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Giant Recurrent Dermatofibrosarcoma Protuberans of the Back: A Case Report

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Abstract **Case Report**

Dermatofibrosarcoma protuberans (DFSP), is a rare, slow-growing cutaneous sarcoma with high local recurrence and low metastatic potential. We report the case of a 39-year-old male presented with a recurrent, painless mass in the left scapular region, previously excised at a non-specialized center. MRI showed a well-defined subfascial lesion without muscle invasion. Histology and immunohistochemistry confirmed DFSP with strong CD34 positivity. The patient underwent wide local excision, including resection of the muscle aponeurosis. Skin coverage was achieved with a split-thickness graft, and recovery was uneventful. In this presentation we aim to highlight the importance of early recognition and multidisciplinary management to prevent and minimize recurrence risk.

Keywords: Dermatofibrosarcoma Protuberans, Soft Tissue Sarcoma, Surgical Excision, Recurrence, Case Report.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), is a rare cutaneous soft tissue tumor characterized by slow progression, high local aggressiveness, and a significant risk of recurrence. These features classify DFSP as a tumor of intermediate malignancy [1]. Histological and immunohistochemical (IHC) analysis remain essential for confirming the diagnosis [2]. In recent years, molecular diagnosis techniques have become increasingly important, as over 90% of DFSP cases are associated with a characteristic chromosomal translocation, t(17;22) (q22;q13) [3]. Complete surgical excision remains the treatment of choice [4], with imaging, particularly MRI, playing a key role in preoperative planning [5].

OBSERVATION

A 39 year old male construction worker presented with a painless dorsal swelling. He had undergone prior surgery for a tumoral mass in the left upper back region at a non-specialized center approximately 18 months earlier. The patient reported that the swelling had reappeared over a couple of months after surgery and had gradually increased in size.

On clinical examination a poorly defined polylobulated mass was observed in the scapular region of the left shoulder. The mass measured approximately 200 cm² in surface area and included an exophytic violaceous component. The mass was firm in consistency and adherent to both superficial and deep tissue planes. Loco-regional examination revealed no palpable axillary lymphadenopathy [Figure 1].

MRI of the left scapular region showed a well-defined mass within the fascial plane, with predominantly low signal on STIR and T2, and iso- to hypointense signal on T1. Post-contrast images revealed heterogeneous enhancement. No invasion of adjacent muscles or surrounding tissue was observed.

An Ultrasound Guided biopsy was performed. Histological and immunohistochemical findings confirmed the diagnosis of DFSP with tumor cells showing a diffuse positivity for CD34 and negativity for S100 and factor XIIIa. A Chest CT scan was performed ruling out any pulmonary metastases.

Under general anesthesia, A wide surgical excision was performed with a 5 cm margin from the tumor, inclusion resection of the scapular muscle

aponeurosis and extension into the muscular plane, all removed en bloc [figure 2].

The surgical specimen, was sent for histopathological analysis. Postoperatively, the patient was managed with fatty dressings, and directed wound healing was achieved within one month.

Upon confirmation of the diagnosis and clear surgical margins from histological analysis, the patient subsequently underwent a split-thickness skin graft, which healed with good functional and aesthetic outcomes [Figure 3].

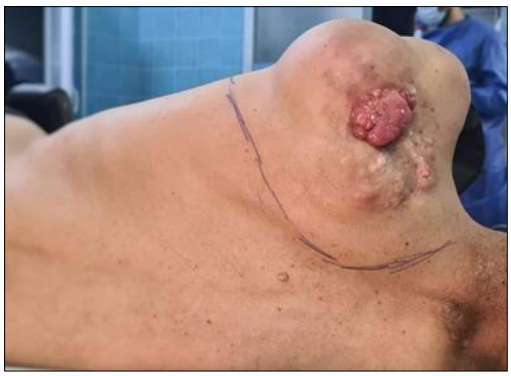

Figure 1: Clinical appearance of the tumor prior to surgical excision, showing a large, polylobulated violaceous mass in the left scapular region

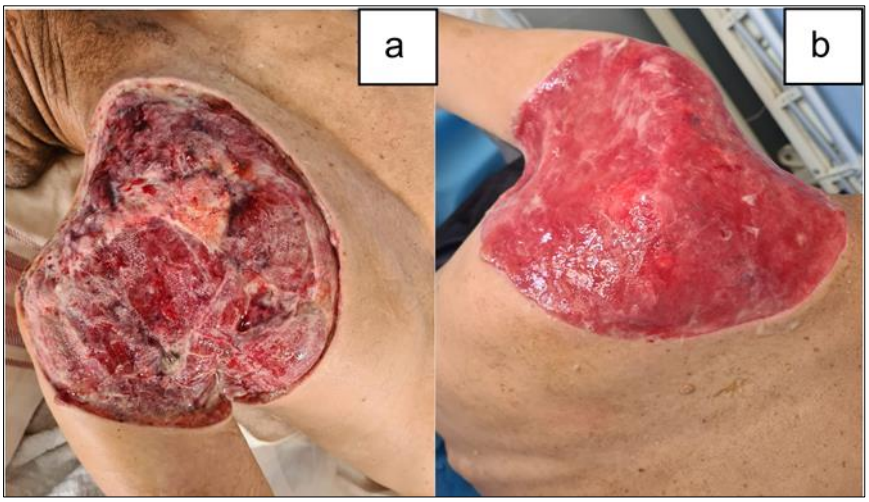

Figure 2: Postoperative appearance of the wound site following wide excision (a) and during the process of directed wound healing (b)

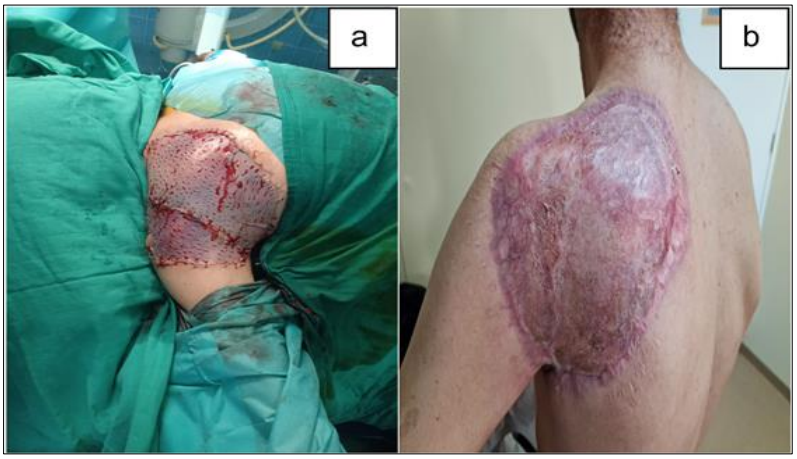

Figure 3: Intraoperative view showing the application of a split-thickness skin graft (a) and its final result after successful graft take (b)

DISCUSSION

The term Dermatofibrosarcoma protuberans (DFSP) was established in 1925 by Hoffman [6-8] it represents less than 2% of all soft tissue sarcomas [9-11], it is mostly observed in adults of 20-40 years old but can occur in any age, with no gender difference [5-12].

Clinically the DFSP is typically presented as an asymptomatic, slowly enlarging violaceous nodule, it is fixed to the superficial skin and mobile over deeper tissues. Although DFSP lesions are usually small, ranging between 1 to 5 cm they can grow significantly, and if left untreated, may exceed 20 cm in size [5]. Such is the case in our presentation. The trunk, upper and lower extremities are the most common sites for DFSP, followed by the scalp and neck regions [4, 5], rarer locations include the breast and vulvar regions [13, 14]. Therefore DFSP may develop in any region of the body with cutaneous tissue.

The diagnosis of DFSP is confirmed with histopathological and IHC findings, it is presented as a cellular neoplasm composed of storiform spindle cells with ovoid to elongated nuclei, minimal cytological atypia and a low mitotic count within a collagenous stroma. tumor cells are generally centered within dermis or subcutis, and is also associated with strong and diffuse expression of CD34 [14], in some cases, the use of molecular biology helps in confirming the diagnosis, since the chromosomal translocation t(17;22) (q22;q13) is present in approximately 90% of cases [14].

When it comes to imaging in DFSP, it does not play an essential role in the diagnostic work up but is essential for planning therapeutic interventions. Since its findings are not specific [5]. MRI, in DFSP, typically shows a lesion with low signal intensity on T1 and high intensity on T2, with homogeneous to heterogeneous mild contrast enhancement [5]. Although DFSP is known to have minimal metastatic potential a preoperative chest CT scan is recommended, particularly, to rule out pulmonary metastases [14].

Surgical excision remains the standard treatment of choice for DFSP, with the primary objective of achieving negative margins to prevent recurrence [15]. As observed in our case the patient had undergone a non-cancerogenic surgery resulting in early recurrence and aggravation of his symptomatology. In the literature, two main surgical techniques are described for managing DFSP: Wide Local Excision (WLE) and Mohs Micrographic Surgery (MMS). Although MMS is more complex and technically demanding, it offers superior margin control and preserve healthy tissue, with reported recurrence rates of less than 2% [15]. In our practice, WLE is the preferred approach, employing peripheral margins of 3 to 5 cm and the excision of a healthy anatomical barrier in depth.

In some cases surgical management of DFSP can pose significant challenges, depending on the size and location of the tumor, thus achieving negative surgical margins without causing substantial aesthetics or, functional compromise may be difficult if not possible [4, 15]. For such cases and due to the presence of the t(17;22) (q22;q13) translocation in DFSP, Imatinib has shown significant efficacy in slowing tumor progression or as a neoadjuvant therapy to reduce tumor size prior to surgery in large or advanced lesions [14].

This case highlights the need for thorough clinical evaluation and accurate diagnosis to guide appropriate treatment and prevent recurrence. Despite their rarity and heterogeneity, soft tissue tumors require histological confirmation to avoid misdiagnosis and inappropriate management.

CONCLUSION

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous sarcoma with slow growth, local aggressiveness, and high recurrence risk. Diagnosis relies on histology, immunohistochemistry, and molecular testing. MRI aids surgical planning, but wide excision with negative margins remains essential for effective management and recurrence prevention.

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