

From Diagnosis to Treatment: Dermatofibrosarcoma Protuberance – A Case Report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP), also known as Darier–Ferrand dermatofibrosarcoma, is a rare soft tissue neoplasm and the most common type of cutaneous sarcoma. It is characterized by slow growth and a low metastatic potential, although it presents a high risk of local recurrence, mainly related to the quality of surgical excision. The standard treatment is wide local excision performed by an experienced multidisciplinary team. In locally advanced, unresectable, or metastatic forms, radiotherapy and targeted therapy with imatinib may be considered. We report the case of a 27-year-old man who underwent surgical treatment for dermatofibrosarcoma protuberans located in the left hypochondriac region

Keywords: Dermatofibrosarcoma protuberans, cutaneous sarcoma, wide local excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), also known as Darier-Ferrand tumour, is a rare, locally aggressive soft tissue neoplasm of dermal origin, representing the most common subtype of cutaneous sarcoma ^[1]. It has an infiltrative growth pattern with slow growth and low metastatic potential, but high incidence of local recurrence, when incompletely excised ^[2]. DFSP is most common in young and middle-aged adults, usually involving the trunk and proximal extremities ^[3]. Clinically it manifests as an indurated plaque or multinodular mass that slowly enlarges and may ultimately become protuberant ^[4]. Histologically DFSP is characterised by a proliferation of monomorphic spindle-shaped cells infiltrating the dermis and subcutaneous tissue in a storiform or cartwheel pattern ^[5]. Immunohistochemical expression of CD34 is usually present in tumour cells and this aids in the differentiation of DFSP from other fibrohistiocytic tumours ^[6]. Cytogenetically, most of them are associated with the t(17;22)(q22;q13) translocation resulting in the COL1A1–PDGFB fusion gene that accounts for the sensitivity of advanced forms to the targeted therapy with imatinib ^[7].

Wide local excision or Mohs micrographic surgery with negative margins is the standard treatment for localised disease ^[8]. In cases of locally advanced unresectable recurrent or metastatic disease, radiotherapy and targeted therapy may be considered ^[9]. The delayed diagnosis and poor surgical management of DFSP, a low-grade sarcoma, can lead to multiple recurrences and significant morbidity, emphasising the importance of early recognition and appropriate treatment ^[10].

Case Report

We report a 27 year old man, with no significant medical history, who has 04 nodules in the left hypochondrium, over a period of 03 months, followed by a dermatologist. The physical examination revealed a patient is in good health, present in left hypochondriac region, 04 purplish pink, indurate, fixed nodules, two measured 02cm and two measured 01cm, painless. Patient was followed by a dermatologist who has prescribed a medical treatment; as result of no improvement and also nodules' size increase, a biopsy was performed, which revealed: a cutaneous fibro-histiocytic mesenchymal proliferation, and on immune histochemical stains revealed CD34 antigen positivity, the DFSP was confirmed. The patient was referred to our surgical department, performed a wide resection with safety margins of 03 cm and wound closure without tension (figure 1). On the immediate postoperative, there were no side effects

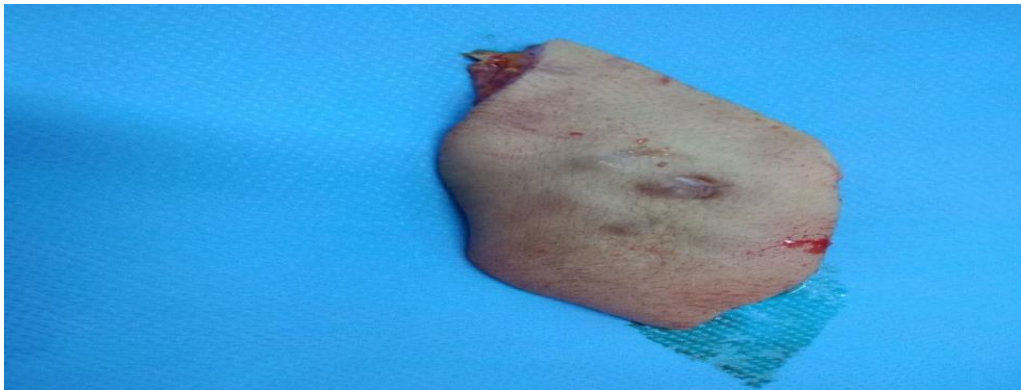


Figure 1. Surgical specimen of tumour with resection margins of 03 cm

On day 05 post-operative; the patient presented with an 8 cm subcutaneous haematoma; treated by its puncture and medical treatment with antibiotic and NSAIDs (figure 2).



Figure 2. The wound condition on the twentieth day after surgery

The histopathology report; confirmed the DFSP diagnostic; the dermis and the hypodermis were infiltrated, also revealed that all the surgical margins are free. Every six months for at least five years; it is planned that the patient is being clinically evaluated.

DISCUSSION

Le Dermatofibrosarcome is a tumor of soft tissues of the dermis, fibroblastic origin, rare, 0,8 to 5 cases/millions inhabitants per year^[11]. DFSP is the most common type of cutaneous sarcoma, about 0,1% of malignant cutaneous tumor. Characterized by slow evolution, and it is a locally aggressive and rarely metastatic occurs around 3 %, it is an intermediate malignancy tumor^[12, 13]; recurrence rate is high about 20 à 40 %. DFSP is diagnosed between 30 and 50 years old, a slight male predominance is reported. Less than 6% of DFSP are rare congenital and pediatric forms^[14,15]. Clinically, DFSP are presented as cutaneous and subcutaneous lesions, and it can spread further deep down, as a mobile, fixed, an indurated, nodules or plaques red, violaceous, bluish, regular or not with a different sizes^[16]. The predilection Sites of DFSPs include the trunk (40 à 50 % des cas), the extremities (30 à 40 %), head and neck (10– 15 %) ^[11, 14]. Fibrosarcomatous is the multi nodular form due to the late evolution^[16], and it presents a higher rate of metastasis. the positive diagnosis of DFPS based on histopathology and immunohistochemical marker CD34 antigen that expressed by the tumor. A reciprocal translocation results in a fusion of the *COL1A1* and *PDGF-β* genes in 90% DFSP^[17]. The MRI imaging is consider as the choice exam for the evolution of DFSP, also MRI assessed the depth and local extend. The main DFSP treatment is surgery, resection tumor with free margin from 01cm to 05cm^[18,19], Mohs micrographic surgical technique; which consists of an excision with immediate histological examination of margins, limiting the parietal sacrifice^[20]. DFSP is sensitive to radiation and chemoresistant tumor. The radiotherapie may be considered after R1 resection without second surgery, also recurrence or inoperable tumor^[18]. Imatinib is a targeted therapy for dermatofibrosarcoma protuberans (DFSP) that inhibits PDGFB-R and shows a response in approximately 50%.^[19]. Imatinib indicated for patients with inoperable, locally advanced recurrent or

metastatic DFSP ^[21]. DFSPs characterized by high local recurrence rates, for that, it is recommended that every six months for at least five years, patients are clinically evaluated for the primary tumor, and every three months for two years, there after every six months for at least three years by clinical examination and imaging studies for lymph node metastases ^[22]. DFSP is tumor with good carcinological prognosis , overall survive about 92 % at 05 years, which is related tumor size, its local characteristics, evolution, et the quality of management ^[23].

CONCLUSION

DFSP is rare tumor, locally aggressive, its management is mainly surgical, Requires rigorous surveillance due to high recurrence rate. DFSP is rarely metastasizing tumor for that it is considered as good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that the name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest

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