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Case Report

Surgery Section

Dermatofibrosarcoma Protuberans Presenting as a Long-standing Infraclavicular Mass in a 79-year-old Male: A Case Report

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ABSTRACT

Dermatofibrosarcoma Protuberans (DFSP) is a rare, low to intermediate-grade cutaneous soft-tissue sarcoma that is characterised by slow growth, local aggressiveness, and a high recurrence rate if inadequately excised. The case reports a 79-year-old Indian male who presented with a long-standing left infraclavicular swelling for the last 35 years, which later exhibited rapid enlargement over a four-month period. Clinical examination of the patient showed a firm, non tender, non fixed mass with non pinchable overlying skin. Imaging of the patient also showed a lobulated, heterogeneously enhancing lesion without any signs of distant metastasis. The patient then underwent Wide Local Excision (WLE); the tumour was also removed completely without infiltration of muscle. Histopathology report and Immunohistochemistry (IHC) finally confirmed the diagnosis of DFSP with negative margins. The postoperative course of the patient was uneventful, along with satisfactory recovery. The case outlines that early identification and surgical management of DFSP are necessary for preventing its recurrence.

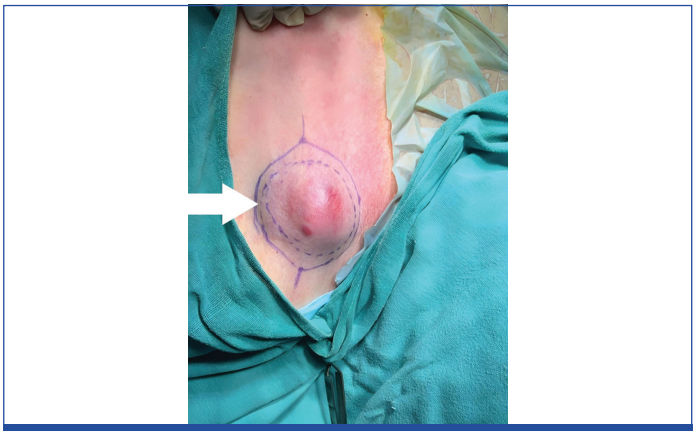
Keywords: Cutaneous sarcoma, Immunohistochemistry, Infraclavicular region, Surgical excision, Wide local excision

CASE REPORT

A 79-year-old Indian male with 20 years of hypertension on regular medication, presented with a single, firm, non tender, non fixed swelling of 8x6 cm in size located in the left infraclavicular region [Table/Fig-1]. The swelling had been present for the last 35 years, with slow, indolent enlargement, but had shown a rapid increase in size over the past four months. Although this occurred after Fine Needle Aspiration Cytology (FNAC), the temporal association was considered incidental rather than causative. The overlying skin was non pinchable and lacked ulceration or pigmentation, with no local rise in temperature. The swelling was not accompanied by pain, fever, weight loss, or systemic complaints.

There was no history of nausea, vomiting, pain in the abdomen, bowel or bladder issues, trauma, or underlying chronic disease, including diabetes, tuberculosis, or ischaemic heart disease. The past history was significant for right inguinal hernia repair with hydrocele surgery, left orchidectomy, multiple transurethral resections of the prostate for benign prostatic hyperplasia, and suprapubic catheterisation.

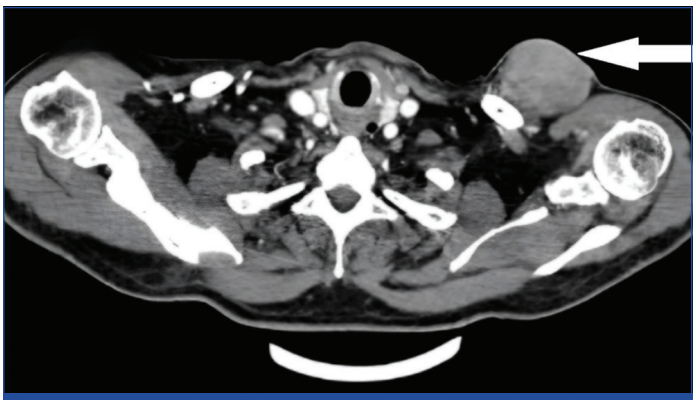
Systemic examination of the patient was within normal limits. Routine laboratory tests were also unremarkable. FNAC of the lesion revealed cellular smears composed of plump ovoid-to-spindle cells arranged singly and loosely adherent to capillary fragments, with mild pleomorphism. These findings suggested a malignant spindle cell neoplasm with possible vascular origin; however, a definitive diagnosis could not be established on cytology alone because of overlapping spindle cell features. Excisional biopsy with histopathological and immunohistochemical evaluation was advised for definitive diagnosis while core needle biopsy was avoided because of the suspected vascular nature of the lesion. Ultrasonography (USG) of the given local area showed a well-defined hypoechoic lesion with internal vascularity in the subcutaneous plane. Contrast-Enhanced Computed Tomography (CECT) of the thorax showed a lobulated, heterogeneously enhancing soft-tissue density lesion of the left anterosuperior chest wall, bordering on the clavicle, pectoralis major and deltoid muscles, supplied by branches



[Table/Fig-1]: Single, firm, non tender, non fixed swelling located in the left infraclavicular region (white arrow).

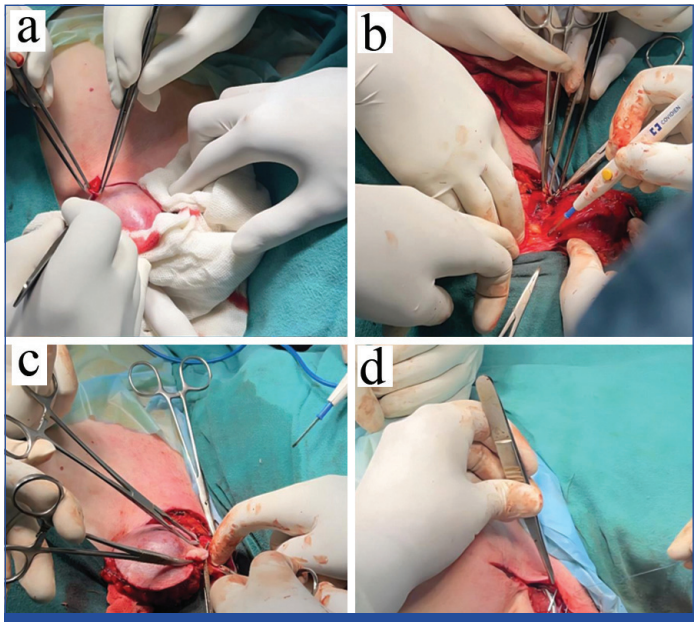
of the subclavian artery. There were no signs of distant metastasis of the given swelling [Table/Fig-2].

The patient was placed under general anaesthesia in a supine position. An elliptical skin incision was made with a 3 cm margin around the swelling. The dissection was done in layers, and the

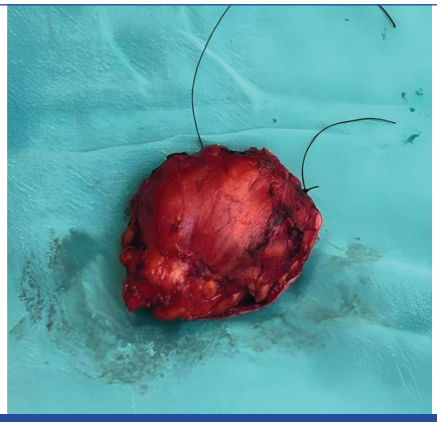


[Table/Fig-2]: CECT thorax of patient showed a lobulated, heterogeneously enhancing soft-tissue density lesion (white arrow).

tumour was accurately separated after excising the surrounding tissues. The lesion was completely excised in toto, without any evidence of infiltration into the underlying muscle planes. Betadine was used to irrigate the surgical bed, thereby achieving meticulous haemostasis. A Romovac drain was placed to prevent postoperative collection. Layered closure was done, with the subcutaneous tissue approximated using absorbable sutures and skin closed with non absorbable interrupted sutures [Table/Fig-3a-d]. The excised specimen sample of size 8.5×6.5×3.5 cm is shown in [Table/Fig-4].



[Table/Fig-3]: Intraoperative steps of Wide Local Excision (WLE) of infraclavicular mass; (a) Elliptical skin incision; (b) Layer-by-layer dissection of tumour region; (c) Complete excision of the lesion in toto, showing intact underlying muscle planes without infiltration; (d) Meticulous haemostasis was achieved, Romovac drain placed, and layered closure performed.



[Table/Fig-4]: The excised specimen sample of size 8.5×6.5×3.5 cm.

Histopathology revealed a spindle cell mesenchymal neoplasm with negative margins, mitotic activity of 9-10/High-Power Field (HPF), and absence of tumour necrosis. No definite histopathological features of fibrosarcomatous transformation, including marked cytological atypia or herringbone architecture, were identified. IHC revealed weak positivity for CD34 and focal Smooth Muscle Actin (SMA), Signal Transducer and Activator of Transcription 6 (STAT6), S-100 Protein (S-100), Desmin (DES), Epithelial Membrane Antigen (EMA), Cluster of Differentiation 31 (CD31), and ETS-Related Gene (ERG) were negative. The overall histomorphological features together with the immunohistochemical profile supported the diagnosis of DFSP and helped exclude other spindle cell neoplasms, including solitary fibrous tumour, vascular sarcoma, dermatofibroma, and fibrosarcoma. The patient was transferred into the recovery unit in good condition. His postoperative hospital stay was uneventful; the drain was removed on day 5, the wound was healing well, and he was discharged in good, stable condition on postoperative day 10 with follow-up recommendations. The patient was reviewed on postoperative day 15, at which time wound healing was satisfactory and there were no immediate postoperative complications.

DISCUSSION

The DFSP is an uncommon, low to intermediate-grade cutaneous soft-tissue sarcoma that arises from the dermal layer, also characterised by aggressive local infiltration, having a high rate of recurrence despite its indolent growth pattern [1]. The recurrence rate ranges from 0% to 41% in cases treated by WLE with at least 2-3 cm of free margin [2].

Clinically, DFSP most commonly presents as a painless, firm, and slowly growing plaque or nodular lesion that remains stable for a few years, then accelerates in growth [3]. DFSP usually progresses slowly over several years, although periods of relatively rapid enlargement can occur during its natural course [3]. Current literature does not establish FNAC or biopsy as a proven cause of tumour acceleration in DFSP. In the present case, the temporal association between FNAC and rapid enlargement was therefore considered incidental rather than causative. Imaging of the lesion further helps in adequately defining the local extent [4]. FNAC findings in DFSP may be non specific because of overlapping cytological features with other spindle cell neoplasms, including vascular sarcomas. Therefore, definitive diagnosis generally requires histopathological examination with immunohistochemical correlation. WLE with adequate margins and Mohs micrographic surgery are standard treatment approaches to minimise the risk of recurrence [5]. Although the present case demonstrated relatively increased mitotic activity, no definitive histopathological evidence of fibrosarcomatous transformation was identified. A comparative summary of reported DFSPs cases, including the present case, is depicted in [Table/Fig-5] [6-9].

Features	Present case	Gangadhar SM et al., [6]	Pardo D et al., [7]	Pathak B and Maimoon S [8]	Zeaiter N et al., [9]
Demographic details	79-year-old Indian male; hypertensive	57-year-old Indian male	62-year-old American male	32-year-old Indian female	59-year-old Lebanese female
Duration and history	35-year history of infraclavicular swelling with slow indolent growth; rapid enlargement over last 4 months post-FNAC	4-year abdominal wall mass; twice-recurrent	>24-month facial mass; gradual enlargement and pruritus	1-year enlarging lumbar swelling; prior cyst excision 4 years earlier	3-year chronic non-healing shoulder wound; initial benign excision; recurrence after 1 year
Clinical presentation	Single, firm, non tender, non fixed 8×6 cm swelling; non pinchable skin; no ulceration, pigmentation, temperature rise, or systemic symptoms	Fixed, ulcerated 10×8 cm lesion with slough, necrosis, bleeding	Two lesions: 3×3 cm firm mass+2 cm multinodular lesion; no nerve deficit	3×3 cm firm, non tender, non mobile lump; normal overlying skin	Ulcerating lesion with scar contracture; restricted shoulder movement
Imaging features	USG: hypoechoic lesion with vascularity. CECT: lobulated heterogeneously enhancing anterior chest wall mass abutting clavicle, pectoralis major & deltoid; arterial supply from subclavian branches; no metastasis	MRI: 6.6×6.9×6.3 cm exophytic mass abutting rectus muscle; no lymphadenopathy	CT: 3 enhancing subcutaneous masses superficial to parotid; no parotid involvement	USG: 3.8×2.1×3.5 cm hypoechoic lesion with septations and vascularity	No advanced imaging reported
Preoperative cytology/ biopsy	FNAC: malignant spindle cell tumour suggesting vascular sarcoma	Incisional biopsy: spindle cells suggestive of DFSP	Biopsy: spindle cell neoplasm; CD34+, PDGFβ rearrangement	FNAC: spindle cell neoplasm	Frozen section intraoperatively: negative margins

Surgery performed	Wide Local Excision (WLE) with 3 cm margins; complete excision in toto; Romovac drain; layered closure	Wide Local Excision (WLE) (R0)+ALT flap+STSG	WLE with 2.5 cm margins+superficial parotidectomy+ALT free flap	WLE including fibroadipose tissue and skin	Full-thickness excision with 1 cm margins+STSG
Tumour size on excision	8.5×6.5×3.5 cm specimen	— (clinical size 10×8 cm)	Lesions 2-3 cm, multifocal	8×3.5×2.5 cm	7×4×2.8 cm
Histopathology Findings	Intermediate-grade mesenchymal tumour on frozen section; Final HPE and IHC consistent with DFSP without fibrosarcomatous transformation; negative margins	Spindle cells, CD34+, negative margins	High-grade DFSP with fibrosarcomatous transformation; CD34+, PDGFβ rearrangement	Storiform pattern, honeycomb fat infiltration, grenz zone	Spindle cell neoplasm consistent with DFSP; negative margins
Postoperative management	Uneventful recovery; drain removed day 5; no adjuvant therapy	No adjuvant therapy	Completed adjuvant radiotherapy	Uneventful recovery; no adjuvant therapy	Dressings twice weekly; complete graft healing in 2 months
Outcome and follow-up	Discharged in stable condition; good postoperative recovery	Disease-free for 2 years	Good recovery; on adjuvant RT	Pain/swelling resolved; positive follow-up	Good healing with restored shoulder function

[Table/Fig-5]: Comparative summary of reported Dermatofibrosarcoma Protuberans (DFSP) cases including the present case [6-9].
ALT: Anterolateral thigh; HPE: Histopathological examination; PDGFβ: Platelet-derived growth factor beta; R0: Complete resection with negative microscopic margins; RT: Radiotherapy; STSG: Split-thickness skin graft.

CONCLUSION(S)

The DFSP is a rare, locally aggressive cutaneous sarcoma that can remain indolent for decades before suddenly becoming aggressive. The present case is noteworthy for the unusually prolonged 35-year duration of the lesion and rapid enlargement, which was temporally associated but considered incidental to the prior FNAC. Early diagnosis and timely intervention are essential for preventing recurrence and improving functional outcomes in patients.

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