Original Research Paper

Volume - 15 | Issue - 04 | April - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Surgery

A RARE CASE OF DERMATOFIBROSARCOMA PROTUBERANCE IN A YOUNG MALE

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ABSTRACT Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia. The tumor typically presents as a slowly growing, firm plaque on the trunk of young adults. The cause of dermatofibrosarcoma protuberans is not clearly understood. Studies have implicated a chromosomal translocation, resulting in the fusion protein COL1A1-PDGFB, which promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF). Diagnosis is made via biopsy.

KEYWORDS : Dermatofibrosarcoma, Excision, mesenchymal

INTRODUCTION:

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia.

Case Report:

A 20 year old male patient present in OPD with complaints of mass lesion over left pectoral region, 11 cm*7 cm in size.

The mesenchymal tumor is thought to originate from a dermal stem cell or undifferentiated mesenchymal cell with fibroblastic, muscular, features. Dermatofibrosarcoma protuberans usually extends into the subcutaneous fat but rarely involves the fascia, muscle, or bone unless it is longstanding or recurrent.

It was not associated with pain, fever, night sweats or loss of weight. Mass was firm and non tender in nature, with violaceous hue and dilated vessels on overlying skin..

Investigation:

CXR, AXR: No abnormality noted

USG(A+P+T): approx. 95*54*47mm sized well defined heterogeneously hypoechoic lesion with significant vascularity, abutting subclavian artery posteriorly, subclavian vein laterally, no mediastinal lymph node enlargement, no lung metastasis. s/p/o neoplastic mass lesion

MR Angiography(u/l): lesion in subcutaneous plane and supplied by a branch of first part of left subclavian artery.

Management:

Wide Local Excision Done: Intra-Operatively, approx. 11 cmx7 cmx3 cm sized mass was excised out keeping margin of 2 cms and the raw area was covered with split thickness skin grafting.

Post Op: postoperative period was uneventful. oral diet started on day 1. Patient was discharged on postoperative day7.

Histopathology:

Microscopic examination was suggestive of low grade dermatofibrosarcoma protuberance.

DISCUSSION:

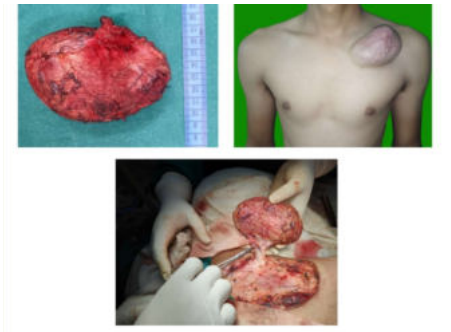
Dermatofibrosarcoma protuberans is considered an intermediate-grade malignancy with a low likelihood of metastasis but a high local recurrence rate.

Dermatofibrosarcoma protuberans is known to have relentless growth with asymmetric, root-like projections which cannot be appreciated on clinical exam. Thus, local recurrence following excision is common. The treatment of dermatofibrosarcoma protuberans is surgical removal, ideally with Mohs micrographic surgery (MMS) or wide local excision depending on location.

CONCLUSION:

Dermatofibrosarcoma is rare but can cause complications. Overall, prognosis is good.

Photos:



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