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Original article

Diagnostic and Therapeutic Challenges of Dermatofibrosarcoma Protuberans (DFSP): Insights from Libyan Medical Practitioners

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Abstract
Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue tumor that poses significant diagnostic and management challenges. In resource-limited settings such as Libya, insufficient awareness and limited diagnostic facilities may contribute to frequent misdiagnosis and delayed treatment. This study aimed to assess healthcare professionals' experience, diagnostic approaches, and management strategies for DFSP in Libya. A cross-sectional survey was conducted among 213 healthcare professionals, including dermatologists, surgeons, oncologists, pathologists, radiologists, and general practitioners from various regions of Libya. Data were collected using a standardized questionnaire covering demographic characteristics, experience with DFSP, diagnostic methods, treatment practices, perceived challenges, and recommendations for improvement. Among participants, 65.7% had previously encountered DFSP cases, and 79.8% reported familiarity with the disease. Misdiagnosis was common, with lipoma (39.9%), keloid (23.5%), and dermatofibroma (18.8%) being the most frequent initial incorrect diagnoses. Biopsy with histopathological examination was the primary diagnostic method (61.0%), while 43.2% routinely used immunohistochemistry (IHC), most commonly CD34 (37.6%). The majority of respondents (65.7%) believed that DFSP is often misdiagnosed in Libya. Preferred treatment modalities included wide local excision (56.3%) and Mohs micrographic surgery (23.5%). Major challenges identified were late diagnosis (46.9%), limited diagnostic resources (37.6%), and insufficient clinician awareness (32.9%). The most frequently suggested improvements were organizing training programs and workshops (70.4%) and enhancing diagnostic facilities (56.3%). Specialty and years of professional experience were significantly associated with disease familiarity and use of IHC ($p < 0.05$). This study highlights substantial diagnostic and management challenges related to DFSP in Libya, primarily due to limited awareness and inadequate diagnostic infrastructure. Targeted training initiatives, improved access to diagnostic tools, and the development of national management guidelines are essential to promote early diagnosis and optimal patient care.
Keywords. Dermatofibrosarcoma Protuberans, Misdiagnosis, Immunohistochemistry, Libya.

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare, slow-growing soft tissue sarcoma of fibroblastic origin that begins in the dermis and frequently spreads to subcutaneous tissue and deeper tissues. DFSP accounts for between 1–6% of all soft tissue sarcomas, with an estimated yearly incidence of 0.8–4.5 cases per million people [1,2]. The tumor was initially identified in the late nineteenth century and later described by Hoffman in 1925 using the present name. It is characterized as a low-to-intermediate grade sarcoma with a high local recurrence rate but a low metastatic risk. DFSP primarily affects people aged 30 to 50, while occurrences in children and the elderly have been described [3]. There is no clear gender preference, although certain populations show a slight male predominance [4]. The trunk is the most common anatomical location (42–72%), followed by the proximal limbs (16–30%) and the head and neck region (10–16%) [5].

Epidemiological data from African and North African populations are scarce. However, studies from Tunisia and Egypt indicate that delayed presentation and misdiagnosis are common due to limited awareness and diagnostic infrastructure [6,7]. Libya currently lacks published data on DFSP incidence, diagnostic procedures, and management outcomes, highlighting a significant knowledge gap that this study aims to address. Clinically, DFSP typically presents as a firm, indurated, skin-colored or violaceous plaque that slowly enlarges over the years. Its indolent appearance often leads to misdiagnosis as benign lesions such as lipoma, keloid, dermatofibroma, or epidermal cyst [8]. In the early stages, the lesion is usually asymptomatic, further contributing to delayed clinical suspicion.

Histologically, DFSP is composed of spindle cells arranged in a storiform pattern, infiltrating subcutaneous fat in a "honeycomb" fashion. Immunohistochemically, DFSP shows strong diffuse CD34 positivity and is typically negative for S100 and P100, aiding in differentiation from other soft tissue tumors [9,10]. The presence of the COL1A1:PDGFB gene fusion t(17;22) (q22;q13) is a characteristic molecular feature identified in more than 90% of cases [11].

Despite these well-defined features, misdiagnosis remains common due to overlapping histological patterns and the lack of routine immunohistochemical testing in resource-limited settings. Pathologists in Libya and other developing countries may face shortages of immunostaining reagents, molecular diagnostic tools, and specialized dermatopathology expertise, leading to diagnostic delays.

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The cornerstone of DFSP management is complete surgical excision with histologically clear margins. Historically, wide local excision (WLE) with margins of 2–3 cm was the standard approach; however, recurrence rates remained high when margins were inadequate, reaching up to 50% [12]. The introduction of Mohs micrographic surgery (MMS) has significantly reduced recurrence rates to approximately 1–3% [13]. For patients with unresectable, recurrent, or metastatic disease, targeted therapy with imatinib mesylate, a tyrosine kinase inhibitor targeting PDGFR-β, has become an important therapeutic option [14]. Radiotherapy may be used as an adjuvant treatment in cases with positive or close surgical margins or when surgical resection may result in significant functional or cosmetic impairment.

In low- and middle-income countries, DFSP poses specific diagnostic and management challenges due to limited access to advanced imaging modalities (CT/MRI), immunohistochemical markers (CD34, S100, P100, Ki-67), and multidisciplinary cancer care centers. In such settings, misdiagnosis as a benign lesion often leads to incomplete excision, increasing recurrence risk.

In Libya, challenges including limited pathology infrastructure, a shortage of trained dermatopathologists, a lack of molecular testing for COL1A1-PDGFB fusion, and weak referral pathways between dermatology, surgery, and oncology services contribute to delayed diagnosis and suboptimal outcomes. Additionally, the absence of national treatment guidelines and limited interdisciplinary coordination further complicate DFSP management. Given these challenges, assessing clinician awareness, diagnostic experience, and management practices related to DFSP is essential. This study aims to evaluate misdiagnosis frequency in Libya, assess access to diagnostic tools, and identify gaps in multidisciplinary collaboration. The findings may support the development of national guidelines, targeted training programs, and improved interdisciplinary coordination to enhance DFSP patient outcomes in Libya and similar resource-limited settings.

Methods

Study Design and Setting

This cross-sectional study was conducted among healthcare professionals in Libya to assess the misdiagnosis and management challenges of Dermatofibrosarcoma Protuberans (DFSP). Data were collected by dermatologists, general surgeons, surgical oncologists, medical oncologists, pathologists, radiologists, and general physicians.

Study Population

The target population included specialists and general physicians working in clinical and hospital settings across Libya. A total of 216 responses were received. 213 was accepted after cleaned.

Inclusion and Exclusion Criteria

Inclusion criteria comprised healthcare professionals who are actively practicing within relevant specialties and who possess experience or knowledge related to dermatofibrosarcoma protuberans (DFSP). Exclusion criteria applied to individuals who declined to provide consent or who did not complete the questionnaire in its entirety.

Data Collection Tool

A structured, self-administered questionnaire was developed, comprising 11 items covering demographics, professional experience, knowledge, diagnostic practices, and challenges related to DFSP. The questionnaire included the following sections: The survey instrument was developed to assess clinicians' knowledge, diagnostic practices, and management strategies related to dermatofibrosarcoma protuberans (DFSP). Participants were asked to indicate their medical specialty, including dermatology, general surgery, surgical oncology, medical oncology, pathology, radiology, or general practice. Information on years of professional experience was collected in categorical ranges (<5, 5–10, 11–20, and >20 years). Respondents were further queried regarding prior encounters with DFSP cases (Yes/No) and their level of familiarity with the disease (very familiar, somewhat familiar, or not familiar).

To explore diagnostic challenges, participants were asked to identify the most common misdiagnoses associated with DFSP, such as lipoma, keloid, dermatofibroma, epidermal cyst, or other conditions. They were also requested to specify their primary diagnostic tools, including clinical examination, biopsy/histopathology, immunohistochemistry, imaging (CT/MRI), or other modalities. The availability and use of immunohistochemistry were examined, with emphasis on commonly employed markers such as CD34, S100, P100, Ki-67, and others. Perceptions of misdiagnosis within the Libyan healthcare context were assessed (Yes, Sometimes, No), alongside preferred treatment and management approaches, which included wide local excision, Mohs micrographic surgery, radiotherapy, targeted therapy, or multidisciplinary management. Finally, participants were asked to identify the main challenges in DFSP management, such as late diagnosis, limited pathology or imaging resources, lack of clinician awareness, restricted access to specialized treatment, limited surgical expertise, or other barriers. Recommendations for improvement were solicited, focusing on training and workshops, enhanced diagnostic facilities, the establishment of national guidelines, and improved inter-specialty collaboration.

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Data Collection Procedure
The questionnaire was distributed electronically via email and social media platforms targeting professional groups. Participation was voluntary, and informed consent was obtained from all respondents. Responses were anonymized to maintain confidentiality.

Data Analysis
Data were coded and entered into Microsoft Excel. Descriptive statistics were used to summarize participants' characteristics, experience, familiarity with DFSP, diagnostic practices, and management approaches. Categorical variables were presented as frequencies and percentages.

Ethical Considerations
The study was reviewed and approved by the Ethical Committee of the National Cancer Institute–Misurata (meeting No. 03, held on September 30, 2025; Reference No. NBC: 021.H.25.01). The research was conducted in accordance with the Declaration of Helsinki (2013). Participation was voluntary, and confidentiality was strictly maintained.

Results
Participant Characteristics
A total of 213 healthcare professionals participated in the study. (Table 1) summarizes the participants' specialties and years of experience.

Table 1. Participant Demographics (n = 213)

Variable	Frequency	Percentage (%)
Specialty		
Dermatologist	50	23.5
General Surgeon	42	19.7
Surgical Oncologist	28	13.1
Medical Oncologist	25	11.7
Pathologist	30	14.1
Radiologist	20	9.4
General Physician	18	8.5
Years of Experience		
<5	40	18.8
5–10	60	28.2
11–20	70	32.9
>20	43	20.1

Experience with DFSP
Among participants, 140 (65.7%) reported encountering a DFSP case, while 73 (34.3%) had not. Regarding familiarity, 60 (28.2%) were very familiar, 110 (51.6%) somewhat familiar, and 43 (20.2%) not familiar with DFSP.

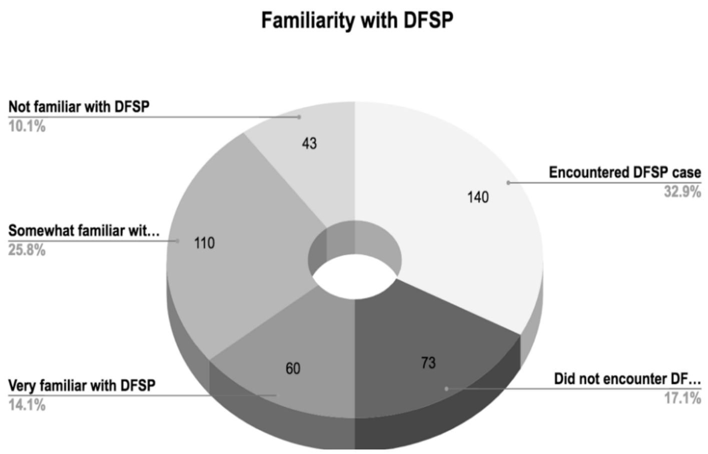

Figure 1. Experience with DFSP

Common Misdiagnoses
The analysis of reported misdiagnoses revealed that lipoma was the most common, accounting for 85 cases (39.9%). Keloid was the second most frequent misdiagnosis, reported in 50 cases (23.5%), followed by dermatofibroma in 40 cases (18.8%). Epidermal cyst was identified in 25 cases (11.7%), while other conditions collectively represented 13 cases (6.1%). These findings underscore the diagnostic challenges

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associated with dermatofibrosarcoma protuberans, particularly its tendency to be mistaken for more prevalent benign lesions.

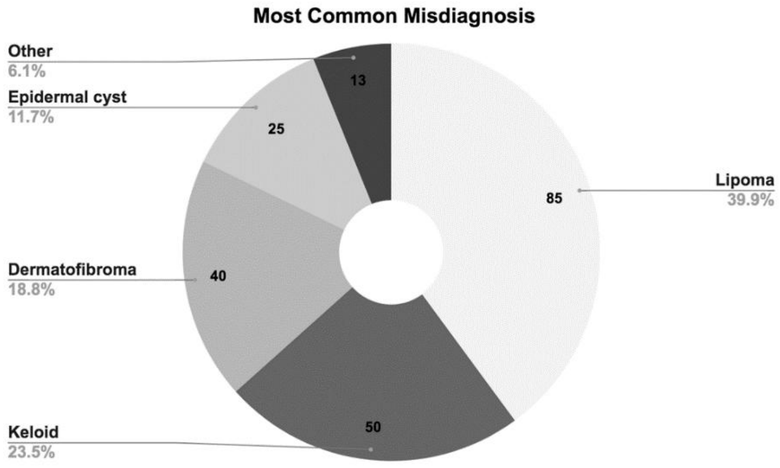

Figure 2. Common Misdiagnoses

Diagnostic Approaches
Participants reported using the following main diagnostic tools (Table 2). Regarding immunohistochemistry, 92 participants (43.2%) reported that IHC is routinely used in their workplace. Among IHC users, the most commonly used markers were CD34 (80; 87.0%), Ki-67 (25; 27.2%), S100 (20; 21.7%), and P100 (15; 16.3%).

Table 2. Main Diagnostic Tools for DFSP

Diagnostic Tool	Frequency	Percentage (%)
Clinical examination	60	28.2
Biopsy/histopathology	130	61.0
Immunohistochemistry	90	43.2
Imaging (CT/MRI)	50	23.5
Other	10	4.7

Perception of Misdiagnosis of Dermatofibrosarcoma Protuberans (DFSP) in Libya
The results indicate a prevalent perception of DFSP misdiagnosis among the surveyed participants (N=213). The vast majority reported that this disease is 'Often misdiagnosed', accounting for 65.7% of responses. A significant portion also reported that misdiagnosis occurs 'Sometimes' (23.5%), which further reinforces the diagnostic complexity. In contrast, only 10.8% of respondents felt that misdiagnosis was not an issue. This data underscores an urgent need to enhance diagnostic accuracy at a national level.

Table 3. Perception of Misdiagnosis

Perception of Misdiagnosis	Count (N=213)	Percentage (%)
Often misdiagnosed	140	65.7%
Sometimes	50	23.5%
No	23	10.8%

Preferred Management and Treatment Approaches
Participants were surveyed regarding their preferred management strategies for dermatofibrosarcoma protuberans (DFSP), with multiple responses permitted, as DFSP management often involves more than one therapeutic modality. Wide local excision was the most commonly selected approach, reported by 120 participants (56.3%). Mohs micrographic surgery was selected by 50 respondents (23.5%). In addition, multidisciplinary management was reported by 35 participants (16.4%), while radiotherapy and targeted therapy were selected by 30 (14.1%) and 20 (9.4%) respondents, respectively. These findings indicate that, although surgical excision remains the cornerstone of DFSP treatment, adjuvant and multidisciplinary approaches are also frequently considered in clinical practice. Percentages exceed 100% because participants were allowed to select more than one management option.

Table 4. Management Strategy

Management Strategy	Count (N=213)	Percentage (%)
Wide local excision	120	**56.3%**
Mohs micrographic surgery	50	23.5%
Radiotherapy	30	14.1%
Targeted therapy	20	9.4%
Multidisciplinary management	35	16.4%

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Key Challenges in DFSP Diagnosis and Management
The study clearly identified several operational challenges that currently hinder the effective diagnosis and management of DFSP in Libya. Late diagnosis was reported as the most prominent obstacle, cited by 46.9% of participants. Other significant challenges include limited pathology or imaging resources (37.6%) and a substantial lack of clinician awareness regarding the disease (32.9%). Limited access to specialized treatment (18.8%) and a scarcity of specialized surgical expertise (16.4%) were also noted as contributing factors to poor outcomes.

Table 5. Challenges in DFSP Diagnosis and Management

Challenge	Count (N=213)	Percentage (%)
Late diagnosis	100	**46.9%**
Limited pathology or imaging resources	80	37.6%
Lack of clinician awareness	70	32.9%
Limited access to specialized treatment	40	18.8%
Limited surgical expertise	35	16.4%

Recommendations to Improve DFSP Diagnosis and Management
In response to the identified systemic issues, participants offered a clear set of recommendations aimed at elevating the standard of care for DFSP patients. The call for training and workshops was the most pressing recommendation, cited by a commanding 70.4% of respondents. This was followed by the crucial need for better diagnostic/imaging facilities (56.3%) and the formal establishment of national management guidelines (46.9%) to standardize care. Finally, improved collaboration between specialties was emphasized by 42.3% to ensure comprehensive case management.

Table 6. Recommendations to Improve DFSP Diagnosis and Management

Recommendation	Count (N=213)	Percentage (%)
Training and workshops	150	70.4%
Better diagnostic/imaging facilities	120	56.3%
National management guidelines	100	46.9%
Improved collaboration between specialties	90	42.3%

Discussion
Our study reveals a significant prevalence of misdiagnosis of Dermatofibrosarcoma Protuberans (DFSP) among healthcare professionals in Libya. The most commonly reported misdiagnoses were lipoma (39.9%) and keloid (23.5%), which is consistent with findings reported in previous studies. Srikumar et al. reported that 52.3% of DFSP patients had received an initial incorrect diagnosis, most frequently keloids, lipomas, or cysts [15]. These findings highlight the diagnostic difficulty posed by the slow-growing and clinically subtle nature of DFSP and emphasize the need for increased clinical suspicion among healthcare providers.
Our findings indicate that although biopsy and histopathological examination are widely recognized as essential diagnostic tools, immunohistochemistry (IHC) is not routinely employed in many Libyan healthcare settings. This is concerning, as IHC—particularly CD34 staining—is critical for confirming DFSP and differentiating it from other spindle cell tumors. Haycox et al. demonstrated the diagnostic value of CD34 immunostaining in distinguishing DFSP from dermatofibroma and other soft tissue lesions [16]. Limited availability of IHC services may therefore contribute to delayed or inaccurate diagnoses in Libya.
Wide local excision (WLE) was the most commonly preferred treatment approach among participants (56.3%), which aligns with established international treatment recommendations. However, DFSP is known for its high local recurrence rate, making careful surgical margin control and long-term follow-up essential. Deng et al. emphasized that inadequate excision margins are associated with recurrence rates of up to 60%, underscoring the importance of regular postoperative surveillance [17].
The primary challenges identified by participants included late diagnosis (46.9%), limited pathology or imaging resources (37.6%), and insufficient clinician awareness (32.9%). These challenges are consistent with reports from other regions, where DFSP is frequently mistaken for benign lesions such as cysts, lipomas, dermatofibromas, scars, or keloids. David et al. highlighted that this clinical similarity is a major factor contributing to diagnostic delay and inappropriate initial management [18].
Participants proposed several strategies to improve DFSP diagnosis and management, including targeted training programs and workshops (70.4%), improved diagnostic and imaging facilities (56.3%), and the development of national management guidelines (46.9%). Implementing these measures could enhance early detection, ensure appropriate surgical management, and reduce recurrence rates in Libya.

Limitations
This study has several limitations. The cross-sectional design limits causal inference, and reliance on self-reported data may introduce response bias. Additionally, the study was conducted within a single country,

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which may limit generalizability to other healthcare systems. Finally, the lack of histopathological confirmation for some reported cases may affect the accuracy of the misdiagnosis data.

Conclusion

This study highlights the urgent need to improve awareness, diagnostic accuracy, and management of DFSP among healthcare professionals in Libya. Routine use of immunohistochemical markers, enhanced access to specialized diagnostic services, and the establishment of national clinical guidelines are essential steps toward improving patient outcomes and reducing the burden of DFSP misdiagnosis.

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Conflict of interest. Nil

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