CASE REPORT

Dermatofibrosarcoma protuberans: a case report and review of the literature

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Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) represents less than 0.1% of all tumors, but it is considered the most common skin sarcoma. Wide local excision (≥5 cm) has been largely replaced by Mohs micrographic surgery; however, recurrence is not rare.

Description of the case: A 35-year-old man presented with a large tumor on the upper side of his back and underwent local excision with the possible preoperative diagnosis of lipoma. Upon histological examination, the diagnosis of DFSP was made, and the patient underwent wide local excision with skin flap reconstruction and was referred for adjuvant radiotherapy. On twenty months follow-up, no recurrence has been observed.

Conclusion: DFSP is the most common cutaneous sarcoma. It originates in the dermis and tends to infiltrate underlying structures, including muscles, tendons, fascia and bone. In our case, the tumor was confined to the skin and subcutaneous tissue, however, our patient underwent adjuvant radiotherapy to avoid a possible relapse that would infiltrate deeper structures. Long-term follow-up is strongly recommended. Hippokratia 2016, 20(1): 80-83

Keywords: Dermatofibrosarcoma, local excision, flap reconstruction, adjuvant radiotherapy, recurrence

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Introduction

Dermatofibrosarcoma protuberans (DFSP) was first described by Darier and Ferrand in 1924, but the definition of "DFSP" was established by Hoffman in 1925[1]. It is a rare, low to intermediate grade soft tissue sarcoma deriving from the dermal layer of the skin[2]. Lesions tend to grow slowly and may originally present as a painless, skin-colored plaque with possible dark red or blue discoloration[3]. At latter stages, DFSPs can increase in size and become protuberant or ulcerative[2]. They are usually characterized by locally aggressive behavior[4] and tend to infiltrate adjacent structures, such as the subcutaneous tissue, muscles, tendons, and even bone structures[5]. Metastasis, however, is rarely reported[4].

The annual incidence is reported to be 0.8-4.5 cases per million in USA[3,6], and the incidence among African-Americans is almost double[6]. Different series have shown slight male or female predominance[6]. Although it appears mostly in adults (20-50 years)[7], various cases series report an incidence of 6-20 % in childhood and can even be congenital[8]. It occurs mostly sporadically in children with adenosine deaminase-deficient severe combined immunodeficiency (ADA-SCID)[9].

The most common location of DFSP is the trunk (42-72 %) followed by proximal extremities (20-30 %), and head and neck (10-16 %)[10]. DFSP sites include surgical scars, old burns, trauma, radiation dermatitis, vaccination sites, central venous line puncture sites and even insect bites[10,11].

Case report

A 35-year-old Caucasian man was admitted to our department with a large protuberant mass located at the right side of his upper back (Figure 1). The patient reported an increasing size of the tumor during the preceding six months. The patient denied any recent weight loss, fever, night sweats or chills.

On physical examination, a large, firm, painless, multinodular mass was found with no sign of localized heat or redness. There were no palpable cervical or axillary lymph nodes. There was neither personal nor familial history of malignancy.

A soft tissue ultrasound was performed, and a poorly defined, heterogeneous cutaneous tumor was described, measuring 10 x 8 cm. Magnetic resonance imaging (MRI) demonstrated an 11.5 x 9.5 x 1.5 cm heterogeneous tumor with peripheral enhancement, extending into the subcuta-

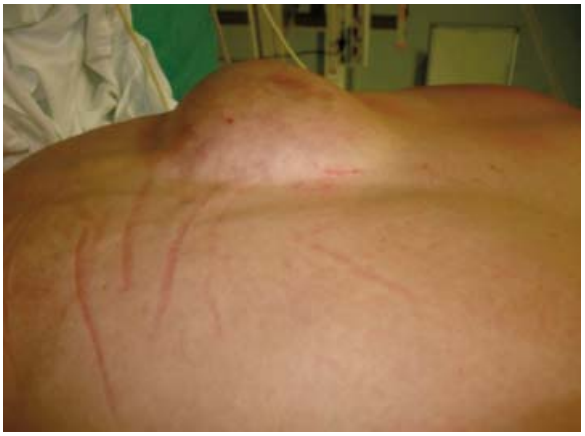

Figure 1: A large protuberant mass located at the right side of his upper back of a 35-year-old man.

neous layer but without infiltration of the adjacent muscular or bony structures (Figure 2).

With the possible diagnosis of a large lipoma, the patient underwent local excision on an outpatient basis. Upon histological examination, a well-circumscribed tumor measuring 14 x 12 x 2.5 cm (Figure 3) was described. Hematoxylin and eosin stained sections showed a cellular spindle cell neoplasm with vague cellular borders and relatively uniform elongated nuclei. On immunohistochemical stains, the spindle cells showed diffuse positivity for vimentin (Figure 4) and CD34 antigen (Figure 5). There was no positivity for smooth muscle actin, desmin, S100 protein, CD68, CD57, CD117 and keratins 8/18. Based on the histological and immunohistochemical findings, the diagnosis of DFSP was made.

The patient underwent an additional wide excision under local anesthesia, with 5 cm lateral and deep resection margins, combined with subsequent cutaneous flap reconstruction. The new histology reported no evidence of tumor cells in the specimen. The patient's postoperative course was uneventful and he was discharged on the second postoperative day, after being referred for adjuvant radiotherapy. Twenty months after surgery, no local recurrence is evident.

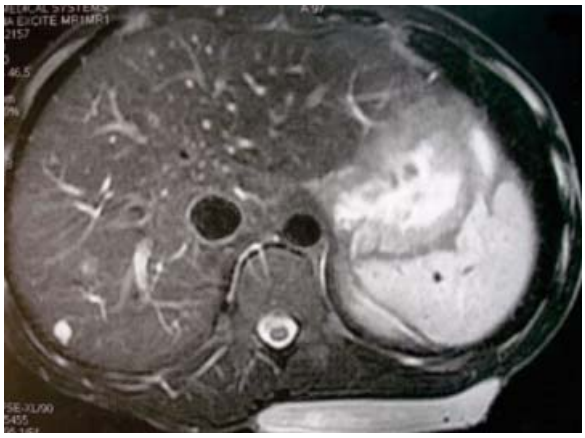

Figure 2: Axial magnetic resonance imaging demonstrating a heterogeneous tumor located at the upper back with peripheral enhancement and without infiltration of muscles or bony structures.

Discussion

DFSP is a rare, slow-growing malignant fibroblastic mesenchymal skin tumor which constitutes less than 0.1% of all malignant neoplasms and 1% of all soft tissue sarcomas[12]. DFSP usually appears as a violaceous, pink or reddish-brown plaque that develops slowly, initially limited to the skin. With time, the tumor evolves into multiple "protuberant" nodules that may infiltrate the subcutaneous tissue, fascia, muscles and even bone[10,11]. In our case, no infiltration of the adjacent muscular or bony structures was evident. Martin et al reported that in almost 50 % of their patients the tumor presented at first as a "non-protuberant" DFSP, with a mean period of 7.6 (± 9.3) years before developing into a protuberant DFSP[13].

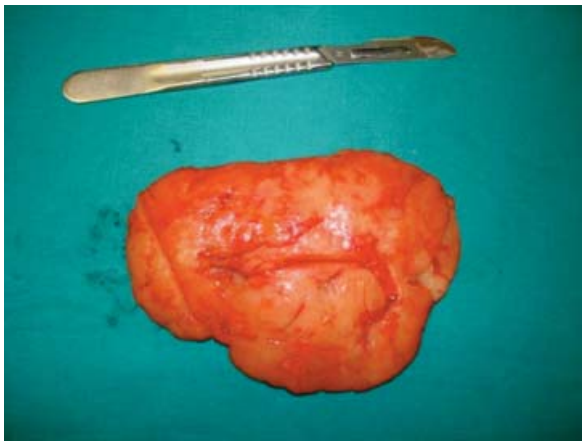

Figure 3: Macroscopic appearance of the surgical specimen is that of a large tumor (14 x 12 x 2.5 cm) with fatty appearance.

In the early stages, DFSP should be differentiated from lipomas, epidermal cysts, keloids, dermatofibroma, and nodular fasciitis. In the later stages, the differential diagnosis should consider pyogenic granuloma, Kaposi sarcoma, and other soft tissue sarcomas[14].

On ultrasound, DFSPs have been found to be mostly hypoechoic or mixed hyperechoic, with mostly well-defined margins or irregular, with projections similar to pseudopodia[15]. Vascularity of DFSP, which is a marker of malignancy, varies as well[16,17]. Since lipomas may also present with similar features, a distinction is not always possible[18]. This happened in our case, where a preoperative diagnosis of lipoma was made.

MRI studies are also not specific since they may not always distinguish DFSPs from other soft tissue sarcomas[19]. Therefore, histological examination is the only definitive diagnostic method.

Microscopically, DFSP is characterized by diffuse infiltration of the dermis and subcutis, usually sparing the epidermis and skin appendages. It grows along preexisting fibrous septa while infiltrating fat lobules giving a typical honeycomb pattern. Rarely DFSP might present

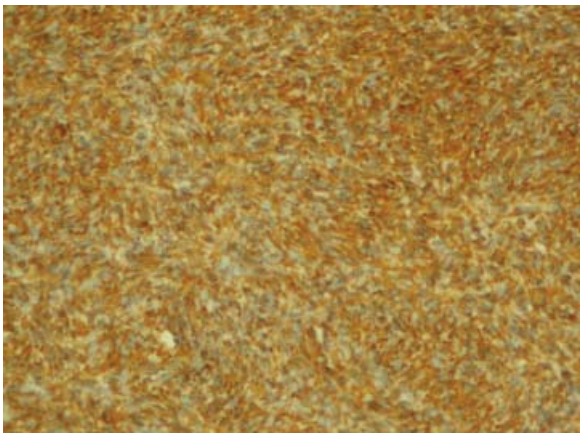

Figure 4: Immunohistochemistry of the tumor cells showing diffuse positivity for vimentin (vimentin, x200).

as an infiltrative subcutaneous mass. Atypia is minimal, and mitoses are rare[9]. The superficial part of the neoplasm might be less cellular causing problems in the differential diagnosis on small biopsies. It rarely shows prominent vessels and granular cell change[9]. Mitotic count, necrosis, and areas of fibrosarcomatous change should be stated in the histopathology report as they have been shown to be correlated with aggressive clinical behavior and lower overall survival[20]. The lack of epidermal hyperplasia, relative cellular homogeneity, a lesser amount of collagenous matrix, and diffuse subcutaneous infiltration distinguish DFSP from benign and cellular fibrous histiocytoma (dermatofibroma).

Other types of DFSP are myxoid DFSP, in which myxoid characteristics predominate and Bednar tumor, which is a pigmented DFSP characterized by the presence of dendritic cells that produce melanin[11].

Immunohistochemically, tumor cells stain for vimentin, CD34, apolipoprotein D, nestin, and may be for EMA. Desmin, S100 protein, FXIIIa, stromelysin III, HMGA1&2, tenascin, D2-40, CD163, and keratins are negative. In myoid nodules, tumor cells stain for SMA. Fibrosarcomatous DFSP may show loss of CD34 positivity and increased expression of TP53[9,20,21]. In our case, tumor cells tested positive for vimentin and CD34, thereby setting the diagnosis of DFSP.

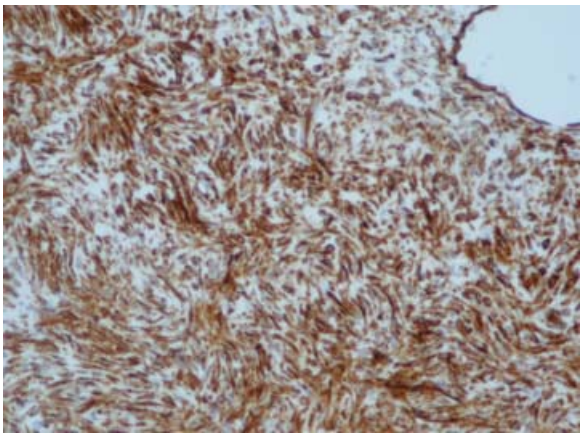

Figure 5: Immunohistochemistry of the tumor cells showing diffuse positivity for CD34 antigen (CD34, x200).

Treatment of choice is wide local excision, with negative margins of 3-5 cm from the tumor edge including the skin, the subcutaneous tissue, and the underlying fascia[22]. In cases with possible bone involvement, the periosteum or even a portion of the bone may also need to be excised to achieve negative resection margins[23]. The rate of recurrence depends on the resection margins[24]. In series where resection margins of five cm were used, recurrence rates were less than 5 %[23]. Reconstructive surgery may be required to restore tissue defects after excision using a local skin flap, skin graft or myocutaneous flap[2]. In our case, a local skin flap reconstruction was chosen.

An alternative to wide surgical resection is Mohs micrographic surgery which is considered by many as the treatment of choice for DFSP[3,25]. The technique consists of successive horizontal sectioning (5-7 μm) during resection and immediate frozen microscopic examination until a tumor-free margin is succeeded[10]. There are reports of local cure rates of 93-100 %[26,27].

Regarding adjuvant treatment, imatinib mesylate, a tyrosine kinase inhibitor, is used in the treatment of unresectable, recurrent and/or metastatic disease. Imatinib inhibits the tyrosine kinase of PDGF and seems effective in treating DFSP in patients with t (17; 22) translocation[3]. Radiotherapy should be considered in cases of positive or inadequate margins, in cases of recurrence or cases of unacceptable functional or cosmetic results after wide excision, in combination with surgery[28,29]. Due to the large size of the original tumor we referred our patient for adjuvant radiotherapy, to avoid potential relapse. Post-operative radiotherapy is reported to have a cure rate of 85 %[30]. Combination of conservative excision and adjuvant radiotherapy has demonstrated a reduced local recurrence rate of 5 %[31].

The recurrence rate is high. Most local recurrences appear within the first three postoperative years, with 50 % presenting within the first year of surgery. However, recurrences after five years are also reported[31]. Thus, it is important to follow-up these patients for long-term.

Conflict of interest

Authors report no conflict of interest.

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