

Case Report

Dermatofibrosarcoma protuberance: a recrudescence dermal malignancy

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ABSTRACT

Dermatofibrosarcoma protuberance is a rare soft tissue tumour of low malignant potential, commonly seen on the trunk and extremities. It is a slow growing sarcoma, with a tendency for recurrence. Rarely, it may be a high-grade tumour. It responds well to wide local excision. Radiotherapy is beneficial for margin positive and high-grade tumours. We present a case of a 22 years old male with a dermatofibrosarcoma protuberance presenting as a swelling over the left inguinal region since, 4 years, that was excised with a 2cm margin. Patient was followed up for six months without any recurrence.

Keywords: Dermatofibrosarcoma protuberance, Sarcoma, Wide local excision, Imatinib, Spindle cells, Storiform pattern

INTRODUCTION

Sarcomas are soft tissue tumours, originating from any part of the body, but most often affect the extremities followed by trunk, retroperitoneum, internal organs and head and neck.¹ Dermatofibrosarcomas are rare sarcomas of the skin, with a high tendency for recurrence. Lymph node involvement and distant metastasis is infrequent and if present, may involve the lungs.² Treatment is wide local excision. Moh's micrographic surgery with continuous histological margin control is propagated to reduce local recurrence. Radiotherapy, both pre- and post-operative, reduces recurrence. Imatinib has been tried to achieve partial and complete remission.³

CASE REPORT

A 22 years old male, presented to the out-patient department with a swelling over the left inguinal region since, 4 years. Swelling was insidious in onset, gradually

progressive and increased to the present size over the course of 4 years. Swelling was a cause of embarrassment to the patient, who concealed it for a long period of time and didn't seek any medical attention for the same. It was not associated with pain, ulceration or any discharge. He did not have any other swelling on his body or any similar swelling in the past. There was no contributory family history. Patient did not have any other complaints or co morbidities. On examination, there was a swelling in the left inguinal region of approximately 15cm length and 5 cm breadth at its widest part, with a height of approximately 3 to 5 cms from skin surface. Surface was bosselated, with skin hyperpigmentation and multiple hard nodules, largest of which was approximately 3×3 cm dimension, felt within the substance of the swelling. There were no palpable lymph nodes. General and systemic examination were unremarkable.

Ultrasound of local part revealed, multiple well-defined rounds to oval lesions in the subcutaneous plane of left

inguinal region. They were heterogeneously hyperechoic with increased vascularity within. Underlying femoral vessels appeared normal while a few blood vessels were seen arising from external iliac/ common femoral vein and extending to the subcutaneous plane to the aforementioned lesion, likely suggestive of neoplastic etiology.

Patient was posted for wide local excision under spinal anaesthesia after taking proper consent. Wide local excision with 2 cm margin was done. Suction drain was kept in situ for 4 days, due to the large size of the defect. Patient was discharged on post-operative day 7 and sutures were removed on day 12.

Histopathology report revealed the mass to be a conventional dermatofibrosarcoma protuberance, with tumour cells arranged in storiform pattern, spindle cells admixed with collagen and fibroblasts, inconspicuous vasculature, scant nuclear pleiomorphism and less than 5-10/HPF mitotic activity. Tumour was 1 mm away from skin, lateral and deeper margins were negative.

Patient was counselled about possible recurrence. As the tumour was low grade and margins were negative, adjuvant therapy was not given. Patient followed up for 6 months without any recurrence.

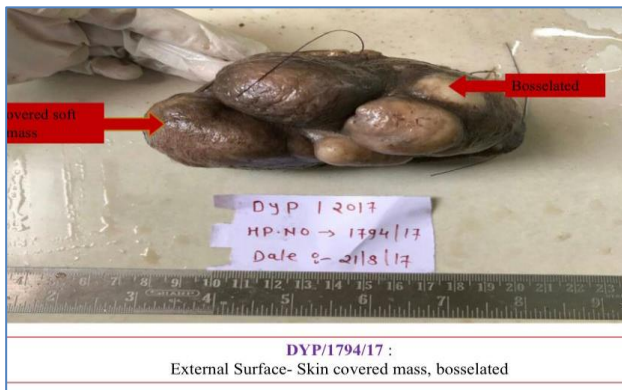


Figure 1: Excised DFSP.



Figure 2: Cut surface of DFSP.

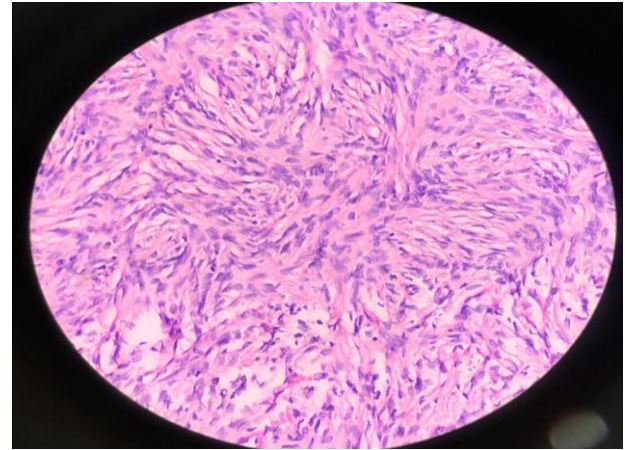


Figure 3: 10X spindle shaped cells in storiform pattern in a low-grade tumour.

DISCUSSION

Sarcomas are rare mesenchymal tumours (1% of all tumours), having more than 70 histological types. Mastrangelo et al concluded that in 1558 patients, the most common visceral sarcomas were GIST (62.3%), leiomyosarcoma (16.2%), endometrial stromal sarcoma (5.5%), while the most common soft tissue sarcomas were liposarcoma (26.2%), leiomyosarcoma (16.1%) and dermatofibrosarcoma protuberance (10.1%).⁴ in the United States, sarcomas account for 0.63% of cases and 1.15% of cancer related deaths.⁵ Dermatofibrosarcoma is a rare cutaneous variant of sarcoma, first described in 1890 by Taylor.⁶ The term 'dermatofibrosarcoma' was coined by Hoffman in 1925.⁷ These are red- violet plaques, with or without surrounding tissue telangiectasia and are most often low grade tumours, with approximately 5% being high grade tumours known as DFSP-FSSs. These are slowly infiltrative tumours with low metastatic potential but high recurrence rates. They are usually fixed to overlying dermis but not to underlying tissue, unless it is an advanced or recurrent disease.⁸ They are seen at any age, but most often in the 2nd and 5th decade.⁹

Symptoms may be present for a few months to a few years. Trunk is the most common site of involvement followed by lower, then upper extremity, but can occur anywhere on the body and most tumours being superficial in location. Clinical examination is done to ascertain the size, consistency and fixity and examination of regional lymph nodes. MRI is done for large, atypical and recurrent tumours. MRI of tumours in T1 images shows isointense or slightly hypointense tumour compared to skeletal muscle and T1 images show lower signal than subcutaneous fat, while T2 images show tumour with higher signal than fat.¹⁰ CT scan offers no valuable insight for local disease, unless it involves bone or there are pulmonary or other metastases. FDG- PET can be done to look for distant metastases, and its local uptake indicates an aggressive disease.¹¹ FNAC can diagnose soft tissue sarcomas to an accuracy of 95% in the hands

of an expert pathologist, but due to the possibility of misdiagnosis, it isn't routinely done.¹² However, it can aid in giving preoperative radiotherapy and decrease the chances of local recurrence. Treatment is wide local excision. Patient needs to be followed up for recurrence, as they may occur beyond 5 years as well.¹³ Some studies have reported recurrence rates of upto 50%, but many factors like positive margins, grade of tumour and follow up play an important role in it. Ideally 2.5-3 cm margin of surrounding tissue, including underlying fascia is needed to prevent recurrence.¹⁴ Radiotherapy brings down the chances of recurrence, and can be given both pre and post operatively.¹⁵ Radiotherapy is recommended for positive margins. Immunotherapy with Imatinib, a tyrosine kinase inhibitor that causes inhibition of oncogenic pathways and modulation of immunological processes is under trial.¹⁶ It is useful in recurrent and high-grade disease, although resistance has been reported.¹⁷ Positive margin and high grade on histology are associated with recurrence.

CONCLUSION

Dermatofibrosarcomas are rare soft tissue tumours, usually of low-grade malignancy but with a very high propensity to recur. They need diligent follow up and management. Adjuvant radiotherapy prevents recurrence and is of great value in high grade and margin positive tumours.

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