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Case Report

Dermatofibrosarcoma protuberans - an atypical tumor with a typical diagnostic dilemma: a case report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare tumor of dermal fibroblast. Diagnosis poses a challenge to every surgeon. Its propensity to recur can be attributed to its characteristic finger-like projections, misdiagnosis, and incomplete excision. Wide local excision with adequate margins may lead to functional defects and cosmetic blemishes. Most cases undergoing second surgery warrants reconstructive procedure which might further complicate the management. We report a case of DFSP, initially misdiagnosed and underwent incomplete excision, later presented with recurrence and atypical features. High-grade suspicion is of paramount importance for ascertaining the diagnosis of DFSP and its appropriate treatment.

Keywords: Dermatofibrosarcoma protuberans, Recurrence, Misdiagnosis, Wide local excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing, low to intermediate grade neoplasm originating putatively from dermal fibroblast. Among all soft tissue sarcoma DFSP accounts for 1%, reported incidence ranges from 0.8 to 5 cases in a million population per year with higher incidence among black races, male sex and old age.[1] Though atypical presentation is not uncommon, majority of patients lies between 20 to 59 years of age.[2] Clinical diagnosis is often misguided due to other differentials and high-grade suspicion is of paramount importance in the proper management of the patient. Local aggressive behavior and recurrence are the two important salient features of DFSP.[3] Recurrence is attributed to misdiagnosis and incomplete resection as well as finger-like projections which may lead to residual disease and incomplete removal. Here, the index case misdiagnosed at first and underwent incomplete excision of the tumor. Upon recurrence diagnosis of spindle cell cancer was made on fine needle aspiration cytology (FNAC), which finally turned out to be dermatofibrosarcoma protuberans on histopathology. This case is being reported in consideration of its atypical presentation and typical diagnostic dilemma in such cases. This case report is in line with surgical case report (SCARE) criteria.[4]

CASE REPORT

A 36-year-old male presented to the outpatient department with a history of recurrent lump over his left shoulder for the past 2 years. He had a history of lump excision 3 years ago which recured within 1 year at the same site, with the appearance of two more similar small lumps in proximity. Physical examination revealed well defined lobulated lump in the left supraclavicular region of size approximately 5×4×4 cm, two other lumps of size 2×2×1 cm with similar characteristics noted anterior to the above-mentioned lump (Figure 1a). These were hard in consistency with a smooth surface and erythematous skin but restricted in mobility.

Ultrasonography revealed a solid lobulated mass in the subcutaneous plane of the left supraclavicular fossa with edema and minimal vascularity. Fine needle aspiration cytology suggested spindle cell neoplasm. Magnetic

resonance imaging revealed a large lobulated exophytic lesion of size 9.4×8.8×4.4 cm in the subcutaneous plane with involvement of the left trapezius muscle. He underwent wide local excision with a 2 cm margin all around and the resultant defect was closed with pectoralis major myocutaneous (PMMC) flap from the ipsilateral side (Figure 1b).

Histopathology suggested of DFSP and margins were free of tumor (Figure 2). Immunohistochemistry revealed strong and diffuse positivity for vimentin and CD34, SMA was negative (Figure 3). The patient was followed for 1 year with no signs of recurrence and satisfactory cosmesis (Figure 1c).

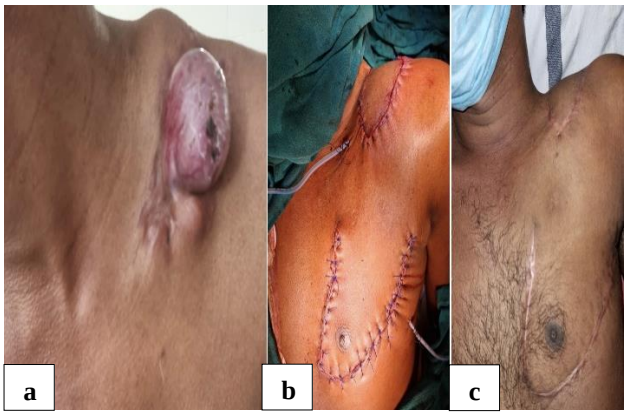

Figure 1: Patient images (a) 5×4×4 cm lump with smooth surface and erythematous skin, (b) defect closure with pectoralis major myocutaneous flap, and (c) post operative image showing complete healing and no recurrence at follow up.

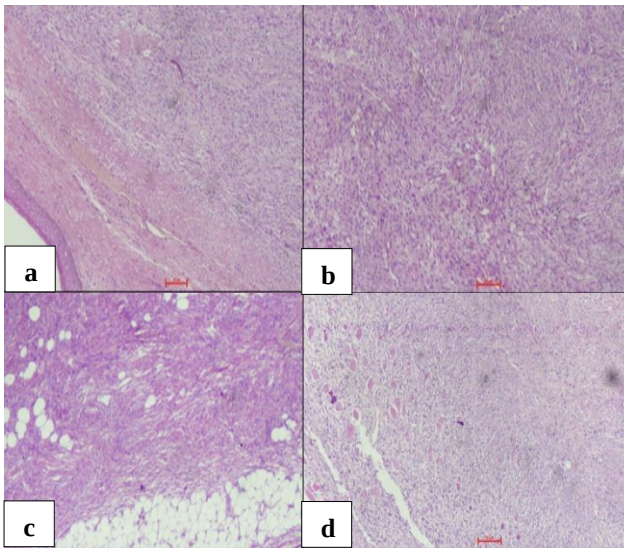

Figure 2: Histopathology slides (a) dermal based circumscribed tumor with atrophic epidermis, (b) spindle shaped cells arranged in storiform pattern, (c) tumor cells infiltrating the underlying fat tissue, and (d) underlying skeletal muscle involvement with lymphoid aggregates.

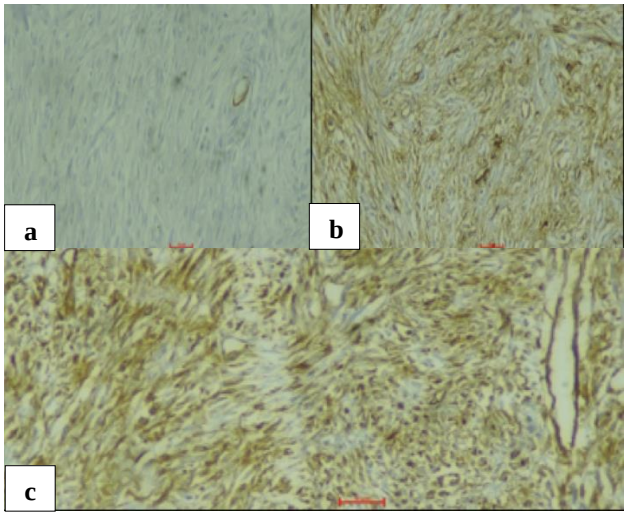

Figure 3: Immunohistochemistry (a) SMA negative, (b) CD 34 positive, and (c) vimentin positive.

DISCUSSION

In 1924 Darier and Ferrand first described DFSP as a rare, cutaneous fibro histiocytic neoplasm. Later the term DFSP was coined in 1925 by Hoffman.[5] Chromosomes 17 and 22 translocations leads to the fusion of gene collagen type 1 alpha 1 (COL1A1) and platelet-derived growth factor subunit β (PDGFB) resulting in the formation of a fusion protein which has autocrine function leading to tumorigenesis.[6,7] Considering this the use of tyrosine kinase inhibitors seems justified as an important component of the multidisciplinary approach in the treatment of malignant, non-resectable, and recurrent lesions.[6,7]

Clinically DSFP generally presents as asymptomatic gradually progressive protuberant lesion with an underlying background of bluish or brownish erythematous skin. Common site of occurrence is trunk and extremities and occurrence above neck is rare. Differential diagnosis for DFSP includes benign lesions like lipoma, keloids, epidermal cyst in the initial stages whereas kaposi and other soft tissue sarcomas in late presentation.[8]

Dermis is the most common location for DSFP but at times it can infiltrate the surrounding fatty tissue as well as the muscle and bone. Histology depicts either a pastry pattern where tumor cells lie parallel to epidermis or a honey comb pattern in which the adipocytes islets are marked between neoplastic cells. DSFP is known for its low metastatic potential with possibility of metastatic spread to regional or distant places <5%.

The principle for treatment for DFSP is complete surgical excision with negative margins. DSFP is notorious due to its pseudopodia-like extensions primarily responsible for a high recurrence rate. Recurrence is most common in the head and neck region attributed to the anatomical

consideration and is reported up to 50%.[9] Surgical options include WLE with at least 2 cm margin all around.[10] Excessive tissue resection can lead to poor cosmetic and functional outcomes which further necessitates flap reconstruction as in index case. Radiation therapy is indicated when negative margins cannot be achieved, or as adjuvant therapy after resection in recurrent DFSP.

CONCLUSION

Succinctly summarising, the two key concerns in a patient of DFSP is recurrence and misdiagnosis. For the patient recurrence is most troublesome both physically and mentally and misdiagnosis adds to his miseries. Diagnosis at first instance is often challenging to every physician and awareness of such cases will be useful in appropriate management.

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