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The Transformation of Dermatofibrosarcoma Protuberans into a Fibrosarcoma (A case Report)

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I. Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare, low- to intermediate-grade soft-tissue tumor originating in the cutaneous dermis. Representing around 0.1% of malignant skin tumors and 2-6% of soft tissue sarcomas, it mainly affects young adults aged between 20 and 50, with no significant gender predominance (1) .

DFSP usually manifests as a nodular mass on the skin, frequently localized on the trunk and extremities. Its main characteristic is local aggressiveness, with no lymphatic spread, and a low risk of metastasis (less than 5%) (2).

Treatment is based on surgical excision with wide margins of 3 to 5 cm to limit recurrence, which is around 40% in the case of marginal excision (1,2).

The malignant fibrosarcomatous transformation of dermatofibrosarcoma protuberans (DFSP) represents an exceptional form of DFSP with a higher risk of local recurrence and distant metastases than ordinary DFSP.

Herein, we report an exceptional case of A 55-year-old woman, presented with a 10-year history of a gluteal skin tumor suggestive of a transformed dermatofibrosarcoma.

II. Case Report

We report a case of a 55-year-old adult woman, with a history of cholecystectomy 4 years ago, admitted to our facility for management of a skin tumor in the gluteal region that had been evolving for 10 years and was suggestive of dermatofibrosarcoma.

On inspection, a swollen mass was found in the upper left buttock quadrant, measuring approximately 15 cm in length. It shows an ulcerated and protruding lesion. The center of the mass has a necrotic appearance .

The mass is irregular on palpation, hard and painful with multiple hard nodular formations, attached to the superficial and deep planes.

Inguinal, cervical, supra-clavicular and axillary lymph nodes were free.

The rest of the clinical examination was unremarkable.

No change in general condition was noted.

A CTAP scan was performed, revealing no distant secondary localization.

An MRI of the gluteal region was performed, which showed a suspicious cutaneous tissue mass in the upper-external quadrant of the left buttock, infiltrating the skin and contacting the gluteus maximus muscle, with no clear evidence of muscular infiltration .

After multidisciplinary consultation, including plastic surgeons, oncologists, and radiologists, a wide excision was decided upon. The patient underwent wide excision with a 3cm margin, removing the first anatomically intact barrier which was the muscle aponeurosis.

The defect was subsequently grafted with a split thickness graft.

Anatomopathological study of the specimen revealed a morphological presentation and immunohistochemical profile of a transformed dermato-fibrosarcoma showing a tumoral proliferation in spindle-shaped cells

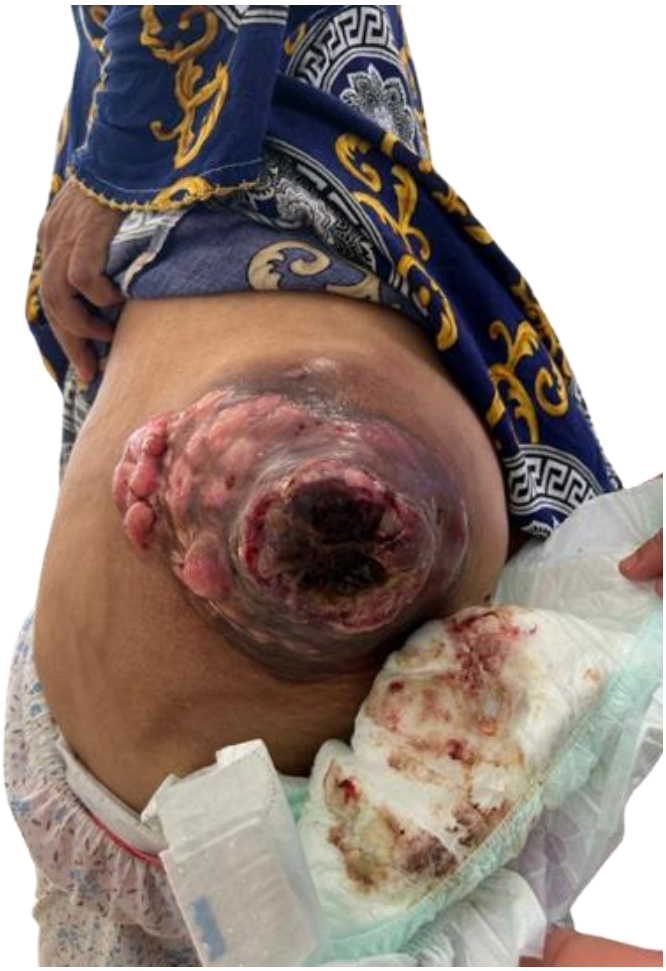

Figure 1 Macroscopic appearance of the ulcerative-exophytic mass

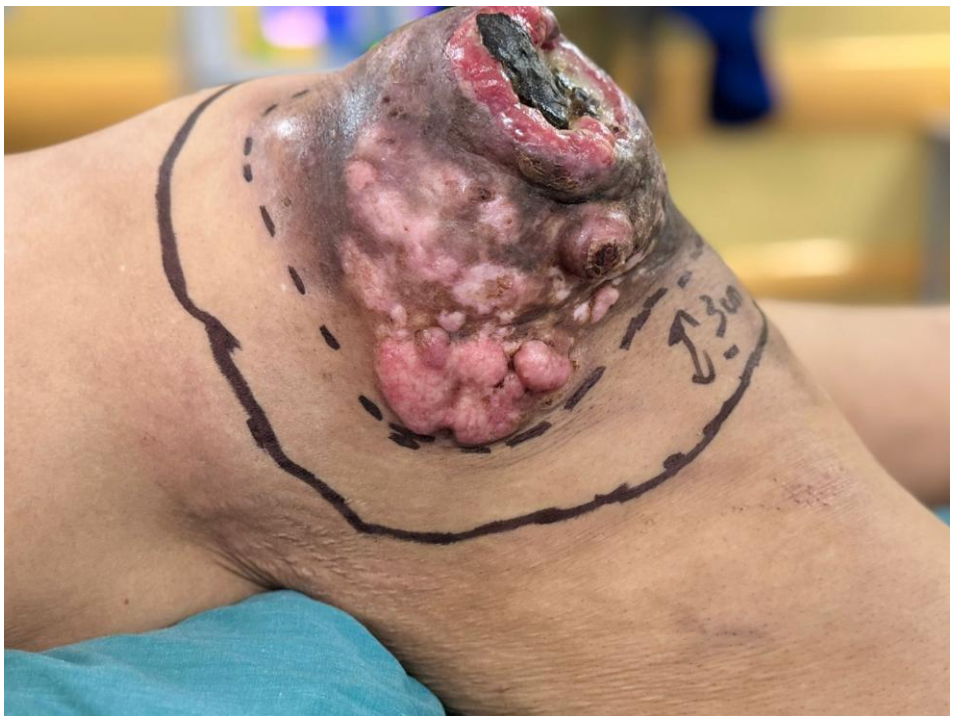

Figure 2Intraoperative lateral view of the ulcerative-exophytic gluteal mass

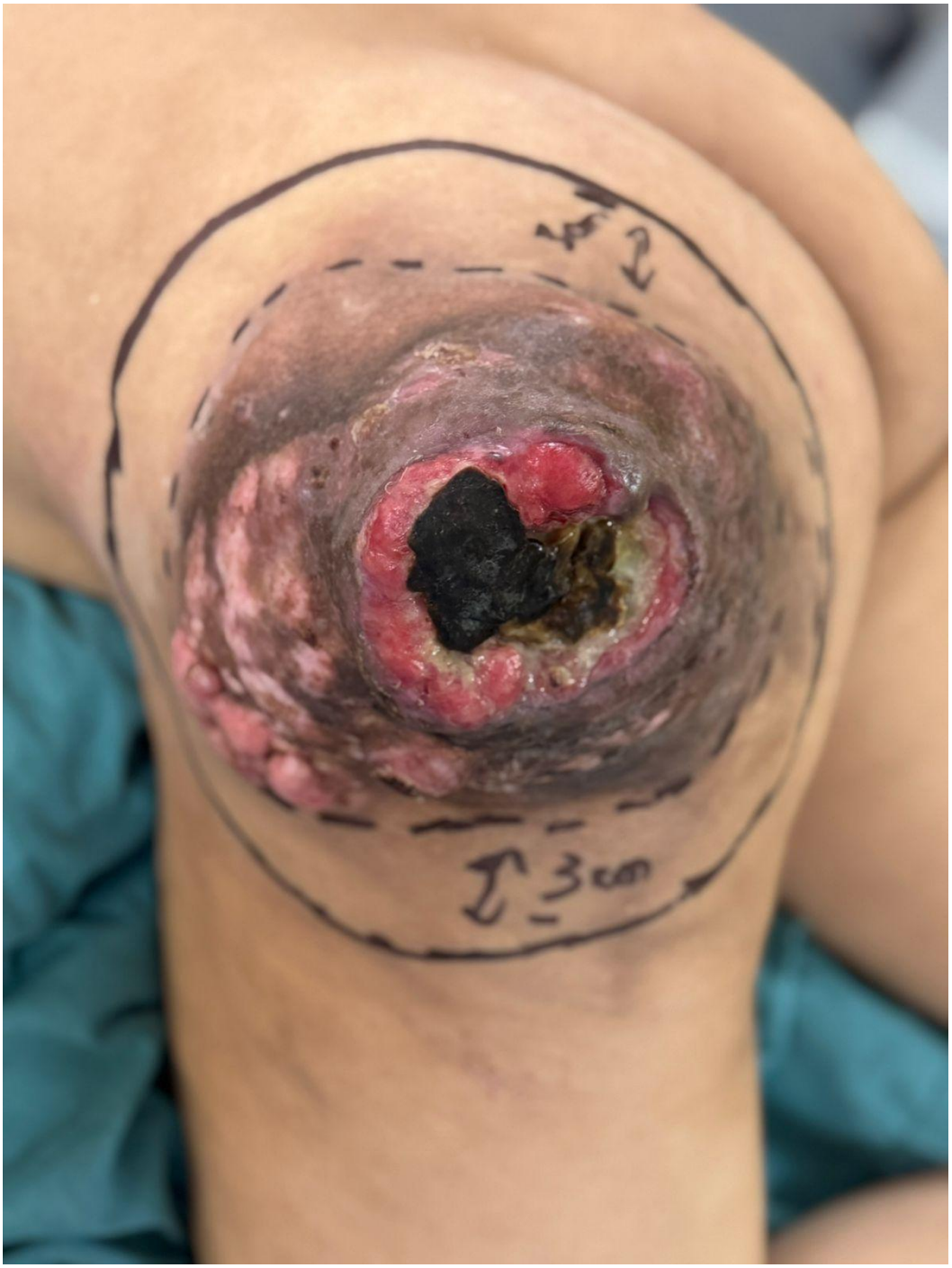

Figure 3Intraoperative superior view showing the necrotic aspect of the tumor

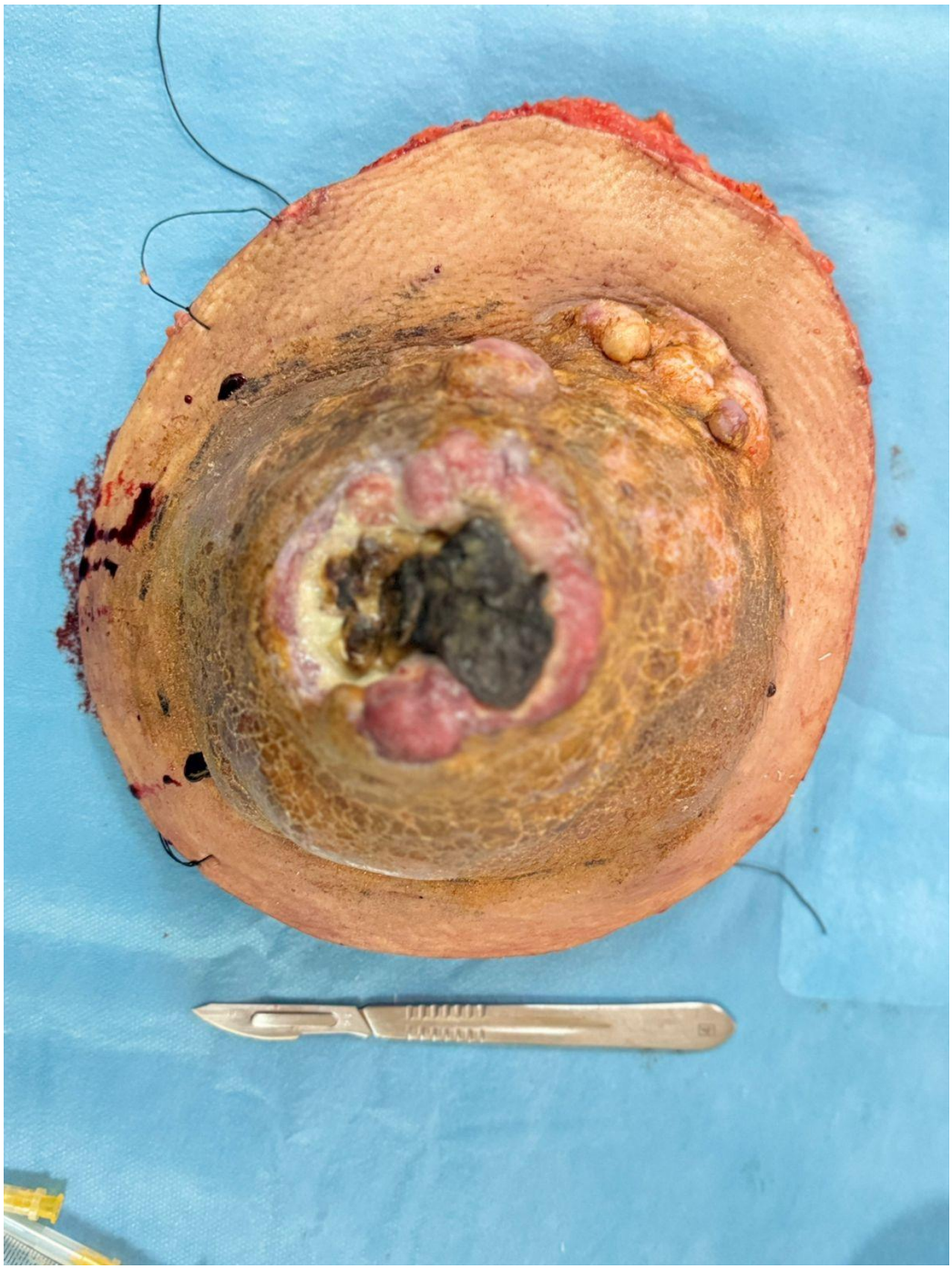

Figure 4 Surgical specimen of the tissue mass

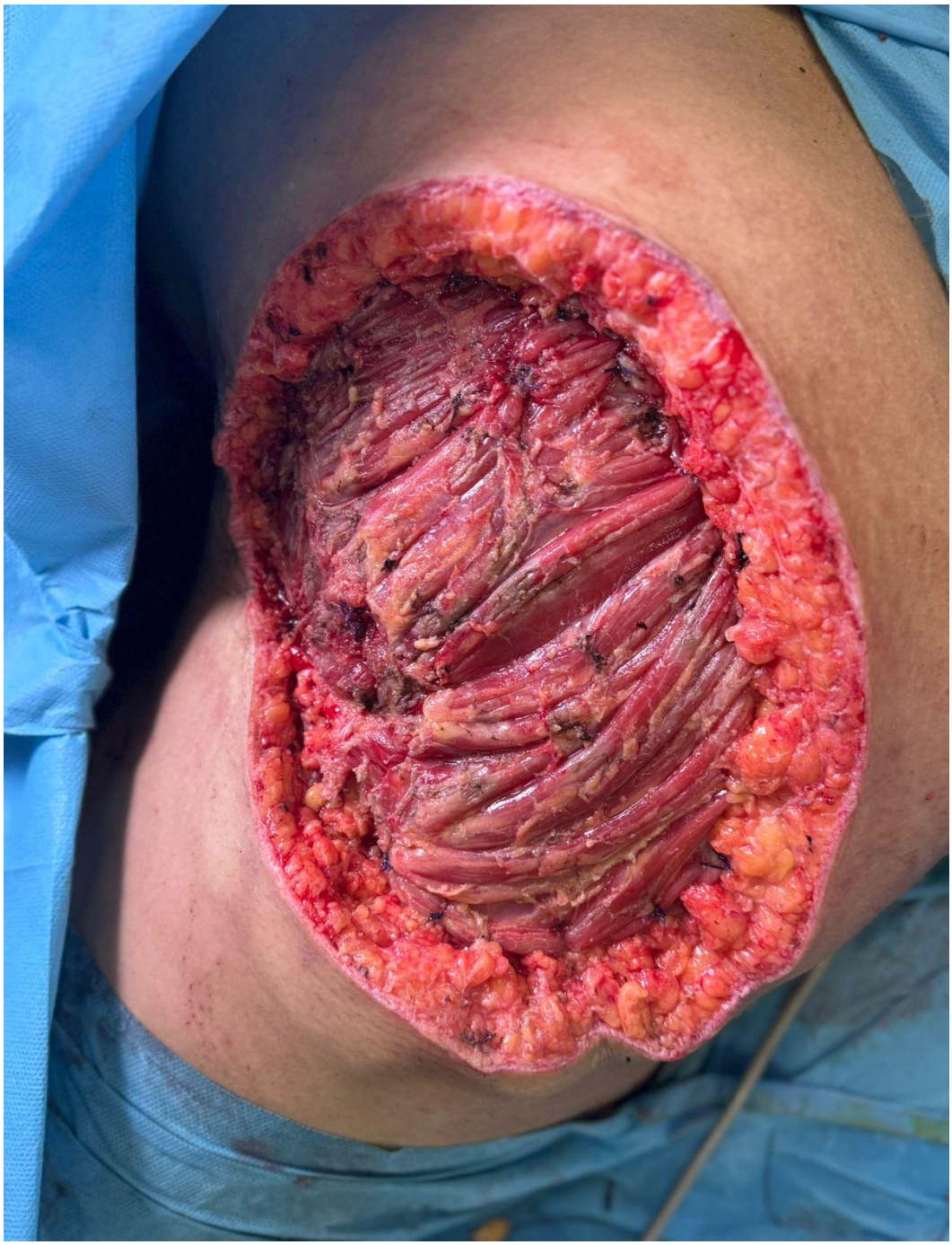

Figure 5 Intraoperative macroscopic appearance after excision of the mass

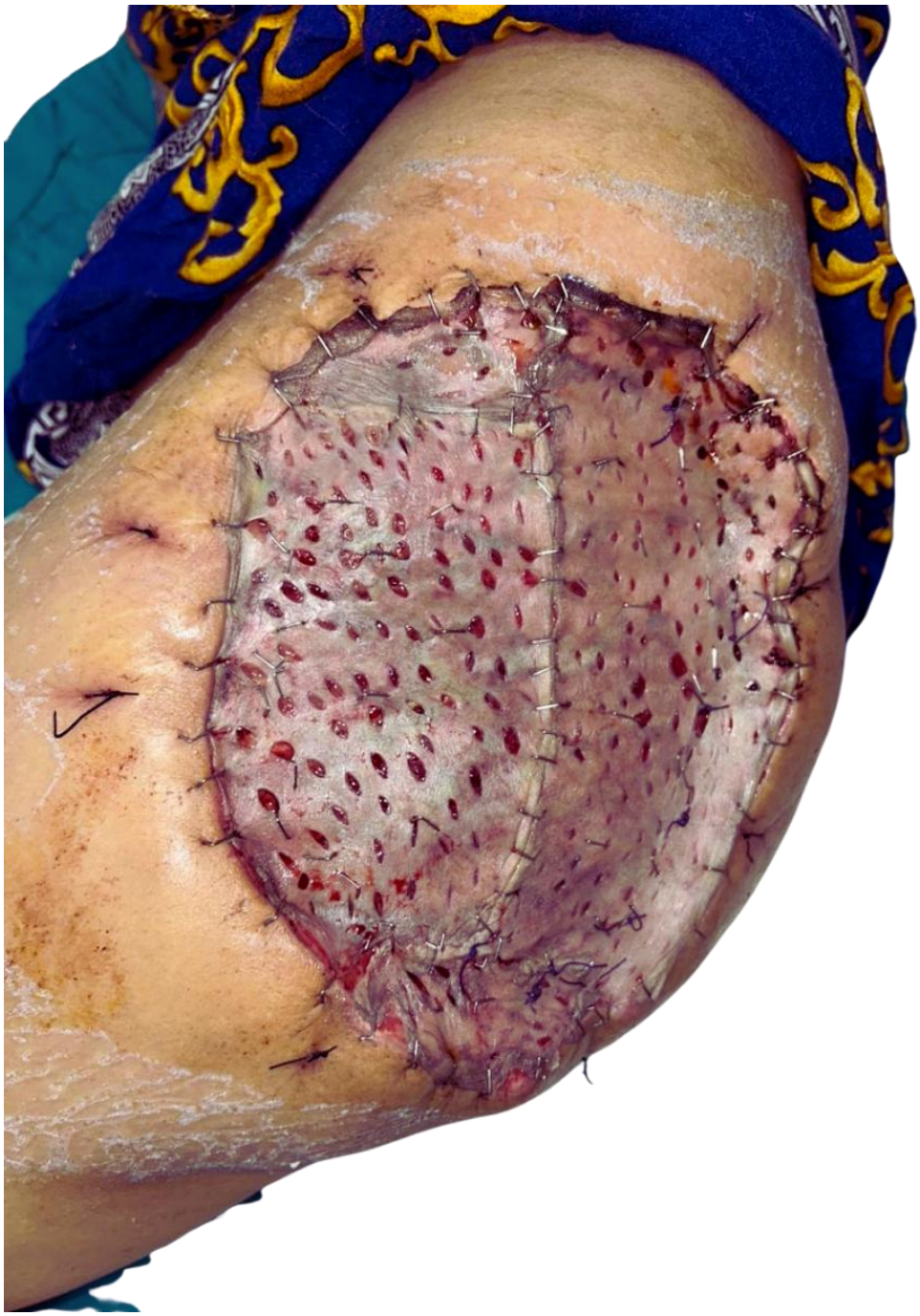

Figure 6 Macroscopic appearance after split-thickness skin graft over the tissue defect

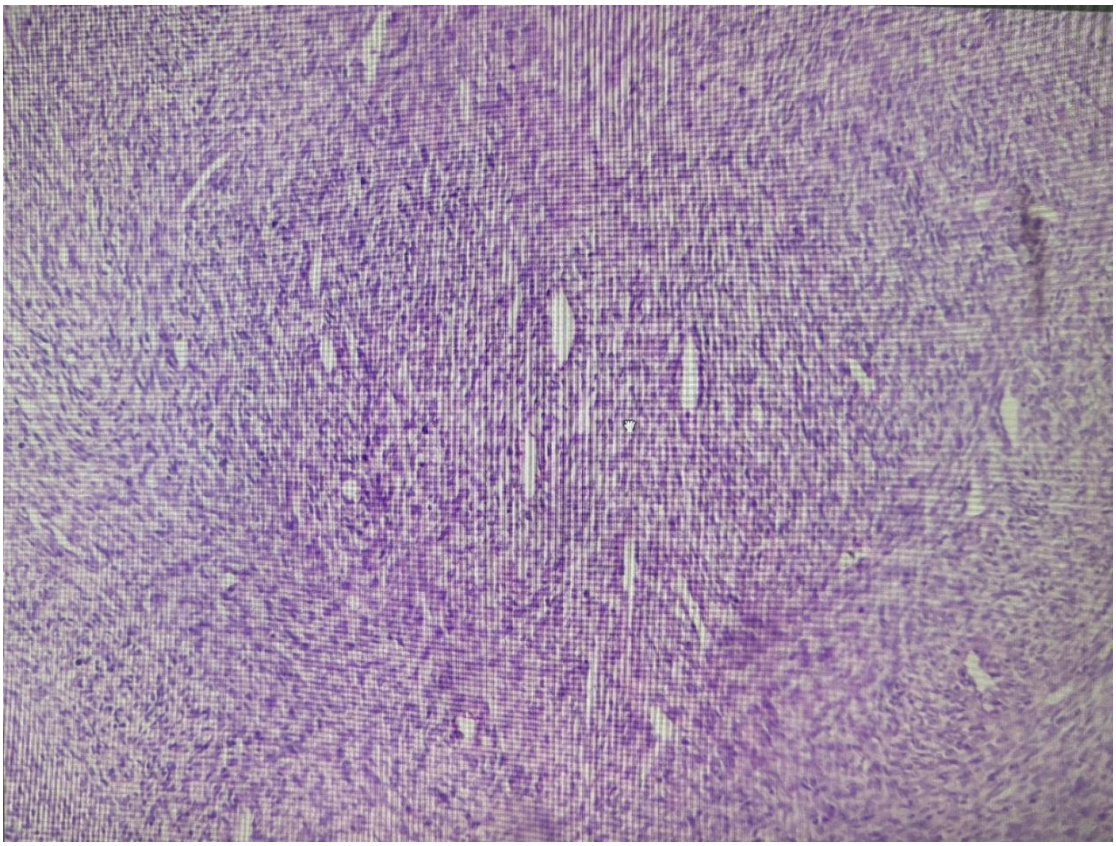

Figure 7 ×10 magnification showing hypercellular transformed areas

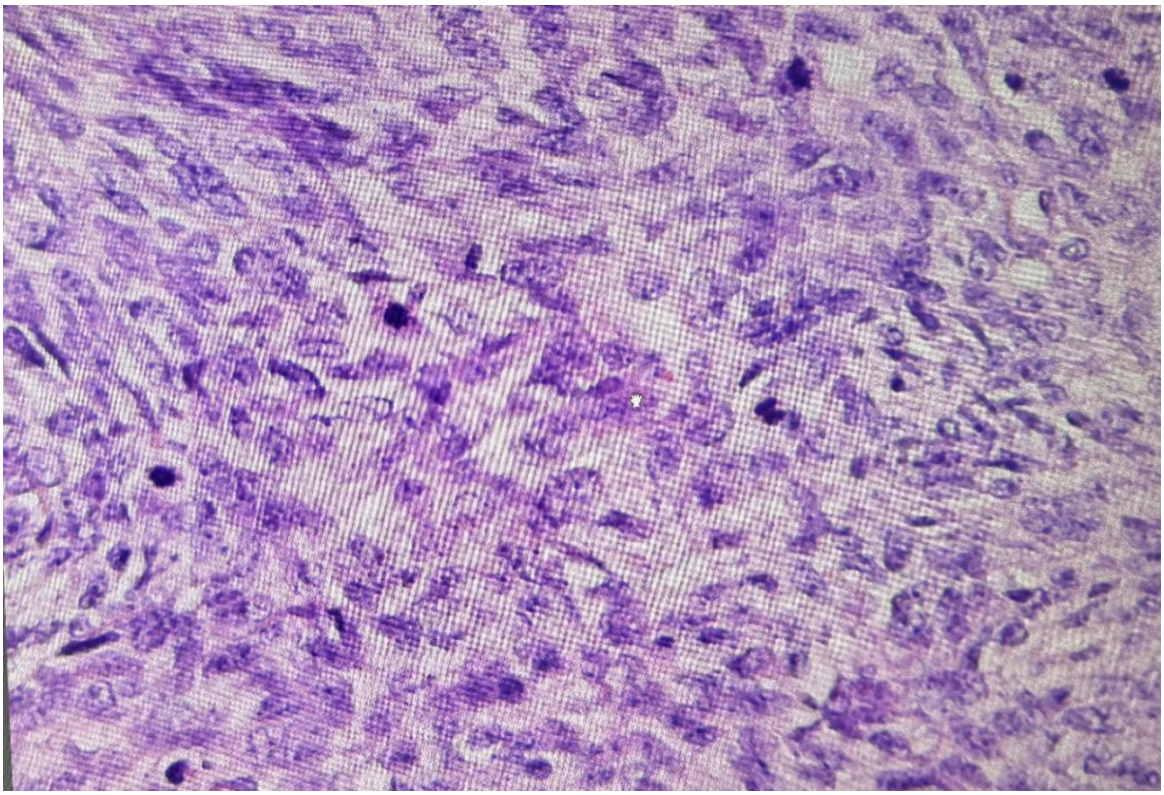

Figure 8 High magnification (×40) showing very high mitotic activity in the hypercellular areas

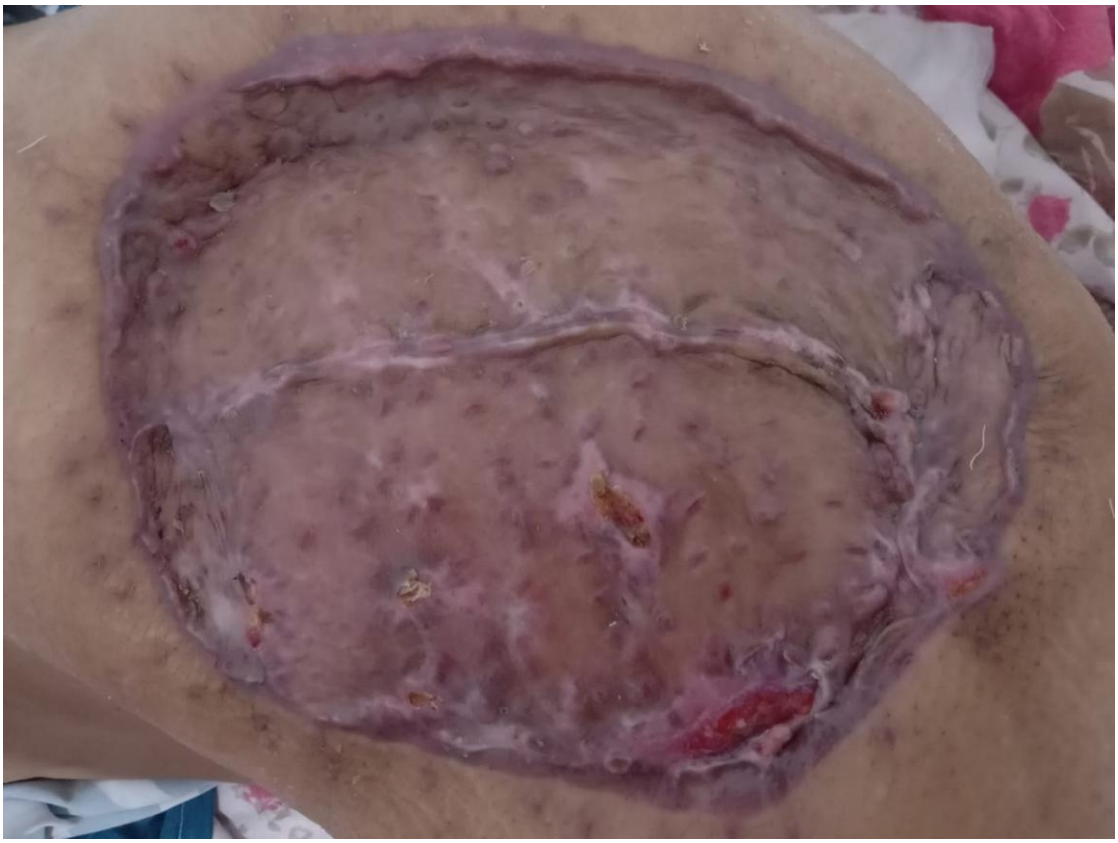

Figure 9 Macroscopic appearance showing the split-thickness skin graft

III. Discussion

Cutaneous sarcomas account for less than 1% of all malignant neoplasia.

Dermato-fibrosarcoma protuberans (DFSP) generally occurs in young adults, with a predominance of onset between the ages of 20 and 40. A slight male prevalence is observed. Transformation of DFSP into fibrosarcoma (FS-DFSP) remains an extremely rare event. Since the first description of a metastatic case of DFSP by Penner in 1953, fewer than 120 cases of FS-DFSP have been documented in the medical literature (3).

Dermatofibrosarcoma protuberans (DFSP) preferentially localizes in the trunk (47%), upper limbs (38%) and cervicocephalic region (15%). The tumor typically begins as a single, locally indurated, reddish-brown to purplish swelling. After several years, sometimes decades, protuberant nodules may gradually emerge. After a prolonged phase of superficial growth, the tumor infiltrates deeper structures, following fascial planes.

No significant clinical differences were observed between the classic form of DFSP and its transformation into a fibrosarcoma (FS-DFSP). Histologically, FS-DFSP is characterized by a proliferation of spindle-shaped cells with little atypia or mitosis. Peripherally, tumor cells are organized in wavy, flexuous bundles with no parallel orientation. At depth, the tumor infiltrates the subdermal tissues, dissociating the fatty lobules and adopting an interlobular septal organization. It may also extend from the primary lesion to invade the underlying fascia or fascial layer.

FSD tumor cells express CD34 in 90% of cases, and factor XIII. PS100 is not expressed (3).

FS-DFSP is defined histologically by the presence in a cellular DFSP of areas oriented towards an FS, the transition being either abrupt or progressive. The cellular areas are characterized by a fascicular herringbone architecture and high mitotic activity.

Surgery is a key component in the curative treatment of DFSP. In over 90% of cases, tumour control is achieved by two surgical techniques: conventional wide excision and Mohs micrographic surgery, which allows tumor excision with margin reduction under histological control of the absence of tumour cells from the excision margins (5,6).

Adjuvant radiotherapy can be used to achieve local control when resection margins are microscopically positive, or when wide excision results in significant functional or aesthetic deficit (7). The locoregional recurrence rate of dermatofibrosarcoma-derived fibrosarcoma protuberans (FS-DFSP) is closely related to the quality of surgical resection, with no significant difference compared to the classic form of DFSP.

However, the interval to recurrence tends to be shorter in FS-DFSP, generally less than two years, compared with the traditional form (8)

A number of factors contributing to poor prognosis have been identified in the various series published: incomplete excision, location in the cephalic extremity, where the requirements of wide excision are more difficult to meet, the existence of fibrosarcoma patches within the tumor, and tumor depth [8,9].

A largescale study published by Mentzel et al[10] demonstrated an association between the presence of fibrosarcomatous components and an increased risk of metastasis. In this cohort, around 15% of patients developed metastases, mainly in the lung, with no significant correlation with conventional prognostic factors such as tumor size, mitotic index or the presence of tumor necrosis.

IV. Conclusion

Dermatofibrosarcoma protuberans (DFSP) is a rare skin tumor characterized by slow local growth over several years.

The disease is difficult to diagnose, has a high recurrence rate and rarely metastasizes, mainly to the lungs. Diagnosis is usually based on clinical examination and confirmed by histological analysis. In case of doubt, immunohistochemistry may be used to detect CD34 antigen expression.

Treatment is based on surgical excision, with the aim of achieving a wide resection including a margin of 3 to 5 cm, depending on the location and whether the tumour is primary or recurrent. Deep excision must include any healthy tissue. Management of the resulting defect requires appropriate reconstruction to ensure adequate coverage.

Transformation of a dermatofibrosarcoma protuberans (DFSP) into a fibrosarcoma (FS) remains a very rare event. This evolution is associated with a significant metastatic risk, estimated at around 15%. If a fibrosarcomatous component is detected, a full extension workup is required, together with close clinical and radiological follow-up.

References

[1]. Marks LB, Suit HD, Rosenberg AE, Wood WC. Dermatofibrosarcoma protuberans treated with radiation therapy. *Int J Radiat Oncol Biol Phys*. 1989;17(2):379-384. doi:https://doi.org/10.1016/0360-3016(89)90454-9

[2]. Rutgers E, Kroon B, Albus-Lutter C, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. *Eur J Surg Oncol J Eur Soc Surg Oncol Br Assoc Surg Oncol*. 1992;18(3):241-248.

[3]. Thway K, Noujaim J, Jones RL, Fisher C. Dermatofibrosarcoma protuberans: pathology, genetics, and potential therapeutic strategies. Ann Diagn Pathol. 2016 Dec;25:64-71. doi: 10.1016/j.anndiagpath.2016.09.013. Epub 2016 Sep 27. PMID: 27806849.

[4]. Angouridakis N, Kafas P, Jerjes W, et al. Dermatofibrosarcoma protuberans with fibrosarcomatous transformation of the head and neck. *Head Neck Oncol*. 2011;3:1-7. doi:10.1186/1758-3284-3-5

[5]. Nouri K, Lodha R, Jimenez G, Robins P. Mohs micrographic surgery for dermatofibrosarcoma protuberans: University of Miami and NYU experience. *Dermatol Surg*. 2002;28(11):1060-1064. doi:28(11):p 1060-1064

[6]. Franks JW. A precision machine for mounting tissue for Mohs micrographic surgery. *Dermatol Surg*. 1998;24(9):989-993. doi:10.1111/j.1524-4725.1998.tb04292.x

[7]. Krishnamurthy A, Majhi U. Fibrosarcomatous dermatofibrosarcoma protuberans: An unusual tumor in the facial skin. *Indian J Dermatol*. 2014;59(1):105. doi:10.4103/0019-5154.123540

[8]. Farma JM, Ammori JB, Zager JS, et al. Dermatofibrosarcoma protuberans: how wide should we resect? *Ann Surg Oncol*. 2010;17:2112-2118. doi:10.1245/s10434-010-1046-8.

[9]. *Mentzel T, Beham A, Katenkamp D, Dei Tos AP, Fletcher CD. Fibrosarcomatous ("high-grade") dermatofibrosarcoma protuberans: Clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance. Am J Surg Pathol. 1998;22:576–87. doi: 10.1097/00000478-199805000-00009. [DOI] [PubMed] [Google Scholar]*