

Dermatofibrosarcoma Protuberans Arising From a Keloid-like Benign Sclerotic Plaque

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is an intermediate-grade tumor originating from the dermal fibroblasts. It's a locally invasive tumor with a potential of distant site metastasis. It usually forms after a long plaque which can be in an atrophic or a keloid-like sclerotic character. The lesion very rarely settles in the breast. Here we report a 52 year-old Caucasian woman with a dermatofibrosarcoma protuberans in her right breast. Patient's lesion arose from a long time keloid-like scleroatrophic looking plaque. Histopathologic examination did not reveal any malignant cells in the sclerotic plaque where the nodule arose from. Cells from nodular part of the lesion were CD34 positive immunohistochemically but there was no positive staining in the sclerotic plaque. The lesion was excised together with the pectoralis fascia. For reconstruction, an elliptically shaped latissimus dorsi musculocutaneous flap was designed. There were no recurrences in the postoperative 2nd year follow-up. To our knowledge this is the first reported case of dermatofibrosarcoma protuberans of a breast arising from a benign keloid-like sclerotic plaque.

Introduction

DFSP can be encountered in various parts of the body, but mainly from the trunk, extremities, and the neck region [1]. DFSP is typified by an initial plaque stage and a subsequent nodular stage. These plaques sometimes may be atrophic or sclerotic [1, 2].

Case Report

A-46-year old Caucasian woman was admitted due to a slowly growing mass in the upper inner quadrant of her right breast. The lesion first appeared 10 years ago as a small, red macula. The lesion enlarged, hardened and discolored slowly in time. For the last 4 years, inferior end of the lesion protruded and became nodular. Dermatologic examination revealed a 6x4 cm light livid coloured, atrophic looking, keloid-like sclerotic plaque fixed

to the underlying breast tissue and a brown-red colored, and a painless 2x2.5 cm nodule arising from the inferior tip of this plaque in the upper medial quadrant of the right breast [Figure 1A]. There were no other masses in the breast. Other systematic examinations did not reveal any other abnormalities. There was no lymphadenopathy. Total blood count, erythrocyte sedimentation rate and laboratory results were all within normal ranges.

T2-weighted MRI sequences demonstrated a hyperintense mass extending from the skin through the breast parenchyma [Figure 1B]. Thereafter, a tru-cut biopsy was performed where pathologic examination revealed monomorphic spindle shaped cells. Proliferating cells were extended to the underlying fatty tissue forming a lace-like pattern. These spindle cells were strongly positive for CD34 at the nodular region. Pathologic diagnosis was a DFPS. On the other hand, there

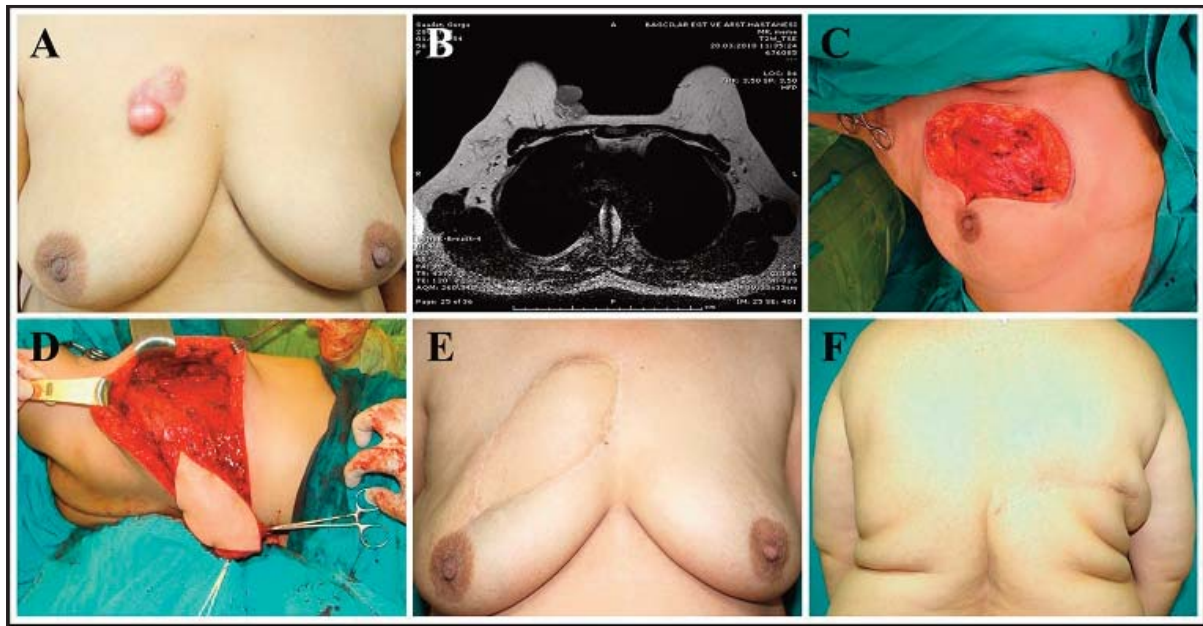


Figure 1. 1A. A 2x2.5cm nodular lesion on the right upper medial quadrant of the breast.
 1B. MRI view of the patient. Note the skin based well-defined mass protruding in the breast parenchyma.
 1C. The defect following excision of the tumor with 5-cm safety margins.
 1D. Harvesting of the latissimus dorsi musculocutaneous flap.
 1E, F. Post-operative photograph of the patient 2-years postoperatively without a recurrence and a satisfactory aesthetic result.

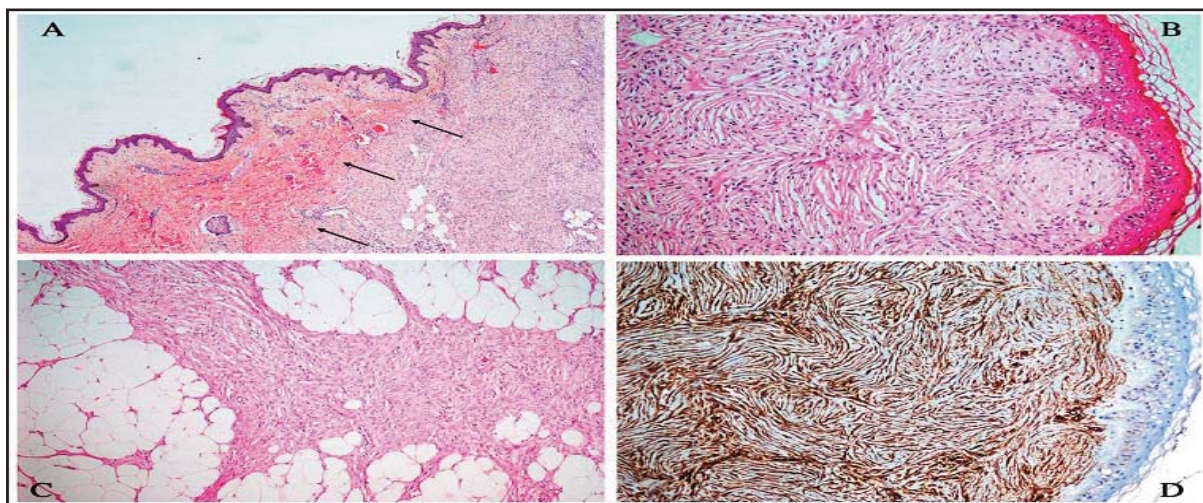


Figure 2. 2A. Spread of the tumor preexisting sclerotic area (40xHE)
 2B. Spindle cells arranged in storiform pattern (200xHE)
 2C. At the deep margin the tumor infiltrating fat tissue (100xHE)
 2D. Strong CD 34 immunoreactivity in dermatofibrosarcoma protuberans (200xCD34)

was significant sclerosis in the plaque starting from boundary of the nodule and expanding peripherally. It was negative for CD34 (Figure 2A, B, C, D).

The lesion was excised with 5-cm safety margins, including the pectoralis fascia. For reconstruction, an elliptically shaped latissimus dorsi musculocutaneous flap was designed (Figure 1C, 1D). At postoperative 2nd year follow-up patient was

recurrence free and satisfied with the aesthetic results [Figure 1E, F].

Discussion

DFSP first described by *Darier and Ferrand* in 1924 as “progressive recurrent dermatofibroma” is a locally aggressive stromal tumor with

potential of distant site metastasis [2, 3]. Its incidence is 4 per million. It is more common in blacks, females and 3rd&4th decades of life [1, 3]. Although most commonly seen in the trunk, extremities and neck, very rare locations like hands, feet, digits and breasts have been reported in literature [4, 5, 6]. It was reported that lesions may arise from burn scars, vaccination sites or previously traumatized regions or very rarely from old radiotherapy sites [7, 8]. In our case, age and gender was consistent with the previously reported studies. She had no predisposing lesions or history of trauma.

DFSP usually starts as a slowly growing plaque and it's rarely atrophic [1, 2]. Martin et al. described four different clinical appearances of early stage DFSP and these are confluent nodular lesions, keloid-like sclerotic plaques, atrophic plaques and tumoral lesions [2]. Although these very early stage lesions have different clinical appearances, they share similar epidemiological, histological and behavioral characteristics with protuberant lesions. Most of the lesions raise and become apparent after a long time. In the following period some of them may become painful and ulcerated [1]. In our case, the lesion started as a small plaque 10 years prior to admission. Afterwards, it was discolored and the base became rigid. Inferior tip of this scleroatrophic looking hard plaque became nodular. DFSP arising from an atrophic plaque are uncommon. Atrophic plaques can be the early lesions in the growth process of DFSP [3]. There are a number of DFSP cases arising from breast in the literature [3, 9, 10, 11]. First DFSP case of the breast which started as an atrophic plaque was reported by Cavuşoğlu et al. [12]. Our case's nodular lesion also arose from a scleroatrophic-looking plaque and there were no predisposing skin conditions. This successive development was consistent with the literature.

Ultrasonography, mammography and magnetic resonance imaging (MRI) are useful to reveal the extent of the disease. In mammography, it's usually seen as a dense lesion without fat density or calcification. USG helps to reveal dermis or subcutaneous tissue invasion of the lesion and MRI can help to detect the thickness of tumor infiltration [3]. MRI was used in our patient to assess invasion and it showed a hyperintense nodular mass, protruding into the breast parenchyma. However, definitive di-

agnosis depends on the pathologic examination due to the resemblance of the lesion with other skin tumors [3, 4].

In histopathological examination, dermis and subcutaneous tissue are replaced by bundles of uniform, spindle-shaped cells with little cytoplasm and elongated hyperchromatic nuclei. When there is deeper involvement, tumor cells infiltrate widely between collagen bundles of the deeper dermis and blend into the normal dermis that is described as a "storiform" pattern. Subcutis is extensively infiltrated and replaced in a typical lace-like pattern [3, 13]. Usually there is low mitosis [2].

Although the lesion was scleroatrophic looking, there were no epidermal or dermal atrophy in histopathological examination but there was a sclerotic plaque, starting from the nodular boundary and extending into periphery. Monomorphic spindle shaped cells formed a lace-like pattern. Most but not all of DFSP specimens are strongly positive for CD34 [13, 14, 15]. Our patient's lesion was also strongly positive for CD34. Nodular site consisted CD34 positive cells. However the lesion starting from nodule-plaque boundary and extending into the periphery had densely populated fibroblasts but it was CD34 negative and lacking spindle cells. Therefore this plaque was different from the previously reported early stage DFSP lesions described as "keloid-like sclerotic". To our knowledge there is no other DFSP cases reported in the literature that arose from a sclerotic plaque with benign characteristics.

It's very important to differentiate DFSP from other tumors in the early plaque phase. Morphea, morpheaform basaloma, atrophic dermatofibroma, atrophic scar, lipoatrophy, atrophoderma, anetoderma, neurofibroma, steroid atrophy, benign fibrous histiocytoma, keloid ve medallion-like dermal dendrocyte hamartoma should be considered [1, 2, 3, 15]. The nodular lesions that should be considered in differential diagnosis are dermatofibroma, fibrosarcoma, malignant fibrohistiocytoma, solitary fibrous tumor, Kaposi sarcoma and desmoplastic melanoma [3, 6, 14]. Different immunohistochemical staining properties of DFSP helps to rule out other lesions [16].

The mainstay of the treatment of DFSP is wide excision. Due to a relatively considerable risk of a recurrence, excision margins of even 5 cm has been recommended [17]. In addition, ra-

diotherapy can be used for recurrences or as an adjunct to the surgery [18]. Recently, an immunomodulator agent, a thryosine kinase receptor, imatinib has been proposed as a new treatment option for patients inamenable to surgery [1]. Although wide excision has been the mainstay of the treatment, it can cause aesthetic concerns, particularly for the breasts. Female breast DFSP cases have been reported previously. However, not much has been emphasized about aesthetic outcomes following tumor excisions. For example, Park et al., emphasized the reconstructive challenge of the breast DFSP following a wide excision [19]. In a case presented by Cavusoglu et al., reconstruction was achieved by an altered key-hole pattern, a technique commonly used in aesthetic breast reductions. For equalizing the breasts, augmentations on the tumor-excised breast or reductions on the contralateral breast were considered. They proposed other breast reduction patterns depending on the lesion localizations, as well [12].

In the case we presented, the tumor excision resulted a large defect requiring a reconstruction instead of performing a 'breast reduction type' of tumor excision pattern. For this purpose, we used the latissimus dorsi muscle-skin flap.

In conclusion, it is reported that DFSP can start as a keloid-like long-lasting plaque. However these early-stage plaques that reported previously, had shared similar histological characteristics with malign nodular lesions as mentioned above. Because of the nodule of our patient arose from surface of the keloid-like benign sclerotic plaque, it was different from the previous ones. To our knowledge, the patient is the first reported case with these properties in literature.

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