Volume : 3 | Issue : 5 | May 2014 • ISSN No 2277 - 8179

Recurrent and Metastatic Dermatofibrosarcomaprotuberans (DFSP): A Relatively Rare Case Report

Medical Science

KEYWORDS :

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a very rare slow-growing, malignant mesenchymal tumor of the dermis accounting for less than 0.01% of all malignancies(1).. Clinically, the malignancy most commonly presents as a painless and a slow-growing subcutaneous polypoidal mass, usually less than 5 cm in size(2) .In many respects, the disease behaves as a benign tumor, but in 2-5% of cases it can metastasize so it should be considered to have malignant potential. Remote metastases occur at the common sites lungs and the regional lymph nodes.[3,4]. According to the NCCN Clinical Practice Guidelines in Oncology, The gold standards treatment is complete surgical excision with appropriate reconstruction.(5) In the present case, the bulky (7× 9 cm) tumor was located in the pelvis. CT Pelvis of the lesion revealed Hetergenously enhancing soft tissue mass lesion of size 73x93x110,causing erosion of right iliac wing and infiltrationof right illiacus muscle .The Mass lesion is causing mass effect in right iliac fossa with medial displacement of adjacent bowel loops.

We report a case of this rare tumor which showed relapse at different location and multiple metastatic lesions in the lung.

Case report

The patient (35-year-old male) Presented in the department of Radiotherapy & Clinical oncology SMS Hospital with recurrent left thigh mass in 2011.The case was discussed in the tumor board and advised wide excision surgery .Wide Excision surgery was done and the histopathology showed Dermatofibrosarcoma protuberance. Radiotherapy and chemotherapy were not used because the incision borders were negative on histopathology. As per decision of Tumor board he was kept on follow up.He lost follow up and then he presented in the department in march 2014 with right sided pelvic mass and lung secondaries.CT Scan of pelvis shows Heterogenously enhancing soft tissue mass lesion of size 73x93x110,causing erosion of right iliac wing and infiltrationof right illiacus muscle The Mass lesion is causing mass effect in right iliac fossa with medial displacement of adjacent bowel loops.

The physical examination revealed surgical scars of recurrent tumors on left side of the thigh[Figure 1] and right sided pelvic mass[Figure 2]. CT pelvis showed enhancing soft tissue mass lesion of size 73x93x110,causing erosion of right iliac wing and infiltrationof right illiacus muscle. [Figure 3].Figure 4].[Figure 5]

Figure 1 Scars of previous surgeries

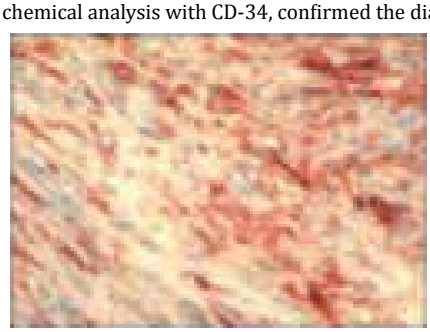
Figure 2 Right sided pelvic mass

Figure 3

CT showing Right sided pelvic mass lesion of size 73x 93 x 110,causing erosion of right iliac wing and infiltrationof right illiacus muscle Rt sided pelvic mass lesion causing erosion of right iliac wing and infiltration of right illiacus muscle

Figure 4

Figure 5
The lung fields showed multiple metastatic lesions on chest x ray.Tru cut biopsy of the pelvis mass was done . Microscopic examination, which included histopathologic and immunohistochemical analysis with CD-34, confirmed the diagnosis of DFSP.



Immunohistochemical appearance (×40 100 HE)

Discussion
It was first described in 1924 as a progressive and recurrent dermatofibroma(6). The trunk and proximal extremities are the most frequent locations of the disease, but it can occur at any other site.

DFSPs rarely progress to a high-grade fibrosarcomatous component(7) Histologically, DFSP is identified by a pattern of monomorphous proliferation of cytological bland spindle cells with a visible storiform or whorled (rushmat-like) architecture (8). Other characteristic features are low mitotic activity and deep, honeycomb infiltration into subcutaneous adipose tissue [9].

The expression of CD34 is almost a consistent finding and it is extremely useful in differentiation of DFSP from benign fibrous histiocytoma , dermatofibroma and other soft tissue tumors . The sensitivity of CD34 staining in DFSP ranges from 84 to 100% [9].

The Metastases occurs mostly to lungs, followed by the regional lymph nodes. Generally, distant metastases is seen after recurrent local relapses.(9,10)

MRI is useful to demonstrate deep tissue invasion, especially in the advanced stages. Nuclear study is useful for ruling out bone invasion.

The preferred therapy of DFSP is radical (wide) surgical excision. The most significant prognostic factor in patients with DFSP has proved to be the extent of surgical resection. The success of the initial surgical excision has a major effect on the outcome as well. In fact, if this procedure fails and hence, the tumor recurs, it could lead to an uncontrollable local growth, as seen in our case.

Due to its infiltrative nature, DFSP is characterized by a high recurrence rate varying in the literature from 10-80% (11)

Radiotherapy is of limited value in the treatment of DFSP. However, it might have some role to play when the resection border is positive or when extensive excision is not possible due to cosmetic or functional difficulties.Imatinib mesylate, a drug used in chronic myelogenous leukemia, has been successful in metastatic disease and/or relapses of the entity.[12]

We discussed this case in our department and patient was kept on Tab imatinib.800mg/Day.

Conclusion
DSFP could be uncontrollable and challenging to cure.

So Aggressive management of the dermatofibro sarcoma protuberans should be done .

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