

Case Report

Fibrosarcomatous Variant of Dermatofibrosarcoma Protuberans: An Aggressive Tumor - A Case Report

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Received: 07/10/2015

Revised: 24/10/2015

Accepted: 28/10/2015

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is an unusual, locally aggressive, cutaneous neoplasm of low to intermediate grade malignant potential. Fibrosarcomatous DFSP (FS-DFSP), a rare variant of DFSP, has got a higher tendency for recurrence and metastasis resulting in significant morbidity and reduced survival. We present a case of this rare malignancy in a 23-year-old female who presented with swelling over the dorsum of the right hand. We emphasize that since FS-DFSP is associated with aggressive biologic behavior, hence careful assessment for the presence of a fibrosarcomatous component in DFSP should be done.

Key words: cutaneous, dermatofibrosarcoma, fibrosarcomatous.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an unusual, locally aggressive cutaneous tumor of low to intermediate grade malignant potential. It accounts for about 6% of all soft-tissue sarcomas. ⁽¹⁾ Exact histogenetic origin of DFSP is still debatable. ⁽²⁾ Out of many histologic variants, fibrosarcomatous DFSP (FS-DFSP) is rare and known for higher tendency for recurrence and metastasis thereby resulting in significant morbidity and reduced survival. FS-DFSP constitutes around 10% of all the cases of DFSP. ⁽³⁾ According to literature nearly 225 cases of FS-DFSP have been described. ⁽¹⁾

CASE REPORT

A 23-year-old female presented with swelling over the dorsum of the right hand since 8 months. There was no history of any prior trauma at the site. The mass

was excised twice but recurred. Present swelling appeared 2 months back, progressive in nature, not associated with any pain or discharge. On examination a 6x 4 cm oval swelling over the dorsum of right wrist extending from MCP joints of 2nd, 3rd and 4th to the dorsal wrist crease. Swelling was firm in consistency, lobulated with ill defined margins and was not freely mobile. X-ray done for the hand showed prominent soft-tissue opacity along the second metacarpal without obvious bone irregularity or joint effusion (Fig 1). Wide excision and posterior interosseous artery based flap was performed. Grossly two nodular tumor tissue fragments with grey white cut section were received (Fig 2). Histology showed an unencapsulated tumor composed of interlacing bundles and fascicles of spindle shaped cells with elongated pleomorphic nuclei, coarse

chromatin, moderate cytoplasm arranged in storiform pattern and focally in herringbone pattern. Tumor showed mitosis of $>10/10\text{HPF}$, areas of necrosis and seen infiltrating the underlying fibrofatty stroma (Fig 3,4,5). On immunohistochemistry tumor cells were positive for Vimentin and CD34 (Fig 6,7). Focal areas of CD34 loss were noted which corresponded with the fibrosarcomatous areas seen on hematoxylin and eosin stain (Fig 8). A diagnosis of fibrosarcomatous dermatofibrosarcoma protuberans was given. A repeat surgery for wide margin clearance was done but the tumor tissue extension on the palmar side was found involved with the tumor. Patient was referred to oncology for adjuvant radiotherapy. CT scan study of thorax showed two non enhancing soft tissue nodules along right oblique fissure - likely metastasis, follow up was suggested (Fig 9). She was planned for radiation 60 GY/30# over 6 weeks. She took the therapy and was on regular follow up. Eight months post surgery mobilization of the hand was done. Repeat X-ray hand was within normal limits (Fig 10).



Fig 1- X-ray Rt hand shows prominent soft-tissue opacity along the second metacarpal without obvious bone irregularity or joint effusion



Fig 2- Skin covered lobulated tumor with grey white areas on cut section

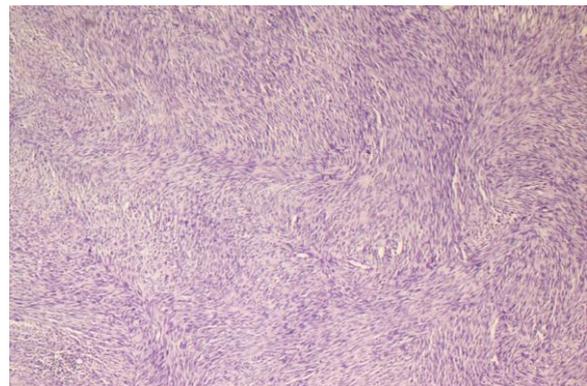


Fig 3- Interlacing bundles and fascicles of spindle shaped cells arranged in storiform pattern H&E X 40

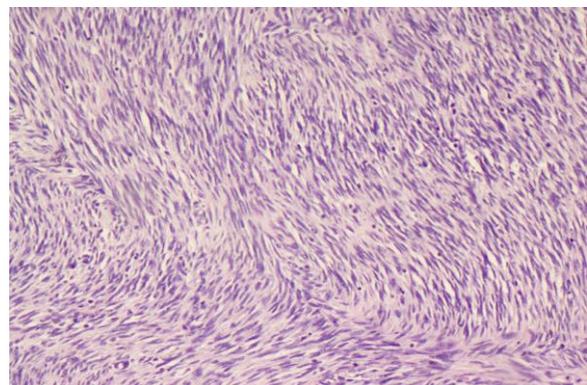


Fig 4- Focal herringbone pattern H&E X 100

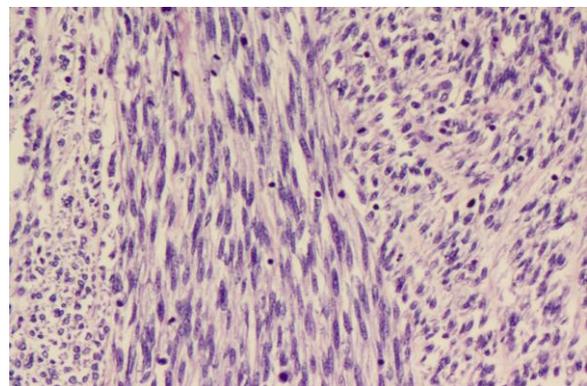


Fig 5- Cells with elongated pleomorphic nuclei, coarse chromatin, and moderate cytoplasm with mitotic figures H&E X 200

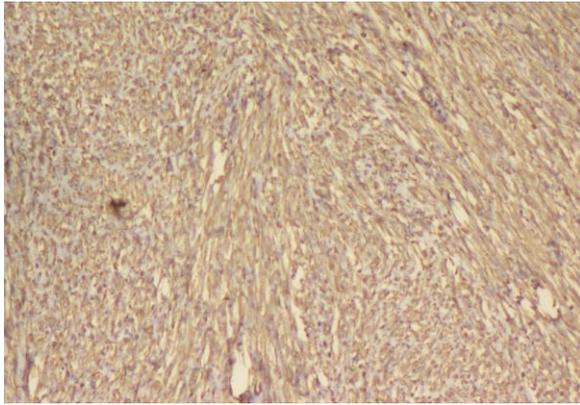


Fig 6- Tumor cell positivity for Vimentin X 100

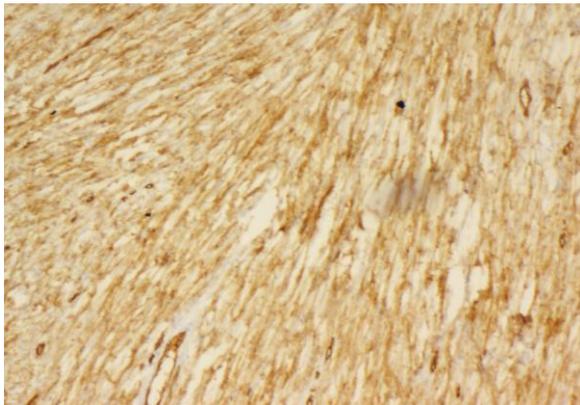


Fig 7- Tumor cell strong positivity for CD34 X100

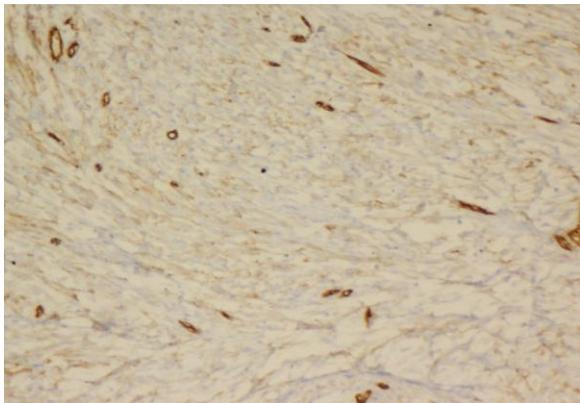


Fig 8- Focal loss of CD34 positivity X100



Fig 9- CT scan thorax showing two non enhancing soft tissue nodules along right oblique fissure



Fig 10- Follow-up X Ray Rt hand showing partial resection of shaft of second metacarpal with mild soft tissue swelling and remodelling of distal metacarpal fragment

DISCUSSION

DFSP was first recognized by Taylor ⁽⁴⁾ in the year 1890, further described by Darrier and Ferrand ⁽⁵⁾ in 1924. Hoffman ⁽⁶⁾ coined the term “dermatofibrosarcoma protuberans” in 1925. Penner ⁽⁷⁾ in 1951 reported a more aggressive subtype indistinguishable from fibrosarcoma representing a fibrosarcomatous progression called “fibrosarcomatous dermatofibrosarcoma” (FS-DFSP).

DFSP has male preponderance and age distribution ranges from 6 to 87 years with a peak incidence during third decade. However FS-DFSP has a female predominance in FS-DFSP. ^(1,2) A transformation of DFSP to FS-DFSP is believed to represent some form of dedifferentiation probably with the acquisition of an aggressive biological behavior. ⁽⁸⁾ However, the precise histogenesis of FS-DFSP remains controversial. Wrotnowski et al. ⁽⁹⁾ believed that fibrosarcomatous areas represent the development of a second neoplasm which was separate and distinct from surrounding DFSP. Grouls and Hienz ⁽¹⁰⁾ considered the neoplasm to represent a transition between DFSP and fibrosarcoma. However, most of the authors believe that FS-DFSP is a peculiar variant of DFSP which has the potential to progress to a higher malignant neoplasm. ⁽²⁾ This was further substantiated by recent

genetic research by Wang et al. ⁽¹¹⁾ showing that DFSP and fibrosarcomatous areas in FS-DFSP have similar gene fusion of the chromosome regions 17q22 and 22q13.

DFSP initially starts as a single, red to bluish, firm nodule ranging from 1 - 25 cm in size, most commonly involving the trunk, extremities followed by head and neck. ⁽¹⁾ This is a slow growing painless tumor leading to delay in diagnosis and clinical misdiagnosis of the initial lesion. However, pain and tenderness may be observed in 10-25% of the patients. ⁽¹²⁾ Histology shows a monotonous storiform growth pattern composed of uniform and cytologically bland tumor cells with hyperchromatic spindle-shaped nuclei. The tumor has a characteristic honeycomb pattern of infiltration into the subcutaneous fat. CD34 which is called as human hematopoietic progenitor cell antigen, is the most specific IHC marker for its diagnosis. ⁽²⁾ Fibrosarcomatous change is characterized as mitotically active cellular areas composed of spindle cells arranged in a clearly fascicular, often herringbone-like growth pattern. The tumor cells in the fibrosarcomatous areas of FS-DFSP show less or no CD34 staining contrary to classical DFSP. Mentzel et al. ⁽¹³⁾ used the term FS-DFSP when fibrosarcomatous areas accounted for at least 5% of the whole lesion. In our case, fibrosarcomatous areas constituted more than 5% of the entire lesion.

The treatment for DFSP is wide local excision with histologically negative margins. DFSP has an infiltrative growth pattern with finger-like projections, thus spreading beyond the visible margins. This stresses the importance of a wide excision, defined as more than or at least equal to 3 cms of clinically uninvolved skin margin. ⁽¹⁴⁾ Distant metastasis is rare (<5%), however local recurrence rates can be as high as 33-60% if surgical resections are performed with inadequate margins. Most

local recurrences of DFSP are noted within 3 years of excision. ⁽²⁾ According to Bowne et al. ⁽¹⁵⁾ advanced age, increased mitotic activity (>10 mitotic figures/HPF), increased cellularity, positive or close microscopic margins of resection, and histologic subtype (DFSP-FS) were associated with a poor clinical outcome. In FS-DFSP, local recurrence rates and distant metastasis rates were found to be 73-89% and 14-63% respectively. ⁽²⁾ According to Mentzel et al. ⁽¹³⁾ the additional finding of necrosis and pleomorphic tumor areas in FS-DFSP favored a poorer prognosis. Distant metastases are rare, and typically preceded by multiple local recurrences. However a case of a FS-DFSP metastasizing to the head of the pancreas without any preceding local recurrence has been reported. ⁽¹⁴⁾ Tumor-related mortality was observed in 5.8% cases of FS-DFSP. ⁽³⁾ Adjuvant radio therapy has been used to achieve local control when resection margins are microscopically positive. But whether this can provide long-term local control remains unknown and further studies are needed for elaboration. Also knowledge of specific chromosomal translocations has resulted in the implementation of targeted therapy with imatinib mesylate, a tyrosine kinase inhibitor, in the management of inoperable, recurrent and metastatic cases of DFSP. ⁽⁸⁾

CONCLUSION

FS-DFSP is a rare form of DFSP, which can lead to significant morbidity and a reduced survival rate. This distinct subtype has more aggressive biologic behavior; hence careful assessment for the presence of a fibrosarcomatous component in DFSP should be done.

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How to cite this article: Sharma S, Kudva R, Valiathan M et al. Fibrosarcomatous variant of dermatofibrosarcoma protuberans: An aggressive tumor - a case report. Int J Health Sci Res. 2015; 5(11):434-438.
