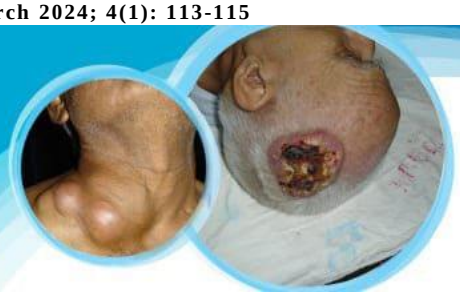
International Journal of Oncology Research



**ISSN Print:** 2664-665X
**ISSN Online:** 2664-6668
IJOR 2024; 4(1): 113-115
www.oncologyjournal.in
Received: 21-05-2024
Accepted: 26-06-2024

**S Khunda**
FRCS (Ed), FRCOG, Professor Emeritus, Baghdad University, Iraq

**Dr. Nawfal Azzo**
FIBOG, Consultant Gynecologist and Director, Department of Gynecology and Oncology, Baghdad Teaching Hospital, Medical City, Baghdad, Iraq

**Dr. Nagham Ibraheem**
CABOG, Fellowship in Gynecologic Oncology, Baghdad Teaching Hospital, Iraq

**Dr. Rasha Fadhil Abas**
MRCOG, CABOG, JMCOG, Fellowship in Gynecologic Oncology, Baghdad Teaching Hospital, Iraq

**Corresponding Author:**
**Dr. Rasha Fadhil Abas**
MRCOG, CABOG, JMCOG, Fellowship in Gynecologic Oncology, Baghdad Teaching Hospital, Iraq

# Treatment of dermatofibrosarcoma protuberans and childbirth

**S Khunda, Nawfal Azzo, Nagham Ibraheem and Rasha Fadhil Abas**

**DOI:** https://doi.org/10.33545/2664665X.2024.v4.i1b.24

**Abstract**
Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor often presenting as a vulval nodule with intact skin but subcutaneous involvement. Initial resections typically have limited tumor-free margins due to misdiagnosis as benign. Once confirmed histologically, wide excision with 2–3 cm margins is the standard treatment to minimize local recurrence, which is common when margins are inadequate. DFSP rarely metastasizes early, and regional lymphatic involvement is uncommon, making routine lymphadenectomy unnecessary. Histological variants, such as fibrosarcomatous changes, may increase aggressiveness. Microscopically, DFSP infiltrates the dermis with projections extending beyond visible margins. Imatinib, a tyrosine kinase inhibitor, is effective in metastatic cases but does not improve overall survival. Despite the rarity of vulval DFSP, treatment principles align with those for other skin sites, though vulvar excisions may disrupt anatomy. Literature suggests low psychosexual impact post-surgery. In our 25-year-old patient, initial excision failed to achieve clear margins, prompting vulvectomy and bilateral lymphadenectomy. Despite this, her sexual activity was unaffected, and she delivered a child via cesarean section two years later. Four years post-treatment, she remains disease-free, reflecting the importance of early diagnosis, adequate margins, and tailored surgical approaches in DFSP management.

**Keywords:** Dermatofibrosarcoma protuberans (DFSP), vulval nodule, soft tissue tumor wide excision

## Introduction

## Case History

A 25-year-old woman, gravida 1, delivered via cesarean section, presented at the oncology clinic of Medical City Hospital in March 2019 with a complaint of a small (3 cm) subcutaneous mass on the right side of the labia majora. The mass was rounded, firm, mobile, and non-tender, with a healed skin incision covering it. The patient provided a biopsy report from a previously excised tumor, which was diagnosed as dermatofibrosarcoma protuberans (DFSP) with no margin free of the resected tumor mass.

## Histopathology Report

The biopsy confirmed DFSP, incompletely excised with no tumor-free margins.
On examination, a new mass measuring approximately 10 x 15 mm was observed at the site of the previous excision. MRI imaging revealed multiple right-side inguinal lymph nodes.

## Investigations

- Renal and liver function tests: Normal
- Complete blood count: Normal
- Chest X-ray: Normal
- Abdominal and pelvic ultrasound and MRI: Normal except for right inguinal lymphadenopathy

The case was discussed at the oncology multidisciplinary team (MDT) meeting at Baghdad Teaching Hospital. A decision was made to proceed with radical surgery.
On April 24, 2019, the oncology team at Baghdad Teaching Hospital performed a vulvectomy with bilateral inguinal lymphadenectomy. The intraoperative and postoperative periods were uneventful, with good wound healing.

A Foley catheter was removed 48 hours after surgery, and urinary function was normal. The patient was discharged six days postoperatively.

The histology report of the radical surgery revealed the following findings:

- **Lesion site:** Dermatofibrosarcoma protuberans with tumor-free margins.
- **Lymph nodes:** All lymph nodes were free of tumor involvement.

The patient has been on regular follow-up for the past four years without the need for further treatment. She conceived spontaneously and delivered a healthy female baby via cesarean section at 37 weeks on October 30, 2020.

## Discussion

Primary sarcoma of the vulva is a rare disease, accounting for about 1.3% of vulval malignancies (Disaia *et al*., 1971) [1]. Among these, dermatofibrosarcoma protuberans (DFSP) is considered one of the most common vulval sarcomas, comprising approximately 27% of vulval sarcoma cases as reported by Sarah Jonson (2020) [2]. However, Canas *et al*. (1996) [3] consider DFSP to be among the least common vulval sarcomas. Aartsen *et al*. (1994) [4], in a study of 74 cases of vulval sarcomas, identified only 9 cases of DFSP.

DFSP typically presents as a nodule or polyp projecting from the vulval skin, attached to subcutaneous tissue, but with intact overlying skin. Initially, these lesions are often resected with limited tumor-free margins, as they are mistakenly treated as benign lesions. Once a histological diagnosis of DFSP is confirmed, a wider excision with 2–3 cm tumor-free margins is recommended.

The natural history of this rare disease is marked by striking local recurrence (Canas *et al*., 1996) [3]. However, distant metastases are uncommon in the early stages of the disease. Local recurrence largely depends on achieving adequate tumor-free margins during the primary excision. Some DFSP tumors may show histological variations, such as leiomyosarcoma or fibrosarcoma components, which render them more aggressive (Rayan *et al*., 2015) [5].

The histological characteristics of DFSP remain controversial. Immunohistochemical studies over the years have suggested a fibroblastic origin, as evidenced by tumor reactivity to vimentin and negativity for histiocytic markers (Canas *et al*., 1996) [3]. Microscopically, DFSP infiltrates the dermis and appears poorly delineated, with projections extending into surrounding dermal tissue, far from the macroscopically visible mass. Canas *et al*. (1996) [3] noted fewer than 50 cases of vulval DFSP reported in the literature, as summarized by Edelweiss *et al*. (2010) [6]. This rarity makes establishing a standard treatment challenging, although DFSP is characterized by a low incidence of distant metastases, which generally occur late.

Regional lymphatic involvement is uncommon and may not necessitate lymphadenectomy during primary treatment. Robert *et al*. (2019) [7] reported on 18 patients who underwent some form of inguinal lymphadenectomy, finding inguinal lymph node involvement in only 5 cases. As metastasis is typically hematogenous and infrequent, involvement of the inguinal glands is rare. The standard treatment for DFSP is wide local excision with 2–3 cm tumor-free margins. A study of 913 cases of DFSP of the skin found a 50% recurrence rate when the resection margins were not tumor-free, which dropped to 13% when margins were clear. Tumor behavior in the vulva is similar to that in other skin locations, although wide excisions in the vulva may disrupt its anatomical structure.

Chemotherapy for distant metastases in the form of the tyrosine kinase inhibitor (TKI) imatinib at a dose of 400 mg daily orally has been shown to be effective in metastatic disease. However, it does not significantly affect overall survival (Rutk *et al*., 2010) [8], (Heinrich MC *et al*., 2008) [9].

In our patient, the primary resection identified the tumor but did not achieve margin-free excision. Magnetic resonance imaging (MRI) revealed scattered right inguinal lymphadenopathy. Consequently, vulvectomy and bilateral inguinal lymphadenectomy were performed. These surgical decisions were influenced by her age and her desire to have more children.

No universally accepted staging system exists for this tumor, though it may be considered under the TNM classification for skin tumors. Importantly, tumor grading did not significantly influence the disease course (Robert *et al*., 2019) [7]. A review of the literature revealed only one reported case by Aartsen *et al*. (1994) [4], involving a 15-year-old patient who underwent local excision and radiation therapy. This patient was able to lead a normal life and later gave birth to two children via cesarean section. We now report on our 25-year-old patient, who is enjoying life with her baby daughter and remains well, four years after her initial treatment in March 2019.

The treatment of choice for this condition is primary local excision with a microscopically clear surgical margin of 3 cm or more, as recommended by Roses *et al*. (1986) [10]. Due to the low incidence of lymphatic spread, routine regional lymphadenectomy is not warranted. Psychosexual function is seldom documented in cases of dermatofibrosarcoma protuberans (DFSP) discussed in the literature. Despite vulvar mutilation, especially in cases involving clitorectomy, psychosexual function was rarely discussed and was not significantly impaired (Banhill *et al*., 1988) [11], (Leak *et al*., 1991) [12]. It appears that sexual activity is not invariably compromised following regional surgery.

In our case, vulvectomy did not affect the patient's sexual activity. She delivered a child two years after her initial treatment via cesarean section and has remained disease-free for over four years.

- The case report was approved by the Ethical Committee of the Medical City.
- No conflicts of interest were declared.
- No financial support was received for this study.

## References

1. DiSaia PJ, Rutledge F, Smith JP. Sarcoma of the vulva: Report of 12 patients. Obstet Gynecol. 1971;38(2):180-184. [PubMed] [Google Scholar].
2. Johnson S. Int J Gynecol Cancer. 2020 Aug.
3. Álvarez-Canas M. Acta Obstet Gynecol Scand. 1996;75:82-85.
4. Aartsen EJ, Albus-Lutter ChE. Vulval sarcoma. Eur J Obstet Gynecol Reprod Biol. 1994;56(2):181-189.
5. Fields RC, Hameed M, Brennan MF. Dermatofibrosarcoma protuberans: Clinical and pathological aspects. Ann Surg Oncol. 2011;18(2):328-336.
6. Edelweiss M, Malpica A. Dermatofibrosarcoma protuberans of the vulva: A clinicopathologic and

immunohistochemical study of 13 cases. Am J Surg Pathol. 2010;34(3):393-400.
7. Neff R. Gynecol Oncol Rep. 2019 May;28:9-11. DOI: 10.1016/j.gore.2019.01.006.
8. Rutkowski P, Van Glabeke M, Rankin CJ, *et al*. Imatinib mesylate in advanced dermatofibrosarcoma protuberans: Pooled analysis of two phase II clinical trials. J Clin Oncol. 2010;28(10):1772-1779.
9. Heinrich MC, Joensuu H, Demetri GD, *et al*. Phase II, open-label study evaluating the activity of imatinib in treating life-threatening malignancies known to be associated with imatinib-sensitive tyrosine kinases. Clin Cancer Res. 2008;14(9):2717-2725.
10. Roses DF, Valensi Q, LaTrenta G, Harris MN. Surgical treatment of dermatofibrosarcoma protuberans. Surg Gynecol Obstet. 1986;162:449-452.
11. Barnhill DR, Boling R, Nobles W, Crooks L, Burke T. Vulvar dermatofibrosarcoma protuberans. Gynecol Oncol. 1988;30:149-293.
12. Leak JF, Buscema J, Cho KR, *et al*. Dermatofibrosarcoma protuberans of the vulva. Gynecol Oncol. 1991;41(3):245-249.

**How to Cite This Article**
Kunda S, Azzo N, Ibraheem N, Abas RF. Treatment of dermatofibrosarcoma protuberans and childbirth. International Journal of Oncology Research. 2024;4(1):113-115.