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Recurrent Dermatofibrosarcoma Protuberans on Anterior Abdominal Wall A Case Report and Review of Literature

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Abstract

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon, slow growing, locally aggressive cutaneous neoplasm. Incidence of DFSP is 0.8 to 4 cases per 1 million per year. Dermatofibrosarcoma protuberans accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas.

In our case a young male presented with recurrent DFSP on his left flank region which highlights two noteworthy features. Firstly, DFSP is a very rare cutaneous soft tissue sarcoma . Secondly, it describes a local recurrence of this entity and its management.

Keywords: *Skin tumour, Dermatofibrosarcoma Protuberans, Recurrence, Wide local excision.*

INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) is a very rare fibroblastic mesenchymal tumour arising from the dermis of skin. It was first described by Darier and Ferrand in 1924, as "progressive and recurrent dermatofibroma. In 1925 Hoffmann termed the lesion as "dermatofibrosarcoma protuberans" [1]. DFSP

usually present in 3rd decade of life with slight male preponderance. Most common location is trunk followed by extremities, but it can affect any parts of the body. It is a low grade tumour which rarely progress to high grade fibrosarcomatous component. Radical surgical resection is the treatment of choice for DFSP [2]. Mohs micrographic surgery (MMS) emerges as an

alternative approach to the use of wide resection surgery with tumor-free margins^[3]. Postoperative radiotherapy is often administered to reduce the risk of local recurrence^[4]. In most DFSP patients (around 90%), t(17;22) chromosomal translocation leads to up regulation of the platelet-derived growth factor B pathway, resulting in uncontrolled cell division. Imatinib, acts through inhibition of tyrosine kinase, thereby blocking the platelet-derived growth factor B(PDGFB) pathway and causing tumour cell apoptosis. Some recently published case series have been reported favourable responses in patients with recurrent or metastatic disease. Presence of t(17;22) chromosomal translocation associated with the favourable response to Imatinib^[5].

CASE REPORT

A 26 years male presented to surgery OPD with complaints of a recurrent, painless swelling over

left flank region. He first noticed the lesion over abdomen 2 yrs back which undergoes excision in rural hospital but no histopathological examination was done. 3-4 months after excision, there was recurrence of swelling with rapid increase in size. There was no H/O weight loss. On clinical examination the swelling was 6cm×7cm in size, firm in consistency, non-tender with lobulated appearance. The skin over the swelling was adherent, tense glossy and telangiectatic [Figure 1(a)]. There was no regional lymphadenopathy. His chest X-ray was normal and abdominal CT scan suggested that it was a parietal abdominal wall lesion with intact fat plane between tumour and abdominal wall musculature. Core biopsy failed to reach a diagnosis due to the copious vascularity. Our differential diagnosis was between soft tissue tumour and desmoid tumour.



Figure 1(a)



Figure 1(b)

Figure 1: (a) DFSP Lesion on left flank region. (b) Lesion excised with 2cm margin.

Assuming a recurrent malignancy, radical excisional surgery was performed with 2cm grossly clear margin and resulting large defect

was reconstructed by Rhomboid (Limberg) flap [Figure 2(a)]. Post-operative outcome was uneventful including the flap. Patient was

discharged after 7 days of hospital stay. Post operative histopathology report suggestive of Dermatofibrosarcoma Protuberans with clear margins. Immunohistochemistry was strongly

positive for CD 34. Our tumour board advised for adjuvant radiotherapy. Patient is under follow up for last 14 months with no signs of local or distal recurrence.



Figure 2 (a)

Figure 2 (b)

Figure 2: (a) Reconstruction with Limberg flap. (b) Low power-magnification photomicrograph showing bundles of uniform, spindle cells, arranged in a prominent storiform or whorled pattern (Haematoxylin and Eosin stain; original magnification $\times 100$).

DISCUSSION

DFSPs initially present as single, raised, red to bluish, firm cutaneous lesion. The lesion is painless and indurated but is extremely infiltrative, that can invade the underlying structures such as fascia, muscles or bones. The tumour is often covered by a red-tinged, sclerodermiform or telangiectatic and atrophic skin. In some cases H/O trauma, burn or surgical scar may be found. At advanced cases tumour may ulcerate, bleed, and become painful. More than 90% of DFSPs are considered to be of low grade tumours. Histologically, DFSP is identified by a pattern of monomorphous proliferation of spindle cells with a visible storiform or whorled

(rhubarb-like) architecture [Figure 2(b)]. Other characteristic features are low mitotic activity and deep, honeycomb infiltration into subcutaneous fat [6]. Immunohistochemical studies confirm the diagnosis, where DFSP tumour cells are strongly positive for CD34 [7]. DFSP can rarely transform into fibrosarcomatous DFSP (FS-DFSP). The extent of surgical resection is the most important prognostic factor. If deep fascia or muscles are involved, then these must be excised. It has high recurrence rate varying in the literature from 10 to 80%. Distant metastasis is very rare (1- 4%), predominantly to the lungs after many years of initial presentation [8].

CONCLUSION

In conclusion DFSP is a very rare and low grade skin malignancy albeit its management may be challenging. As it grows finger like projections in the subdermal tissue, it has a high recurrence rate specially if resection is inadequate. Wide local excision with ≥ 2 cm. margin is recommended. Although radiotherapy is often used to improve local control. Chemotherapy plays a limited role, used only in unresectable and metastatic disease. Close surveillance is mandatory for 5 years or longer to identify late recurrence like other malignancies.

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Conflict of Interests:

The author(s) declare(s) that there is no conflict of interests

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