

CASE REPORT

Lessons learned from an unusually aggressive recurrent dermatofibrosarcoma protuberans with chest wall involvement: A case report

Shoon Mya Aye^{1*}  and Wah Wah Myint Zu²

¹Myanmar Medical Association, Yangon, Myanmar

²Department of Radiation Oncology, Yangon General Hospital, Yangon, Myanmar

Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing soft-tissue sarcoma with low metastatic potential. This report describes an unusual case of aggressive, recurrent DFSP in a 50-year-old female, characterized by extensive chest wall involvement despite multiple surgeries, radiation, and targeted therapy. The clinical course was complicated by poor wound healing, infection, and severe ulceration. This case highlights the potential for aggressive local progression, the difficulty of achieving durable local control in recurrent DFSP, and the limitations of systemic therapies for locally advanced disease. The case also emphasizes the importance of a multidisciplinary approach, patient education, and palliative care, especially in resource-limited settings. This report underscores the need for continued research into new treatments for aggressive DFSP.

*Corresponding author:

Shoon Mya Aye
(drshoonmyaaye89@gmail.com)

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1. Background

Dermatofibrosarcoma protuberans (DFSP) is a rare soft-tissue sarcoma, often presenting as a firm, violet-red, or blue plaque on the trunk and extremities. Its slow growth frequently leads to delayed diagnosis, sometimes over several years.¹

DFSP generally occurs in patients between 30 and 50 years of age and affects both genders equally. This rare tumor accounts for a small minority of soft-tissue sarcomas (1–6%) and nearly one-fifth of cutaneous sarcomas. The underlying genetic driver is a characteristic translocation in which the *COL1A1* gene on chromosome 17 fuses with the *PDGFB* gene on chromosome 22, creating the *COL1A1-PDGFB* fusion protein. Other, less common translocations involving *PDGFB* have also been reported.¹

As an intermediate-grade tumor, DFSP rarely progresses to a high-grade sarcoma. Tumor cells, arranged in a storiform or “woven” pattern, infiltrate the subcutaneous tissue, often entrapping adipocytes. DFSP typically involves deeper structures, such as muscle or bone, only when recurrent. Lesions grow slowly and may resemble keloids or dermatofibromas; they can reach several centimeters in size and may be associated with pain, ulceration, and misdiagnosis. While typically fixed to the skin, the tumor can also extend into deeper tissues and may become fixed to underlying structures.

Nearly half of DFSP cases appear on the trunk, with fewer occurrences on the extremities or the head and neck. The diagnosis of DFSP is typically supported by diffuse immunohistochemical positivity for CD34.^{1,2}

The standard treatment for localized DFSP is surgical removal with clear margins, an approach guided by the tumor's size and location. Because of its low metastatic risk, lymph node dissection is generally not indicated. Delayed diagnosis and the tumor's infiltrative growth pattern often result in inadequate initial resections. Approximately 50% of patients experience local recurrence after simple excision, and recurrent tumors are more likely to invade deeper tissues and, in rare cases, metastasize. Both wide local excision and Mohs micrographic surgery (MMS) are effective options, with research indicating that MMS may offer better local control.¹

The National Comprehensive Cancer Network guidelines recommend adjuvant radiotherapy in the following settings:

- (i) Positive margins or gross disease.
- (ii) Negative but narrow margins, as judged by the treating physician when MMS is not used.
- (iii) Recurrent or metastatic disease, when further resection is not feasible.³

For unresectable, recurrent, or metastatic DFSP, imatinib is a targeted therapy that inhibits the platelet-derived growth factor receptor and other tyrosine kinases such as c-KIT. Its mechanism of action involves blocking ATP binding to the PDGFR receptor, thereby suppressing kinase activity, inhibiting tumor proliferation, and inducing cell death. The presence of the t(17;22)(q22;q13) translocation is associated with a more effective response to this drug. Common side effects include gastrointestinal issues, edema, fatigue, anemia, and rash. However, tumors may acquire resistance after initial imatinib treatment.^{1,4-6}

In imatinib-resistant DFSP, other treatments may be considered, including second- or third-generation tyrosine kinase inhibitors (TKIs), immunotherapy, or surgery. Several studies suggest that combining imatinib with immunotherapy may be an effective strategy.⁶ Sunitinib is a second-line targeted therapy, and other novel TKIs are being investigated for imatinib-refractory or progressive cases.^{6,7}

In some cases, radiotherapy has been reported to induce an abscopal effect, with antitumor activity observed outside the irradiated field in imatinib-resistant DFSP.⁸

Patients treated for DFSP should regularly perform self-examinations and have the primary site examined every 6–12 months.³ The overall prognosis is particularly favorable, with a reported 10-year survival rate of 99%.

However, once the disease has metastasized, survival is significantly reduced, with a mean survival of approximately 2 years.¹

2. Case presentation

A 50-year-old female was initially diagnosed in 2018 with a breast mass, later confirmed as DFSP, showing diffuse CD34 positivity. She underwent a total mastectomy followed by adjuvant chemotherapy with Adriamycin in a district hospital.

In 2019, a chest wall recurrence was identified. She received chest wall radiation therapy consisting of 45 Gy in 15 fractions, with a boost of 10 Gy in 5 fractions, resulting in complete regression. She was then lost to follow-up.

In early 2021, she presented with a chest wall nodule after applying traditional topical remedies, which was followed by a tetanus infection. After recovery from the wound infection, her primary attending surgeon decided to perform excision again. Histopathology confirmed recurrent DFSP, and surgical margins were positive; thus, imatinib 400 mg daily was initiated.

In September 2021, a 0.5-cm nodule reappeared and was then excised. She was subsequently referred to the palliative care team of the Myanmar Medical Association, Yangon, Myanmar, for further treatment. Imatinib 400 mg twice daily and chest wall intensity-modulated radiation therapy (50 Gy in 25 fractions) were administered (Figure 1).

In February 2023, a fourth recurrence developed as an ulcer over the chest wall scar. The patient, feeling discouraged, delayed seeking appropriate treatment and applied traditional topical remedies to the ulcer. Consequently, the ulcer enlarged, became infected with



Figure 1. The operated chest wall was irradiated with intensity-modulated radiation therapy (2021)

purulent discharge, and showed poor response despite culture-guided antibiotic therapy. Her consulting surgeon decided on another excision. The biopsy revealed recurrent DFSP, with no evidence of lymphovascular invasion. The ineffective imatinib was replaced with sunitinib 25 mg daily, resulting in improved ulcer healing and size reduction.

She was again lost to follow-up for approximately 7 months. In September 2023, she returned with a large, aggressive, infected chest wall ulcer with foul odor and cutaneous air leakage (Figure 2). Culture revealed *Citrobacter* species. Computed tomography imaging revealed a chest wall defect with a pleurocutaneous fistula and a small pneumothorax. Despite these findings, the patient—now experiencing significant psychological distress—declined further intervention and missed appointments.

In January 2024, she requested conservative treatment only and was transitioned to palliative care with supportive measures. Sunitinib 50 mg daily was restarted. Her wound remained frequently infected, and education regarding wound care, chest physiotherapy, and psychosocial support was provided to her and her caregivers.

Over the following year, the ulcer progressively enlarged, exposing the underlying ribs; a midsternal nodule and additional small nodules appeared on the surrounding skin (Figure 3). The patient experienced significant anxiety and pain. Pain management was initiated with tramadol 50 mg twice daily and gabapentin 100 mg at night. Medications were titrated according to her needs, and her pain was eventually controlled with morphine 5 mg every 6 h and as needed, along with pregabalin 75 mg twice daily. Psychological distress was addressed with counseling, citalopram, and alprazolam. Social support was provided for caregiving and financial assistance. However, providing comprehensive palliative care remained challenging due to limited resources in her remote village.

3. Discussion

This unusually aggressive case of recurrent DFSP, characterized by extensive chest wall involvement, provides several critical insights for clinicians managing this rare sarcoma.

This case illustrates that, despite typically being an intermediate-grade sarcoma with low metastatic risk, DFSP can exhibit highly aggressive local behavior, particularly after multiple recurrences. Incomplete surgical resection can lead to rapid regrowth, and a history of prior recurrence may further increase the risk of subsequent postoperative recurrence. Tumor location may also indirectly influence this risk.⁹ Larger tumor size is a significant risk factor for local recurrence.¹⁰ Fibrosarcomatous DFSP is a more



Figure 2. Large, aggressive chest wall ulcer on the right chest wall (2023)



Figure 3. Progressively aggressive ulcer with exposed ribs, a midsternal nodule, and small nodules on the surrounding skin (2025)

aggressive subtype, characterized by faster tumor cell growth and a higher degree of malignancy compared to typical DFSP.² The rapid, deep invasion into the chest wall, resulting in rib exposure and a pleurocutaneous fistula, highlights the potential for aggressive progression in this case.

Despite repeated surgeries and multiple courses of radiotherapy, including boost doses and intensity-modulated radiation therapy, durable local control was not attained. This highlights the difficulty of eradicating DFSP because of its infiltrative nature and the challenges of achieving clear margins in recurrent cases with distorted local anatomy.⁹

While imatinib initially provided a response, resistance ultimately developed. Sunitinib offered temporary local improvement but was unable to halt the aggressive progression. Even targeted therapies directed against the

COL1A1-PDGFB fusion may not ensure long-term control, especially in the setting of aggressive local spread.^{7,11}

The patient's use of traditional topical remedies contributed to complications and delayed access to appropriate care.¹² This emphasizes the need for patient education and timely access to evidence-based treatment.

Complications often arise in DFSP due to the extent of the surgical defect and the challenge of managing local recurrence. These include wound infections, difficulty with defect coverage requiring skin grafts or complex plastic reconstructive procedures, contractures, keloid formation, and poor cosmetic outcomes.¹³ The development of a pleurocutaneous fistula and pneumothorax as a consequence of the tumor's invasion into the chest wall highlights the potential for significant morbidity associated with locally advanced DFSP.

A collaborative, multidisciplinary approach involving surgical, radiation, and medical oncology, as well as chest and plastic surgery, is crucial for effectively managing complex cases and associated complications. This teamwork ensures the optimization of treatment strategies tailored to patient needs.¹⁴

Gaps in follow-up likely delayed timely treatment of recurrences, potentially contributing to locally advanced progression. Therefore, it is important to emphasize the need for consistent follow-up and strict adherence to treatment plans for the effective management of DFSP.¹

In advanced, refractory cases where intensive treatments are ineffective or declined, a comprehensive palliative care approach is particularly vital. Such care focuses on managing pain and symptoms, ensuring proper wound care, offering psychosocial support, and addressing emotional and spiritual needs. However, providing comprehensive palliative care in resource-constrained settings remains challenging because of limited access to adequate pain control, wound care supplies, psychosocial resources, and trained palliative care professionals. Addressing these gaps is particularly crucial.

Moreover, this case highlights the need for ongoing research to explore resistance mechanisms to targeted therapies and to develop new treatment strategies for aggressive and refractory DFSP, especially for patients who have exhausted standard therapeutic options.

4. Conclusion

This case serves as a stark reminder of the potential for aggressive local behavior in DFSP, the challenges in achieving durable control in recurrent disease, and the limitations of current systemic therapies in locally advanced cases. It also underscores the critical importance

of a comprehensive, patient-centered approach, including timely intervention and robust palliative care when curative options are exhausted.

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Conflict of interest

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Author contributions

Conceptualization: All authors

Formal analysis: Shoon Mya Aye

Investigation: Shoon Mya Aye

Methodology: Shoon Mya Aye

Writing—original draft: Shoon Mya Aye

Writing—review & editing: All authors

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent for publication was obtained from the patient.

Availability of data

Data used in this work are available from the corresponding author upon reasonable request.

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