SAR Journal of Surgery

Abbreviated Key Title: *SAR J Surg*
Home page: https://sarpublication.com/journal/sarjs/home
DOI: 10.36346/sarjs.2022.v03i01.001

ISSN 2707-7748 (P)
ISSN 2709-6912 (O)

Case Report

A Reminder of a Rare Benign but Aggressive Tumor: Dermatofibrosarcoma Protuberans

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Article History: | Received: 19.01.2022 | Accepted: 25.02.2022 | Published: 28.02.2022 |

Abstract: Dermatofibrosarcoma protuberans (DFSP) is a slowly growing fibrous tumor of the skin with a very high risk of local recurrence, but with low metastatic potential. Its malignant sarcomatous transformation with metastasis is exceptional. We report a rare case, of a 30-year-old patient, who presents dermatofibrosarcoma protuberans in the left supraclavicular region. Through this case we will underline the clinical presentation as well as the management of this benign but aggressive tumor.

Keywords: Dermatofibrosarcoma protuberans, supraclavicular, reconstruction, case report.

INTRODUCTION

Dermatofibrosarcoma is a mesenchymal dermal tumor of intermediate malignancy [1]. It is a rare but not exceptional tumor, representing between 0.1% and 1% of malignant skin tumors [2]. It is a tumor with a very high risk of local recurrence, but with low metastatic potential. We report this case to highlight the clinical presentation as well as the management of this tumor.

CASE REPORT

We report the case of a 30-year-old patient who presented with a mass in the left supraclavicular region that had been gradually increasing in volume for 6 months. The skin covering the mass was inflamed with a purplish-red appearance (nodular-looking mass) (figure 1). The tumor is fixed the skin, mobile relative to the deep plane and painless. Cervical examination found no associated lymphadenopathy. A cervicothoracic CT scan showed the presence of a tissular mass in the left supraclavicular region without infiltration of the underlying muscle (figure 2). A biopsy was performed; the histological study confirmed the diagnosis of dermatofibrosarcoma protuberans.

The patient underwent surgery, we did a large excision with a marge of 5cm from the tumor, and the loss of substance was reconstructed by a skin graft.

Histopathological examination of the surgical specimen confirmed the diagnosis and resection was complete with margins passing 5cm from the tumor. The patient is under regular surveillance, she shows no sign of recurrence and she is very satisfied with the result.

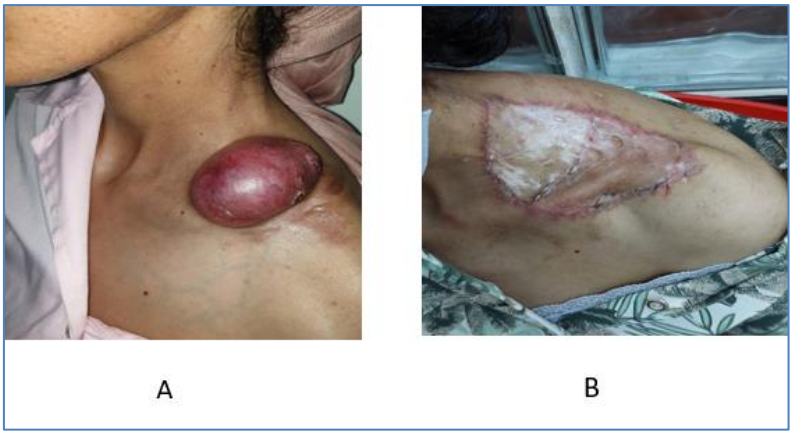

Fig-1, A: picture showing the supraclavicular mass, purplish-red skin and the nodular appearance of the tumor
Fig.1, B: postoperative picture

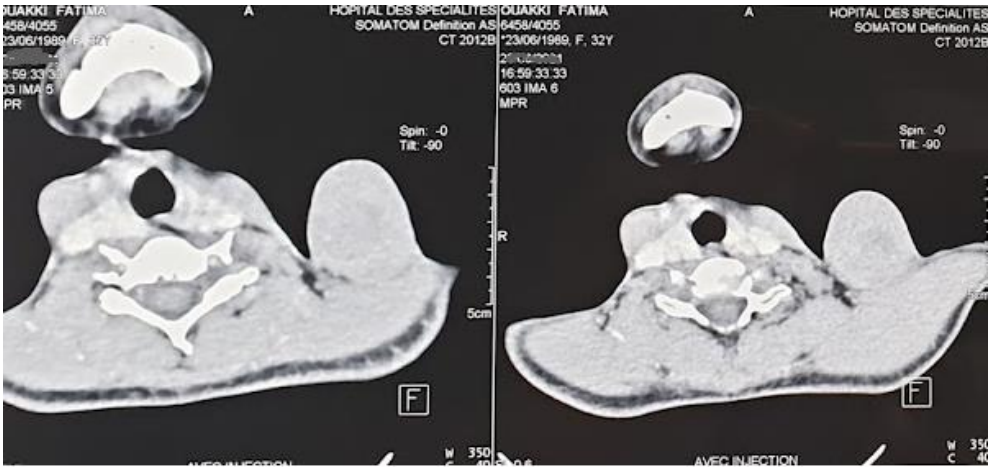

Fig-2: Cervicothoracic CT showing the presence of a tissue mass in the supraclavicular region without infiltration of the underlying muscle

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a mesenchymal cutaneous tumor with intradermal development described for the first time by Taylor [1] as a sarcomatous tumor. It is a tumor, located between the pole of benignity of the very common and harmless cutaneous fibroma and the pole of malignancy of true cutaneous fibrosarcoma. Its malignant sarcomatous transformation with metastasis is exceptional.

DFSP can affect any part of the body. According to the literature, there is a predilection for the trunk which is affected in 50 to 60% of cases. Limbs account for 20-30% of locations and 15-20% are attributed to the head and neck [2, 3].

This tumor can occur at any age. In the literature, the age of diagnosis varied between 28 and 47 years [3,4]. DFSP is rare in children under 15 and the congenital form is exceptional [5].

Clinical diagnosis is difficult. At an early stage, the lesion appears as an indurated plaque. At a more advanced stage (nodular stage), a few months to a few years, it presents as a nodular or multinodular mass. Untreated, these lesions can become very large, or ulcerate to become painful and hemorrhagic [2, 6].

Histological examination is essential for diagnosis. The tumor is made of a dense cell proliferation, poorly limited, not encapsulated, occupying the dermis, most often in its entirety. In general, the histological appearance helps to guide the diagnosis. In doubtful cases, immunohistochemistry can distinguish DFSP from other spindle cell tumors [4,5].

Wide surgical excision is the reference treatment with safety margins of 4 to 5 cm. Postoperative radiotherapy is recommended by some authers, from the second recurrence [7]. Systemic chemotherapy is not recommended. Rigorous clinical monitoring must be maintained, due to the slow evolution and high recurrence potential of this tumor [7].

CONCLUSION

DFSP is a tumor whose prognosis and evolutionary risk are mainly related to the diagnostic time and the quality of the first excision. Late diagnosis makes excision and reconstruction surgery difficult.

Improving the prognosis requires early and codified multidisciplinary management.

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