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# Dermatofibrosarcoma Protuberans: A Case Report

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**Abstract:** ***Background: Dermatofibrosarcoma protuberance (DFSP) is rare slow growing cutaneous fibro-histiocystic neoplasm with high local recurrence and infiltration but low metastatic potential. Wide local excision with tumor free margin is the most acceptable treatment modality. Case description: A 57 years lady presented with large protruding mass on the left side of upper neck and underwent wide local excision with 1 cm free skin margin. On histological examination the diagnosis of DFSP was made and was referred for adjuvant radiotherapy. Conclusion: DFSP is cutaneous sarcoma which arises from dermal layer of skin with tendency to infiltrate into deeper structure like fascia, muscle, tendon and even bone. It appears mostly in adults between 20-50 years age group. Wide local excision is treatment of choice particularly in head and neck region. In our case, tumor was confined to the skin and subcutaneous tissue only however patient was sent for radiotherapy to avoid a possible relapse.***

**Keywords:** DFSP, adjuvant radiotherapy, IHC, Mohs micrographic surgery

## 1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is rare cutaneous fibrohistiocystic neoplasm, the term was coined by Hoffman in 1925 [1]. It is deriving from the dermal layer of the skin either from healthy skin or chronically damaged areas include trauma, surgical scar, radiation dermatitis, old burn, vaccination site and insect bite. It account for 1% of all soft tissue sarcoma. Although it appears mostly in adult between the ages of 20 to 50 but it has been diagnosed in people of all ages. The most common location of DFSP is the trunk (42-72 %) followed by proximal extremities (20-30 %), and head and neck (10-16 %). [2] DFSP typically follow an indolent clinical course and have locally aggressive behavior. It is sometimes described as having tentacles that can grow and infiltrate adjacent structures, such as the surrounding fat, muscles, tendons, and even bone with high rate of localrecurrence but low metastatic potential. [3]In early stage DFSP presents as a slow growing, painless, skin-colored plaque like lesion with possible dark red or blue dis-coloration. At latter stages, it can increase in size and become protuberant or ulcerative. [4]

## 2. Case Report

A 57-year-old female was presented with large protuberant mass located at the left side of her upper neck. The tumour was increasing in size in last two years. The patient denied any recent weight loss, fever, night sweats or chills. There was neither personal nor familial history of malignancy.

On physical examination, a multinodular, large, smooth surface, firm, non-tender, red discoloration mass was found in the left cervical region at level II and V with no sign of inflammation and regional lymphadenopathy. Contrast Enhanced computed tomography (CECT) scan demonstrated heterogeneous enhancing soft tissue density mass measuring approximately 59X51mm is seen involving subcutaneous plane of left side of neck without infiltration of muscular and bony structures.
With the possible diagnosis of cutaneous neoplastic mass, the patient underwent excision of mass with 1 cm surrounding free skin as a margin of mass. Intraoperativelythe mass was extended into subcutaneous fat plane with sparing of ipsilateral sterno-cleido-mastoid muscle. Wound margin was closed by primary suturing. On histo-pathological examination Hematoxylin and eosin stained sections showed a cellular spindle cell neoplasm with vague cellular borders and elongated nuclei relatively uniformed. Viamentin and CD34 antigen was diffusely positive and focal positive S100 protein in spindle cell on immunohistochemical stains. There was no positivity for other smooth muscle marker like actin, desmin, CD57, CD68, CD117 and keratin 8/18. Finally Dermatofibrosarcoma protuberance diagnosis made on the basis of histological finding and immunohostochemical marker.

The patient's postoperative days were uneventful with removal of skin suture on 14$^{th}$ days of surgery, after that patient was referred for adjuvant radiotherapy in regional cancer center of our institute.

## 3. Discussion

DFSC is anuncommon indolent course fibroblastic malignant mesenchymal dermal tumor with low metastatic potential. Initially tumor limited to dermal layer and appear as reddish-brown plaque like lesion. With time it presents as protuberant mass due to lesion evolvementand infiltration into the subcutaneous tissue, fascia, muscle and even bone. Untreated lesion usually progresses to nodule and may ulcerate and/or bleed. DFSC has very less metastatic tendency and when occur they are found in regional lymph node and distant metastasis in lung in less than 5 % cases. [5]In our case no infiltration to ipsilateral sterno-cleido-mastoid muscle and any other adjacent muscle and bony structure was seen. DFSP should be differentiated from other lesion like keloid, epidermal

cyst, nodular fasciitis, lipoma in early stage and Kaposi sarcoma, and pyogenic granuloma in the later stages.

The histolpathologic features of DFSP are usually characteristic but immunohistochemical staining help to differentiated from other smooth muscle sarcoma. DFSP shows CD34 positive in 92% to 100% cases. [3]The expression of PDGFB protein is also a marker for DFSP, with 1 study PDGFB expression was absent in 47 (98%) of 48 patients in non-DFSP mesenchymal tumors. [6]

Treatment of choice for DFSP is wide local excision with histologically 3-5 cm negative margin from tumor edge including the skin, subcutaneous tissue and fascia as recurrence rate depends on the negative resection margin. Farma et all reported a recurrence rate of less than 1% in large series with the negative surgical margin of 1cm to 2 cm, although 20% of tumors required multiple excision to achieve clear margin. [7]Chang CK et all found that recurrence rate was less than 5% in series where resection margin of 5 mm was used. In case of involvement of deeper structure possible periosteum with and without bone may also need to be removed to achieve tumor free margin. [8] Reconstructive procedure may be required to maintain tissue contour after excision using local skin flap, skin graft or myocutaneous flap [4]. In our case, tumor was excised by wide local excision with 1 cm free margin and primary closure of surgical wound.

Mohs micrographic surgery is an alternative to wide local excision, as it allows the clinician to analyze 100% of the tumor margins, identify microscopic extension and remove them while conserving healthy tissue. Recurrence rate for DFSP treated with MMS has been reported at 0% to 8.3% [9].

Radiotherapy for DFSP is reserved in case of positive or inadequate margin, recurrent of tumor or case of unacceptable functional or cosmetic result after WLE [10]. Imatinib mesylate, a tyrosine kinase inhibitor is used as a adjuvant treatment in unresectable, recurrent and/or metastatic disease.

DFSP tendencyof localrecurrence after five years of surgery has also been reported. Most local recurrences presented in first three postoperative years. Thus, it is important to follow up these patients for long term.

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**Figure No.1:** DFSP. Pink to red coloured Multi-lobulated lesion in the left upper neck with area of ulceration.

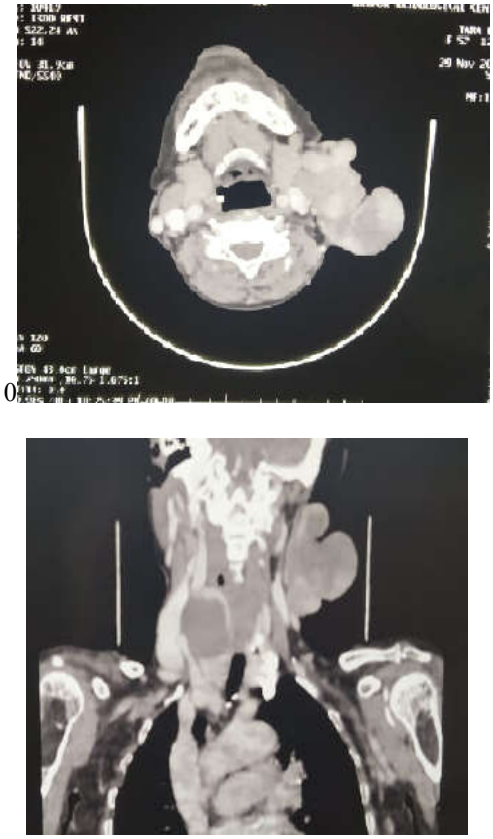

**Figure No.2:** Axial and coronal CT scan views of patient.

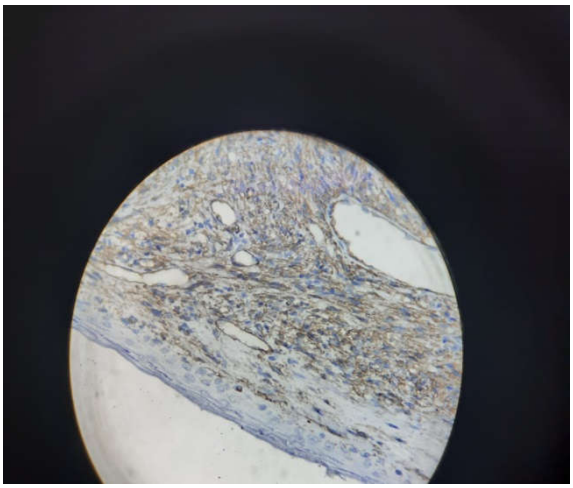

**Figure No.3:** Immuno-histochemistry of the tumor cells showing diffuse positivity for CD 34 antigen