

Dermatofibrosarcoma Protuberans (DFSP): An Uncommon Skin Cancer

Saumya Prakash Srivastava

Vivekanand Polyclinic College of Nursing, Lucknow, India

Email address:

Saumya12.aiims@gmail.com

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Abstract: Dermatofibrosarcoma Protuberans (DFSP) is a very uncommon type of skin cancer that begins in the connective tissues of skin and extend to nearby structures in the form of lump. The cause of cutaneous fibroids is unknown, although a suggested risk factor includes skin lesions at the affected site. DFSP tends to affect people between the age of 20 and 50, but it has been diagnosed in people of all ages. This case report describes a highly recurrent DFSP disease condition of 25 years old female, who got admitted in a tertiary care cancer centre with complaints of painless swelling in the back, fever, and loss of weight since last 1 month. Surgical history revealed three past surgical excisions of the mass over the same location. CT scan and tissue biopsy revealed recurrent Dermatofibrosarcoma protuberans. Initially, the patient was on imatinib 600 mg/PO/OD, after which there was a subjective decrease in size of mass. But as the patient was unable to tolerate high doses of imatinib, she then underwent wide local excision of the mass with split skin graft taken from right thigh under general anesthesia. Surgery was uneventful and she was discharged thereafter. As local recurrence is common, patients required close clinical follow-up after completing treatment.

Keywords: Dermatofibrosarcoma Protuberans, Surgical Excisions, Soft Tissue Sarcoma, Rare Cancers

1. Key Messages

Dermatofibrosarcoma protuberans (DFSP) is a rare type of soft tissue sarcoma that begins as a small lesion or hypertrophic scar. Benign DFSP can be treated well with wide local excisions and taking adequate margins. Newer surgical excisional procedures such as Mohs micrographic surgery is still into investigation, literature suggests its use in critical locations. Recurrent and metastatic DFSP is usually managed with imatinib along with excisional procedures. Role of radiation therapy is more in post-operative phase for achieving clear margins.

2. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow growing, soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia. The tumor typically presents as a slowly growing, firm plaque on the torso and can also be found on the arms, legs, head, or neck. of young adults. It accounts for between 1% and 6% of all soft tissue sarcomas and 18% of all cutaneous soft tissue

sarcomas. [1] DFSP is extremely rare soft tissue sarcoma and is estimated to occur in 1 in 100,000 to 1 in 1 million people per year. [2]

Appearance of a purplish, reddish or flesh- coloured thin, firm patch of skin is typically the first sign of a tumor. The tumor develops slowly in most cases, but it has a proclivity for recurrence after removal. It is very rare for it to spread to other areas of the body. Although the cause of DFSP is unclear, damage to the affected skin may be a risk factor. [3]

Tumour formation can occur due to excessive proliferation of cells secondary to stimulation by an abnormal fusion protein like the PDGFB. The abnormally fused COL1A1-PDGFB gene provides instructions for making an abnormal fusion protein. [3]

According to the NCCN Clinical Practice Guidelines in Oncology, full surgical excision with adequate reconstruction is the gold standard treatment, and Imatinib, a tyrosine kinase inhibitor is a drug that has been approved for the treatment of advanced cancer. Adjuvant radiation therapy has a role in case of positive surgical margins. [4] According to many experts, Mohs micrographic surgery can be one of the ideal treatment option for DFSP as it involves microscopical

examination of each removed tissue for cancer cells. Therefore, it will be helpful for obtaining clear margins with greater sparing of healthy tissues; still its role is inconsistent in literature. [5]

The overall prognosis of DFSP is good, with a 10-year survival rate of 99.1%. [6] Regional and distant recurrences are infrequent in DFSP. The overall recurrence rate of DFSP is about 50%. Wide re-excision is a promising treatment option for recurrent DFSP with a lesser recurrence rate of 12% after re-excision. [7] Metastasis is linked to a poor prognosis. However, recent findings with Imatinib in patients with metastasis are promising. [8]

3. Case Description

A 25 years old female presented with a painless swelling on the right lower back. Physical examination revealed a multilobulated, soft, non-tender and mobile mass. CECT chest and abdomen reported a multinodular soft tissue mass lesion epicentered in subcutaneous plane in right lower back abutting the posterior chest wall muscles without intraperitoneal extension. Tissue biopsy showed monomorphic, benign-appearing spindle cells arranged in an irregularly storiform, cart-wheel like pattern. It was later confirmed as DFSP on histopathological examination and immunohistochemistry findings which reported the presence of CD34+. Patient gave the history of presence of mass since birth for which she underwent multiple surgical resections. First excision was done at an age of 8 years, the lump then recurred after 2 years, which was gradually progressive in nature and was painless. She underwent a second surgery followed by several other relapses and re-resections. After a series of painless recurrences, fourth recurrence was painful, itchy and has blood and pus discharge, following which she was again resected and received 2 courses of radiation therapy and was started on imatinib 600 mg daily. The response was positive with a subjective decrease in the size of the lump. But, it lasted for few months, after which it again progressed. Upon progression, imatinib dose was escalated to 800 mg daily. The increase in the dose didn't resulted in any positive response and has caused side effects such as nausea and vomiting. Following which, the patient was again maintained on 600 mg daily dosage of imatinib. It did not resulted in much benefit this time and the disease continued to progress. As a final treatment, patient underwent wide local excision of the mass with split skin graft taken from right thigh under general anaesthesia.

There is no visible progression of the disease, CECT chest and abdomen also revealed a dramatic reduction in the size of tumor. And, the patient is continued to be monitored for any recurrences.

4. Discussion

DFSP is a rare soft tissue sarcoma, which is rubbery soft and non-tender in consistency. It is more commonly seen in males as compared to females. Highest incidence is seen in

males of around 30 years. Lesions can vary in size and can have variable presentation. DFSP is a very slow growing tumor and has an indolent course; initially it appears as hypertrophic scars or soft tissue tumors without any alarming symptoms. [9] Risk factors predisposing DFSP is still not clear, but literature suggests a strong association between previous injury or trauma and development of DFSP scar. [10, 11] Wiesmueller, et al [9] in their study found that 6 out of 19 patients developed tumor growth over the skin that previously had been exposed to trauma, such as chronic irritation, laceration, or burns.

In majority of cases, the tumor is limited to the dermis. But it can also present as an infiltrative growth to the subcutaneous fatty tissue and underlying muscles. It rarely metastasizes to lungs and lymph nodes and has less than 5% probability for regional or distant metastases. [12] DFSP is diagnosed on the basis of histopathology findings which are typical of elongated nuclei arranged in storiform pattern. The differential diagnoses includes hypertrophic scars, keloid, metaplastic carcinoma, fibromatosis or other underlying breast lesion. [13]

Gold standard Treatment for DFSP consists of surgical excision, mainly wide excision or Mohs micrographic surgery with 2- to 4-cm margins if wide local excision is applied. (as recommended by National Comprehensive Cancer Network). Wiesmueller et al [9] suggested using a mean margin width of 1.67 cm, as it resulted in negative margins in all patients with a median recurrence-free survival of 84 months. Mohs micrographic surgery is also recommended by some authors as it provides sufficient histopathological control during the excision and has lower chance of recurrences when compared to wide excisions. [14, 15] However, many experts deny its use as it involves utilizing well-trained staff and sophisticated equipments, which might not be available in every institution. Therefore, its use as a treatment for DFSP can be considered for the tumors located at critical locations such as head and neck, to minimize the risk of complications. [16]

Imatinib, a tyrosine kinase inhibitor was approved by the Food and Drug Administration (FDA) in year 2006 for the treatment of unresectable, metastatic, or recurrent DFSP. Use of radiation therapy is more promising after the surgery, in cases of positive surgical margins and unresectable disease [17].

Follow-up should be done at an interval of every 6 to 12 months for the examination of suspected site followed by histopathological examination in cases of suspected recurrences. [17]

The prognosis of DFSP is excellent for low grade lesions. Overall mortality is less than 3% at 10 years. [18] Criscito et al. [19] In their study found that, age at diagnosis, male sex, and DFSP tumor size and tumor characteristics are important prognostic factors.

5. Conclusion

Dermatofibrosarcoma Protuberans is a type of rare skin

cancer which grows out like a root of a tree. This extensive growth makes it difficult for complete excision. Excisional and Mohs micrographic surgery is considered a gold standard treatment for DFSP. Imatinib, a tyrosine kinase inhibitor is recommended for unresectable, recurrent and metastatic DFSP. Local recurrence is relatively common with extensive local resection even with clear surgical boundaries.

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