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Giant Dermatofibrosarcoma Protuberans Originating from a Keloid Scar-A Case Report and Review

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous sarcoma derived from dermal fibroblasts and accounts for 1-6% of all cutaneous soft tissue sarcomas. These low-grade tumors typically follow an indolent early course marked by slow growth but have a small potential to exhibit rapid growth with high-grade transformation. DFSPs are clinically similar to benign fibro proliferative skin tumors such as keloids, which may lead to misdiagnosis and delays in treatment. We present a case of DFSP originating from a keloid scar on the right breast. Here, we highlight the difficulty of timely diagnosis and need for multidisciplinary care. Excision of large tumors often leave significant defects requiring local tissue rearrangements, skin grafts, or complex reconstruction. Further, we review pertinent literature and outline the nuanced challenges of DFSP management.

Keywords: Case Report; Dermatofibrosarcoma Protuberans; Keloid; Surgical Resection; Literature Review.

Introduction

Although rare, dermatofibrosarcoma protuberans (DFSP) is the second most common cutaneous soft tissue sarcoma. It is derived from dermal fibroblasts and typically has low to moderate nuclear grade and low metastatic potential. However, they can be locally aggressive and have a propensity for local recurrence [1]. DFSPs often present as asymptomatic, slow-growing, flesh-coloured plaques or nodules with a 2-5% chance of fibrosarcomatous transformation (FS), hallmarked by rapid enlargement, marked infiltration and distant metastasis [1,2]. Unfortunately, DFSP can mimic various pathologies, including abscesses, schwannoma, cutaneous neurofibroma, dermatofibroma, lipoma, and keloids [3]. Because of low clinical suspicion, the diagnosis of DFSP can be challenging, leading to delays in definitive diagnosis. In this case report, we present an example of a DFSP arising from a keloid that evaded timely diagnosis and management.

Case Presentation

A healthy 29-year-old Black American female presented to our outpatient clinic with a painful, progressively enlarging right breast mass. She reported that the mass had originated from a keloid, a

result of a stab wound to her right chest ten years ago. Five years ago, she noticed a change in the keloid's appearance as it started enlarging. Over the next four years, it continued to grow, causing her significant pain and leading to multiple emergency department (ED) visits. She sought medical attention one year before her surgical oncology referral. She was initially given a course of antibiotics, followed by a targeted right breast ultrasound that revealed a 6.6 x 4.7 x 5.9 cm well-circumscribed, heterogeneous mass with internal vascularity, underlying edema, and involvement of the overlying skin. She was referred to a plastic surgeon for keloid excision, but the surgical intervention was delayed until she committed to smoking cessation. She returned to the ED months later with uncontrolled pain and a large, fungating mass with serosanguinous drainage (Figure 1A).

She was referred to our surgical oncology office, and a core needle biopsy was performed. A repeat-targeted right breast ultrasound demonstrated a well-circumscribed, hypervascular mass with similar echogenic features, now measuring 11.2 x 5.7 x 8.0 cm. Histopathology from the core needle biopsy showed spindle cells arranged in short fascicles with variable cellularity in a richly vascular stroma (Figure 2A and B). Atypical mitoses were present, 2 per 10 high power fields. Immunohistochemistry revealed that the tumor cells were patchy and positive for CD34 (Figure 2C) and negative for desmin, smooth muscle actin, and ALK. STAT6 and MUC4 were also negative, ruling out a solitary fibrous tumor and low-grade fibromyxoid sarcoma, respectively. Carcinoma was excluded with negative staining for cytokeratin AE1/AE3, cytokeratin 5/6, and p63. Interestingly, the tumor cells were focally positive for SOX10 (Figure 2D) but negative for Melan-A. Beta-catenin exhibited wild type membranocytoplasmic staining pattern (not shown), against desmoid-type fibromatosis.

Given the morphologic and immunophenotypic features, including weak CD34 and unexpected SOX10 expression, along with the rapid growth, an intermediate-grade spindle cell sarcoma with broad differentials were considered, including DFSP with fibrosarcomatous transformation and NTRK-rearranged neoplasms. However, when in-house next-generation sequencing (NGS) studies (based on Thermo Fisher Scientific Oncomine Comprehensive Assay v3) were performed, no NTRK rearrangements or other pathognomonic alterations detected by the panel were observed. Because of the lack of a definitive diagnosis, further investigation with a comprehensive NGS assay (CARIS MI Profile Comprehensive Testing Platform)

Computed tomography (CT) of the chest with intravenous contrast showed extensive skin involvement as well as extension close to the pectoralis major muscle, but without signs of metastatic disease (Figure 3A). Her case was discussed during our multidisciplinary tumor board. Considering that she had a high-grade, rapidly enlarging sarcoma of the right chest wall with a recent ED visit for bleeding from the mass, we recommended preoperative radiation therapy with 50 Gy in 25 fractions. She returned to the clinic for operative management after completion of radiation. The mass was unchanged in size and appeared more necrotic with continued serous drainage and foul odor (Figure 1B). Restaging CT showed stable size of the mass and no evidence of metastatic disease (Figure 3B). Comprehensive NGS profiling revealed a COL1A1::PDGFB pathologic fusion, confirming the tumor origin as a DFSP. Given the size and location of the mass, a wide local resection with a right mastectomy was planned. The patient requested to undergo a contralateral mastectomy without reconstruction. Given the expected large defect size, the plastic surgery team was enlisted to perform the right-sided wound closure. She underwent surgical intervention six weeks after the completion of radiotherapy.

A large defect was made to ensure wide margins; the superior aspect was to the level of the right clavicle (Figure 4A). The wound was not able to be closed primarily. Local tissue rearrangements (LTRs) were designed using the upper abdomen (Ryan flap) and the lateral chest skin [4]. A back-cut was made along the inferolateral chest wall, and progressive tension sutures were placed to reduce tension on the closure. Perfusion to the skin was evaluated using laser-assisted indocyanine green angiography via the SPY Intraoperative Perfusion Assessment System (manufactured by Novadaq Technologies Inc., Richmond, British Columbia, Canada). An additional margin of lower breast skin that was not well perfused was sharply excised and sent to pathology as an additional specimen. The resulting soft tissue defect required additional local tissue rearrangement, and the skin over the lateral chest was dissected to the border of the latissimus dorsi. Progressive tension sutures were placed, and skin perfusion was re-evaluated using SPY. The pocket was opened and irrigated, and two drains were placed. The skin was then closed using layered absorbable sutures and non-absorbable sutures to the skin for reinforcement (Figure 4B). The incisions and skin flaps were coated with a thin layer of topical nitroglycerin paste and covered with Tegaderm dressings.

Gross examination of the mastectomy specimen reveals a large, firm, tan-white to tan-gray fun gating mass protruding through the dermis (Figure 5). The subcutaneous cut surface of the lesion is heterogeneous, tan-white, and well-circumscribed, grossly involving the epidermis and extending into the subcutaneous tissue and approaching the surgical margins. Histologic examination revealed a 14.4 cm post-treatment DFSP with a suboptimal treatment effect of approximately 25%. Foci of incipient necrosis were present, but overall reduced atypia and cellularity apparent in the post-treatment sample as compared to the core biopsy (Figure 2E-F) such that no definitive residual fibrosarcomatous component remained post-treatment. However, a small segment of the anterior superior margin was positive. Ten axillary lymph nodes were retrieved from the final specimen, all of which were negative for

metastases. She was discharged home on the first postoperative day and was last seen during her 1-month follow-up, revealing a well-healing wound with no signs of infection, hematoma, or lymphedema (Figure 6). She now continues with close clinical follow-up.

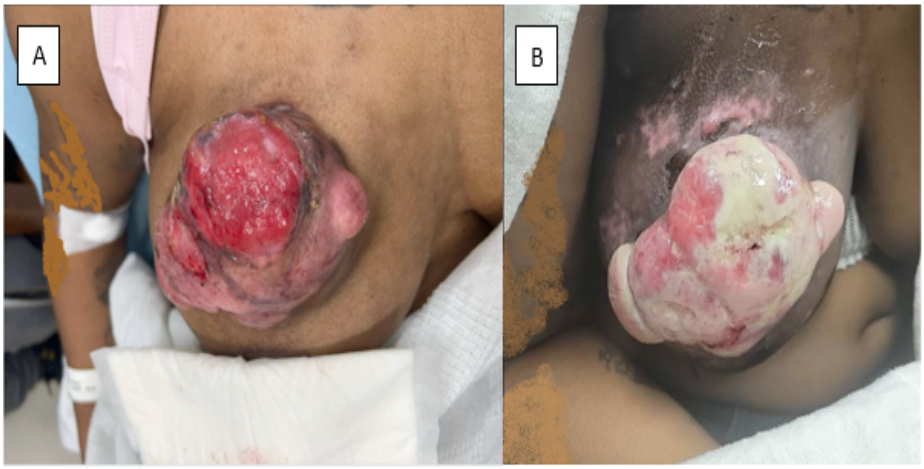

Figure 1: Large dermatofibrosarcoma protuberans of the right breast. (A) DFSP that was initially mistaken for a keloid. (B) Post-radiation treatment changes of the right breast DFSP. Images were altered to de-identify the patient.

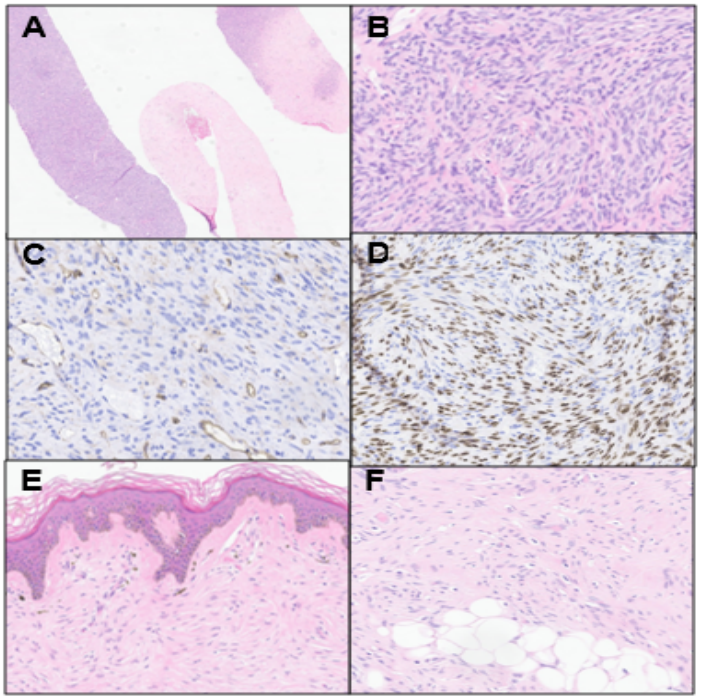

Figure 2: (A) Low-power view (4x) of the biopsy specimen showing predominantly viable spindle-shaped tumor within a richly vascular stroma and areas of sclerosis. (B) Higher magnification (40x) reveals tumor cells arranged in short fascicles with variable cellularity. (C) Immunohistochemistry demonstrates focal loss of CD34 expression in the tumor cells (40x). (D) Tumor cells exhibit focal expression of SOX10 (40x). (E) Histologic examination of the post treatment mastectomy specimen shows the tumor involving the dermis (20x). (F) The tumor extends to the deep dermis and subcutaneous tissue, showing a similar fascicular pattern of tumor cells as observed in the initial biopsy, but with less atypia and cellularity (40x).

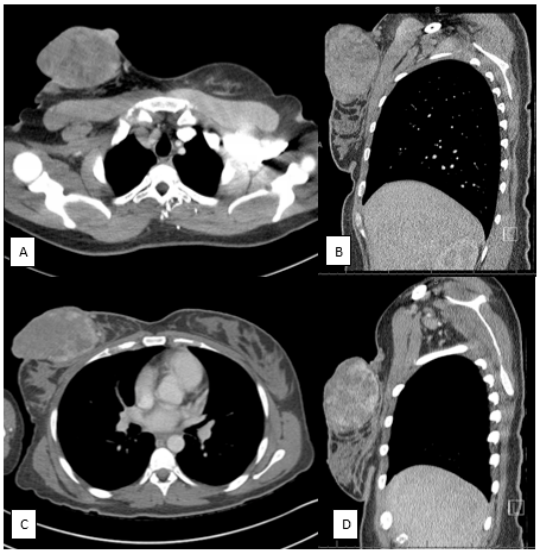

Figure 3: Computed Tomography of right breast DFSP. (A) Axial and (B) sagittal images depicting DFSP before radiation therapy. Repeat (C) axial and (D) sagittal images taken after completion of radiation therapy showing the stable size of the tumor.

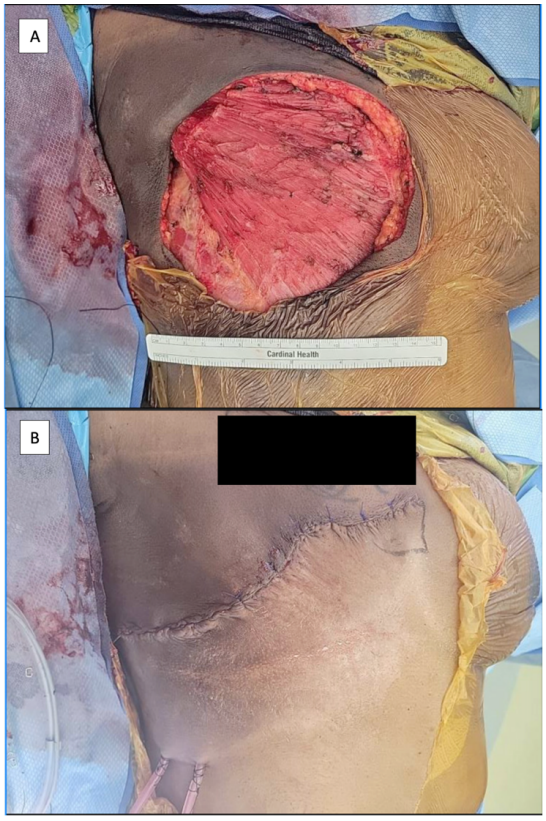

Figure 4: A) Right chest defect with exposed pectoralis muscle after dermatofibrosarcoma protuberans resection. B) Right chest wall closure after removal of additional non-viable skin and extensive adjacent tissue transfer. Image was edited to de-identify the patient

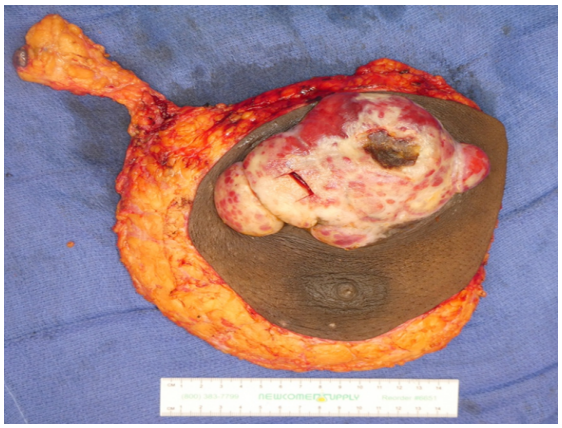

Figure 5: Gross right mastectomy specimen depicting the Dermatofibrosarcoma protuberans.

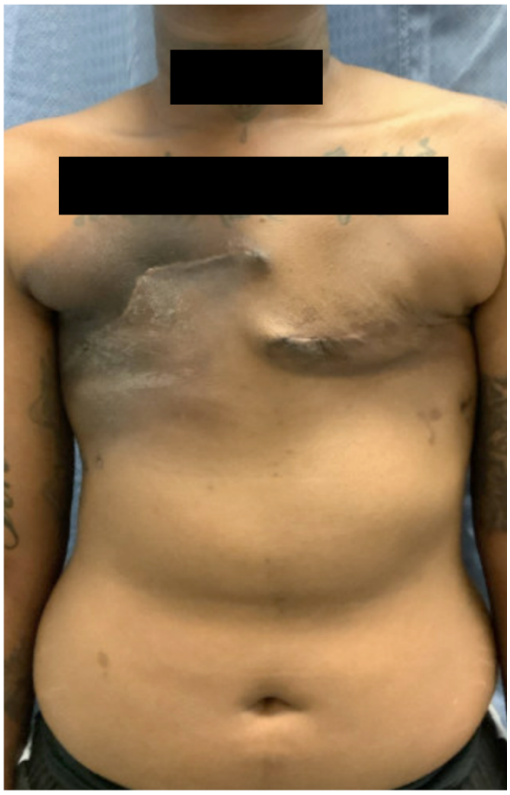

Figure 6: Two-month post-operative image of right wide local excision with local tissue rearrangement and left mastectomy closure. Image was edited to de-identify the patient.

Discussion

Dermatofibrosarcoma was first described by Danrier and Ferrand-Drake in 1924 as a progressive recurring dermatofibroma but was defined as a keloid sarcoma by Hoffman in 1925 [5,6]. Although DFSPs are rare, representing 0.1% of all cutaneous cancer diagnoses, they account for 1-6% of soft tissue sarcomas and 18% of cutaneous soft tissue sarcomas, with an incidence of approximately 4.2 cases per million people in the United States [7-10]. It commonly affects individuals of African descent, with a peak incidence between the ages of 20 and 50, and has a roughly equal sex distribution [5,8,11]. The clinical risk factors are not well understood and are thought to be associated with prior trauma or scar. Most occur on the trunk (40-50%) but can also occur on the extremities (30-40%), head, and neck (10-16%) [1,12]. Clinically, they present as flesh-coloured, pink, or violaceous protuberant plaques or nodules that progress slowly in radial and vertical directions. They usually develop as single lesions or with multiple satellite nodules [1,14]. Despite periods of prolonged size stability or stable growth, 2-5% can have sudden, rapid enlargement.

Delays in diagnosing DFSP are common, and patients may receive multiple treatments before a definitive diagnosis is made. Clinically, it is difficult to differentiate between DFSPs and keloids. Domonkes highlighted that "dermatofibrosarcoma protuberans is characterized by bulky, protuberant tumor masses that look like infected keloids [14]." Furthermore, Ju cautioned that a fibro sarcoma arising in a surgical scar might be mistaken for a keloid [15]. If there are atypical focal areas of firmness upon palpation or a concern about a keloid growing rapidly, DFSP should be considered as a potential diagnosis, and a biopsy is necessary for further evaluation [16].

In our case, a DFSP developed from a keloid scar. Reviewing the literature, there are few case reports documenting this phenomenon (Table 1), with some reporting up to 25 years before concerning clinical features arise. Our patient also presented with a long-standing keloid, which suddenly and rapidly progressed. Furthermore, there are a few documented cases of various sarcomas arising from keloids, including low-grade myofibroblastic sarcomas, undifferentiated or high-grade pleomorphic sarcomas, and epithelial sarcomas [21-24].

Keloids are primarily diagnosed clinically and occur in the same clinical context as DFSPs. Despite their clinical similarities, they are histologically distinct. Keloids typically have large, disorganized, hyalinized collagen bundles with haphazard, small, dilated blood vessels [16]. On the other hand, DFSPs are composed of uniform spindle cells with low mitotic rates and pleomorphisms. These cells are arranged in a storiform pattern and diffusely infiltrate the dermis and subcutaneous fat. They stain strongly positive for CD34 and Vimentin approximately 80-100% of the time, but the expression of CD34 can be seen in other mesenchymal tumors [25, 26]. There are multiple low-grade DFSP subtypes, including Giant cell fibroblastoma, pigmented (Bednar), atrophic, sclerosing, and myxoid. A small focus of fibrosarcomatous transformation (FS) can be seen in up to 20% of any subtype and is considered high grade, characterized by cytologic and nuclear atypia, increased cellularity and mitotic rate, loss of CD34 expression, and a fascicular "herringbone" growth pattern [26,27]. The SOX10 positivity observed in this case is of uncertain significance, though it has been recently observed in DFSP showing aberrant melanocytic differentiation [28]. Genetic testing can also increase diagnostic accuracy. Over 90% of DFSPs harbor a collagen type 1 alpha 1 and platelet-derived growth factor B-type (COL1A1-PDGFB) fusion gene resulting from a (17;22) (q22;q13) translocation, causing an upregulation of PDGFB and over activation of the PDGFR [29,30]. DFSP with FS has also been found to have additional p53 mutations, microsatellite instability, and additional copies of COL1A1-PDGFB [31].

Once confirmed, the cornerstone of management is an R0 resection. There is ongoing debate about the best surgical technique, with American and European guidelines favouring Mohs microsurgery (MSS) over wide local excision (WLE). The most recent National Comprehensive Cancer Network (NCCN) guidelines recommend using Mohs surgery or other forms of peripheral and deep enface margin assessment (PDEMA). If these techniques are unavailable, WLE can be considered [32]. The 2015 European consensus guidelines favour MSS over WLE but recommend 3 cm gross margins if WLE is used [33]. However, no level 1 evidence shows the superiority of MSS over conventional WLE [1,13]. Regardless of the approach, having an experienced dermatopathologist carefully assess the margins is crucial to ensure complete resection, thereby giving patients the best opportunity for local control. Patients with incomplete resections should undergo re-excision to obtain negative margins when possible. The soft tissue defect after extirpation of DFSP, particularly of the chest, can be substantial and often requires reconstruction with LTR or pedicled flaps. In some cases, mesh placement is required to reconstruct an anterior thoracic wall defect, and a free flap may be needed for soft tissue coverage [34,35]. Irrespective of the reconstruction method, it should only occur after confirming negative margins.

Table 1: Reports of patients with dermatofibrosarcoma protuberans arising from keloid scars and their management.

Author	Year of publication	Age at DFSP diagnosis (years)	Time from initial keloid diagnosis to development of DFSP (years)	Biopsy confirming prior keloid diagnosis	Size of DFSP (cm)	Location	Surgical Technique	Wound closure
Manalan et al. [18]	1974	29	3	no	Unkn	Anterior chest	WLE	STSG
Hardy [19]	1987	26	16	yes	Unkn	Neck	WLE	Unkn
Kimura et al. [20]	2014	46	25	no	10	Anterior chest	WLE	Reconstruction
Ward and Odili [2]	2018	41	10	no	3	Anterior chest	WLE	Reconstruction
Ahmed et al. [21]	2024	57	25	yes	4.5	Anterior chest	WLE	FTSG

DFSP = dermatofibrosarcoma protuberans, STSG = split thickness skin graft, FTSG = full thickness skin graft, WLE = wide local excision, Unkn = unknown

Although complete surgical resection is the mainstay of treatment for respectable disease, targeted therapy and radiation have been utilized for unresectable and metastatic disease. Conventional cytotoxic chemotherapy is ineffective for DFSP and has a minimal role in treatment. Imatinib mesylate, a tyrosine kinase inhibitor (TKI) with activity against PDGFR, is effective in treating unresectable and metastatic disease; its use for DFSP management was approved by the FDA in 2006. A recent systematic review by Henry et al. showed that 70 to 95% of patients treated with TKIs achieved complete response, partial response, or stable disease [36]. Similarly, Nacarrete-Dechent et al. showed that Imatinib could produce a complete and partial response rate of 50% to 60% [37]. TKIs have also shown promise in the neoadjuvant setting in phase II clinical trials, showing complete or partial response rates of 36 to 57% [38,39], and, in select cases, patients may be considered for curative surgical resection. The role of radiation therapy in managing DFSP is limited as there are mixed results on its effectiveness and small patient volumes [1,13,37]. The NCCN guidelines recommend adjuvant radiation therapy for managing positive margins, <1 cm margin in patients not undergoing PDEMA, and indeterminate or positive margins. Doses of 50-60 Gy are utilized, but higher doses of up to 66 Gy can be used for gross residual tumors with fields extending widely beyond the surgical margins if feasible [32].

After an R0 resection, the prognosis for DFSP is excellent, with a 15-year overall survival approaching 100% and a 10-year disease-specific survival of 99%. However, DFSP is locally aggressive, and approximately 11-50% of primary DFSPs recur locally, with a 33% recurrence rate even after additional resections. Despite the locally aggressive behaviour, most tumors are low-grade. As such, distant metastatic disease is rare, with a distant and regional metastatic rate of 1% and 6%, respectively [1,13]. Risk factors for recurrence include tumor depth, tumor size > 5 cm, positive margins, age over 50, and histologic subtypes, specifically FS. In a systematic review of 24 studies, DFSP with FS was associated with a worse prognosis compared to other subtypes [26]. While the Bednar subtype has the lowest recurrence rate of 11-13%, FS has a 55% local recurrence rate and a 14% incidence of metastatic disease [27,40]. Because of the high local recurrence rates, long-term follow-up is necessary. The 2015 European guidelines recommend clinical examinations every six months for five years, followed by annual exams until ten years post resection, with imaging reserved for DFSP with evidence of FS [35]. Similarly, the NCCN guidelines recommend a clinical exam every six to twelve months after resection with consideration of magnetic resonance imaging surveillance [34].

Conclusion

This case emphasizes the importance of the high degree of suspicion required for timely management of DFSP, as they can be misidentified as or develop from keloids. Differentiating DFSPs

from keloids requires a thorough clinical examination, including palpation and observing the lesion's growth pattern, followed by a biopsy for histological confirmation. DFSP should be considered in the differential diagnosis, especially when a keloid shows rapid growth after a long period of stability. Since DFSP is locally aggressive with a low potential for metastatic disease, it can often be treated with resection and careful margin assessment by an experienced dermatopathologist. However, the emergence of targeted therapy with TKIs has highlighted the necessity of a multidisciplinary approach to managing DFSP, emphasizing the importance of each professional's role in the process. Finally, close follow-up is essential, particularly in cases of FS.

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