

Dermatofibrosarcoma protuberans – a wolf in sheep’s clothing: a case report and review of the literature

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a locally invasive and slow growing tumor of the subcutaneous tissue. It rarely metastasizes but progressive and recurs frequently after surgical excision. The trunk and proximal extremities are the most common sites of the disease. A 53-year-old woman presented to dermatology outpatient department with a 3-cm, firm, violaceous and multinodular mass located on the left upper shoulder. Diagnostic work-up including magnetic resonance imaging and histopathological findings of biopsy were consistent with DFSP. The patient underwent wide local excision with skin flap reconstruction. No recurrence has been observed during ten months of follow-up.

Key words: *Dermatofibrosarcoma protuberans, neoplasm, violaceous, multinodular, wide excision, flap reconstruction, recurrence.*

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, low to intermediate grade soft-tissue sarcoma that was originally described in 1924 by Darier and Ferrand. The term dermatofibrosarcoma protuberans was named by Hoffman in 1925.¹ It accounts for less than 5% of soft-tissue tumors and 0.1% of all malignancies with an annual incidence of 0.8 to 4.5 per million individuals.² It is most commonly seen among young adults in their third or fourth decades. The commonest site of involvement is trunk (40-60%), followed by the proximal extremities (20-30%) and the head and neck (10-16%).³ A slight male predominance has been reported among patients with DFSP.⁴ The tumor has a low chance of metastasis, either

to regional lymph nodes or distantly but is aggressive locally.⁵ The tumors are most commonly slow-growing and remain asymptomatic. Sometimes it presents as skin colored, indurated, firm plaque that eventually progresses to violaceous and red-brown nodules with varying size from one to several centimeters in diameter. At the later stage, DFSP can increase in size and become protuberant or ulcerative.⁶ Also DFSP may present as an atrophic plaque resembling morphea and is often misdiagnosed as such.⁷ This case is reported here for its rarity.

CASE REPORT

A 53-year-old woman presented with a 3-year history of raised, firm, violaceous lesion over left shoulder. The lesion was progressively increasing in size, more so during the preceding six months. The patient denied any recent weight loss, fever, night sweats or chills. On examination, there was firm, tender, multinodular mass with violaceous color measuring about 3 cm in diameter (Figure 1). There was no palpable cervical or axillary lymph node. No other lesion was noted elsewhere. There was neither personal nor family history of malignancy. She denied any treatment of the lesion in the past.

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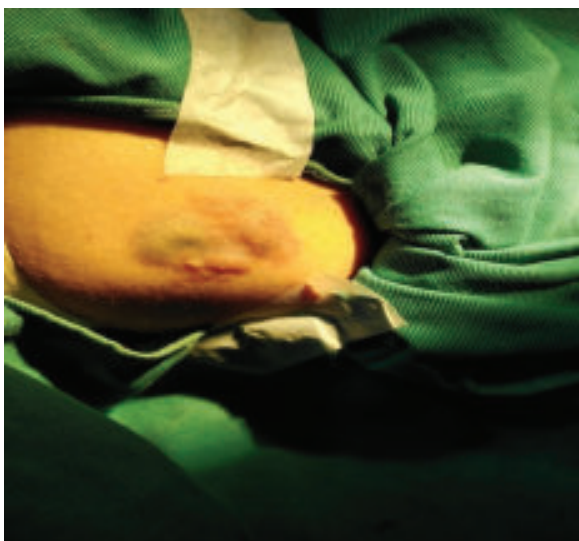


Figure 1 Photograph showing erythematous, multi-nodular, violaceous plaque located on left upper shoulder

Magnetic resonance imaging (MRI) demonstrated a 3 cm x 2 cm heterogeneous tumor with peripheral enhancement, extending into the subcutaneous layer of postero-lateral aspect of left shoulder without infiltration of the adjacent muscular or bony structures (Figure 2). Histopathology of skin biopsy showed a spindle cell neoplasm located primarily in the dermis



Figure 2 MRI scan left shoulder (contrast) showing ill-defined area about 3 cm x 2 cm within skin and subcutaneous fat but separated from muscle

with infiltration of the subcutaneous fat in a lace like pattern. The neoplastic cells showed uniform slender and elongated nuclei with scanty pale cytoplasm. Also, the spindle cells were arranged as intersecting fascicles and as whorls in a storiform pattern (Figure 3). Biopsy findings were consistent with the diagnosis of DFSP. Routine hematological and biochemical investigation reports were within normal limits.

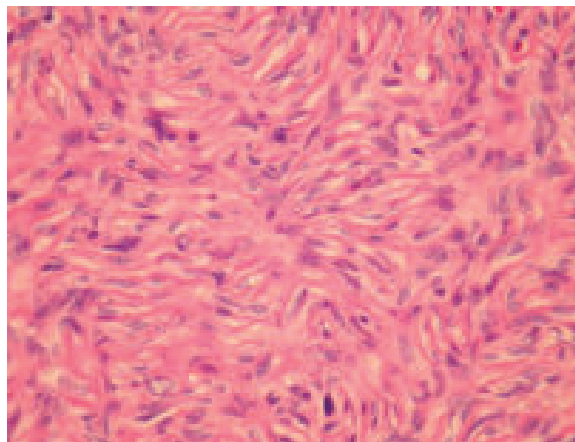


Figure 3 Histopathological features showing proliferating spindle shaped fibroblastic cells arranged in storiform pattern with elongated pleomorphic nuclei, occasional mitotic figures are also present

The patient was referred to plastic surgery department, where under general anesthesia, with 2 cm lateral and deep resection margins, combined with subsequent cutaneous flap reconstruction was done. The patient's postoperative course was uneventful and she was discharged on the second postoperative day advising regular follow up. Ten months after surgery, no local recurrence is evident.

DISCUSSION

The involvement of DFSP is initially limited to skin; with time, the tumor evolves into multiple 'protuberant' nodules that may infiltrate the subcutaneous tissue, fascia, muscles and even bone. The tumor has a low chance of metastasis, either to regional lymph nodes or distantly but is aggressive locally. A local recurrence rate of DFSP of up to 60% has been reported.⁸ Its high recurrence rate is due to its strong capacity to infiltrate subcutaneous tissue, fascia and underlying muscle (infiltration in the form of pseudopods).^{9,10} 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 ‘nonprotuberant’ DFSP, with a mean period of 7 to 8 years before developing into a protuberant DFSP.¹¹ Also, there are reports of lesions in areas that suffered previous trauma or surgery. Our patient had no such history.

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.¹²

Vascularity of DFSP, which is a marker of malignancy, varies as well. MRI studies are also not specific since they may not always distinguish DFSP from other soft tissue sarcomas. Therefore, histological examination is the only definitive diagnostic method. Microscopically, DFSP is characterized by diffuse infiltration of the dermis and sub-cutis, 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 as an infiltrative subcutaneous mass. Atypia is minimal and mitoses are rare.¹³

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.^{14, 15} Reconstructive surgery may be required to restore tissue defects after excision using a local skin flap, skin graft or myocutaneous flap. In our case, a local skin flap reconstruction was chosen. An alternative to wide surgical excision is Mohs micrographic surgery which is considered by many as the treatment of choice for DFSP.¹⁵ But unfortunately, this technique is not available here.

The rate of recurrence depends on the resection margins. In series where resection margins of 5 cm were used, recurrence rates were less than 5%. Most local recurrences appear within the first 3 years of surgery, with 50% presenting within the first year. However, recurrences after 5 years are also reported. Thus, it is important to follow-up these patients for long-term.

Radiotherapy is an adjuvant therapy in cases where adequate surgical margins are not easily reached or result in cosmetic /functional defect or in cases of positive margins, even after maximum resection. It is also

indicated for inoperable macroscopic lesions. In our case, all margins of resected specimen were histologically free from tumor.

The factors associated with high rates of recurrence are histological subtype, cellularity, size, location on the head and neck and high mitotic rate.¹⁶ Since, the tumor projects in multiple directions, reaching deep structures, not even wide excision can remove all residual tumor in single or multiple focus and this would explain lesion recurrence.

This case inspired us to review the most important aspects of this rare tumor. Dermatologists and surgeons must know about the frequent recurrence of this tumor, sometimes even when excised with wide margins. These patients must be observed periodically after surgery for a long period.

Authors’ contribution: MKA diagnosed the case, did literature review and drafted the manuscript. MRI did surgical management of the case and helped in drafting the manuscript.

Consent: Informed written consent was obtained from the patient for the publication of this case report and any accompanying images.

Conflict of interest: Nothing to declare.

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