# GIANT DERMATOFIBROSARCOMA PROTUBERANS OVER AN ANTERIOR CHEST WALL: A RARE CASE REPORT

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**ABSTRACT:**

*Dermatofibrosarcoma protuberans (DFSP) is a rare, slow growing fibro histiocytic neoplasm with intermediate to low grade malignancy. It is locally invasive tumor but rarely metastasis with high rate of recurrence. It commonly occurs on the trunk, proximal extremities. We report a rare case of a 58 year old male who presented with very huge mass over anterior chest wall. Wide local surgical resection with surrounding surgical margins was performed. Histopathological examination along with immunohistochemistry confirmed the diagnosis of DFSP. We present this case due to its rarity and its unusual gross appearance.*

**Keywords:** *Dermatofibrosarcoma protuberans, fibrohistiocytic, anterior chest wall.*

## INTRODUCTION

DFSP is a rare, slow growing soft tissue tumor with low to intermediate grade malignant potential, initially localized to the skin then invades deeper subcutaneous tissue (fat, fascia, muscle and bone).[1] Although metastasis rarely occurs, DFSP is a locally aggressive tumor with a high rate of recurrence.[2] It accounts for less than 0.01% of all malignancies and less than 0.1% of all the cutaneous neoplasm.[3,4] The most common presentation of disease is painless, slow growing subcutaneous nodule and mainly occurs in second and fifth decades of life.[4]
Herein, we present a case of giant size DFSP without recurrence in a 58 year old male due to its rarity and uncommon gross features.

## CASE HISTORY

A 58 year-old-man presented with swelling over anterior chest wall since 2 years. Initially swelling was small, slow growing and painless. Since last 6 months swelling increased to present size with complaints of pain and discomfort. There was no history of abdominal pain and distension. He had no history of previous trauma or any surgery. Past, personal and family history was not significant. Suddenly since last week the swelling started bleeding on touch.Local examination revealed extremely huge multinodular tumor mass measuring 20x18x16 cm over anterior chest wall and ulcerated at one place[Fig 1] which was clinically diagnosed as malignant soft tissue tumor. All the routine investigations were within normal limits. Wide local excision of the mass with wide surgical margins (3cm) was carried out and the specimen was sent for histopathological examination. Grossly we received excised huge mass measuring 20x18x14 cm from anterior chest wall. The overlying skin was ulcerated at one side. External surface was irregular, bosselated with multinodular appearance. Cut section showed solid, firm and grey white tumor mass. Areas of hemorrhage and necrosis were seen.[Fig 2] Microscopically multiple sections studied showed stratified squamous epithelium with underlying well circumscribed tumor. The tumor was composed of monomorphic population of spindle cells arranged in storiform pattern. The tumor cells were large spindle shaped nuclei with mild pleomorphism and scanty eosinophilic cytoplasm. Overlying skin was free of tumor. [Fig 3-4]

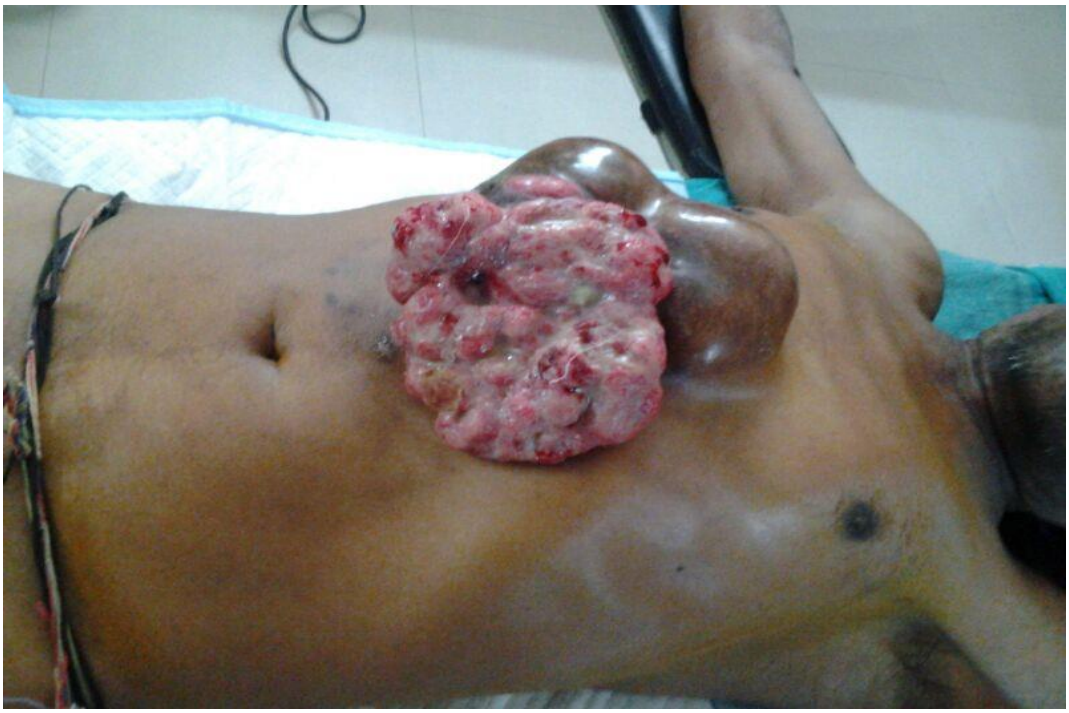

**Figure 1:** Local examination revealed extremely huge multinodular tumor mass measuring 20x18x16 cm over anterior chest wall and ulcerated at one area.

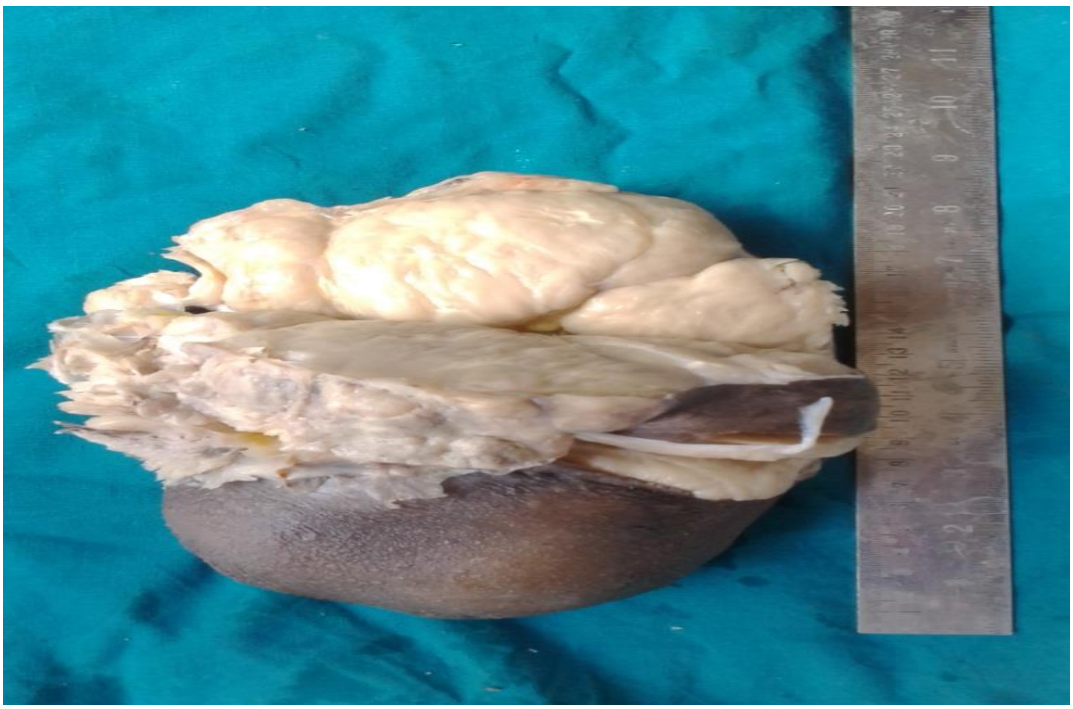

**Figure 2:** External surface was irregular, bosselated with multinodular appearance. On cutting open showed solid, firm and grey white tumor mass.

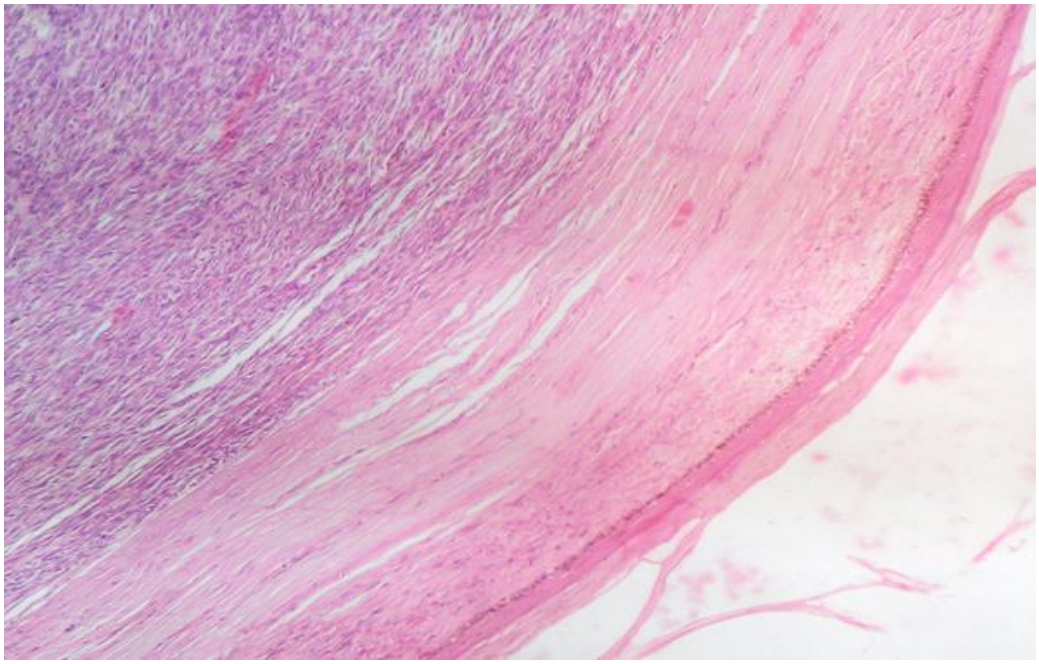

**Figure 3:** Microscopy showed squamous epithelium with underlying well circumscribed tumor composed of monomorphic population of spindle cells. [H & E, X 100]

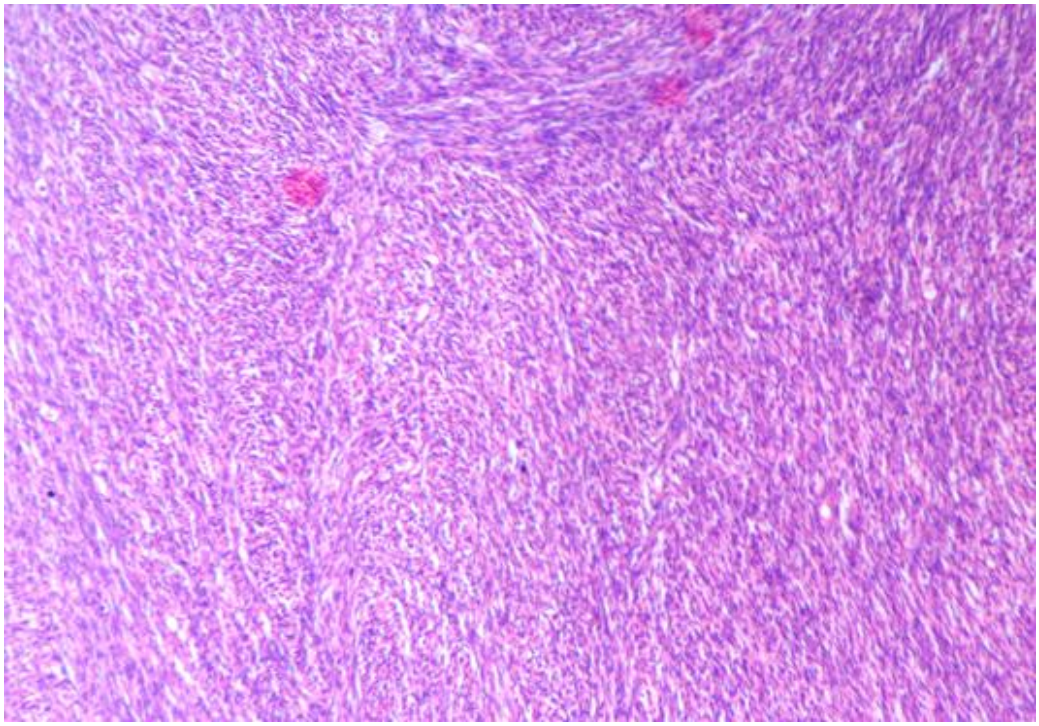

**Figure 4:** The tumor composed of monomorphic population of spindle cells arranged in storiform pattern. The tumor cells were large spindle shaped nuclei with mild pleomorphism and scanty eosinophilic cytoplasm. Overlying skin was free of tumor.[H & E, X 400]

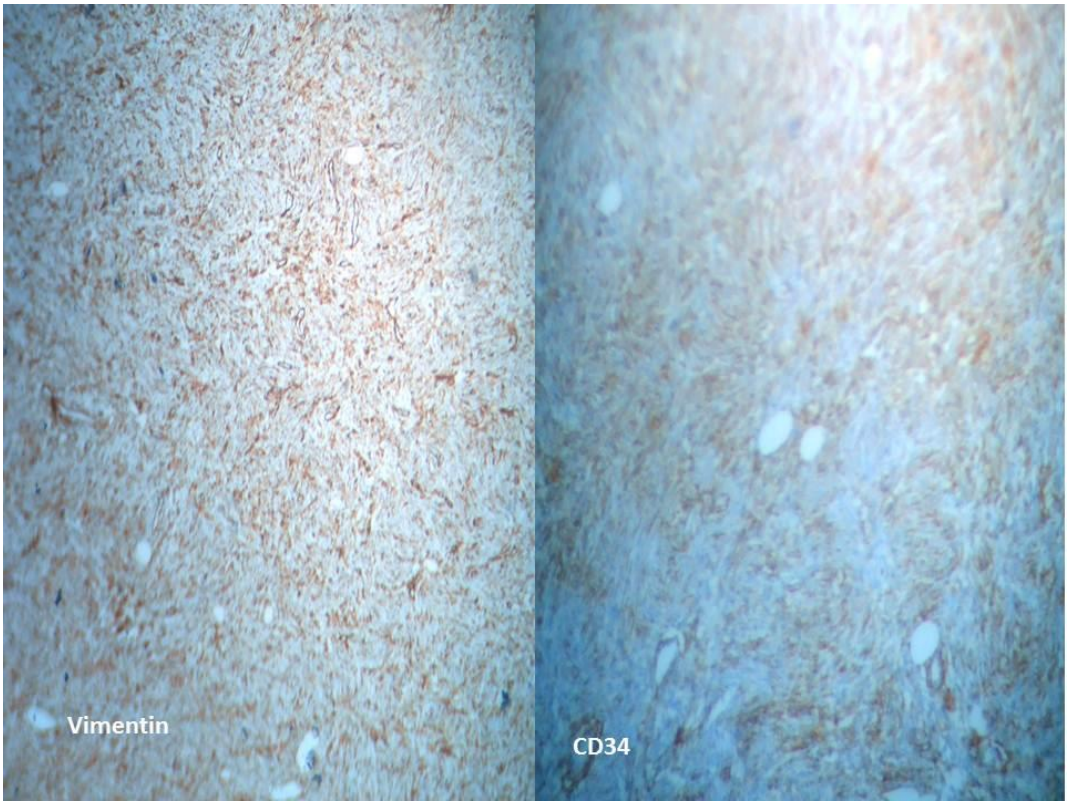


**Figure 5:** Immunohistochemistry study showed tumor cells positive for CD34 and vimentin.

Based on morphological features diagnosis of DFSP was done. Immunohistochemistry study showed tumor cells positive for CD34 and vimentin.[Fig.5] The combined histologic and immunohistochemical findings were diagnostic of DFSP over anterior chest wall. The patient is free from local recurrence or metastasis in 6-months follow-up after surgery.

**DISCUSSION**

DFSP is a rare soft tissue tumor of cutaneous origin characterized by locally infiltrative and recurring properties. It may occur at almost any site but commonly occurs on the trunk and extremities. Rarely it has been reported in the breast.[5,6] It represents 1 to 6 % of all soft tissue sarcomas. It accounts for 0.01% of all malignancies and less than 0.1% of all the cutaneous neoplasms with reported incidence being 0.8 cases per million persons a year.[3,4,7] Darier and Ferrand first described DFSP in 1924 as a progressive and recurring dermatofibroma. In 1925, based on the tendency of the tumor to develop protruding nodules, Hoffman termed it as 'DFSP'.[8] The histogenesis of DFSP is unclear. The possible origin may be fibroblastic, histiocytic or neuroectodermal. [9] Over a 90% of DFSP cases present a translocation in chromosome 17 and 22. There is t(17,22) involving COL1A1( collagen type 1a1gene) and PDGFb (platelet derived growth factors gene) respectively.[10]

A history of trauma is a possible etiological factor in development of DFSP and in 10-20% cases of DFSP history of trauma is reported.[11] Our patient had no history of trauma or surgery. Initially these cases present as a discrete asymptomatic plaque with reddish-brown or pink appearance with irregular borders, mimicking hemangioma and may be confused with localized scleroderma or solitary fibromatosis.[6]

During the later stage, it presents as a painless, multiple nodule protruding from the skin and usually less than 5 cm in size.[12] In the present case tumor size was 20x18x16 which is largest tumor size reported in the literature until now. Histologically DFSP is characterized by

multiple spindle cells arranged in a storiform pattern with elongated nuclei without significant cytological atypia or pleomorphism. In 1962, Taylor and Helwing reviewed 115 cases of DFSP and described the histological characteristics of the lesion and characterized a fibroblastic growth appearing as a low-grade sarcoma in which the tumor cells were organized in fascicles with a spiral or cartwheel arrangement. [13]Similar features were seen in our case. Immunohistochemical studies are highly sensitive for diagnosis of DFSP. In 1992 firstly CD34 were demonstrated for DFSP tumor and to be the main marker for diagnosis of the tumor.[14]DFSP show immunopositivity to CD34 and vimentin in 84-100% and negative for S100 protein and factor XIIIa.[9] In our case, the tumor cells were diffusely positive for CD34 and vimentin. The differential diagnoses include dermatofibroma, malignant fibrous histiocytoma, neurofibroma, hypertrophic scars, keloid, fibromatosis, fibrosarcoma, inflammatory myofibroblastic tumor. Deeper involvement of subcutaneous tissue ,absence of inflammatory cells,pigment laden macrophages ,giant cells and CD34 diffuse positivity ruled out dematofibroma in this case. CD34 immunopositivity and characteristic storiform pattern ruled out fibromatosis. Inflammatory myofibroblastic tumor was ruled out due to absence of inflammatory infiltrate. Absence of marked pleomorphism, mitotic activity, necrosis ruled out malignant fibrous histiocytoma. Lack of neural differentiation and presence of highly cellular tumor with mitotic activity ruled out neurofibroma morphologically. The characteristic histopathological and immunohistochemical studies ruled out other lesions like keloid and hypertrophic scar. Based on review of literature, the standard treatment of DFSP is radical wide local resection of tumor with surgical margin of 2-3 cm and three dimensional resections including skin, subcutaneous tissue and underlying fascia.[3] This was done in our case. Local recurrence occur in 20-25% cases so must be observed periodically even after surgery for a long time. The local recurrence rate decreases with wider margin of excision.[3]

Recently a newly approved molecular targeted drug, Imatinib mesylate is an effective drug for unresectable, recurrent and metastatic DFSP.[8] Local recurrence did not occur in our patient though the tumor was large. The role of post resection radiotherapy also has been reported in the literature. [14]

**CONCLUSION**

DFSP is a rare soft tissue tumor of cutaneous origin with an intermediate level of malignancy. Although metastasis rarely occurs but high rate of recurrence may occur so special attention has to be given to these patients even after surgery.

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