



**ORIGINAL RESEARCH PAPER**

**General Surgery**

**DERMATOFIBROSARCOMA PROTUBERANS IN PREGNANCY : A NOT SO COMMON ENCOUNTER**

**KEY WORDS:** protuberant, locally infiltrating, hormone receptor, recurrence

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**ABSTRACT**

**Background :** Dermatofibrosarcoma protuberans is a rare cutaneous soft tissue neoplasm of disputed histogenesis, with sparse reports of new onset or gross enlargement during pregnancy. **Description :** This case report is about a 34 year old, postpartum female who presented with recurrent soft tissue lesion of her left thigh following excision for a suspicious benign growth during her pregnancy. **Conclusion:** A high index of suspicion is to be maintained for neo-occurrence of progressively enlarging soft tissue mass during pregnancy to rule out Dermatofibrosarcoma protuberans and to provide appropriate surgical management to avoid recurrence and the associated morbidity.

**INTRODUCTION**

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, low-grade mesenchymal tumour of fibroblastic origin from dermis. The lesion is notorious for local recurrence and misdiagnosis as benign tumour due to it's non-specific appearance and slow growth. To highlight the significance of the need for high index of preoperative suspicion to offer appropriate wide local excision, we present a case of recurrent lower extremity DFSP in a post-natal mother.

**CASE REPORT**

A 34 year old post-natal mother presented with slow-growing, progressive swelling of her left thigh for the past 6 months. Patient had underwent excision of a swelling at the same site at 3 months of her gestation with no further follow-up. She noticed recurrence 4 months later. Examination revealed 2x2x4cm non-tender, protuberant, firm lesion at middle-third of left thigh with overlying skin ulceration and no involvement of underlying adductor muscles. Laboratory investigations were normal. Magnetic resonance imaging revealed features of cutaneous soft tissue sarcoma. Core needle biopsy showed Dermatofibrosarcoma protuberans. Computed tomography of lung showed no metastases. Wide local excision of the lesion with 3cm margin clearance and primary closure was done. Postoperative Histopathology confirmed the diagnosis of DFSP with free resection margins; immunostaining was CD34 positive with negative ER,PR. 1 year into follow-up, patient is still free of recurrence.

**DISCUSSION**

DFSP is a low-grade, locally aggressive, rare soft tissue tumour with low metastatic potential accounting for 2-5% of cases<sup>[1]</sup>. It was first described by Darier and Ferrand. The term was however officially coined by Hoffman<sup>[3]</sup>

There are only few reports in literature of DFSP in pregnancy. It shows a male predominance with average age of occurrence being 6 - 65 years<sup>[2]</sup>. It often starts as a painless, firm cutaneous nodule with a pink or violaceous hue and exhibits expansive growth. However, in late stages, it forms a protuberant mass<sup>[4]</sup>. The most common site is trunk ( 42-72%), followed by upper extremities (16-30%), and then lower extremities (13%). Head and neck region is less commonly involved. Risk of distant metastases is 2-5% and occurs commonly to lungs<sup>[5]</sup> by hematogenous spread.

The driving pathophysiology of this tumour is still not clearly known. Recent studies however have revealed that t(17;22)(qq21;q13) fusion gene product COL1A1 ( collagen

type 1 alpha 1 gene) – PDGF beta (platelet derived growth factor beta) codes for a carcinogenic protein, which plays a pivotal role in tumourigenesis of DFSP<sup>[6]</sup>.

Fine Needle Aspiration Cytology or core needle biopsy can often be inconclusive due to inadequate representation of lesion. Tumour is histiocytic or neurogenic in origin. Histopathology examination reveals monomorphic, slender, bland spindle cells with elongated nuclei and scanty, pale cytoplasm arranged in storiform or cart-wheel pattern, with finger-like infiltration into subcutaneous tissue as pseudoseptate or lace-like pattern known as honeycomb appearance<sup>[7]</sup>. Immunohistochemistry staining shows CD34 positivity and factor XIIIa negative. As with other stromal neoplasms, low levels of hormone receptor expression is observed in DFSP. This accounts for new occurrence and often rapid enlargement during pregnancy<sup>[11]</sup>.

Computed tomography reveals well-defined, nodular or globular, soft tissue lesion involving skin and subcutaneous tissue; with pathognomonic protrusion from skin surface. With contrast imaging, moderate enhancement is seen due to mild to moderate hypervascularity of the lesion<sup>[6]</sup>.

American Joint Committee on Cancer has not developed a staging system for DFSP. However, Short German Guidelines<sup>[10]</sup> for DFSP identifies 3 stages for clinical use:

1. Stage I – primary, localized disease
2. Stage II – lymph node metastases
3. Stage III – distant metastases

Wide local excision of lesion is the standard treatment with 3 – 5 cm margin of uninvolved skin with an aim of obtaining oncologic clearance with maximum functional preservation. Any previous biopsy scar must also be completely excised with the lesion. Margins less than 2cm are often associated with recurrence. Mohs micrographic surgery (MMS) when available, is a better option. Radiotherapy can be used both as adjuvant therapy and for palliation. In recurrent cases, and margin positivity post excision, radiotherapy along with Imatinib mesylate, tyrosine kinase inhibitor can help achieve long term local disease control. Imatinib is also to be considered for neo-adjuvant therapy in borderline respectable cases<sup>[9]</sup>.

Local recurrences are seen in 20-55% of cases<sup>[7]</sup>. Most recurrences are observed in first 5 years. Follow-up must be strictly endured every 3 – 6 months, atleast for the first 3 years.

Care must be taken to examine scar site and regional lymph nodes.

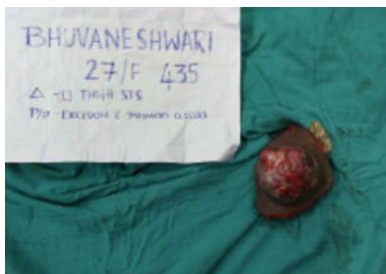
**CONCLUSION**

DFSP, though an uncommon extremity cutaneous tumour, must be considered a differential in progressive, protuberant soft tissue lesions especially during pregnancy. Surgical excision with adequate margins is the mainstay of treatment. Though prone for local recurrence, radiotherapy and tyrosine kinase inhibitors can offer disease control.

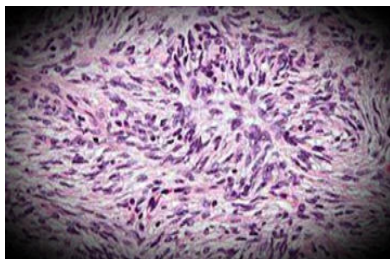
**Pre-operative image**



**Excised specimen**



**Histopathology**



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