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Case Report

Dermatofibrosarcoma Protuberans During Pregnancy: A Case Report

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Abstract: Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade soft tissue sarcoma characterized by local aggressiveness and high recurrence after excision. A 23-year-old primigravida at 32 weeks' gestation presented with a progressively enlarging natal cleft mass. Biopsy confirmed DFSP (CD34 positive). Definitive surgery was performed postpartum with wide local excision and flap reconstruction, achieving clear margins. DFSP during pregnancy requires careful timing of intervention. Wide local excision with clear margins remains the treatment of choice.

Keywords: Dermatofibrosarcoma Protuberans; Pregnancy; Natal Cleft; Wide Local Excision; Reconstruction.

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1. Introduction

Dermatofibrosarcoma protuberans is a rare, low-grade malignant soft tissue tumor arising from the dermis with potential extension into subcutaneous tissue, fascia, and muscle. It is known for local aggression and high recurrence rates but rarely metastasizes. Occurrence during pregnancy is uncommon and may present management challenges [1].

2. Case Report

A 23-year-old female, primigravida, nullipara, at 32 weeks' gestation, presented to the general surgery clinic with a progressively enlarging mass at the natal cleft. The mass initially appeared as a small, firm, painless nodule early in her pregnancy and had grown rapidly over the following months. The patient reported no history of trauma, infection, or prior similar lesions. There was no significant past medical history or family history of malignancies.

On examination, a firm, mobile, non-tender, skin-colored nodule approximately 4 cm in diameter was noted with apical ulceration at the midline of the natal cleft. No inguinal lymphadenopathy was noted. The lesion was initially considered in the differential diagnosis of pilonidal disease and hidradenitis suppurativa due to its location in the natal cleft. However, the absence of sinus formation, discharge, or inflammatory changes prompted further evaluation. Diagnostic radiological assessment by high-resolution superficial ultrasound showed a hypoechoic mass with irregular borders, suggestive of a soft tissue tumor (Figure 1).

Due to its rapid enlargement, a biopsy was performed, revealing spindle-shaped tumor cells arranged in a storiform pattern, infiltrating the dermis and subcutaneous tissue. Immunohistochemistry was positive for CD34, negative for S-100, and smooth muscle acting (SMA), confirming the diagnosis. High-power histological examination did not demonstrate features of fibrosarcomatous transformation, such as increased cellular atypia, mitotic activity, or herringbone pattern architecture (Figure 2).

Figure 1: Sonographic assessment of the natal cleft mass demonstrating a hypoechoic lesion with irregular borders suggestive of a soft tissue tumor.

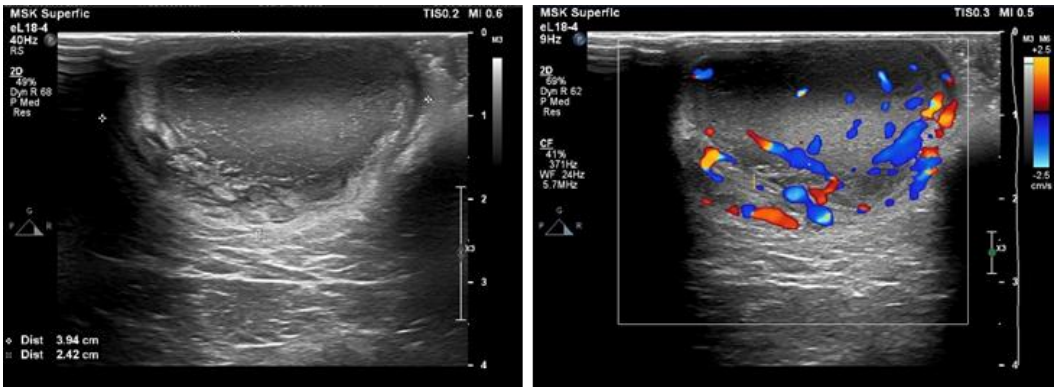
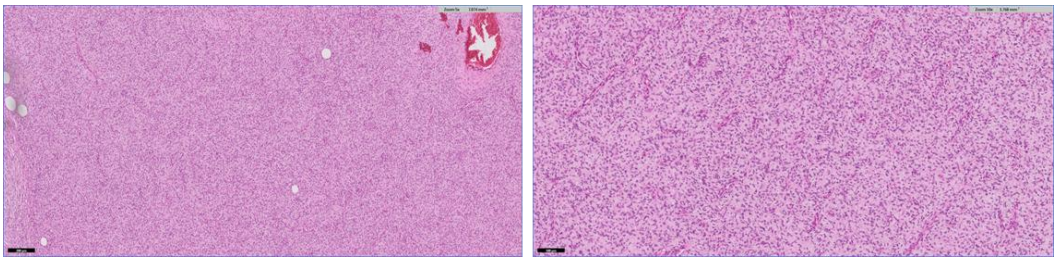


Figure 2: Histopathological examination showing spindle-shaped tumor cells arranged in a storiform pattern infiltrating the dermis and subcutaneous tissue (Hematoxylin and Eosin stain).



Following histopathological confirmation, the case was discussed in a multidisciplinary setting involving obstetrics and surgical teams. Given the absence of rapid progression, lack of high-grade features, and the well-documented low metastatic potential of dermatofibrosarcoma protuberans, a decision was made to defer definitive surgery until the postpartum period to minimize maternal and fetal risk. Regular follow-up was arranged to monitor the lesion for signs of increased growth or complications. At initial presentation (32 weeks' gestation), the lesion measured approximately 3.94 × 2.42 cm.

At 39 weeks' gestation, the patient underwent an uncomplicated vaginal delivery of a healthy infant. At the time of surgical excision, 6 weeks postpartum, the lesion measured 4.23 × 2.51 cm, with mild increase in size consistent with the known slow-growing nature of DFSP. Wide local excision of the DFSP was performed under general anesthesia. A margin of 2-3 cm of healthy tissue around the lesion was excised to ensure complete removal of the tumor. Excision included the underlying fascia over the sacrococcygeal region to ensure complete oncologic clearance, given the infiltrative nature of DFSP. Due to the anatomical location and the extent of tissue removal, local tissue reconstruction was necessary.

Reconstruction was performed using a local rotational gluteal flap to close the defect in the natal cleft. The flap provided sufficient adequate tension-free closure with good vascularity and acceptable cosmetic outcome. Histopathological examination of the excised tissue confirmed clear margins. The postoperative course was uneventful, and the patient recovered well. She was discharged on postoperative day 2 with advice on wound care and was seen in follow-up clinics for two weeks and six weeks' post-surgery. The flap healed without complications, and there was no evidence of recurrence at six months' follow-up. Given the potential for DFSP recurrence, the patient will be followed with regular clinical examinations every 6 months for the first 3 years, then annually up to 10 years. Imaging with MRI will be done if recurrence is clinically suspected.

3. Discussion

DFSP is rare, accounting for less than 1% of all soft tissue sarcomas. Its incidence is 0.8-4.5 cases per million individuals annually, DFSP occurring in the natal cleft is uncommon, with limited cases reported in the literature. Compared to more typical sites such as the trunk and proximal extremities, lesions in the natal cleft may present later due to misdiagnosis as benign conditions such as pilonidal disease. However, recurrence rates appear to be more closely related to margin status rather than anatomical location. Adequate surgical excision with clear margins remains the most important prognostic factor regardless of site [1]. In our case the lesion was initially considered in the differential diagnosis of pilonidal disease and hidradenitis suppurativa due to its location in the natal cleft. However, the absence of sinus formation, discharge, or inflammatory changes prompted further evaluation and biopsy, which established the diagnosis.

Immunohistochemical staining for estrogen and progesterone receptors was not performed in this case. While hormonal influence has been hypothesized in cases demonstrating rapid growth during pregnancy, current evidence remains inconclusive, and routine receptor testing is not yet standardized in DFSP [2]. Although DFSP is locally aggressive, it is characterized by slow growth and a low metastatic rate. In selected cases during late pregnancy, short-term postponement of definitive surgical management may be considered after multidisciplinary evaluation, particularly when there is no evidence of rapid progression or high-grade transformation. This approach balances oncologic safety with obstetric considerations [2].

Excision with clear margins remains the treatment of choice to prevent recurrence. [3] In cases where large defects are created, local or regional flap reconstruction is often necessary for functional and cosmetic restoration [4]. In our case wide local excision (WLE) with 2–3 cm margins were selected over Mohs micrographic surgery (MMS) due to limited availability of MMS in our institution and the need to balance oncologic clearance with timely intervention in a postpartum patient. Although MMS offers superior margin control and tissue preservation, WLE remains an accepted standard treatment for dermatofibrosarcoma protuberans, particularly in settings where MMS is not readily accessible. Additionally, the lesion location within the natal cleft allowed for adequate margin acquisition with acceptable reconstructive options.

Given the known risk of late local recurrence in DFSP, long-term follow-up is essential. by clinical examination every 6 months for the first 3 years, then annually up to 10 years. Imaging with MRI will be reserved for cases with clinical suspicion of recurrence. [4].

4. Conclusion

Early recognition and biopsy of suspicious lesions during pregnancy are essential. Surgical delay should not be routine but may be considered in highly selected cases after multidisciplinary evaluation. Wide local excision with clear margins provides optimal outcomes. long-term follow-up is essential.

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Conflicts of Interest: All other authors declare no conflicts of interest.

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