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Case Report

# Dermatofibrosarcoma Protuberans of the Suprapubic Region: A Rare Clinical Entity

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**Abstract**

**Introduction:** Dermatofibrosarcoma Protuberans (DFSP), an incompletely understood disease entity, has been reported in literature at different sites. However, the suprapubic region as its location further adds to its unusual persona.

**Case:** A 51 year old gentleman presented with a slow growing suprapubic lesion, which on clinical and laboratory evaluation was confirmed to be a Dermatofibrosarcoma Protuberans, subsequently managed adequately with wide excision and groin flap coverage.

**Conclusion:** Dermatofibrosarcoma Protuberans, in itself, is a rare entity and the suprapubic region as its location further adds to the unknown persona of the condition. Thus, the importance of maintaining a vigil for DFSP even at the most unusual sites, for optimal and timely diagnosis and management. As is rightly said, “What the mind knows, is what the eyes see”.

**Keywords:** Dermatofibrosarcoma Protuberans; Suprapubic region; groin flap; unusual site
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**Consent:** We confirm that family members of the patients have given their informed consents for the case report to be published.

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# Introduction

Dermatofibrosarcoma protuberans(DFSP), a fibroblastic mesenchymal dermal tumour, has been reported in literature to occur over the trunk, extremities, head and neck and rarely of the genitourinary system, all having minimal potential for distant metastases [1,2]. We describe a patient with a suprapubic DFSP managed with wide excision and groin flap coverage. On account of the lack of literary evidence, ours could be one of the few reported DFSP's of this region, adding to the unusual location of presentation.

# Case Presentation

A 51 year old male of lower socioeconomic status presented to the outpatient department with a lesion over his suprapubic area, which had slowly grown to its current size over a period of several months.

On examination, it appeared well defined, approximately 6x6cm in size, exophytic ulceroproliferative, with punctate haemorrhage and surface necrosis (Figure 1). It was free from the underlying tissues. Superficial inguinal lymph nodes were non palpable and there were no signs of satellitism or local tumour infiltration or similar lesions elsewhere over the body. History and general physical examination were unremarkable and seronegativity was confirmed by ELISA. Other laboratory studies were also within normal limits.

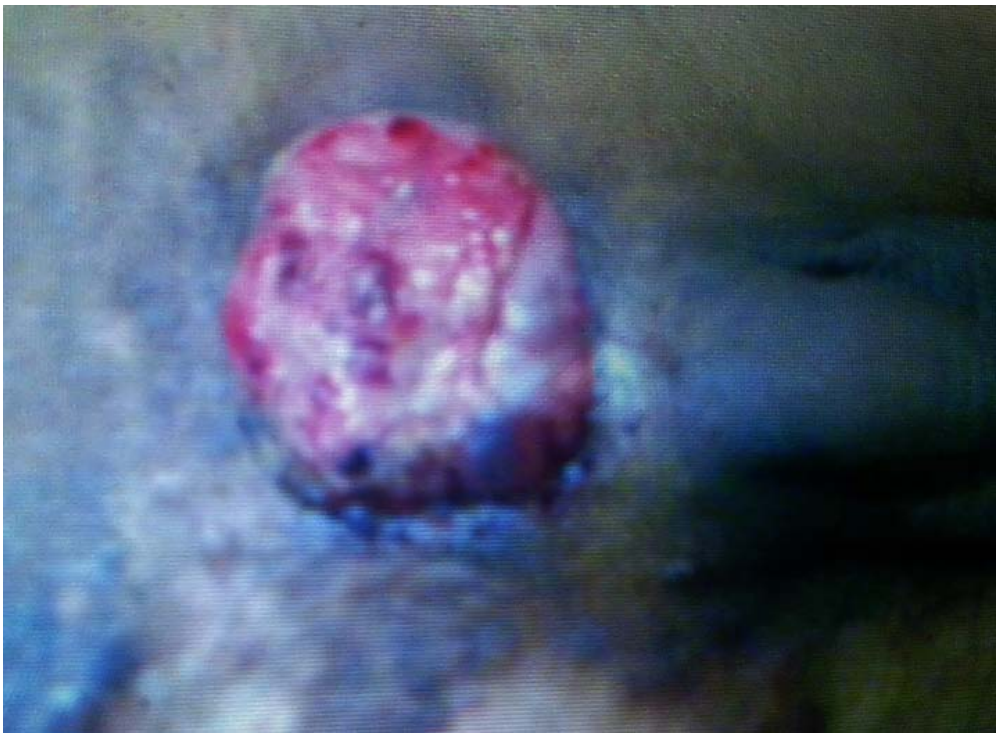

**Figure 1** Gross appearance of the Suprapubic DFSP.

On incisional biopsy the tumour was found to be composed of proliferative neoplastic spindle cells arranged in storiform pattern (Figure 2). Immunohistochemical analysis showed CD34 staining, hence corroborating the diagnosis of DFSP.

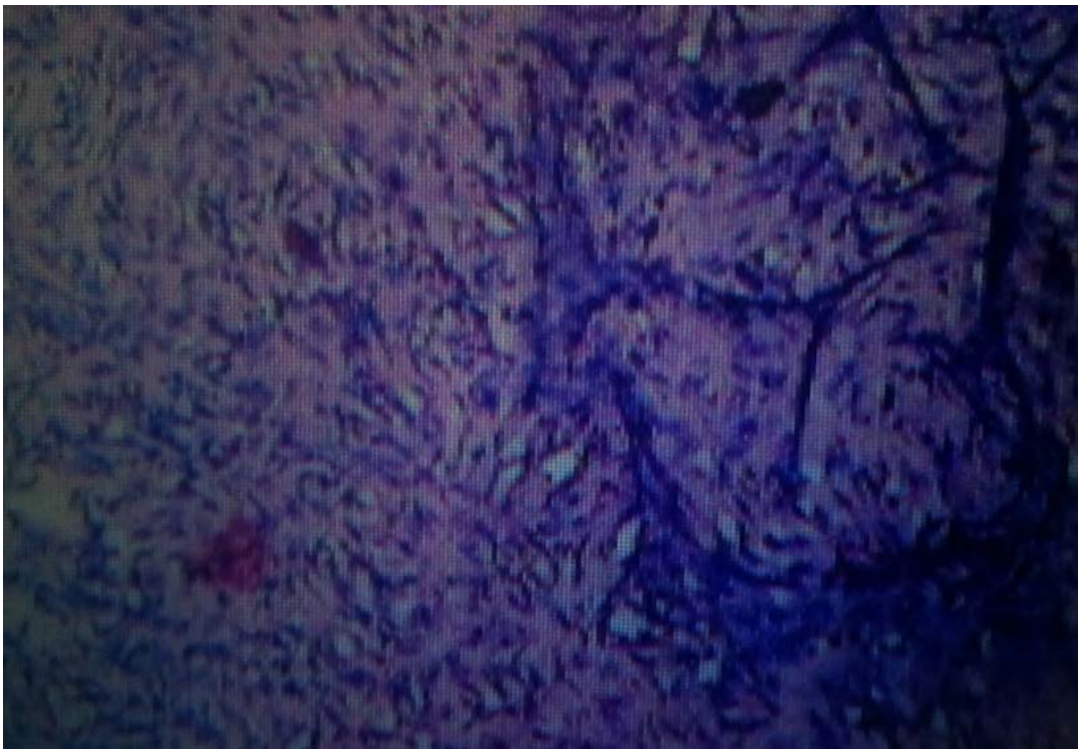

**Figure 2** Microscopic Appearance: Storiform pattern of cells of DFSP.

A wide local excision with 3cm margins and groin flap coverage over the soft tissue defect was performed, without event. Three monthly follow ups till one year postoperatively were uneventful with no evidence of recurrence of the lesion.

## Discussion

"Dermatofibrosarcoma Protuberans" first coined by Hoffman in 1925, accounts for a rare low grade malignancy of the skin and subcutaneous tissues [1-3]. It is most commonly seen in the fourth and fifth decades of life, the overall incidence estimated as 0.8 – 4.2 per million population, with no gender predilection [3].

Around 50 – 70% have been reported to involve trunk and proximal extremities, with head and neck and genitourinary system rarely affected. The presentation of DFSP is duration dependant, the early lesion presents with small, indurated plaques whereas the later stages presents with ulcerative polypoidal nodules [3,4]. The fixity to the deeper structures also depends upon the stage of presentation, with early lesions being freely mobile on the deeper tissues whilst in later stages and in cases of local relapse being fixed.

The exact etiology remains unknown and studies are on to establish the origin of this rare entity. Histopathologically DFSP's have been found to have multiple monomorphic spindle cells arranged in a storiform pattern and also have been found to stain positive for CD34 which helps differentiate them from other differentials such as malignant fibrohistiocytoma, nodular fasciitis, keloids and hypertrophic cicatrix [3,4].

Imaging studies such MRI, CT scans and occasionally angiography add to the armamentarium of

the clinicians to delineate the anatomy, depth of invasion as well as bony involvement of this disease entity [4]. Though MRI's are nonspecific, an arteriogram may reveal moderate hypervascularity [3].

Surgical excision remains the gold standard in the management of DFSP. Wide excision with a minimum of 2-3 cm of tumour free margin, confirmed histopathologically, is the most common procedure employed [3]. However Moh's micrographic surgery in anatomically restricted or cosmetically unacceptable areas, is also sufficient. With complete excision alone, the recurrence rates are less than 10% [3]. The role adjuvant radiotherapy is restricted to an unresectable DFSP or one with persistent positive margin after reasonable surgical attempts [3,4]. A tyrosine kinase inhibitor, imatinib mesylate, the drug of choice in chronic myeloid leukemia has also been used in relapses. There is no conclusive evidence to support the role of chemotherapy in the management of DFSP [4].

Long term follow ups in patients of DFSP are mandatory as recurrences have been reported even five years after surgery, although the vast majority have been seen in the last 2- 3 years [5].
Thus, there are still multiple rifts in our knowledge, that need bridging in order to ascertain the optimal management of this sarcomatous lesion. Till we gain sufficient evidence, literature would have to rely on isolated institutional case reports to manage this disease entity.

## Conclusion

Dermatofibrosarcoma Protuberans, in itself, is a rare entity and the suprapubic region as its location further adds to the unknown persona of the condition. Thus, the importance of maintaining a vigil for DFSP even at the most unusual sites, for optimal and timely diagnosis and management. As is rightly said, "What the mind knows, is what the eyes see".

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