Case Report

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A Rare Case of Dermatofibrosarcoma Protuberans of the Left Arm

Oncology Section

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare form of soft-tissue tumour mostly arising from the dermis layer of the skin, precisely from fibroblastic cells. These are slow growing tumours, usually infiltrating surrounding tissues and are locally invasive. These tumours can infiltrate the subcutaneous tissue, and also the underlying fascia, muscle or deeper structures. These tumours can be neglected due to its initial non-specific and painless clinical presentation. Haemorrhagic and cystic presentations are commonly noticed in large tumours of >5 cm size. It can be classified as low- to intermediate-grade tumours. These are associated with chromosomal translocation, which produces fused COL1A1-PDGFB protein in majority of the cases. Also, dermatofibrosarcoma protuberans has been reported to have higher local recurrences but a favourable prognosis. A regular follow-up is crucial, attributed to its local recurrence risk. These tumours are managed by local incision and adjunct radiation or immunotherapy, if required. This case of 35-year-old male, presented with ulcerative growth over his left arm. The patient was managed by wide local excision and Latissimus Dorsi (LD) flap graft.

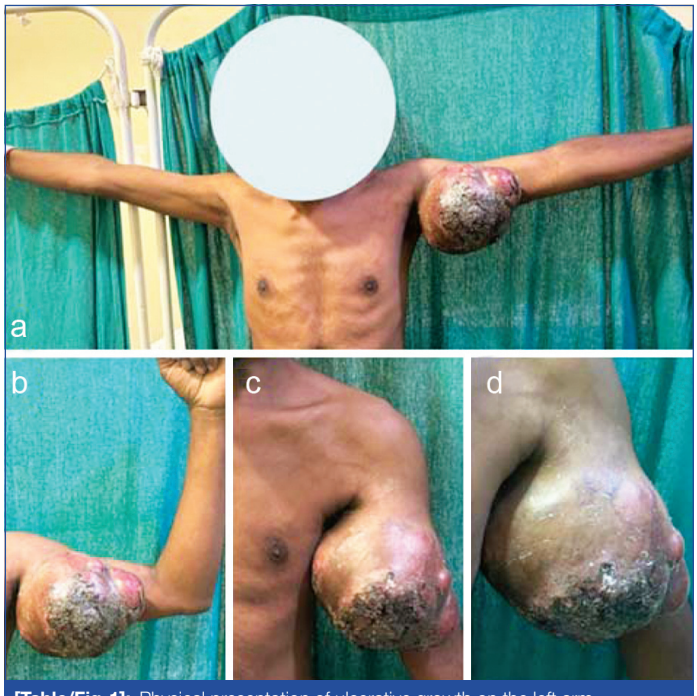
Keywords: COL1A1-PDGFB protein, Dermatofibrosarcoma, Low grade tumours, Slow-growing tumours, Split skin grafting

CASE REPORT

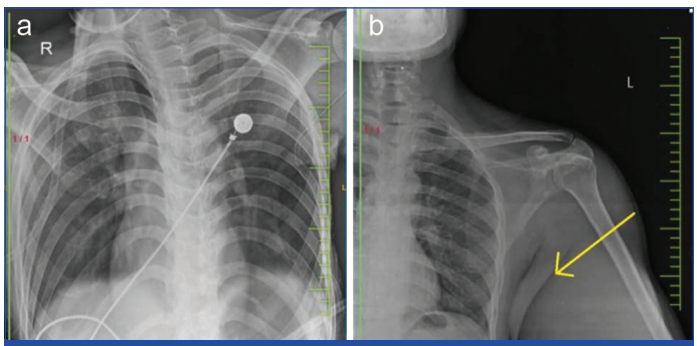
A 35-year-old male presented with a major complaint of rapidly progressive ulcerative swelling on his left arm. The patient was a labourer by profession with no addiction, negative family history, or any other co-morbidities. The swelling was firm, hyper-pigmented and ulcerative growth, which bled on touch [Table/Fig-1]. The patient was further subjected to X-ray imaging of the lump, which was suggestive of the presence of soft-tissue lesion [Table/Fig-2]. The lump was subjected to High Resolution Computed Tomography (HRCT) of the left upper arm that showed a section revealing a large well-defined heterogeneous soft-tissue density mass lesion with few cystic areas measuring approximately 16.0×12.6×15.6 cm with no evidence of calcification. HRCT was further followed by Magnetic Resonance Imaging (MRI), which revealed a large lobulated heterogeneously enhancing soft-tissue lesion with a few non-enhancing areas of necrosis noted in the superficial plane of medial aspect of the left arm. The lesion showed T2/STIR heterointense and few hyperintense areas within T1 hypointense with areas of blooming within the lesion of approximate dimensions 11.5×15.3×12.7 cm [Table/Fig 3]. Diffuse subcutaneous edema with intervening streaks of fluid was also noted with multiple collaterals in subcutaneous plane in the anterior chest wall and proximal one-third and distal one-third of the left arm. Evidence of focal narrowing of brachial vein just above the elbow joint was observed which was noted to be compressed likely by the lesion, though, it recanalised in the distal one-third of the arm.

Since, it was an ulcerated, rapidly progressive growth, soft-tissue sarcoma, neo-adjuvant radiotherapy might worsen the growth, hence, surgical excision was planned with limb amputation SOS. The patient was managed by wide local excision of the soft-tissue sarcoma over the left arm with LD flap and split skin grafting. The excised sample was subjected to biopsy which showed a spindle-shaped cellular arrangement in a whirled/storiform pattern. This was suggestive of malignant mesenchymal tumour (sarcoma) with a possibility of neural/vascular origin with TNM staging mentioned as pT4NxMx and negative margin infiltration [Table/Fig-4,5].

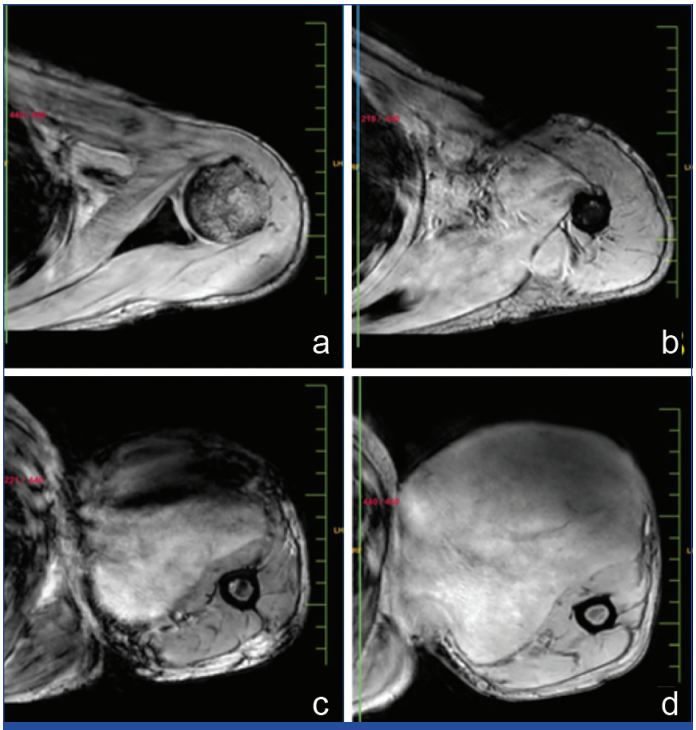
Immunohistochemistry tests for soft-tissue panel were found as Myogen-negative, MYOD1-negative, CD34-positive, SMA-negative, Desmin-positive, S100-negative, and CK-negative. Final diagnosis based on histopathological and immunohistochemical markers was concluded as dermatofibrosarcoma protuberans. The procedure was uneventful and patient was discharged on the 7th Postoperative



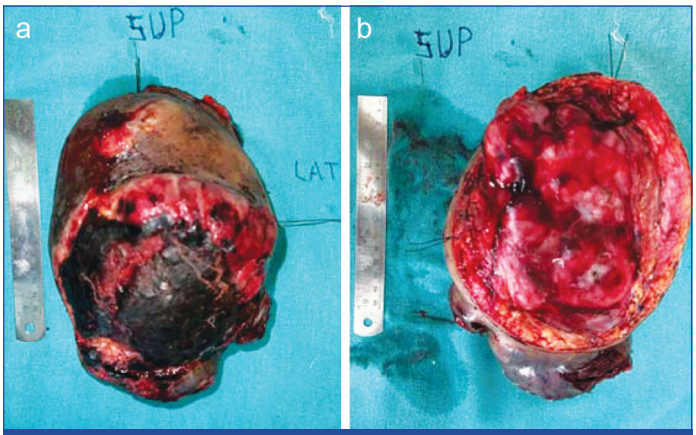
[Table/Fig-1]: Physical presentation of ulcerative growth on the left arm.



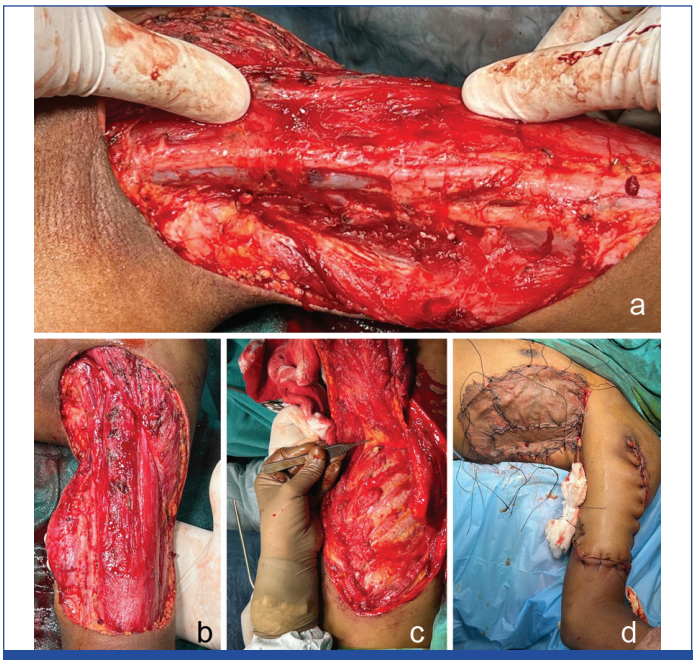
[Table/Fig-2]: Radiographic imaging of the patient's left arm.



[Table/Fig-3]: High Resolution Computed Tomography (HRCT) imaging of the tumour.

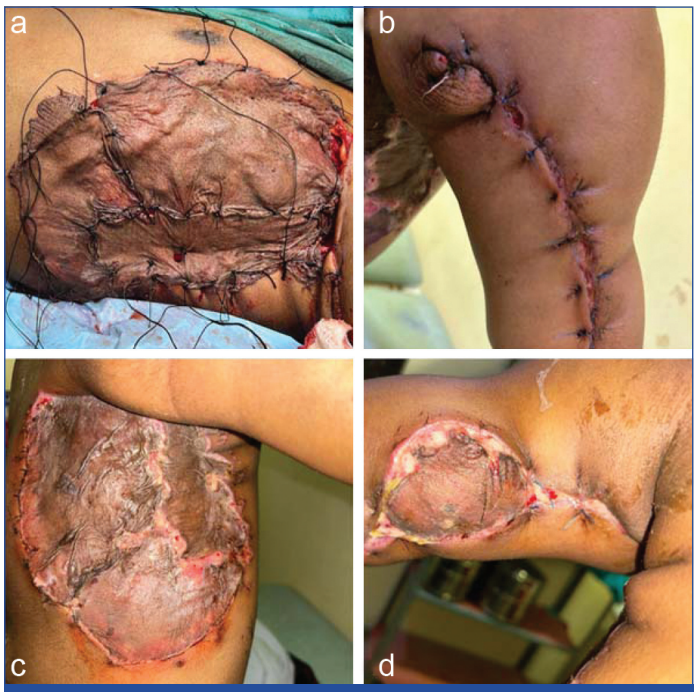


[Table/Fig-4]: Excised oval and lobulated specimen, measuring about 12 cm in the greatest dimension with irregular surface, areas of haemorrhage and necrosis, suggesting tumour necrosis or ulceration. The skin surface is stretched and ulcerated, indicating tumour invasion up to or through the dermis and subcutaneous tissue. The anatomical orientation is shown by labelling Superior (SUP) and Lateral (LAT) positions for the specimen.

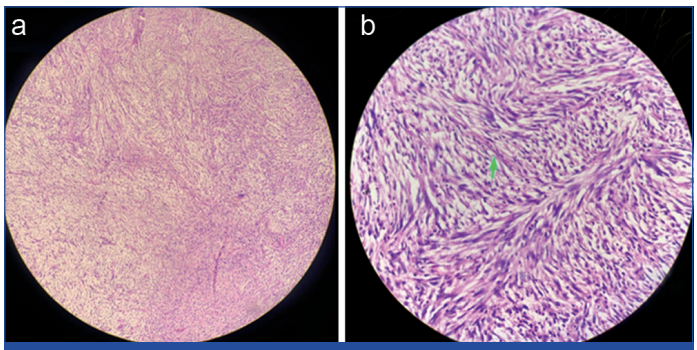


[Table/Fig-5]: Intraoperative image and split skin flap grafting.

Day (POD) [Table/Fig-6], with an advice of adjuvant radiotherapy delivering a total of 60 Gy in 30 fractions using Intensity-Modulated Radiation Therapy (IMRT) technique. Histopathology was suggestive of dermatofibrosarcoma protuberans by exhibiting spindle cell fascicles growing in a storiform pattern [Table/Fig-7]. The patient was found recovering well at six months follow-up.



[Table/Fig-6]: Postoperative image at the time of discharge showing healthy postoperative recovery.



[Table/Fig-7]: Histopathology slide (H & E staining) of the excised specimen: a) 10X magnification; b) 40X magnification.

DISCUSSION

Dermatofibrosarcoma protuberans is a rare and slow-growing tumour arising from the dermis of the skin. This soft-tissue sarcoma has distinct characteristics with specific CD34 expression counted as definitive diagnosis. It is commonly observed in individuals between 30-49 years of age [1]. The underlying etiology of most of these tumours is reported to be connected to a chromosomal translocation changes t(17;22) (q22;q13), resultant of the production of abnormal fusion protein COL1A1-PDGFB, which is noted in 90% of the patients. It is commonly noted in the proximal extremities and trunk area [2]. Most of the dermatofibrosarcoma protuberans tumours are low-grade with only 15% high-grade tumours. These tumours are reported to have high rates of recurrences and some are also associated with distant metastases [3]. Differential diagnosis was dermatofibrosarcoma protuberans, giant cell tumour of tendon sheath, liposarcoma, rhabdomysosarcoma, malignant peripheral nerve sheath tumour, synovial sarcoma, and fibrosarcoma. However, the diagnosis of dermatofibrosarcoma protuberans was made attributed to the histopathology and immunohistochemistry markers. Dermatofibrosarcoma protuberans is mostly noted in head, neck and extremities and only a small percentage is observed in upper extremities. In this patient, dermatofibrosarcoma protuberans in the

upper arm was noted, which had an unusual size, as compared to typical dermatofibrosarcoma protuberans tumours which is often <5 cm. The presentation of these tumours might look benign and be ignored, which might be resultant of adverse outcomes such as amputation [4,5]. Recommended management approaches for dermatofibrosarcoma protuberans is surgical excision either wide local incision or Mohs micrographic surgery and neoadjuvant therapy in the cases with unresectable margins [6]. This is a case of 35-year-old male with progressive ulcerative swelling in his left arm. The patient was diagnosed to have dermatofibrosarcoma protuberans and was managed by wide local incision with skin flap and split skin grafting as recommended as per the standard guidelines [7]. Though dermatofibrosarcoma protuberans is defined by aggressive local infiltration and commonly associated with a high rate of recurrence; on the contrary, there are very long durations associated with dermatofibrosarcoma with some reported to continuously expand for as long as 23 years [8,9]. There was a similar observation noted in this case as well, where the tumour reached its current size. A recent systematic review reported another treatment approach of Mohs micrographic surgery to have lowered rate of recurrences and recommended, if available [10]. Dermatofibrosarcoma protuberans can be misdiagnosed with other common lesions such as fibroma, atrophic plaques, and haemangioma, attributed towards its low prevalence [9]. More cases of dermatofibrosarcoma protuberans need to be reported in order to increase awareness of these tumours and help in timely intervention.

CONCLUSION(S)

Dermatofibrosarcoma protuberans is rare but a significant skin condition, which needs timely intervention and regular monitoring to manage recurrence, which can occur even after post-treatment. It can be misdiagnosed with other common neoplastic presentations, due to its less incidence. Overall, appropriate management and regular follow-ups can be helpful in favourable outcomes.

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