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## **Dermatofibrosarcoma Protuberans: Rare Tumor**

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**Keywords:** Dermatofibrosarcoma protuberans; Chromosomal translocation; PDGF (Platelet-Derived Growth Factor); Metastasis; MRI thorax; Mohs micrographic surgery (MMS).

## **Clinical Image description**

One of the rarest type of skin cancer - DFSP i.e. Dermatofibrosarcoma Protuberans, is uncommon soft tissue tumor involving dermis and subcutaneous but in some rare condition it involves muscles and fascia. Studies have implicated a chromosomal translocation resulting in overproduction of PDGF (Platelet-Derived Growth Factor) that promotes tumor growth. It has high local recurrence rate but a low likelihood of metastasis. This rare tumor has 0.5 to 4.5 cases in every million incidence rate. A male patient aged 72 years came to our hospital with complaint of swelling at chest region without pain and tenderness, having history of operated for same 2 times earlier but recurred again, earlier two operated tumor were of small in nature but at this 3rd recurrence the swelling is enormous. The sarcoma has maximum growth in last 8 days. On examination the size of tumor was 13.5\*12\*6 cm. Patient has no history of hypertension, DM, tuberculosis, thyroid disease. Surgical history-removal of sarcoma 3.5 years back. FNAC on shows hemorrhagic background with marked collection of spindle shape cells with mild inflammatory cells reaction. After evaluating the current situation patient was referred to higher center, there MRI thorax reveals a well-defined altered signal intensity lesion involving subcutaneous fat in the upper left parasternal region of chest wall with overlying skin thickening and extension as describeds/o residual/ recurrent neoplastic lesion. Histopathology report



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Table 1: Dermatofibrosarcoma Protuberans.

reveals cut surface firm. Homogenous white, areas of necrosis or hemorrhage are not seen. The optimal treatment modality for DFSP is Mohs Micrographic Surgery (MMS).

**Differential diagnosis:** Cellular fibrous histiocytoma/dermatofibroma, Solitary fibrous tumor, Spindle cell lipoma, Angiosarcoma, Peripheral nerve sheath tumors, Spindle cell melanoma, Angiomyxoma, Myxoid sarcoma, Synovial sarcoma, Sarcomatoid carcinoma.