

Dermatofibrosarcoma protuberans in the anterior abdominal wall: A case report

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Abstract

Dermatofibrosarcoma protuberans is a rare intermediate grade sarcoma related to the group of fibrohistiocytic tumors that occur predominantly in the dermis. It can occur at any site and characterized by its latency in its early detection, result in large size at time of diagnosis in some cases, as in this case that present in the anterior abdominal wall of a 65 years female which is a rare site of this lesion.

Key Word: dermatofibrosarcoma protuberans, abdominal wall.

Introduction

Historically Darrier and Fernand first recognized DFSP as a clinicopathological entity and Hoffman introduced the term "DFSP".^(1,2) It is a slowly growing, nodular, polypoid neoplasms that are occur almost exclusively in the dermis, but it can occur also in the deeper soft tissues, most commonly found in the trunk and proximal extremities⁽³⁾. It may occur at sites of previous trauma, affecting more commonly men than women and has a peak incidence during the third decade of life, and most of the cases occur in the adults, but they have also been seen in infancy and childhood⁽⁴⁾.

It is locally aggressive tumor with a high rate of recurrence that can occur in 20-55% of cases and with an extremely low rate of distant metastasis⁽⁵⁾. The tumor first appears as a single, red to bluish, bleachable, firm, cutaneous nodule. During the late stage, rate of growth accelerates, producing the characteristic protrusion from the skin⁽⁶⁾. The growth rate is variable and lesions may remain stable for many years or they may grow slowly with periods of accelerated growth⁽⁷⁾. Clinically, it presents as an asymptomatic raised, firm nodular lesion fixed to the skin but mobile over the deep fascia, with a pattern of slow, progressive growth^(8,9). Diagnosis is established after excisional

biopsy or punch biopsy⁽¹⁰⁾. CT-scan is useful to determine the tumor extent of penetration⁽¹¹⁾. The CT appearance of DFSP is well defined, as unmineralized, nodular soft tissue mass involving the skin and subcutaneous adipose tissue. CT scans or MR images are well suited to show this location, the relation of lesion to underlying structures and the distinct lobular or nodular architecture⁽⁶⁾. CT scanning DFSP may be confused with a dermatofibroma, a neurogenic tumor, a fibrosarcoma or a malignant fibrous histiocytoma as the CT findings of these tumors resemble those of DFSP; DFSP displays an almost pathognomonic protrusion from the skin, a feature which distinguishes DFSP from other tumors⁽⁶⁾.

The disease is thought to be of histiocytic origin and characterized by uniform, slender, spindle shaped, fibroblast like cells, arranged in a typical storiform or cartwheel pattern. Other features of diagnostic importance are the high cellularity, monomorphic appearance, moderate to high mitotic activity, lack or inconspicuousness of foamy or hemosiderin laden macrophages and or multinucleated giant cells and entrapment of fat cells when invasion of subcutaneous tissue.⁽¹²⁾ The diagnosis of DFSP can be made or at least suspected on the basis of fine needle aspiration cytology.⁽¹⁶⁾

Immunohistochemical staining was strongly positive for vimentin and CD34, and cytogenetically DFSP commonly has translocation involving PDGF-beta & COL1A1⁽¹⁵⁾. DFSP has two histologic variants: the more typical low-grade tumor, and a high-grade rare fibrosarcomatous variant demonstrating necrosis, high mitotic rate (> 10 mitoses/high-power fields) and presence of pleomorphic areas¹². This last variant is associated with a poor clinical outcome.

Metastasis is rare. Lung metastasis is most common, while lymph node metastasis is exceedingly rare¹². Wide surgical excision using a margin of three centimeters with inclusion of superficial fascia is currently the standard therapy in children¹³. Mohs micrographic surgical excision has wide acceptance among adults cases¹⁴. DFSP is a radioresistant tumor¹⁵.

Case Report

A 65 year woman presented with a supraumbilical abdominal swelling which was slowly progressively increasing in size for 5 years, with a history of bleeding and ulceration. Clinical examination revealed a firm, 12 x 7cm fungating lump in the supraumbilical area shown in figure1. Excision had been done and histopathological result was reported as Dermatofibrosarcoma Protuberance (DFSP). CT examination revealed a large, nodular, well circumscribed, soft tissue mass, centered in the subcutaneous fat of anterior abdominal wall in the midline with cystic component. The overlying skin was stretched over the mass and was not separately identifiable from the mass. The lesion caused a prominent bulge in the abdominal contour. Posteriorly the lesion was invading the rectus muscles and the subcutaneous fat can not be spared if the tumor invades the rectus muscle. The CT findings were suspecting the diagnosis of DFSP.

It was of five years duration slowly growing, and the patient was informed that this mass is recurrent Para umbilical hernia with omentocele, so ignored by the patient until the mass surface ulcerate and bleed, the patient admitted to the hospital, ultrasound was revealed a solid mass in the abdominal wall, invading muscular layers, and the patient prepared for the operation, wide local excision with 3 cm margin was done, the defect closed by Mayo's repair.

In the pathology department the mass was examined and revealed a big mass 13x8 cm with surrounding fibromuscular tissue around, infiltrative margins and solid cut section. Areas of hemorrhage and necrosis are seen focally. Many samples are taken from different areas for histopathological diagnosis, show classical type of DFSP. Quite lack of circumscription is demonstrated, high cellularity, storiform arrangement of cells, with high mitotic activity. (fig 2).

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare, low grade malignant soft tissue tumor of the skin and subcutaneous tissue with a high propensity for local invasion and recurrence. (17) Though mostly seen in middle age, a few cases have been described in infants and children between the ages of 14 months and 12 years (8). The case is middle age, which is the common age, but it is a female that were less commonly affected, in the abdominal wall, a rare site, unlike the desmoid tumor which is most commonly seen in the anterior abdominal wall, while the least frequent is dermatofirbosarcoma Protuberans(2).

The tumor must be excised with a 3cm or greater margin of uninvolved skin, as in this case, because there is a preoperative suspicion of malignancy especially that it is preceded by helpful CT scan for proper decision of the line

of incision and avoiding inadequate excision which leads to local recurrence or metastasis (6). In addition histopathological full assessment was greatly helpful in full diagnosis, assessment of excision margins, and adequacy of excision of safe tissues around. Lack of immunohistochemical markers does not affect the diagnosis and it is needed in differentiating it, in some diagnostic problems.

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Figure (1):Supraumbilical fungating lump

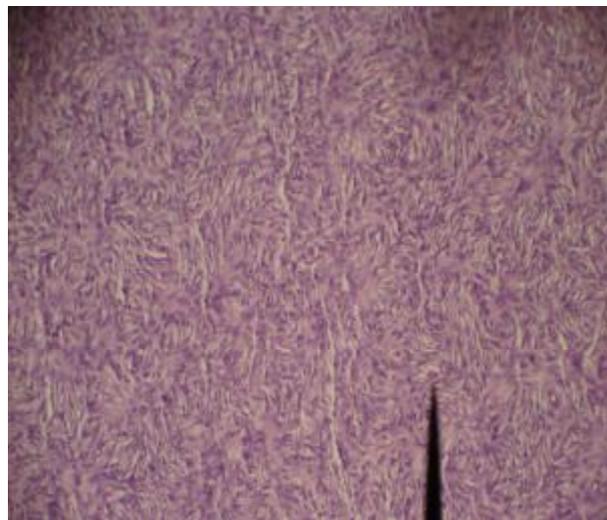


Figure (2) Moderately cellular tumour composed of bland, elongated cells with characteristic storiform arrangement (haematoxylin & eosin, original magnification x 100).

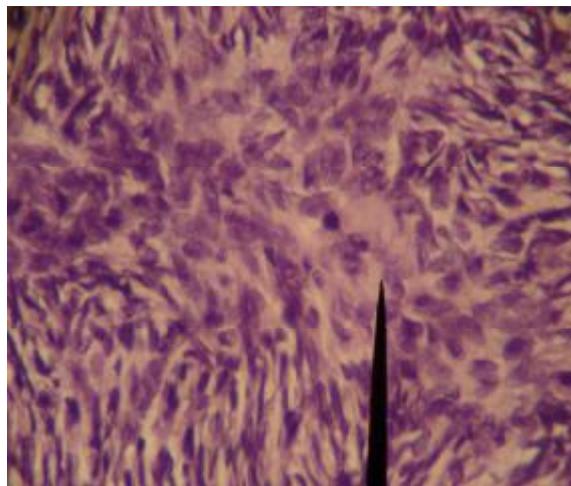


Figure (3) high power view show mitotic figures and large Pleomorphic cells. (haematoxylin & eosin, original magnification x 100).