

Dermatofibrosarcoma Protuberans is Possible Without Skin Involvement

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ABSTRACT Dermatofibrosarcoma protuberans (DFSP) is a painless, slowly growing, rare skin tumor. It is most common in the trunk part of the body. It is frequently seen in the 40s. It is usually a low-grade sarcoma. DFSP, which is a locally benign-aggressive tumor, begins with a small size and may infiltrate the skin, subcutaneous adipose tissues, muscles and bones, and may recur despite extensive excision. It never metastasize without higher grade fibrosarcomatous changes. Although the benefit of chemotherapy and radiotherapy is limited, it can be applied in cases of recurrence or metastasis. We present a case of DFSP without skin lesions in a 46-year-old woman presenting with a slow-growing mass on her back. The mass was removed with a wide surgical margin. We aimed to emphasize that DFSP, which is very rare, can be seen without dermal involvement.

Keywords: Dermatofibrosarcoma protuberans; dermis; subcutaneous tissue

As a rare tumor, dermatofibrosarcoma protuberans (DFSP) is an intermediate malignant skin tumor that probably arises from fibroblasts and is characterized by an infiltrative growth pattern and local aggressive potential. It was first defined by Darier and Ferrand.^{1,2} DFSP is characterized by locally infiltrative and debilitating growth, asymmetric, subclinical extensions in the horizontal direction and infiltration of deeper structures. It mainly affects young and middle-aged adults. It emerges as an asymptomatic bluish or brownish erythematous multinodular plate, and the involved areas are most frequently the trunk, proximal extremities, head and neck. Local recurrences often occur requiring more than one surgical procedure, as the tumor is spread intra- and/or subcutaneously and is often very difficult to distinguish clinically.³⁻⁵ In histopathology, the tumor is located in the dermis and consists of uniform fusiform cells, densely grouped, with an elongated nucleus. The

mass should be excised with a wide surgical margin to maintain an adequate safety distance. Developing a multidisciplinary approach is an expertise that could achieve for the treatment of DFSP lesions.¹⁻⁷

CASE REPORT

A 46-year-old female patient presented with a gradually growing back mass, sometimes accompanied by pain, for the last 6 months. In the physical examination, a mass of 50x40 mm in size was palpated under the scapula in the upper left lateral thorax. There was no dermal involvement, the mass was mobile under the skin. The chest wall was evaluated as a lesion compatible with the mass by superficial ultrasonography. Thoracic tomography was performed and it was reported that there was no lesion bone involvement with smoothly circumscribed soft tissue density of 50x38 mm in the subcutaneous adipose tis-

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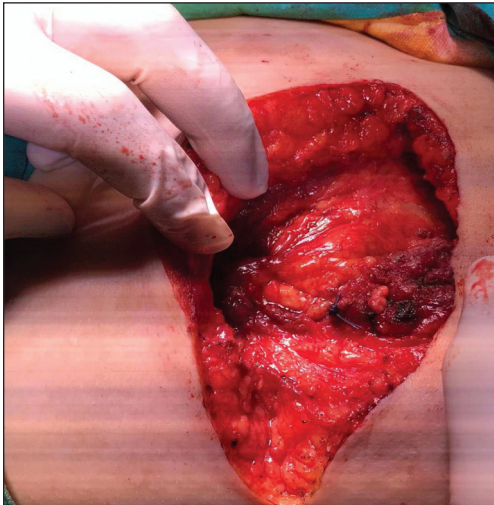


FIGURE 1: The mass in the subcutaneous adipose tissue just adjacent to the deltoid muscle in the lateral part of the left thoracic wall.

sue just adjacent to the deltoid muscle in the lateral part of the left thoracic wall (Figure 1). A diagnostic interventional radiology biopsy was taken. The biopsy result was diagnosed with DFSP. Positron emission tomography/computed tomography (CT) was performed on the patient in terms of distant metastasis. There was diffuse moderately increased fluorodeoxyglucose (FDG) uptake (SUV_{max} value 3.08) of the soft tissue density lesion with smooth borders and encapsulation of the mass. Also, an increased FDG uptake (SUV_{max} 8.31) was reported in the lesion with an irregular border of soft tissue density of 17.3x12.6 mm in the posterior neighbourhood of the right foot and the second finger. The patient was examined by orthopedics and a magnetic resonance imaging (MRI) of the right foot was ordered. A

nodule with a diameter of 1 cm was observed in the right 2nd metatarsophalangeal joint at the flexor level. The patient was operated on when metastasis was not considered by orthopedics. Wide resection was made from the mass itself and the healthy tissue. Large tissue samples were sent for the frozen section examination from the remaining intact tissue from the lower, upper, right, left, skin and lower muscle fascia. When the surgical margin was negative, the skin was closed. The pathology result of the mass was reported as DFSP. On macroscopic examination, the entire excision material consisted of a tumour mass. It was observed that the tumor, measuring 5x4x3 cm, contained a capsule and had a straight border. On microscopic examination, tumor cells showed an arrangement in storiform patterns, characterized by spindle nuclei and a moderate amount of cytoplasm in CD34, H&E, and Ki-67 stains (Figure 2).

With the decision of the tumor council, it was decided to follow up the patient without recommending chemoradiotherapy. The informed consent was obtained from the patient for the case report.

DISCUSSION

DFSP occurs in the dermis of the skin and spreads throughout the epidermis, a local aggressive sarcoma that spreads throughout adipose, muscle, and bone tissues. Usually, there is skin involvement, it can be seen as puffy as multiple lesions on the skin.^{2,5,7,8} In our case, no visible skin involvement was detected, and no skin-related involvement was found in the pathology interpretation. The mass can be confused

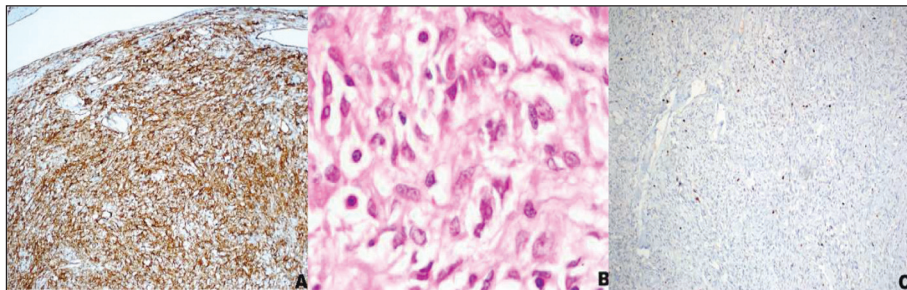


FIGURE 2: The tumor cells were arranged in a swirling pattern, and were characterized by spindle nuclei and a moderate amount of cytoplasm on microscopic examination [A. CD34 Stain (X20), B. H&E Stain, (X100), C. Ki-67 Stain, (X20)].

with many benign and malignant tumors in its external appearance and CT examination. Besides, tomography and MRI are necessary for the relationship of the mass with deep tissues.⁷ An aggressive surgical intervention should be performed for this tumor within safe surgical margins for complete cure.^{2,7} Reconstructive procedures are required in extensive surgeries due to the large size of the tumors. This requirement occurs mostly in head and neck lesions.² In our study, a wide surgical margin was applied, and the surgery was terminated after confirming that the surgical margin was free of tumor with frozen section. There was no need for reconstructive surgery. The tumor may remain silent for years or local recurrence may occur. Local recurrence may occur especially in head and neck tumors. Local recurrence occurs most frequently within 3 years after surgery.^{7,9} In a study, it was reported that local recurrence occurred 2 months after surgery.¹⁰ Distant metastases occur after recurrent tumors, they can reach distant organs through blood. These can be lungs, breasts and other organs.^{2,8} Fibromatous progression in DFSP results in metastasis.¹¹ It is a tumor limited to chemotherapy and radiotherapy but can be performed in the presence of recurrence and metastasis. In limited cases, radiotherapy can be applied to those with positive surgical margins.² We did not apply post-surgical chemoradiotherapy to our patient. Patients should be checked for at least 6 months in the first 5 years and annually for recurrence in the next 5 years.^{5,9,10,12} No recurrence or metastasis has been observed in our patient for 2 years, during our regular follow-up.

In conclusion, DFSP is a rare malignant tumor and may be nonprotuberant at presentation. Extensive surgery is necessary to avoid recurrence. After the entire tumor is removed, the surgical margin should be controlled by intraoperative pathological methods and the surgical margin should be increased according to the frozen result. DFSP may not be seen macroscopically on the skin or it can be palpated as a mass without involving the skin.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Menduh Oruç, Atalay Şahin; **Design:** Menduh Oruç, Atalay Şahin; **Control/Supervision:** Menduh Oruç, Ayşenur Keleş; **Data Collection and/or Processing:** Menduh Oruç, Ayşenur Keleş; **Analysis and/or Interpretation:** Menduh Oruç, Ayşenur Keleş, Atalay Şahin; **Literature Review:** Atalay Şahin, Menduh Oruç; **Writing the Article:** Atalay Şahin, Menduh Oruç, Ayşenur Keleş; **Critical Review:** Atalay Şahin, Menduh Oruç, Ayşenur Keleş; **References and Fundings:** Atalay Şahin, Menduh Oruç, Ayşenur Keleş; **Materials:** Ayşenur Keleş, Menduh Oruç.

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