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Dermatofibrosarcoma protuberans: A rare mesenchymal soft tissue sarcoma of skin

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Abstract
Background: Dermatofibrosarcoma protuberans is soft tissue sarcomas of skin seen less commonly
Case History: A 49-year-old male patient came with the complaints of blackish purple swelling over right side of trunk for 2 years
Intra Operatively: A swelling measuring around 2.5x 2 cm noted over right side of trunk 1 cm deep extending till subcutaneous plane
Discussion: Dermatofibrosarcoma protuberans is an uncommon slowly progressive soft tissue sarcoma involving dermis, in relation with COL1A1 gene with very rare metastatic features
Treatment usually requires wide local excision.
Conclusion: Wide local excision of the tumor is treatment of choice for Dermatofibrosarcoma protuberans.

Keywords: Blackish purple swelling, dermatofibroma sarcoma protuberans, rarely malignant, wide local excision

Introduction

Dermatofibrosarcoma protuberans is a locally aggressive, superficial mesenchymal soft tissue scarcoma of skin which is encountered less commonly, presenting as a swelling which is slowly progressive over months to years and typically noticed over trunk, extremities. It is more common in blacks. Previous burns, scars, tattooing are found to predispose this skin condition. It is associated with PDFGB/ COL1A1 fusion gene affected by the genomic gain.

This condition is seen in all age groups but most commonly seen in individuals of age 20-50 years, both male and females are equally affected. Although rare metastatic potential noted with fibrosarcomatous transformation.

Case History

A 41-year-old male patient came with the complaints of swelling over right side of trunk for 2 years, insidious onset, gradually progressive initial of size gradually progressed to present size of 2.5x2 cm, with hyperpigmentation, no history of alcohol/ tobacco's consumption, no known comorbidities

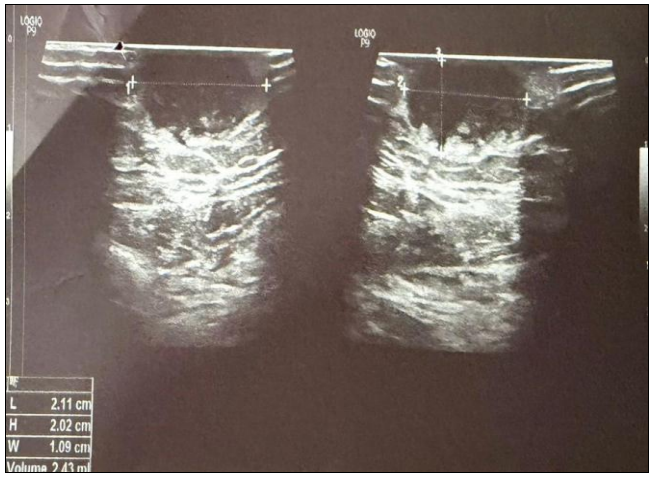
No history of pain, discharge, similar complaints in the past.

On local examination

A solitary swelling approximately 2.5x2 cm noted over right side of the trunk, 2cm below the costal margin in the midclavicular line, blackish / purple colour, ovoid shape, with regular margins, smooth surface, no visible pulsation. On palpation - no local rise of temperature, non-tender, firm in consistency, immobile

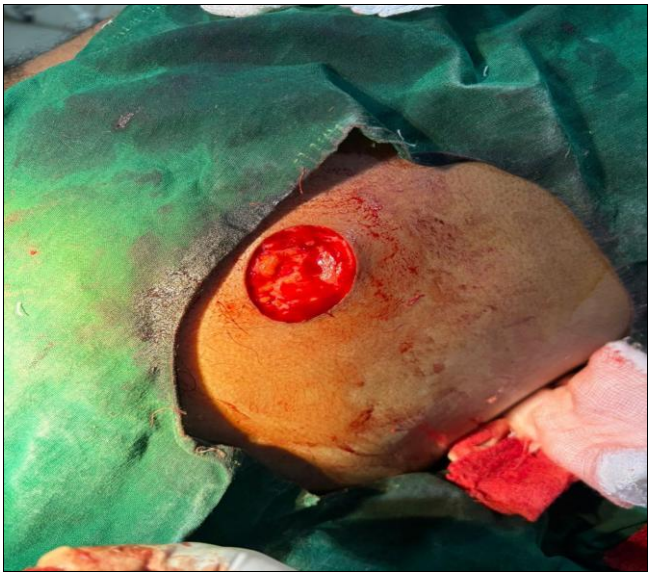
Usg Soft Tissue

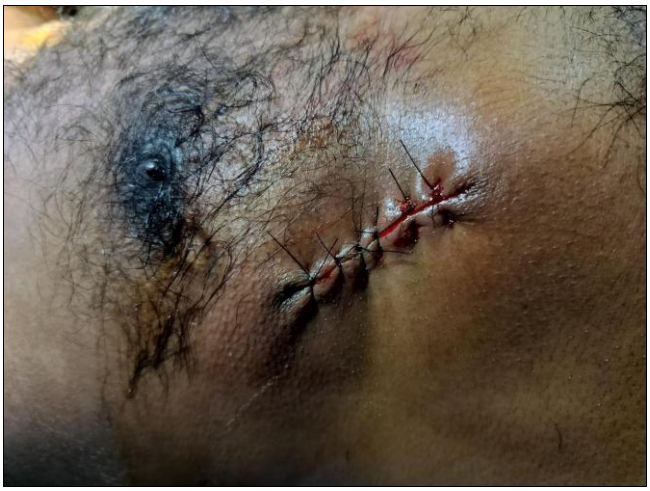
An ill- defined heterogeneousiy hypoechoic iesion measuring 2.1 x 2.0 x 1.0 cm is seen in the subcutaneous plane of right anterior chest wall with a small tract opening into skin surface. Mild surrounding fat stranding is noted -? infected sebaceous cyst. The underlying muscles appear normal

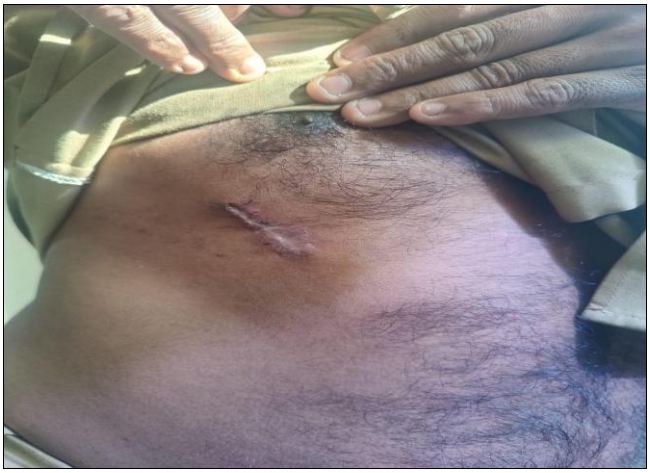


Intraoperative Findings

A 2.5x2 cm swelling noted over right side of trunk under anesthesia, elliptical incision taken and swelling is found to be extended 2cm deeply along subcutaneous plane, bleeding noted intra operatively, dissection done after achieving hemostasis







Histopathological Examination Microscopy

Excision biopsy from swelling over right lower chest.

Received single, partially covered by skin, elliptical soft tissue mass measuring 2.5 x 2 x 1 cm, cut section - solid, grey white area measuring 1.8x x x x cm.

Representative sections studied (A, B) show skin lined by stratified squamous epithelium. Deeper dermis shows an ill-defined lesion composed of spindle cells arranged in interlacing fascicles, whorls and bundles along with myxoid material. Individual cells are plump, spindle shaped, with vesicular chromatin, prominent nucleoli, and eosinophilic cytoplasm. Also seen is thin walled proliferating blood vessels. The periphery of the lesion shows spindle cell infiltrating fat in a honey comb pattern.

Impression: Histomorphological features are suggestive of fibroblastic/ myofibroblastic tumor – dermatofibrosarcoma protuberans*

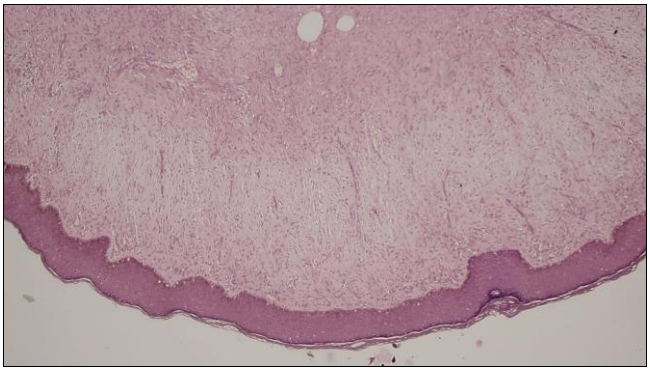


Fig 1: Sections shows skin lined by stratified squamous epithelium and dermis shows ill defined lesion composed of spindle cell

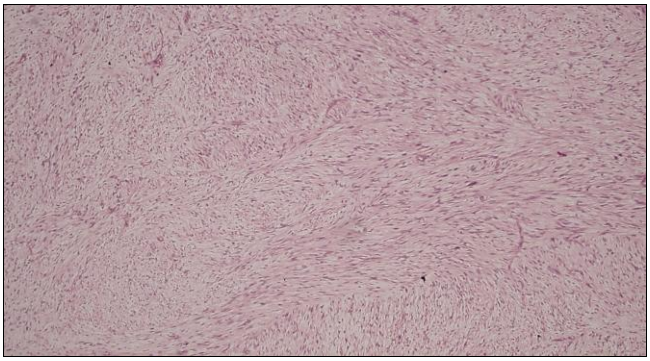


Fig 2: Spindle cells arranged in interlacing fascicles, whorls and bundles

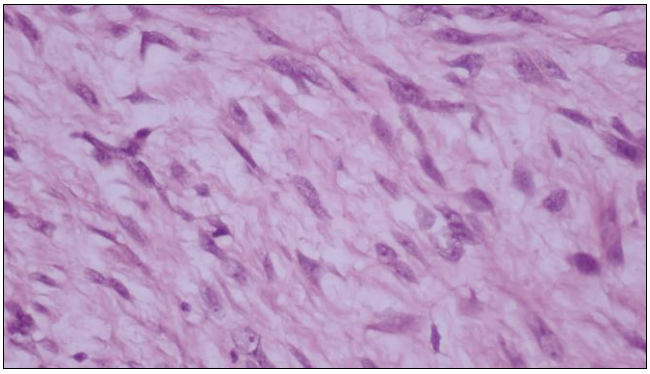


Fig 3: Plump spindle shape cells with vesicular chromatin, prominent nucleoli and eosinophilic cytoplasm

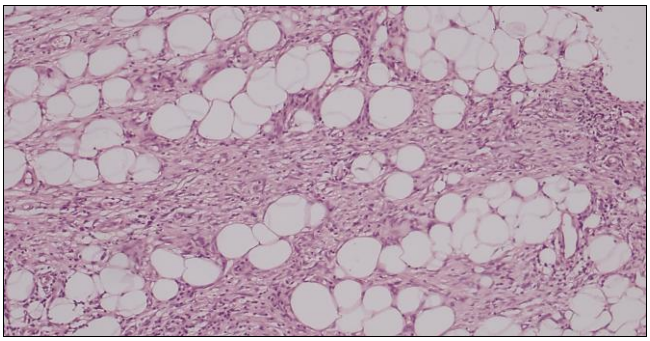


Fig 4: Periphery of the lesion shows spindle cells infiltrating fat

Discussion

DFSP is a very slowly growing tumor and the diagnosis is often delayed for months to years. In addition, misdiagnosis can occur due to inadequate tissue sampling or superficial biopsy. For this reason, National Comprehensive Cancer Network (NCCN) guidelines recommend performing a punch or incisional biopsy, including the deeper subcutaneous layer, for definitive diagnosis. [7]

DFSP is a rare tumor, occurring at a rate of 0.8 to 4.5 cases per million persons annually and represents 1% to 6% of all soft tissue sarcomas and 18% of cutaneous soft tissue sarcomas—affecting both genders equally. Some studies report a slight predominance in med. While most commonly diagnosed in adults between the third and fifth decades, DFSP can manifest across all age groups. Children account for 6% of all cases of DFSP. This tumor may exhibit accelerated growth during pregnancy. [3]

DFSP-FS accounts for 5% to 15% of all DFSPs, and the Bednar or pigmented variant represents fewer than 5% of all DFSP cases. DFSP, including the pigmented variant, is more common in Black individuals than White. [4]

Pathophysiology

The t (17;22) (q22; q13) translocation results in the formation of supernumerary ring chromosomes from chromosome 22 and contains low-level amplified sequences from 17q22-qter and 22q10-q13.1. [4, 5] Less commonly, a linear derivative of chromosome 22 may also be implicated. [6] The ring chromosomes and the translocated linear derivative of chromosome 22 harbor a fusion gene where PDGFB merges with the collagen type 1A1 (COL1A1) gene. The usually suppressed PDGFB now becomes activated by the COL1A1 promoter. This genetic rearrangement induces PDGFB upregulation, resulting in excessive platelet-derived growth factor (PDGF) production, continuous activation of the tyrosine kinase PDGFRB receptor, cellular proliferation, and tumor development. [7]

Clinical features

DFSP typically manifests as an asymptomatic, skin-colored to red-brown firm plaque, eventually progressing to multiple raised nodules with a violaceous to red-brown hue. *The* pigmented variant often presents with irregular brown pigmentation, and the atrophic variant appears as a violaceous plaque resembling morphea or scar tissue. Lesions exhibit slow growth over months to years, occasionally resembling keloids or dermatofibromas, leading to misdiagnosis, particularly in the early stages. As the tumor progresses, it may reach several centimeters in diameter, potentially accompanied by telangiectasia in the surrounding or overlying skin. The tumor is typically fixed to the dermis but can freely move in relation to underlying structures. Later in the disease, the tumor may become fixed to underlying structures. Rarely, DFSP can arise from preexisting scars or tattoos. As lesions enlarge, some may ulcerate and become painful. Nearly 50% of lesions are on the trunk, followed by 35% on the extremities, and 15% on the head and neck. [11]

Because most cases of DFSP are superficial, clinicians can assess the extent of tumor and lymph node involvement by physical examination. Imaging is not a routine part of the diagnostic process. Occasionally, imaging may be helpful for large or recurrent tumors if the clinician suspects bone invasion or needs to define the extent of the disease. Magnetic resonance imaging is the imaging modality of choice. [12, 13, 14, 15] Computed tomography (CT) is only helpful if underlying bone involvement or lung metastases are suspected. The lung is the most common site of metastasis via hematogenous spread, and regional lymph nodes are rarely involved. Due to the unlikely occurrence of lymphatic and hematogenous dissemination, an extensive staging workup is not generally necessary. Some authors suggest obtaining a radiograph or CT scan of the lungs before treatment, though the National Comprehensive Cancer Network does not make specific recommendations regarding a staging evaluation. [16] [17]

DFSP is poorly circumscribed and usually involves the dermis and subcutis, although rare cases can be limited to the dermis. Due to the possibility of distant metastasis and aggressive local invasion, DFSP is considered an intermediate tumor between a benign dermatofibroma and a frank fibrosarcoma. Transformation to a high-grade sarcoma is extremely rare. The overlying epidermis does not show any atypical histological features. [8]

The spindle-shaped tumor cells of DFSP are arranged in a storiform or woven pattern, parallel to the epidermal surface, and have little pleomorphism and scant cytoplasm. The cells are surrounded by collagenous stroma, sometimes associated with hyaline or myxoid changes. The characteristic honeycomb appearance results from irregular tentacle-like projections infiltrating the underlying subcutaneous tissue, traversing the septa and fat, leading to fat entrapment. DFSP typically extends into subcutaneous fat but seldom involves fascia, muscle, or bone unless recurrent or long-standing. Necrosis is uncommon. While mitoses are present, no significant mitotic activity is present, and atypical mitoses are rare. A mitotic count that reaches 10 mitoses per 10 high power field (HPF) and tumor size correlates with metastatic spread. About one-fifth of all DFSPs undergo fibrosarcomatous transformation, which appears as an expansive tumor with fascicular and herringbone patterns and atypical cytological features. [9] [10]

Treatment

The initial treatment for localized DFSP is surgical resection with negative margins. [18]

Wide Local Excision

Wide local excision (WLE) is an option for resection. The resection margins are essential in determining the risk of local recurrence. Tentacle-like tumor projections can extend beyond 3 cm of the primary tumor. Results from one study reveal the local recurrence in patients with a resection margin less than 3 cm is 47% compared to 7% in patients with margins of 3 to 5 cm. [20, 21, 22] The NCCN recommends margins of 2 to 4 cm with clear pathologic margins when clinically feasible

Mohs Micrographic Surgery

Mohs micrographic surgery (MMS) involves the progressive horizontal slicing of tissue during resection, coupled with immediate microscopic evaluation via frozen section analysis. Microscopic evaluation includes prompt immunostaining for CD34 until achieving a clear margin. Real-time microscopic examination of margins reduces the likelihood of positive margins. Some studies propose that MMS may result in lower local recurrence rates than WLE

for DFSP. Notably, DFSP treated with WLE exhibits a recurrence rate of approximately 7.3%, contrasting with the 1% recurrence rate observed with MMS treatment. [23] [24] However, randomized trials and comprehensive long-term data are required to confirm these findings. A significant benefit of MMS is the smaller required lateral margins, resulting in more minor wounds and less complex reconstruction. MMS is a good option for cosmetically sensitive areas, where achieving narrow margins is preferable.

Radiation Therapy

DFSP is radiosensitive tumor. Radiation therapy is rarely used alone in the treatment of DFSP.

Current NCCN guidelines recommend the use of adjuvant radiation in the following settings

- Positive margins after surgical resection
- Negative margins, with the closest margin being less than 1 cm in patients who did not undergo MMS
- For recurrent or metastatic disease when surgical excision is not feasible [25]

Molecularly Targeted Therapy

Imatinib inhibits the PDGF receptor and other receptor tyrosine kinases such as c-KIT. Imatinib has the United States Food and Drug Administration (FDA) approval for unresectable, recurrent, or metastatic DFSP. [27]

Imatinib competitively inhibits adenosine triphosphate (ATP) binding to the PDGFB receptor, slowing down kinase activity, limiting tumor growth, and promoting apoptosis. Patients with the t (17;22) (q22; q13) translocation show a more significant response to imatinib, and thus, screening for this translocation should be performed before initiating therapy.

Adverse effects associated with imatinib include gastrointestinal upset, edema, fatigue, anemia, and rash. Most patients with DFSP with translocation respond favorably to imatinib therapy, with studies suggesting a response rate of approximately 65%. The duration of therapy varies. Some sources recommend 6 months of therapy, which may be extended. [28, 29]

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