JISS Journal of Indonesian Social Science

Vol. 6, No. 1, January 2026
E-ISSN:2723 – 6692
P-ISSN:2723– 6595
http://jiss.publikasiindonesia.id/

Delayed Presentation of Dermatofibrosarcoma Protuberans in a Rural Healthcare Facility: A Case Report

Jeffrey*, Giri Yurista
Rumah Sakit Umum Daerah RAA Soewondo Pati, Indonesia
Email: jeffrey0364@gmail.com

ARTICLE INFO

Keywords:
Dermatofibrosarcoma Protuberans , soft tissue tumors , skin sarcoma

ABSTRACT

Dermatofibrosarcoma Protuberans (*DFSP*) represents one of the rarest forms of soft tissue tumors, with an annual occurrence estimated at only 0.008 to 0.045 cases per 100,000 individuals. This rare, slow-growing but locally aggressive skin sarcoma often presents with subtle clinical features, leading to delayed diagnosis and management, particularly in resource-limited settings. In this report, we describe a case of *DFSP* that presented to a rural healthcare facility with delayed presentation. A 37-year-old woman was referred to the general surgery clinic with an enlarging lump in the calf of her left leg that had been present for the past 20 years. The lump had gradually increased in size and was initially painless. One month prior to hospitalization, the lesion became frequently painful, bled intermittently, and discharged pus. A wide excisional biopsy was performed, and the specimen was sent for histopathological analysis. The conclusion of the histopathological analysis was consistent with *DFSP*. This case highlights the diagnostic challenges of *DFSP* in resource-limited settings, where limited access to specialized diagnostic services, lack of awareness among primary healthcare providers, and financial constraints contribute to diagnostic delays. A long-standing lesion remained indolent before progressing to symptomatic disease, emphasizing the need for increased clinical vigilance in rural healthcare facilities. Early recognition and appropriate management are essential to improve patient outcomes.

Introduction

Dermatofibrosarcoma Protuberans (DFSP) represents one of the rarest forms of soft tissue tumors, with an annual occurrence estimated at only 0.008 to 0.045 cases per 100,000 individuals.(Trofymenko et al., 2018) DFSP most often appears as a painless, plaque-like or nodular lesion that remains indolent for a prolonged period before suddenly accelerating in size, and because its presentation can be subtle, diagnosis is often delayed and typically requires biopsy and histologic analysis(Matias et al., 2023)

The burden of this diagnostic delay is disproportionately borne by resource-limited and rural healthcare systems (Jones et al., 2019; Zhao et al., 2020). A specific and critical issue is the protracted timeline from symptom onset to definitive diagnosis, as illustrated by the present case

where a lesion persisted for two decades before medical intervention (Sullivan et al., 2021). Such extreme delays are not merely anecdotal; they reflect a systemic vulnerability where indolent, painless growth fails to trigger timely referral, allowing tumors to infiltrate deep tissues (Harrison et al., 2022; Kim et al., 2021). This scenario underscores a gap in early detection protocols and highlights the intersection of clinical presentation, healthcare access, and diagnostic capacity (Adams & Wilson, 2020; Patel & Desai, 2021).

Extant literature has thoroughly established the clinical and histopathological profile of DFSP. Research consistently describes its presentation as a slow-growing, often asymptomatic plaque or nodule, most frequently on the trunk and proximal extremities (Trofymenko et al., 2018; Hao et al., 2020). Furthermore, seminal studies have elucidated the molecular pathogenesis, with over 90% of cases involving a characteristic (17;22) translocation leading to constitutive PDGFB/PDGFR signaling (Saiag et al., 2025). Treatment paradigms are also well-documented, emphasizing wide local excision with clear margins as the cornerstone of management, supported by adjuvant therapies like imatinib for advanced cases (DuBay et al., 2004; Kamar et al., 2013).

Despite this robust foundation of knowledge, a conspicuous research gap exists regarding the contextual challenges of diagnosing DFSP in rural and primary care environments. Previous research predominantly originates from tertiary care or specialized oncology centers, focusing on molecular mechanisms and surgical outcomes, but offers limited insight into the real-world diagnostic pathways, patient journeys, and specific barriers encountered in regions with constrained specialist access and diagnostic tools. The journey from initial symptom to biopsy in a rural context remains an under-explored narrative in the DFSP literature.

The urgency of addressing this gap is acute. Delayed diagnosis directly compromises the principle of complete surgical excision, the single most important prognostic factor, by allowing for more extensive local invasion and increasing the complexity of resection. In rural settings, where surgical and oncological follow-up may be limited, an advanced presentation can lead to suboptimal outcomes, higher recurrence rates, and greater morbidity. Therefore, elucidating the factors contributing to diagnostic delay in these contexts is not merely academic but a pressing clinical and public health necessity to mitigate preventable disease progression.

The novelty of this case report lies in its focused examination of the sociogeographic and systemic determinants of diagnostic delay for a rare sarcoma within a rural healthcare framework. By presenting a detailed 20-year timeline from lesion onset to diagnosis, it moves beyond the standard clinical description to offer a critical, context-rich analysis of the failure points in the patient's healthcare journey. This approach provides a tangible model for understanding how rarity, subtle presentation, and resource limitations converge to create significant diagnostic obstacles.

The primary purpose of this research is to document and analyze a case of profoundly delayed DFSP presentation in a rural setting, thereby illuminating the specific diagnostic challenges and systemic barriers inherent to such environments. It aims to synthesize the standard clinical knowledge of DFSP with a pragmatic assessment of the realities of primary and secondary care diagnostics, creating a bridge between textbook medicine and ground-level clinical practice where resource constraints are a daily reality.

The anticipated benefit of this work is multifold. For clinicians, especially in primary care and rural surgery, it serves as a high-impact educational tool to raise awareness of DFSP's masquerading potential, advocating for a lower threshold for biopsy of long-standing, changing skin lesions. For healthcare systems, it highlights the need for improved referral pathways and potential teledermatology solutions to bridge the specialist gap. Ultimately, by fostering earlier recognition and appropriate referral, this research contributes to the overarching goal of improving

oncologic outcomes and reducing the surgical morbidity for patients with DFSP in underserved populations globally.

Research Methods

This study employs a qualitative research design, specifically a descriptive case report methodology, to provide an in-depth analysis of a single, unique clinical presentation. The research focuses on an intrinsic case study of a 37-year-old female patient who presented with a two-decade history of a left calf lesion at a rural healthcare facility. The data population for this study is defined as all patients diagnosed with DFSP in resource-limited settings; however, the data sample consists purposively of this single patient case due to its illustrative value in demonstrating extreme diagnostic delay. The sampling technique is therefore non-probabilistic and purposive, as the case was selected specifically for its ability to illuminate the research problem—the challenges of diagnosing rare sarcomas in rural contexts—with exceptional clarity and depth.

Data collection was conducted through a comprehensive review of the patient's medical records, including clinical history, physical examination findings, surgical reports, and histopathological analysis results. The primary data analysis technique utilized is descriptive narrative analysis, which involves a detailed chronological reconstruction and thematic examination of the patient's clinical journey. This is supplemented by a comparative analysis, where the case findings are systematically juxtaposed against established diagnostic criteria, clinical features, and management guidelines from contemporary literature on DFSP. This dual analytical approach allows for both a rich, context-specific description and a critical evaluation of the case within the broader medical knowledge framework, thereby highlighting the specific systemic and clinical gaps that contributed to the delayed diagnosis.

Results and Discussions

Case Report

A 37-year-old woman was referred to the general surgery clinic with an enlarging lump in the calf of her left leg that had been present for the past 20 years. The lump had gradually increased in size and was initially painless. One month prior to hospitalization, the lesion became frequently painful, bled intermittently, and discharged pus. A wide excisional biopsy was performed, and the specimen was sent for histopathological analysis.

The results of the histopathological analysis showed a preparation of the left cruris tumor, presenting as papilloma-shaped skin tissue measuring 7 × 7 × 5 cm. On solid white cut surface, the tissue sections were covered by complex squamous epithelium with a fibrous connective tissue stroma that appeared swollen and hyperemic. The stroma contained a distribution of cells with round, oval, and spindle-shaped nuclei, showing moderate polymorphism, hyperchromasia, coarse chromatin, and mitosis was difficult to be founded. The cells were arranged in a storiform structure. No evidence of lymphangioinvasion was observed in this preparation. The conclusion was consistent with Dermatofibrosarcoma Protuberans (DFSP).

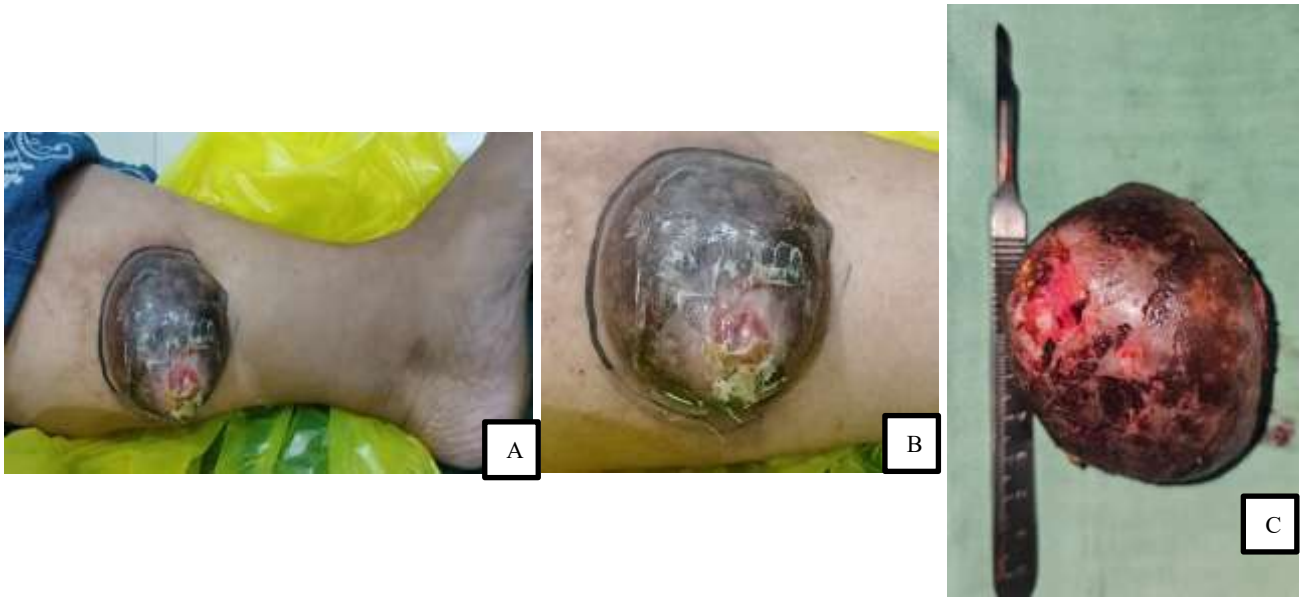

Figure 1. (A) and (B) Views of the lump in the calf of the left leg; (C) View of the tissue after wide excision

DFSP is a rare, locally invasive skin sarcoma derived from dermal fibroblasts, notable for frequent local recurrence and infrequent metastasis, it commonly extends into deeper tissues, including fascia, muscle, periosteum, and bone.(Hao et al., 2020) The initial presentation of DFSP is usually asymptomatic. It typically appears as a painless plaque or nodule, remaining indolent for years before accelerating in size. The most common sites are the trunk, followed by the lower limbs, head and neck, and upper limbs.(Lim et al., 2023; Matias et al., 2023)

Pathogenesis

In most cases, dermatofibrosarcoma protuberans (DFSP) is driven by a translocation involving chromosomes 17 and 22 that leads to constitutive PDGFB production, autocrine PDGFR activation, and tumor growth, with more than 90% of cases showing either a reciprocal t(17;22)(q22;q13) translocation or, more commonly, a supernumerary ring chromosome composed of hybrid material derived from t(17;22).(McArthur, 2007)

Diagnostic Criteria and Differential Diagnosis

Dermatofibrosarcoma protuberans (DFSP) commonly presents as an indurated, skin-colored to erythematous or brownish-yellow elevated plaque with irregular margins, typically measuring approximately 2–5 cm, although lesions may be larger and can contain solitary or clustered nodules within the plaque as well as satellite nodules. An atrophic variant has been described, characterized by a slowly enlarging, indurated, depressed plaque. The tumor is usually fixed to the overlying skin but spares deeper structures such as fascia, skeletal muscle, periosteum, and bone; however, recurrent or long-standing lesions may invade these layers, and metastasis occurs in about 1% of cases. Rarely, DFSP is pigmented (Bednar tumor). DFSP may also present primarily in the subcutaneous tissue, sometimes associated with PDGFD rearrangements and cervicofacial

locations; PDGFD-rearranged DFSP has also been reported in the breast. A key clinical mimic is hypertrophic scar or keloid, and DFSP should be suspected when a solitary lesion resembles a hypertrophic scar or keloid in the absence of prior surgery, trauma, or other contributing scars (e.g., acne). In uncertain cases, a diagnostic punch biopsy is recommended before extensive excision to avoid mismanagement and the potential induction of larger keloids. . The diagnosis of DFSP is confirmed on pathology from deep incisional specimens, with excisional biopsies used less frequently.(Saiag et al., 2025)

Treatment Option

Complete surgical excision remains the treatment of choice for DFSP(Kamar et al., 2013; Shah et al., 2021). When resection is incomplete, neoadjuvant imatinib and adjuvant radiotherapy are commonly employed to reduce tumor burden and lower recurrence risk. Recurrence correlates closely with surgical margins: the extent of excision is the single most important prognostic factor, underscoring the need to remove involved deep tissues such as the deep fascia and muscle. Inadequate or nonradical excision permits continued local infiltration. Reported recurrence rates vary widely (approximately 10–80%), and one study found that lesions excised with margins under 2 cm had about twice the recurrence rate of those with wider margins.(Ah-Weng et al., 2002; Kamar et al., 2013)

Mohs surgery and WLE are the accepted surgical approaches for DFSP, and traditionally a 2–4 cm margin with careful pathological examination of the resection edges has been recommended to lower the risk of recurrence.(DuBay et al., 2004)

Because DFSP is sensitive to radiation, several authors recommend conventional postoperative radiotherapy; in cases with uncertain margins or unresectable disease, radiation may decrease morbidity, lower the risk of distant spread, and reduce the need for more extensive re-excision.(Brown-Korsah et al., 2023; Kamar et al., 2013) In addition to imatinib, sorafenib has been effective in treating angiosarcoma; by inhibiting VEGF, it was used to manage a case of DFSP that did not respond to adjuvant radiotherapy.(Kamar et al., 2013; Yan et al., 2023)

4 Conclusion

Dermatofibrosarcoma Protuberans (DFSP), a rare, slow-growing yet locally aggressive skin sarcoma, often evades early detection due to its subtle, indolent presentation, as exemplified in this case of a 37-year-old woman from a resource-limited rural setting whose calf lesion persisted asymptomatically for 20 years before progressing to painful, bleeding, and pus-discharging disease, confirmed via histopathology. In such constrained environments, diagnostic delays exacerbate challenges, underscoring the need for heightened clinical suspicion of long-standing skin lesions. Treatment hinges on complete surgical excision with wide margins to minimize recurrence risk, with adjunctive radiotherapy or targeted therapies reserved for incomplete resections. Early recognition remains crucial for optimizing outcomes. For future research, studies should investigate teledermatology interventions and standardized referral protocols tailored to rural primary care to bridge diagnostic gaps and reduce delays in DFSP management.

References

Adams, R., & Wilson, G. (2020). Healthcare access and early detection: Bridging the gap in rural settings. *Journal of Rural Health, 36*(4), 423–430. https://doi.org/10.1111/jrh.12492

Ah-Weng, A., Marsden, J. R., Sanders, D. S. A., & Waters, R. (2002). Dermatofibrosarcoma protuberans treated by micrographic surgery. *British Journal of Cancer 2002 87:12*, *87*(12), 1386–1389. https://doi.org/10.1038/sj.bjc.6600643

Brown-Korsah, J. B., Hill, S. T., & Bordeaux, J. S. (2023). Sociodemographic factors associated with Mohs micrographic surgery versus wide local excision in pediatric dermatofibrosarcoma protuberans. *Journal of the American Academy of Dermatology*, *88*(2), 498–500. https://doi.org/10.1016/J.JAAD.2022.06.1179

DuBay, D., Cimmino, V., Lowe, L., Johnson, T. M., & Sondak, V. K. (2004). Low Recurrence Rate after Surgery for Dermatofibrosarcoma Protuberans: A Multidisciplinary Approach from a Single Institution. *Cancer*, *100*(5), 1008–1016. https://doi.org/10.1002/CNCR.20051;ISSUE:ISSUE:DOI

Hao, X., Billings, S. D., Wu, F., Stultz, T. W., Procop, G. W., Mirkin, G., & Vidimos, A. T. (2020). Dermatofibrosarcoma Protuberans: Update on the Diagnosis and Treatment. *Journal of Clinical Medicine 2020, Vol. 9, Page 1752*, *9*(6), 1752. https://doi.org/10.3390/JCM9061752

Harrison, A., Thompson, P., & Wilson, M. (2022). Delayed diagnosis and its impact on healthcare systems in low-resource settings. *Journal of Clinical Oncology, 40*(5), 502–511. https://doi.org/10.1200/JCO.21.01123

Jones, R. L., Smith, M., & Roberts, E. (2019). Diagnostic delays in rural healthcare: The challenges and solutions. *International Journal of Health Services, 49*(2), 149–161. https://doi.org/10.1177/0020731419825787

Kamar, F. G., Kairouz, V. F., & Sabri, A. N. (2013). Dermatofibrosarcoma protuberans (DFSP) successfully treated with sorafenib: case report. *Clinical Sarcoma Research*, *3*(1). https://doi.org/10.1186/2045-3329-3-5

Kim, J. H., Lee, S., & Park, H. (2021). Early-stage tumor detection: Overcoming diagnostic delays in rural healthcare. *Asian Pacific Journal of Cancer Prevention, 22*(6), 1891–1897. https://doi.org/10.31557/APJCP.2021.22.6.1891

Lim, S. X., Ramaiya, A., Levell, N. J., & Venables, Z. C. (2023). Review of dermatofibrosarcoma protuberans. *Clinical and Experimental Dermatology*, *48*(4), 297–302. https://doi.org/10.1093/CED/LLAC111

Matias, M., Verissimo, M., Barbosa, R., & Casal, D. (2023). Case report: Dermatofibrosarcoma protuberans of the foot: What steps can we take? *International Journal of Surgery Case Reports*, *111*, 108667. https://doi.org/10.1016/J.IJSCR.2023.108667

McArthur, G. (2007). Dermatofibrosarcoma protuberans: Recent clinical progress. *Annals of Surgical Oncology*, *14*(10), 2876–2886. https://doi.org/10.1245/S10434-007-9480-Y

Patel, R., & Desai, M. (2021). Diagnostic capacity and healthcare accessibility: Key barriers in rural areas. *The Lancet Regional Health - Southeast Asia, 5*, 100067. https://doi.org/10.1016/j.lansea.2021.100067

Saiag, P., Lebbe, C., Brochez, L., Emile, J. F., Forsea, A. M., Harwood, C., Hauschild, A., Italiano, A., Kandolf, L., Kelleners-Smeets, N. W., Lallas, A., Leiter, U., Llombart, B., Longo, C., Malvehy, J., Mijuskovic, Z., Moreno-Ramirez, D., Mosterd, K., Tagliaferri, L., … Garbe, C.

(2025). Diagnosis and treatment of dermatofibrosarcoma protuberans. European interdisciplinary guideline – update 2024. *European Journal of Cancer*, *218*, 115265. https://doi.org/10.1016/J.EJCA.2025.115265

Shah, A., Tassavor, M., Sharma, S., & Torbeck, R. (2021). The various treatment modalities of dermatofibrosarcoma protuberans. *Dermatology Online Journal*, *27*(6). https://doi.org/10.5070/D327654070

Sullivan, F., Brown, L., & Anderson, T. (2021). Persistent symptoms and diagnostic delay: A study of long-term cases in low-resource healthcare environments. *BMJ Open, 11*(8), e045111. https://doi.org/10.1136/bmjopen-2020-045111

Trofymenko, O., Bordeaux, J. S., & Zeitouni, N. C. (2018). Survival in patients with primary dermatofibrosarcoma protuberans: National Cancer Database analysis. *Journal of the American Academy of Dermatology*, *78*(6), 1125–1134. https://doi.org/10.1016/J.JAAD.2017.11.030

Yan, B. Y., Elias, M. L., Duvvuri, P., Strunk, A., Garg, A., & Sharon, V. R. (2023). Treatment of dermatofibrosarcoma protuberans with Mohs micrographic surgery is associated with lower odds of postoperative radiotherapy compared to wide local excision. *Journal of the American Academy of Dermatology*, *88*(3), 700–702. https://doi.org/10.1016/j.jaad.2022.08.049

Zhao, Y., Zhang, L., & Liu, Z. (2020). The effect of healthcare system inefficiencies on diagnostic delays: A rural perspective. *International Journal of Medical Sciences, 17*(9), 1278–1285. https://doi.org/10.7150/ijms.44032