Journal Homepage: - www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/16408
DOI URL: http://dx.doi.org/10.21474/IJAR01/16408

RESEARCH ARTICLE

VULVAR DARIER-FERRAND DERMATOFIBROSARCOMA: UNUSUAL LOCALIZATION OF A RARE TUMOR

Achraf Jarroug, Soukaina El Aziz, Yasser Ait Benkaddour and Abderaouf Soummani

Manuscript Info

Manuscript History
Received: 10 January 2023
Final Accepted: 14 February 2023
Published: March 2023

Abstract

Dermatofibrosarcoma protuberans (DFSP) is a low-to-intermediate grade sarcoma of dermal origin that rarely presents in the vulva, typically occurring on the trunk of young to middle-aged adults. Even though it has a low potential for distant metastases, it often recurs locally. Therefore, surgical excision is the treatment of choice. We report a case of DFSP with fibrosarcoma transformation involving the vulva treated by large excision with tumor free margins followed by plastic reconstruction. Our object is to highlight this rare disease and through literature evaluate the safety and efficacy of surgical treatment. Early recognition of this rare entity whose localization and the aggressive nature of the fibrosarcomatous component will be an issue in the surgical management.

Introduction:-

Vulvar cancer accounts for about 0.4% of all cancers, with preference for older-aged women in the seventh decade of life. The most common histological type is the squamous cell carcinoma, diagnosed in 90% of cases, followed by melanomas,extra mammary Paget's disease, adenocarcinomas, basocelular carcinoma,sarcomas and undifferentiated carcinomas .

Primary gynecological sarcomas are infrequent and account for 3 to 4% of all the malignant neoplasm of the female genital tract, with anatomical distribution as follows: uterus 85% of the cases, ovary 8%, vulva and vagina 5%, and the remaining 2% in other gynecologic organs, being leiomyosarcoma the most frequently seen histological type, followed by endometrial stromal sarcoma.

The scientific literature has reported fewer than 60 cases of dermatofibrosarcoma protuberans (DFSP) with an extremely rare location in the area of the vulva [1] , according to the new WHO classification of soft tissue tumors, it belongs to the category of fibroblastic/ myofibroblastic tumors with differentiation of intermediate biological potential, being locally aggressive, marked tendency for local recurrence and extremely infrequent distant metastases [2] .

Patient And Observation:-

A 52-year-old woman presented with a growing mass in the vulvar area. She complained of slow but progressive growth for the past 11 months. The patient had no medical or surgical history. She mentioned that the cutaneous lesion had started as a small plaque on the vulva . Given the continued growth of the tumor and the pain associated, the patient consulted our department. Physical examination revealed a 20 cm*18 cm firm mass of elastic

Corresponding Author:- Achraf Jarroug

consistency on all the surface of the vulva . Non-contrast and contrast-enhancing MRI were obtained reporting an expansive growth tumor in the vulva, solid in appearance and infiltrating the subcutaneous tissue . After an excisional biopsy of the vulvar lesion, histological analysis showed a cellular neoplasm composed of plump spindle cells arranged in a storiform pattern, scattered pleomorphic cells. The oncologic gynecology department decided to surgically excise the lesion with circumferential peripheral and deep margins . The tumor was radically excised with free margins of 1cm reconstruction by a lotus petal flap was performed by the plastic surgeons . On sectioning, the tumor was apparently well delineated, brown in color and of firm consistency, measuring 22 x 18 x 12cm .The microscopic examination shows a proliferation of spindle cells with hyperchromatic nuclei, organized in a storiform pattern with frequent mitotic (6 mitoses per 10 high-power fields), infiltrating from the reticular dermis to the subcutaneous cellular tissue. No tumor necrosis was identified, nor lymphovascular or perineural invasion. Margins were lesion free.

Immunohistochemical stains revealed diffuse CD34 positivity. Tumor cells show higher nuclear positivity to Ki67 in hypercellular areas than in DFSP typical areas . Based on the histopathologic and immunohistochemical findings, a diagnosis of dermatofibrosarcomaprotuberans with fibrosarcomatous transformation (FS-DFSP) was made, having tumor-free resection margins.

The patient consulted a month later with a healed incision and good esthetic state. Regular evaluations are being performed for local recurrence and distant metastasis.

Discussion:-

Dermatofibrosarcoma protuberans (DFSP), previously considered in the World Health Organization classification of tumors of soft tissue [3]. As a neoplasm of fibrohistiocytic differentiation, is now listed as a tumor of fibroblastic/myofibroblastic differentiation in the 2013 edition of the classification [4]. It is a superficial low-grade sarcoma of intermediate malignancy that typically arises on the trunk and proximal extremities. Even though it has a low potential for distant metastases [5], it has a high frequency for local recurrence due to diffuse infiltration of the dermis and subcutis [4]. DFSP of the vulva is extremely rare and mostly affects women in the fourth or fifth decades of life. The labia majora was most commonly affected. Patients generally presented with a 4.2cm (average) firm, asymptomatic mass. Lesions presented with erythema, hyperpigmentation, ulceration, or even of orange peel skin appearance. Patients reported slow to minimal change in size (ranging from 3 months to 10 years) [6]. As with all solid tumors, clinical suspicion is confirmed by biopsy. Histopathogically, DFSP shows a distinct "storiform" or "cartwheel" arrangement of uniform appearing fibroblasts. Immunohistochemical staining demonstrates strong positivity for CD34 (sensitivity 84-100%) and vimentin [7]. DFSP can rarely present as a more aggressive fibrosarcomatous variant which occurs in around 10-15% of cases. Areas of fibrosarcoma are characterized by increased cellularity and loss of CD34 imunopositivity. Patients with this more aggressive variant tend to be more prone both to local recurrence and distant metastasis. Currently, only around 150 cases of transformed DFSP have been reported with distant metastasis in 13% [8] . The initial treatment of choice for DFSP is surgery. It must be extensive as to remove the tumor completely, given its proclivity for irregular and often deep subclinical extensions. Re-resection is recommended if the initial surgery did not achieve clear margins [9]. The surgical approach to DFSP must be meticulously planned. Size, location of the tumor and fibrosarcomatous transformation, as well as cosmetic issues, will dictate the most appropriate surgical procedure. Mohs or modified Mohs surgery and traditional wide excision, typically with 2 to 4cm margins to investing fascia that is subsequently verified to be clear by traditional pathologic examination, are all methods to achieve complete histological assessment [10]. A large retrospective series of 204 patients with DFSP showed a very low local recurrence rate (1%) using wide excision with a standardized surgical approach, underscoring the importance of meticulous pathologic margin evaluation with any surgical technique [11]. A systematic review found a lower rate of recurrence with Mohs surgery compared to wide local excision [12].

Radiation has occasionally been used as a primary therapeutic modality for DFSP, but it is more commonly used as adjuvant therapy after surgery [13]. Postoperative radiation therapy is a preferred option for positive surgical margins if further resection is not feasible. If a negative margin was achieved, no adjuvant treatment is necessary. No matter which treatment option is chosen for DFSP, long-term follow up is necessary. NCCN 2018 recommends a physical exam with a focus on primary site every 6-12 months with patient education about regular self-exam. As for DFSP-FS it is recommended that the patient undergoes a physical exam every 3 to 6 months for 2 to 3 years then annually [14]. Since recurrence rates are high, some studies have suggested that follow up should include magnetic resonance imaging (MRI) to monitor for any sign of recurrence closely . Some studies also suggest that patients

should receive routine chest x-rays because multiple local recurrences as well as the fibrosarcomatous transformation can increase the risk for lung metastases [14].

Conclusion:-

By way of conclusion, dermatofibrosarcoma protuberans in the vulva is a rare pathological condition affecting the adult female population locally devastating and with a tendency to local recurrence, therefore . It requires a multidisciplinary approach to identify the right diagnosis and indicate individualized treatment, through highly effective Surgical excision with free margins is the treatment of choice given that the entire prognosis depends on it . The rate of local recurrence is high, but rarely are metastatic lesions present thus a frequent clinical follow-up is imperative in this rare case to detect and diagnose any recurrence of the tumor, especially for cases with a fibrosarcomatous transformation.

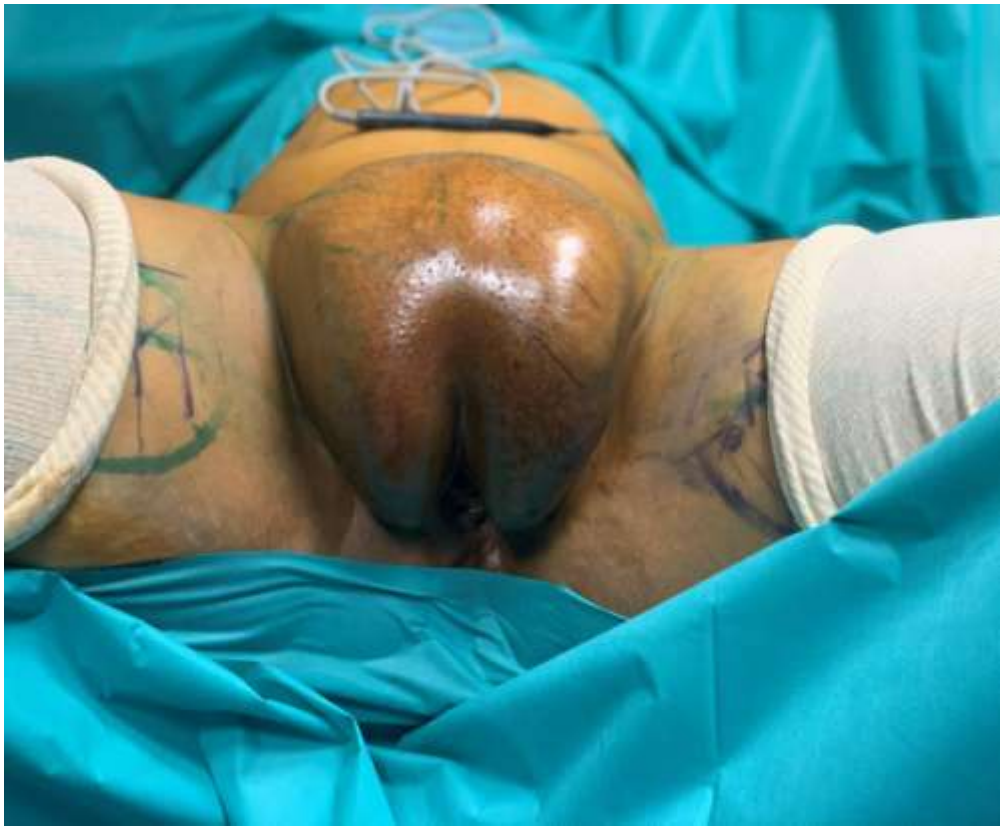

Figure 1:- A 20*18 cm tumor on the total surface of vulva.

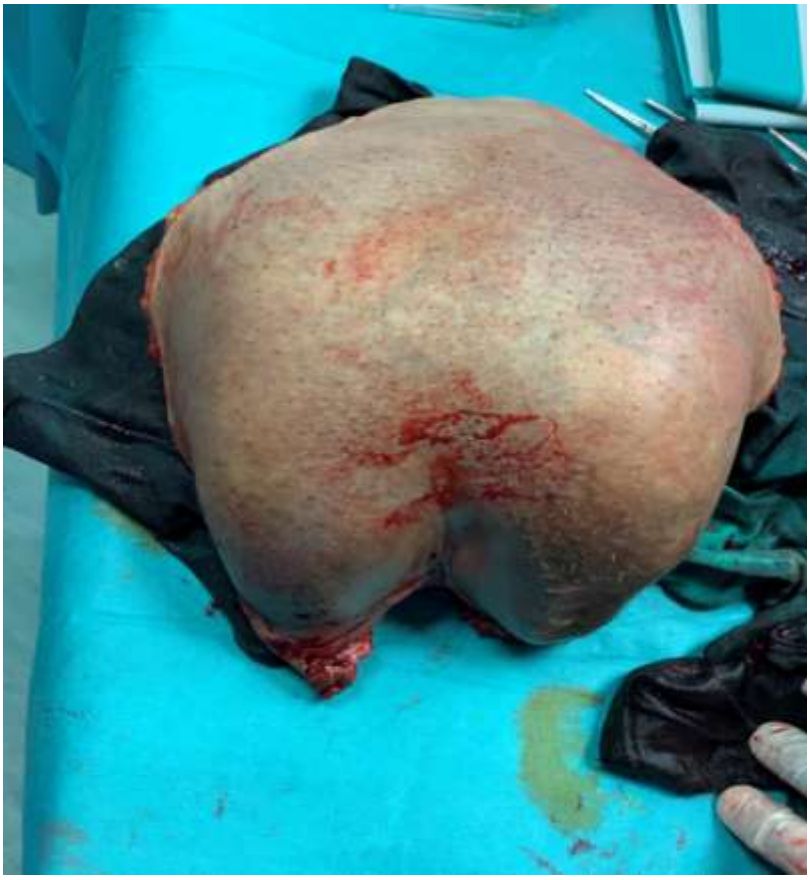

Figure 2:- Total tumor after resection .

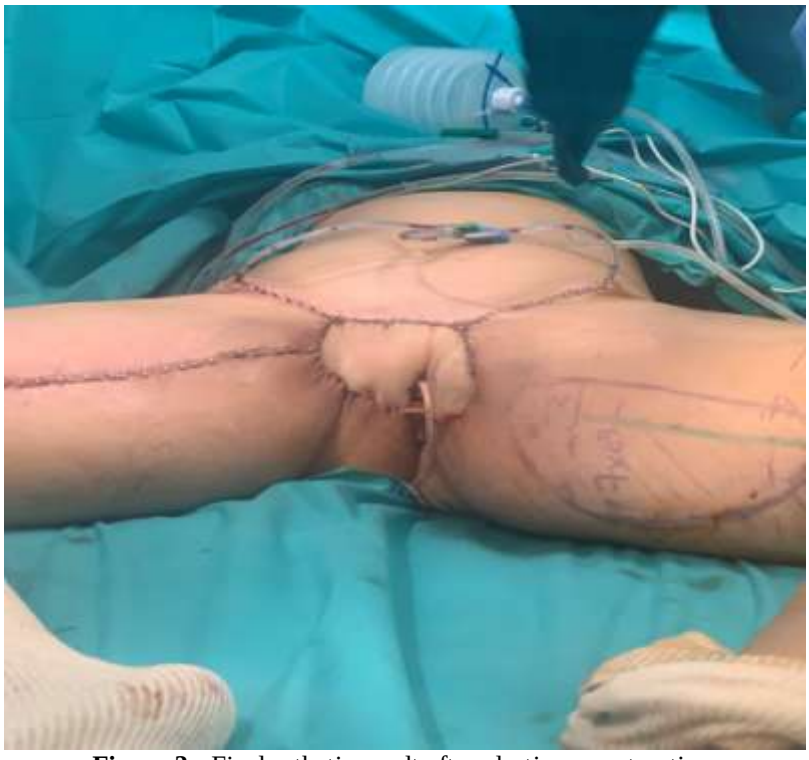

Figure 3:- Final esthetic result after plastic reconstruction.

References:-

1-Jeremic J, Stefanovic A, Jeremic K, et al. Giant dermato fibrosarcoma protuberans vulvae: Rare clinical presentation and literature review. J BUON 2019; 24:1289-95.
2-Neff R, Collins R, Backes F. Gynecologic oncology reports. Dermato fibrosarcoma protuberans: A rare and devastating tumor of the vulva. Gynecol Oncol Rep 2019; 28: 9-11. 9. Allen
3. Fletcher CD, Unni KK, Mertens F. Pathology and genetics of tumours of soft tissue and bone. Iarc. 2002. **Google Scholar**
4. Fletcher C. WHO classification of tumours of soft tissue and bone: [this book reflects the views of a working group that convened for a consensus and editorial meeting at the University of Zurich, Switzerland, 18-20 April 2012]. Internet Agency for Research on Cancer. 2013.
5. Bhambri S, Desai A, Del Rosso JQ, Mobini N. Dermatofibrosarcoma protuberans: a case report and review of the literature. The Journal of clinical and aesthetic dermatology. 2008; 1(1): 34. **PubMed** | **Google Scholar**
6. Nguyen AH, Detty SQ, Gonzaga MI, Huerter C. Clinical Features and Treatment of Dermatofibrosarcoma Protuberans Affecting the Vulva: a Lliterature review. Dermatologic surgery: official publication for American Society for Dermatologic Surgery. 2017; 43(6): 771-4. **PubMed** | **Google Scholar**
7. Gatlin JL, Hosch R, Khan M. Dermatofibrosarcoma protuberans of the scalp with fibrosarcomatous degeneration and pulmonary metastasis. Journal of clinical imaging science. 2011; 1: 55. **PubMed** |**Google Scholar**
8. Voth H, Landsberg J, Hinz T, Wenzel J, Bieber T, Reinhard G et al. Management of dermatofibrosarcoma protuberans with fibrosarcomatous transformation: an evidence-based review of the literature. Journal of the European Academy of Dermatology and Venereology: JEADV. 2011;
9. Oge T, Benedicic C, Tamussino K, Regauer S. Dermatofibrosarcoma protuberans of the vulva: a case report. BMJ case reports. 2009; 2009. **PubMed** | **Google Scholar**
10. Farma JM, Ammori JB, Zager JS, Marzban SS, Bui MM, Bichakjian CK et al . Dermatofibrosarcoma protuberans: how wide should we resect: annals of surgical oncology. 2010; 17(8): 2112- 8. **PubMed**|**Google Scholar**
11. Foroozan M, Sei JF, Amini M, Beauchet A, Saiag P. Efficacy of Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans: systematic review. Arch Dermatol. 2012; 148(9): 1055-63. **PubMed** | **Google Scholar**
12. Hamid R, Hafeez A, Darzi MA, Zaroo I, Rasool A, Rashid H. Outcome of wide local excision in dermatofibrosarcoma protuberans and use of radiotherapy for margin-positive disease. Indian dermatology online journal. 2013; 4(2): 93-6. **PubMed** | **Google Scholar**
13. von Mehren M, Randall RL, Benjamin RS, Boles S, Bui MM, Ganjoo KN et al . Soft Tissue Sarcoma, Version 2 2018, NCCN Clinical Practice Guidelines in Oncology. Journal of the National Comprehensive Cancer Network: JNCCN. 2018; 16(5): 536- 63. **PubMed** | **Google Scholar**
14. Ohlinger R, Kuhl A, Schwesinger G, Bock P, Lorenz G, Kohler G. Dermatofibrosarcoma protuberans of the vulva. Acta obstetricia et gynecologica Scandinavica. 2004; 83(7): 685- 6. **PubMed** | **Google Scholar**