improve local control and reduce the risk of recurrence post-operatively. so, early recognition of DFSP is extremely important because of the excellent prognosis following adequate excision.[6-9]

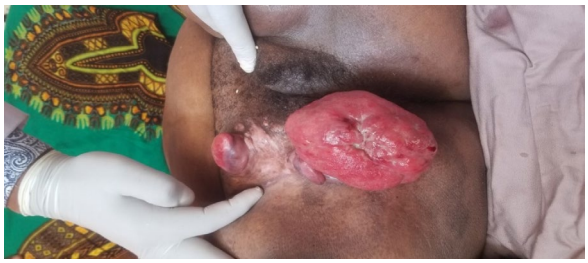
Here we report a new case of DFSP on vulvar area which was pre-operatively diagnosed with incisional histopathology to be Leiomyoma.

CASE PRESENTATION

A 45-years-old Para 4 women from rural area presented in Jimma University Medical Center (JUMC) for complaint of swelling on right vulvar area for the last 17-month duration which increase in size for the last 6-month. She visited local traditional healer 3-month back, ointment given to be applied daily base and started to have discharge with blood from the mass, itching sensation and pain.

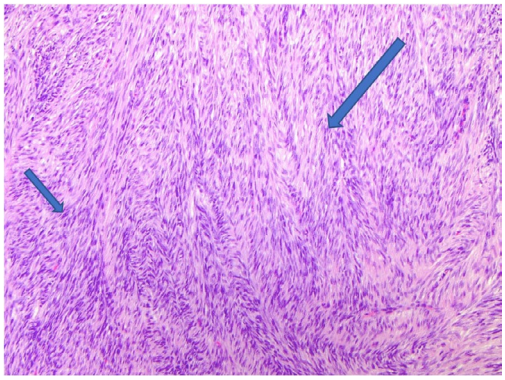
Her Gynecologic evaluation shows stable vital signs and two raised masses on the right part of vulva. The first one is 10 by 7 cm fleshy, non-tender, round mass with broad based pedicle, with discharge on the surface on the right side of vulva, along the labia majora and the second located on the right upper part of the vulva, non-tender measuring 3×3 cm which extends to subcutaneous tissue, but moves separately from underlying structure. The overlying skin of the mass is indurated with hypopigmentation (Figure 1).

Figure 1. *Picture Taken after the Mass has been Cleaned, Shows Mass Arise from Indurated Skin*



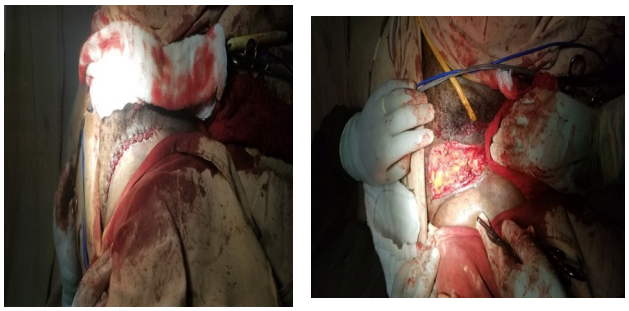
Initially Incisional biopsy taken from the mass and diagnosed as "*Suggestive of leiomyoma*" but due to rare occurrence of myoma on the vulvar area and difficulty of correlation with clinical presentation of our patient; re-biopsy was taken and the histologic examination showed with intersecting fascicles of bland spindle cells and diagnosed as leiomyoma again (Figure 2).

Figure 2. *Intersecting Fascicles of Bland Spindle Cells*



With the diagnosis of vulvar myoma the patient admitted to Gynecology ward, oncology side and she was put on perineal care BID (two times a day), the mass become clean and excision done 3-days after admission. Intra-operatively the mass with indurated skin localized clearly, incision put circumferentially, around both masses together, with 2-3 cm of margin to include indurated skin and there was plane of separation between the mass and underlying tissue, through which removal of the mass completed and sent for pathologic examination. The defect evaluated, 2 cm depth at the center, there was no remaining mass and changed skin, closed in two layers with vicryl no 2/0 in interrupted manner and the skin also closed with the same manner (Figure 3).

Figure 3. *Intraoperative Picture A Shows a Defect where the Mass Removed B, after the Defect Closed in Layer*

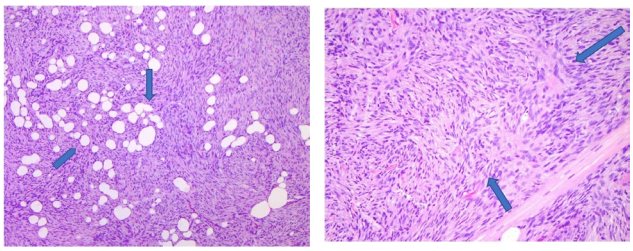


A B

Post-operatively the patient kept at bed rest for 72-hours to reduce tension on the surgical site and discharged on 7th post-operative day after she started ambulation fully and no problem with surgical site.

Histological examination of the excised mass demonstrated a storiform and honeycomb pattern composed of Monomorphic bland fibroblast like spindle cells extending into the surrounding fibro-adipose tissue with areas of Myxoid change and all margins were involved. Based on the morphologic pattern, a diagnosis of dermatofibrosarcoma protuberans with positive margin was rendered. After six-week of the procedure the patient is fine, the surgical site healed and started her regular sexual intercourse (Figure 4).

Figure 4. *Honeycomb Pattern Infiltrating in to Lobules of Subcutaneous fat (H and E,*100) A and B Uniform Population of Slender Fibroblasts Arranged in Storiform Pattern (H and E,*100 and 400) C and D*



A B

DISCUSSION

Dermatofibrosarcoma protuberans is an uncommon soft tissue tumor characterized by a low propensity for metastasis and a high rate of local recurrence, it rarely presents in the vulva.[10]

DFSP usually occurs as a solitary lesion, but can also present with multiple foci.[11] The tumor is often solid and due to its indolent nature, often escapes detection in the early stages.[12] DFSP has a distinctive histologic appearance but can mimic other diseases. The morphologic and molecular pathological findings of vulvar DFSP are similar to DFSP in other sites.[13,14] a variety of diagnoses, such as cellular dermatofibroma, cellular leiomyoma, neurofibroma, low-grade leiomyosarcoma, fibrosarcoma, low-grade

malignant schwannoma, desmoplastic melanoma, cellular neurofibroma, and low-grade malignant peripheral nerve sheet tumor were initially considered in some tumors later diagnosed as DFSP. This finding underscores the importance of proper recognition of the histologic features diagnostic of DFSP and the use of CD34 immunostaining to confirm this diagnosis.[15] Our patient was also diagnosed as leiomyoma initially.

DFSP is characterized by the presence of slender spindle cells arranged in a whorled pattern at the center of the tumor. The superficial portion of the tumor is hypocellular with a mixture of spindle cells and dermal collagen, whereas, in the tumor deep portion, the spindle cells infiltrate the fat creating a honeycomb appearance. Nuclear pleomorphism is absent, and the mitotic index is usually up to 5 mitoses per 10 high power fields (HPFs).[16,17] Our patient's cells demonstrated this classic storiform pattern with monotonous spindle cells and also invaded the subcutaneous fat. Histologically, several variants of DFSP have been described and it is important they are well-characterized to avoid misdiagnosis with other types of tumors. These variants include pigmented, myxoid, myoid, granular cell, sclerotic, atrophic DFSP and giant cell fibroblastoma.[18]

Immunohistochemically, DFSP is diffusely and strongly positive for CD34 and vimentin and negative for S-100, smooth muscle actin, desmin, keratin, and epithelial membrane antigen. In addition, it is usually negative for Factor XIII, although scattered cells can be positive for this marker. Translocation of chromosomes 17 and 22 (t (17:22)) is observed in over 90% of DFSPs.[19]

To ensure a correct diagnosis, initial biopsy samples must be analyzed using comprehensive immunohistochemical studies, such as CD34- staining, that are capable of distinguishing between DFSP and other spindle-cell tumors.[20]

The treatment, for DFSP of vulva, is based on surgery, whether primary or recurrent. The principal aim of surgery is to remove the tumor completely, because of the close relationship between residual tumor and local recurrence.[21]

CONCLUSION

This is a rare case vulvar DFSP that presented with clinical symptoms of vulvar carcinoma that was initially diagnosed as leiomyoma. 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.

INSTITUTIONAL REVIEW BOARD

This study has been approved by the Institutional Review Board (IRB).

CONSENT

We would like to thank our patient who consented to participate in this case report.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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