Case Report

Dermatofibrosarcoma protuberans of penis: Case report and literature review

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Abstract Dermatofibrosarcoma protuberans (DFSP) is a rare mesenchymal skin tumor with intermediate to low grade malignancy and occasional distant metastasis and high rate of recurrence locally. It mostly involves trunk, extremities, scalp, and neck. This article describes a middle-aged married male with a DFSP lesion involving ventral aspect of glans and distal shaft of penis who underwent local excision and primary closure. The patient was tumor-free at three years of follow-up.

Key Words: Dermatofibrosarcoma protuberans, local excision, penis

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INTRODUCTION

It is a rare cutaneous tumor constituting <0.1% of all malignancies.[1] Earliest description of Dermatofibrosarcoma Protuberans (DFSP) was given by Taylor in 1890. Hoffman, in 1925, first employed the term DFSP.[2] Despite their locally aggressive behavior, they rarely metastasize to regional lymph nodes and viscera. Probability of distant metastasis is <5%.[3] DFSP mostly involves trunk, extremities, and scalp. This article describes a case of penile DFSP treated successfully. A brief review of literature including clinical features, histopathological characteristics, treatment modalities is reported.

CASE REPORT

A 45-year-old circumcised, married, fertile male reported to outpatient department with chief complaints of penile swelling since one year. On examination, patient was having an irregular swelling, roughly size of 4 cm × 3.5 cm, with a base arising from the ventral aspect of the glans penis and distal shaft of penis. Overlying skin was adherent, and underlying tissues were free. he swelling had started one year back and progressed to the present size. There was no history of pain, difficulty in urination or any urethral discharge or itching [Figure 1].

Baseline investigations showed hemoglobin - 14 mg/dl. TLC - 9000/ml (Neutrophils 60%), (Lymphocytes 17%), (Eosinophils 6%), ESR- 25, Urea/Creatinine/LFT/coagulograme/Routine urine was normal. Patient was screened for any sexually transmitted disease, which was negative. IgE levels were normal. Fine Needle Aspiration (FNA) of the swelling revealed features of dermatofibrosarcoma protuberans (DFSP). Patient was admitted, and excision of the tumor was done. [Figure 2] The urethra was free of the tumor. The defect could be closed primarily over a Foleys catheter. Margin of the tumor was negative. Histopathological examination of resected specimen revealed characteristic cartwheel appearance confirming the diagnosis of the dermatofibrosarcoma protuberans. [Figure 3] Urinary catheter which was placed preoperatively was removed on 7th postoperative day and patient discharged. After three years of follow-up, patient is doing well with normal sexual activity, voiding, and without any local or distant recurrence.

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Figure 1: Dermatofibrosarcoma protuberans lesion on glans penis

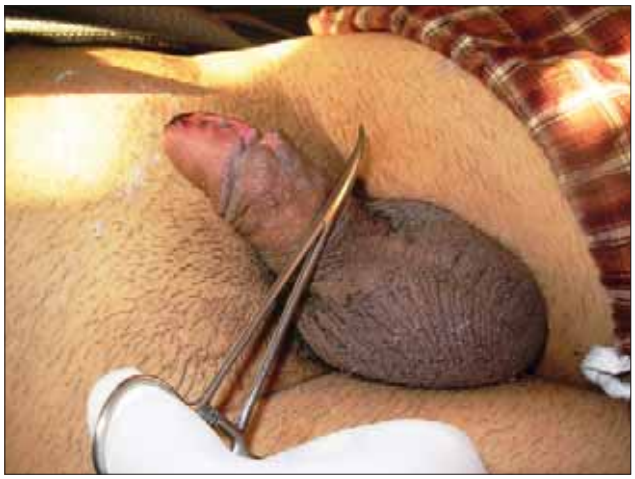

Figure 2: Post-operative picture after excision of lesion

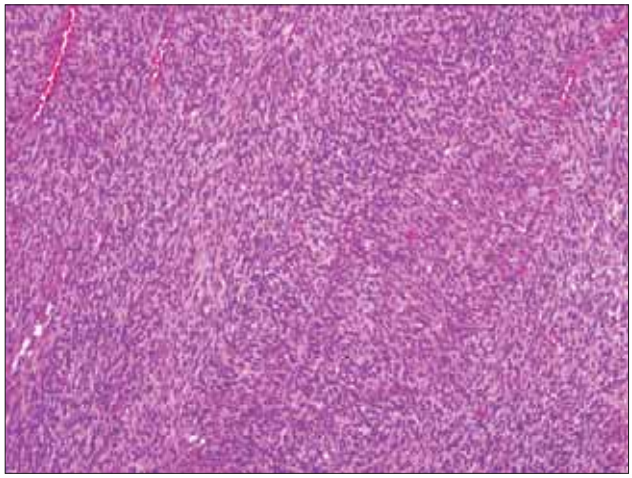

Figure 3: Histopathology of resected specimen; Hematoxylin and eosin staining showing characteristic features of storiform pattern (cartwheel appearance)

DISCUSSION

DFSP is a rare, low to intermediate grade malignancy of subcutaneous origin, with high rate of recurrence locally, but distant metastasis is a rare phenomenon. Mostly occurs between 20-40 years of age. It mainly involves trunk followed by extremities. The site of the lesion described in our case is extremely uncommon. Taib F *et al.* have described a case of a large fungating infantile fibrosarcoma of the penis in a two-year-old Malay boy.[4]

Grossly, tumor is multinodular and with a bluish hue. Overlying skin is fixed to tumor, but underlying muscles are free of tumor as described in our case. CT and MRI may be sometimes needed to look for the deeper extent of the tumor, but in our case, the diagnosis was revealed by fine needle aspiration (FNA) and confirmed after histopathological examination of resected specimen.

Cartwheel appearance is characteristic of DFSP on microscopy as was found in our case also.[5] CD_{34} is a highly specific tumor marker for diagnosis of DFSP.[6] The recommended form of treatment for DFSP is wide local excision of tumor after including subjacent facia and margin of apparently normal tissue in all planes.[7-9] In the case described, the lesion had a base involving corona and part of the ventral glans. Excision was done along with some normal-looking tissue. The defect of glans and corona was re-approximated over the catheter taking care of the urethra. Wacker J recommended the use of paraffin sectioning and three-dimensional histological evaluation as an accurate additional tool for treatment optimization, particularly in recurrent lesions.[10] In addition to surgical methods (recurrent and metastatic lesions), molecular targeted therapy with imatinib mesylate may be considered as a suitable alternative or additional treatment option for DFSP.[5]

Local recurrence rate of 20-50% is reported in cases with incomplete resection. Patients with unresectable or positive margins should be treated with adjuvant radiotherapy to decrease the recurrence rate. It reduces the local recurrence rate.[11] Recurrent lesion can be treated either with re-excision, radiotherapy, or both. Imatinib is currently considered the gold standard in the treatment of inoperable and/or metastatic and/or recurrent cases of DFSP. Therapy with imatinib may potentially facilitate resection or decrease possible disfigurement related to radical surgical procedure.[12] But, in our case, histopathology of resected specimen had negative margins, so our case did not receive any adjuvant therapy.

McGregor, in 1961, recommended wide local excision with planned primary skin grafts as the primary treatment of DFSP.[9] Cai *et al.* analyzed 260 patients of DFSP and concluded that adequate initial resection is important for patients with DFSP, a similar observation was made by Mcpeak .[7,13] In a study by Jambhekar, treatment for DFSP was mainly surgical, and only few patients received radiotherapy.[14]

Suit *et al*. analyzed the success of radiation therapy used postoperatively for margin positive disease due to technical or medical reasons.[15] Using a standardized surgical approach, including meticulous pathologic evaluation of margins, a very low recurrence rate (1%) was achieved with relatively narrow margins (median 2 cm), allowing primary closure in 69% of patients.[16] Paradici *et al*. reported a significantly lower recurrence rate in patients subjected to Modified Moh's Micrographic surgery (MMS) compared with those treated with wide local excision. There is inconclusive evidence for any advantage of MMS in non-primary cases, while MMS was most effective in treating head and neck tumors.[17] Different authors mention different resection margin of normal skin around the tumor ranging from 1 cm to 3 cm, but the goal of the excision should be to achieve negative margin whenever possible.

The trunk, extremities, and head and neck being the usual site of this tumor, the location of the tumor on glans penis represents a very unusual and interesting case. The case is second of its instance, first being reported in 1980.[18] So, DFSP should be kept a possibility when a patient presents with a slow-growing multinodular swelling penis.

Primary sarcoma of the penis is a very rare condition, and to date, there are only around 44 cases described in the medical literature, of which 30 are leiomyosarcomas and 14 are epithelial sarcomas.[19] Penile sarcoma is usually treated by total penectomy with or without associated radiation and chemotherapy.[20] Regardless of the therapeutic modality used, the majority of patients die in less than two years due to systemic dissemination.[20] While only one case of DFSP has been reported so far, DFSP has got less chances of local and distant metastasis than sarcoma, and treatment is local resection as contrary to sarcoma, which may need total penectomy.

From review of literature and experience of our own case, it is clear that the recommended form of treatment of DFSP is wide local excision. Imanitib is an option for recurrent and large lesions. Moh's micrographic surgery is a good option if the facilities are available and useful in areas where extensive resection in not feasible like head and neck. The main objective of local resection should aim at achieving negative margin and simultaneously preserving the functional outcome of the affected organ as described in our case. Patients should be followed regularly for any local recurrence.

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