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NAVIGATING THE COMPLEXITY OF VULVAR DERMATOFIBROSARCOMA PROTUBERANS: A RARE CASE REPORT

Dr. Varsha. S. B* — Junior Resident, Department Of Obstetrics And Gynecology, Pushpagiri Institute Of Medical Science And Research, Thiruvalla, Kerala *Corresponding Author

Dr. Rachel Mathew — Associate Professor, Department Of Obstetrics And Gynecology, Pushpagiri Institute Of Medical Science And Research, Thiruvalla, Kerala

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma originating from the dermis. It is a rare soft tissue neoplasm. The incidence is 0.1% of all malignancies and 1% of all soft tissue sarcomas. It has local aggressiveness with a significant likelihood of local recurrence, although a reduced chance of distant metastasis. It primarily manifests on the trunk or extremities; vulvar involvement is uncommon. This patient presented with a vulvar mass which on excisional biopsy revealed to be a DFSP, underwent wide excision twice to get a tumour free margin. The case highlights the challenges in diagnosing and also achieving clear surgical margins due to the tumor's proximity to delicate structures and emphasizes the importance of multidisciplinary management in optimizing patient outcomes. A review of the existing literature on vulvar DFSP is also conducted to elucidate diagnostic difficulties, treatment strategies, and the need for tailored surgical approaches to minimize recurrence rates and enhance quality of life for affected patients. This report aims to contribute valuable insights into the management of this rare neoplasm, advocating for increased awareness and standardized treatment protocols.

KEYWORDS :

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare sarcoma of dermal origin that infrequently occurs in the vulva. It possesses a significant local recurrence rate. It impacts adults between their 20s to 50s, with the most commonly affected regions being the trunk, proximal extremities, head, and neck.

Due to the elevated local recurrence rate, surgical excision is the preferred treatment, and prompt identification is crucial because of the favourable prognosis following sufficient excision.

Case Report

A 42-year-old para 2 female approached with complaints of swelling in the right vulvar region for the previous 7 months, which has enlarged from the size of a peanut to around 5x5 cm over the last 3 months, accompanied by pricking pain for one month.

The overall examination was uneventful; nevertheless, the local examination identified a 5x5 cm smoothly marginated spherical mass in the upper portion of the right labia majora, exhibiting a firm to hard consistency and a negative slip sign (figure A). The overlying skin appeared normal. An excisional biopsy was conducted.

Histopathological examination revealed characteristics indicative of a fibroblastic/myo fibroblastic tumour of intermediate malignant potential, favouring dermatofibrosarcoma protuberans, with tumour cells extending to the margins. The patient was referred to an oncologist.

Immunohistochemistry performed, confirming diagnosis with CD34 positivity.

The patient was admitted to the Oncology department. A wide excision of the prior scar (excisional) on the right labia majora measuring 5 x 2 cm was performed (figure B), and the defect was repaired using a transposition flap (figure C and D).

She underwent excision for positive margins again and now under follow up.

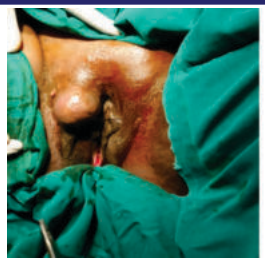
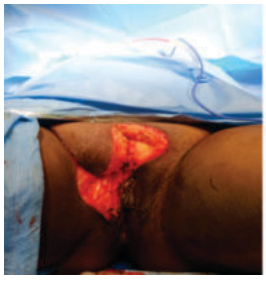


Figure A- Vulvar mass before excisional biopsy
Figure B- rewide excision of the previous excisional scar



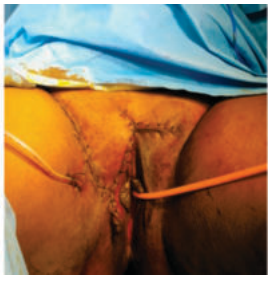


Figure C - Transpositional flap creation
Figure D- Reconstruction

Overview Of Reported Cases

The literature has less than 50 instances of vulvar DFSP, rendering this case especially important for comprehending the presentation, management, and consequences of DFSP in this rare site.

In 2010, Edelweiss et al. presented the most extensive case series with 13 individuals with histologically confirmed dermatofibrosarcoma protuberans (DFSP). Of the 13 patients, 12 exhibited a vulvar mass, with the right labia majora identified as the predominant site of origin in 8 cases, whereas one patient presented with a pigmented skin lesion. All patients underwent surgical management either excision biopsy, extensive local excision, or radical vulvectomy. Among them, 7 individuals experienced local recurrences, with one patient subsequently developing distant metastases to the lungs. All local recurrences were presented with the option of repeat surgery. During follow-up spanning 2 to 444 months, 9

patients remained disease-free, 2 were living with disease, one patient succumbed to metastasis, and another died from a different etiology. Invalid input. Please provide text for rewriting.

Goyal et al. (2021) documented a case with a 35-year-old female who underwent three surgical resections for recurrent DFSP, ultimately obtaining clear margins following a third extensive local excision. The patient was free of recurrence for two years following surgery, underscoring the significance of thorough margin evaluation in treatment approaches. In a separate study, two Iranian women, aged 35 and 37, had nodular masses in the labia majora, which were identified as dermatofibrosarcoma protuberans (DFSP) following excisional biopsy. Both underwent effective surgical excision, and the postoperative recovery was unremarkable. Subsequent imaging indicated no evidence of metastases, confirming the efficacy of surgical intervention in these instances. Hammonds and Hendi (2010) documented a case managed with Mohs micrographic surgery, facilitating accurate margin management and reducing defect size. This approach is especially advantageous for recurrent cancers, as it guarantees total excision while safeguarding adjacent healthy tissue. Their patient exhibited no recurrence following treatment, highlighting the efficacy of Mohs surgery in the management of DFSP.

DISCUSSION

Dermatofibrosarcoma protuberans typically presents as a painless, thickened area of skin (plaque) and/or a nodule that feels rubbery or firm to the touch and is attached to the underlying tissue. The lesion may appear reddish-brown or flesh-toned and usually develops gradually over several months to years. Tumour sizes can range from 0.5 to 25 cm in diameter, with 50 to 60 percent occurring on the trunk, particularly in the shoulder and chest areas. The absence of symptoms often leads to delays in diagnosis, with redness and pain reported in only 15% of cases. This condition is frequently misdiagnosed as other dermatological issues, especially in its early stages. Although uncommon, DFSP of the vulva must be considered in the differential diagnosis of vulvar masses, especially in instances of gradually growing, solid, nodular lesions. The lethargic characteristics of DFSP may result in a postponed diagnosis, frequently misidentified as benign conditions such vulvar cysts or lipomas, highlighting the necessity for heightened clinical vigilance in atypical instances.

Dermoscopy is not diagnostic since the characteristics of dermatofibrosarcoma protuberans are vague. The histopathological analysis of the skin biopsy reveals a distinctive microscopic appearance characterized by densely organized spindle-shaped cells. CD34 immunohistochemical testing yields a conclusive diagnosis. Complete eradication may be challenging to evaluate due to extensive extensions inside the skin and underlying structures. DFSP is characterized by the t (17; 22) (q22; q13) translocation, leading to COL1A1-PDGFB fusion transcripts in over 90% of cases.

Management characteristics encompass aggressive local resection with negative margins. DFSP lacks a standardized staging system. The AJCC guidelines for soft tissue sarcomas are frequently utilized. This method considers tumor size, grading, and nodal or distant metastatic disease. Mohs' microscopic surgery is indicated for minor lesions, however larger lesions necessitate a 2–3 cm margin of healthy tissue. Routine lymph node evaluation has not been documented for DFSP; however, positive lymph nodes carry prognostic significance. Extensive excision of the lesion encompassing the deep fascia, with a 1–3 cm margin of healthy skin. Multiple surgical interventions may be necessary to achieve total excision of a tumour. Radiotherapy is occasionally employed with surgery when a tumour cannot be entirely excised through surgical means. Chemotherapy is unproductive. Adjuvant therapies, including radiation or systemic treatment with tyrosine kinase inhibitors such as imatinib, may be contemplated in instances of positive surgical margins or when total excision is impracticable. Imatinib has demonstrated efficacy in DFSP owing to a translocation between chromosomes 17 and 22, resulting in the creation of the COL1A1-PDGFB fusion gene, which promotes tumour proliferation. In cases of localized vulvar DFSP with clear surgical margins, adjuvant therapy is typically unnecessary.

Local recurrences occur in 11–20% of instances, typically within 3 years post-surgery, therefore making follow-up essential. It is advised to occur biannually for five years, followed by annual assessments thereafter. The tumour metastasizes in only 5% of instances. It disseminates in 1% through lymphatic vessels to the regional lymph nodes and in 4% via the bloodstream, predominantly to the lungs, followed by the brain, bones, and heart.

CONCLUSION

DFSP is an uncommon phenomenon, particularly in the vulvar region. The clinical manifestation of vulvar DFSP can be insidious, frequently presenting as a hard, nodular lesion that may first be misidentified as more prevalent vulvar diseases such as Bartholin cysts, lipomas, or vulvar fibromas. The indolent growth of DFSP can prolong diagnosis, as it may not initially elicit suspicion of malignancy. The patient's lesion was first misdiagnosed, underscoring the necessity of a heightened suspicion for DFSP while assessing unusual vulvar tumours.

The differential diagnosis of vulvar DFSP include various soft tissue sarcomas, dermatofibromas, and severe angiomyxomas. Histopathological analysis, in conjunction with immunohistochemistry, is essential for precise diagnosis. DFSP generally has a storiform arrangement of spindle cells and expresses CD34, aiding in its differentiation from other spindle cell neoplasms.

Surgical excision with clear margins is the preferred treatment, as the overall prognosis is contingent upon it. The incidence of local recurrence is elevated; however, metastatic lesions are infrequently observed. Therefore, regular clinical follow-up is essential to identify and characterize any tumour recurrence. Additional research and case reports are essential to enhance the understanding of DFSP behaviour in this atypical anatomical site and to refine treatment options.

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