

CASE REPORT

DERMATOFIBROSARCOMA PROTUBERANS OF TRUNK IN A MALE PATIENT

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Dermatofibrosarcoma protuberans is a rare cutaneous malignancy. Usual sites of origin are trunk and extremities. Aetiology of the condition is not well understood but a genetic basis is explained as a reciprocal translocation t (17;22) (q22; q13). Distant metastasis is quite rare as it is mostly a locally advancing tumour. Histopathology is the most accurate investigation to make a diagnosis. Imaging is still required to rule out distant metastasis. Surgical excision is the treatment option for stage I tumours. Imatinib, a tyrosine kinase inhibitor, has shown promising results in management of locally advanced and metastatic disease.

Keywords: Dermatofibrosarcoma; Histopathology; Imatinib

Citation: Hashmi JZ, Ghafoor A, Hiraj MRK, Chaudhary WA. Dermatofibrosarcoma protuberans of trunk in a male patient. J Ayub Med Coll Abbottabad 2022;34(2):364-5.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an uncommon tumour that arises from dermis, is locally invasive and metastasis is rare.¹ It is a moderately or poorly differentiated sarcoma that can arise on trunk, extremities, head, neck and is often labelled as cauliflower-like tumour.

Annual incidence of DFSP is reported as 5 in 1,000,000 according to an American study which included cases from 1973 to 2002.² Although it occurs equally in males and females but females are slightly more prone than men. The size of the tumour usually ranges from 1-3 cm but size up to 12 cm has also been seen.³ Some researchers have tried to associate rapidly proliferating damaged tissue and previous history of trauma with DFSP but there is no evidence to support it and aetiology of the tumour is still under investigation.⁴ Genetic basis of the tumour is explained as a reciprocal translocation t(17;22) (q22;q13) which results in upregulation of the platelet derived growth factor subunit B (PDGFB) gene. PDGFB gene expresses itself as COL1A1-PDGFB fusion oncogene.

CASE REPORT

A 27-year-old male reported in surgical outpatient with complaints of a mass in left breast for 4 years, gradually progressing in size and now presenting with bloody discharge due to skin necrosis. He also complained of vomiting for 1 day. There was no history of trauma, fever and weight loss. There was no previous history of pulmonary tuberculosis. Patient was non-smoker and non-addict. Systemic history was not significant.

On clinical examination, he was a lean middle-aged man of average build and height. His pulse rate was 84 beats/min, blood pressure 120/70 mmHg, respiratory rate 16 breaths/min, and temperature 98 °F. There was an 8x12 cm protuberant mass on the left

lower breast region as shown in Figure-1. There was an area of 5x5 cm skin ulceration on the tip of the mass with indurated edges. The mass was not fixed to the underlying structures. It was firm, non-fluctuant and non-trans illuminant. No axillary lymph nodes were palpable. His Hb was 10 g/dl and TLC was 9000 /mm³. Renal and liver function tests were unremarkable. Histopathology of the core biopsy showed it to be a dermatofibrosarcoma protuberans. CT scan chest, abdomen and pelvis did not reveal any distant metastasis.

After appropriate preoperative preparation, wide local excision of tumour along with subcutaneous tissue and superficial fascia with 3 cm margin all around was done under general anaesthesia and sample sent for histopathology. Postoperative course was smooth and patient was found to be doing well on follow up one week after the discharge at surgical outpatient clinic.



Figure-1: Figure showing the specimen of dermatofibrosarcoma

DISCUSSION

Dermatofibrosarcoma protuberans is a rare soft tissue sarcoma of dermis most commonly found on trunk in patients of all ages. As it is locally invasive with only 2–5% chance of distant metastasis, it is labelled by World Health Organization as an intermediate fibroblastic tumour.⁵ Aetiology of the disease is still not defined. Imaging studies only give a high degree of suspicion and it is diagnosed on histopathological findings.

Imaging studies (ultrasound, mammography, computed tomography and MRI) provide a high degree of suspicion and each imaging modality has its own unique findings. Ultrasound and mammographic findings are not very conclusive. Ultrasound may show nonspecific findings such as lobulated, hypoechoic region or irregular mixed echoes. Mammogram may reveal a subcutaneous smooth mass with well-defined margin. CT scan shows an isodense or slightly hypodense region which has well defined margins whereas MRI usually shows prolonged T1 and T2 relaxation phases. Although imaging studies only give nonspecific clues but their real value lies in the differential diagnosis, localization of tumour and estimation of depth of invasion.⁶ Therefore preoperative imaging is necessary for planning safe surgical excision.

Definitive diagnosis of DFSP is made on pathological and immunohistochemical examination of the excised segment. Various histological types of DFSP have been described such as pigmented (Bednar tumour), giant cell fibroblastoma-like, atrophic, sclerosing, granular cell variant, fibrosarcomatous and myxoid DFSP.¹ Fine needle aspiration cytology and core biopsy often don't provide reliable evidence therefore surgical excision is considered more helpful in diagnosis. CD34 is found in 80-100% of the specimens of DFSP and is considered a reliable diagnostic marker.⁷

Dermatofibrosarcoma protuberans is somewhat radiosensitive but resistant to chemotherapeutic agents. Therefore, upfront surgery is the most appropriate treatment. Wide local excision with a margin of 3-5 cm is associated with less chance of recurrence (0–30%).⁸ Moh's surgery is the best technique to be employed in the treatment of DFSP with an added advantage of primary closure of the defect.⁹

Association of DFSP to reciprocal translocation t(17;22) (q22;q13) which results in up regulation of the platelet derived growth factor subunit

B (PDGFB) gene make targeted therapy a good treatment modality for stage 4 and metastatic disease. Imatinib mesylate, a tyrosine kinase inhibitor, was tested in 2005 on 10 patients for treating metastatic and locally advanced stage 4 disease and results were reported to be very promising.¹⁰ Since then it is used regularly for treating complicated tumours.

In conclusion, dermatofibrosarcoma protuberans is a rare sarcomatous tumour. Due to its initial slow progression, it is often taken as a benign disease leading to late diagnosis. Imaging modalities help in the differential diagnosis and planning of surgery while definitive diagnosis is made on histological findings and immunohistochemical markers. Wide local excision is the best treatment modality and Imatinib is reserved for locally advanced and metastatic tumours.

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Submitted: January 12, 2021

Revised: June 18, 2021

Accepted: October 24, 2021

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