



## Case report

## A unique case of Dermatofibrosarcoma Protuberans arising from an inguinal hernial repair scar in a Middle Eastern male - A Case Report

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## ABSTRACT

**Introduction and importance:** Dermatofibrosarcoma Protuberans (DFSP) is a rare and fatal variant of Spindle Cell Sarcoma. It has an annual incidence rate of 0.8 to 4.5 cases per one million individuals. It's locally aggressive and has vague and masquerading clinical presentations. Misdiagnosis is devastating as it can lead to time wasting, expenditure of unnecessary resources, and possibly raise morbidity and mortality for patients. It is warranted to raise preoperative clinical awareness to achieve prompt surgical therapeutic interventions to reach an up-to-par prognosis.

**Case presentation:** We demonstrate the case of a 50-year-old previously healthy Middle Eastern male patient, who was referred to our General Surgery clinic with the chief complaint of an expansive bulge in his left iliac fossa. Preoperative imaging could not exclude a neoplastic cause behind the presentation. Based on the clinical picture, a surgical intervention was decided.

**Clinical discussion:** Our patient's treatment was consummated by means of classical surgical resection of the lesion with adequate negative margins and referring him to an oncologist specialized in DFSP to undergo the necessary adjuvant treatment. Definitive diagnosis was firmly entrenched postoperatively after finalization of the histopathological and immunohistochemical analyses of the resected protuberance.

**Conclusion:** DFSP is an eminently rare entity, especially DFSPs which originate from a surgical scar -as was our patient's- and fluctuates in its clinical presentation, thus, it is our responsibility to depict, study this malignant tumor, and document its incidence, so that we can make ironclad clinical decrees to plummet the morbidity and mortality of this relentless neoplasia.

## 1. Introduction

Darier and Ferrand were the pioneers who clinically portrayed Dermatofibrosarcoma Protuberans (DFSP) in 1924. The definition of DFSP was later solidified by Hoffman in 1925 [1].

This neoplasm is a rare sarcoma of soft tissues which arises from the skin's dermal layer [2]. Initially, the pathological lesions have a sluggish growth pattern and appear as skin-pigmented painless plaques with colors ranging from blue to burgundy [3]. In their advanced stages,

DFSPs can rapidly proliferate, protrude underneath the skin, and sometimes cause overlying skin ulcerations [2]. This form of neoplasia has a vicious locally invasive behavior [4]. Neoplastic cells are notorious in aggressively invading surrounding soft tissue organs, for instance; muscles, ligaments, tendons, subcutaneous tissues, and astoundingly; bone [5].

DFSPs possess an annual incidence rate of 0.8 to 4.5 cases per one million in the United States of America [3,6].

It is intensely complex to denote the preoperative diagnosis since this

**Abbreviations:** DFSP, Dermatofibrosarcoma Protuberans; IV, Intravenous; CT, Computed Tomography; IHC, Immunohistochemistry; SMA, Smooth Muscle Actin; MRI, Magnetic Resonance Imaging.

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neoplasia mimics several other pathological dermatological lesions. This will ultimately result in inadequate treatment of said tumor [7]. It is warranted to maintain high clinical suspicion and establish sensible surgical treatment approaches to reach a satisfactory prognosis [8,9].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [10].

## 2. Presentation of case

### 2.1. Patient information

We demonstrate the case of a 50-year-old previously healthy Middle Eastern male patient, who presented to our General Surgery clinic with the chief complaint of an expansive bulge in his left iliac fossa. His clinical complaint initiated 4 years ago, six months after he had undergone surgery to repair a left inguinal hernia. 4 years ago, he started feeling a bulge at the site of the surgical incision. It was small at first, but later started to gradually proliferate in size. There wasn't any accompanying pain. The patient ignored his symptoms until 5 months prior to his clinical visit because the size of the mass started increasing noticeably. The lesion posed remarkable discomfort for him upon ambulation. However, the overlying skin did not undergo any discolorations, except for mild degrees of erythema. He also denied any overlying skin ulcerations and/or hyper-/hypopigmentation. He didn't report any chills or fever, cold or night sweats, general weakness, malaise, weight changes, history of trauma, or recent infections. Furthermore, he denied any genitourinary symptoms or any changes in his bowel habits. His surgical history consisted solely of the previously mentioned inguinal hernial repair. His previous medical, drug, allergic, family, and history of a similar incidence were all negative. He doesn't consume alcohol and he isn't an active smoker. His BMI was 28 kg/m<sup>2</sup>.

### 2.2. Clinical findings

Physical examination revealed mild tachypnea accommodated by borderline tachycardia. The remaining vital signs were normal. By inspection, the abdomen moved harmoniously with respiration and no increase in abdominal contour was marked. However, an obvious bulging was seen in the left inguinal region. It wasn't movable nor exaggerated by coughing or standing up. It didn't disappear upon lying down. No skin puckering or retraction were seen. No skin changes (i.e., induration, telangiectasia, overlying skin variations, ulcerations, pigmentations) were marked. However, the overlying skin was erythematous (Fig. 1). By palpation, there weren't any guarding or tenderness. A well-demarcated hard immobile mass was felt, the overlying skin was adherent to it, and it was irreducible with pressure.

Laboratory investigations in total yielded normal results.

### 2.3. Diagnostic assessment

Abdominal Ultrasound was performed and established a lesion with hypoechoic constitution located in the left iliac fossa region and it measured (7 × 2.7 cm). The radiological diagnosis leaned towards a diagnosis of Lipoma. Malignant etiologies could not be excluded.

Initial management consisted of establishing an Intravenous (IV) access line, preoperative IV antibiotics, and complete blood work to prepare for surgery. Naturally, blood samples for crossmatch were drawn.

Notable preoperative challenges were the unavailability of a laparoscopic device in the hospital at the time of patient admission, and the unavailability of a Computed Tomography (CT) scanning machine due to the patient's low socioeconomic status, which prevented him from performing it in another medical center.



**Fig. 1.** Preoperative image demarcating the obvious bulging notable in the left inguinal region, it wasn't movable, it didn't disappear upon lying down, no skin puckering, or retraction was seen, no induration nor telangiectasia were noted, and no overlying skin changes were witnessed such as ulceration or hyper-/hypopigmentation. Notable erythema is demonstrated.

### 2.4. Therapeutic intervention

An elliptical incision was the incision of choice based on the given clinical elements. The surgical operation was accomplished at a tertiary hospital, was seen through by a General Surgery Consultant with 10 years of General Surgery experience, and the operative time was 25 min.

The mode of anesthesia was local, however, there were no perioperative anesthetic or surgical complications. Surgery had confirmed the findings noted during the preoperative ultrasound. Through surgical exploration, a hard immobile mass, adherent to the subcutaneous tissue of the abdominal wall, was found and roughly measured (9 × 3.5 cm). Utter excision of the mass was achieved after isolating it from its surrounding tissues. Resection of a 5 cm free margin was accomplished.

Histopathological analysis established the presence of nodular formation composed of ill-demarcated, highly cellular proliferative mass with spindle cell formation, containing oval vascular nuclei with notable elongated cytoplasm and mild atypia without any marked mitosis. The diagnosis conforms with Dermatofibrosarcoma Protuberans (Low Grade Fibrosarcoma) (Fig. 2A-B).

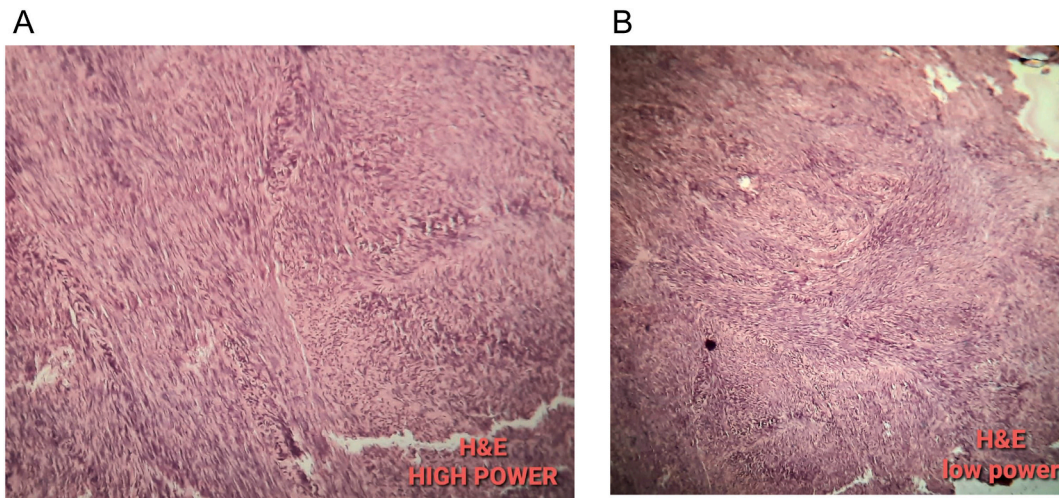
The negative margins of surgical of resection were deemed free of neoplastic involvement.

Immunohistochemistry (IHC) was demanded based on the histopathological analysis. It yielded positive staining for CD34, negative staining for Smooth Muscle Actin (SMA), and a KI67 higher than 14 % (Fig. 3A-B-C).

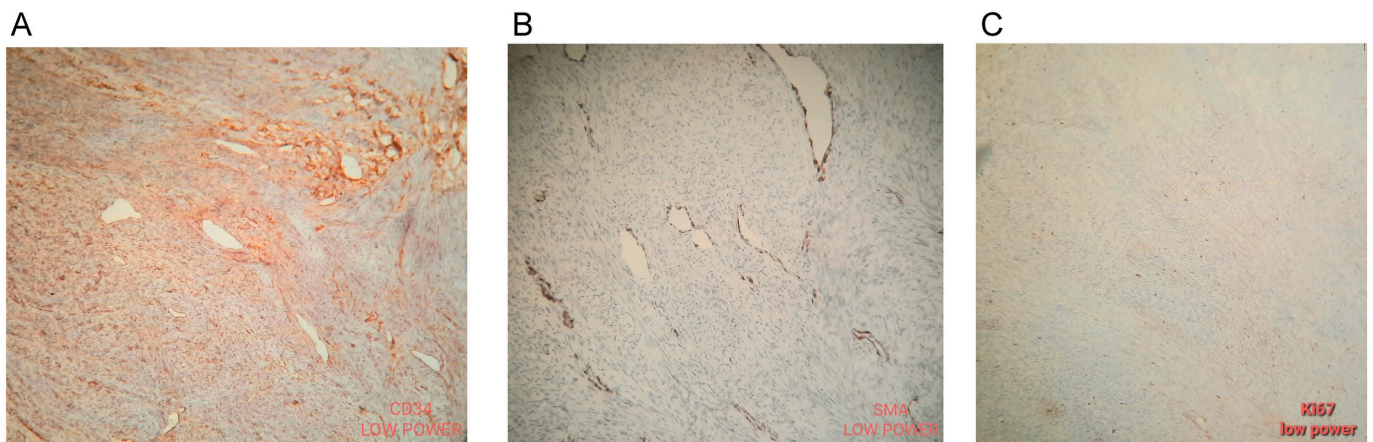
The patient didn't receive any blood transfusions, underwent successful postoperative recovery, and was discharged home within 3 days of the operation.

Within that time, our patient was given full details of the nature of his oncological situation. Moreover, we provided him with informative instructions including a list of multiple lifestyle amendments which assist in a complete recovery, such as consistent sterile wound dressings by a medical provider, reasonable analgesia for pain management, a prescription of postoperative antibiotics for the prophylaxis of wound infection, and a scrupulous postoperative follow-up protocol.

The follow-up regimen was set-up in the outpatient settings for five months thus far and is still currently being conducted. He has had



**Fig. 2.** (A-B): Histopathological analysis results (Hematoxylin and Eosin staining) established the presence of nodular formation composed of ill-demarcated, highly cellular proliferative mass with spindle cell formation containing oval vascular nuclei with notable elongated cytoplasm with mild atypia without any marked mitosis. The diagnosis conforms with Dermatofibrosarcoma Protuberans (Low Grade Fibrosarcoma).



**Fig. 3.** A: Immunohistochemistry revealed CD34 marker to be positive.  
 B: Immunohistochemistry revealed SMA marker to be negative.  
 C: Immunohistochemistry revealed KI67 > 14 %.

systematic scheduled appointments at the Oncology clinic to undergo meticulous clinical postoperative evaluations via physical examination, postoperative CT scanning, and ultrasound imaging. Additionally, surveillance also included panels of laboratory investigations.

Postoperative pan-CT scan demonstrated clear surgical site, lung fields, and abdominal organs.

We referred him to a specialized oncologist for determining the best adjuvant treatment for him. Presently, he is deemed free of any relapsing lesions, metastasis, or neoplastic recurrence at the surgical site.

### 3. Discussion

DFSP is a vastly rare and locally invasive dermal mesenchymal malignancy. Its origin is said to be from cutaneous tissue, which develops into a blunt sarcoma. It constitutes 0.1 % of all malignant neoplasia and makes-up nearly 1 % of all sarcomas of soft tissues. Furthermore, studies have established an annual incidence rate ranging from 0.8 to 4.5 distinct cases per one million individuals in the United States of America [11–15].

It is characterized by a sluggish growth rate and a high affinity towards local recurrence, because of its supreme ability to invade the surrounding subcutaneous tissue, muscles, and fascia [15,16].

The most frequently involved regions by DFSP are the trunk, proximal limbs, and the head and neck [12,14].

When it comes to gender prevalence, several renowned studies have demonstrated a minimal predominance rate in males over females [6].

Regarding age group incidence, studies have clarified that this neoplasm ponderously occurs in adult age groups of ages ranging from 20 to 50 years [17]. In contrast, numerous articles demonstrated a childhood incidence rate of 6 to 20 %. Moreover, it can have a congenital origin [18].

DFSP can occur in multiple body regions with specific tendencies, such as in old burn areas, trauma regions, areas of surgical repair, vaccination injection sites, post-radiation dermatitis, puncture holes of central venous lines, and sometimes even areas of insect bites [19,20].

When shedding light over the metastatic potential of this neoplasia, we must emphasize that studies have found that there is a relatively low capability of DFSP to metastasize. To be specific, the metastatic probability was found to be <5 % with regards to local, distant, or regional metastasis. In the rare instances in which this tumor metastasizes, the lungs are almost the sole metastatic site and to a much lesser degree, to lymph nodes [14,21].

This potentially fatal neoplasia can have multiple diverse preoperative differential diagnoses, such as keloid, hypertrophic scar tissue,

fibromatosis, myofibroblastoma, and even metaplastic carcinomas [14].

Preoperative imaging ultrasound in cases of DFSP has been deeply effective in diagnosis. It demonstrates either extremely hypoechoic appearance or a mixture of hyperechoic and hypoechoic radiological image [22].

In majority of cases, DFSP cannot be definitively differentiated from a Lipoma because of the similar clinical and radiological characteristics [23].

Preoperative radiological evaluations based on Magnetic Resonance Imaging (MRI) cannot accurately differentiate DFSP from other kinds of soft tissue sarcomas [24].

Different variants of DFSP exist and the most predominant subtype is the "Classic" DFSP, which is our patient's, and it constitutes almost 90 % of DFSPs [20].

Decisive diagnosis is reached postoperatively via histopathological analysis, where histopathology sheds light on the typical DFSP traits, such as somehow uniform but closely packed fusiform cells with elongated nuclei. [12]. The ultimate diagnosis is established after immunohistochemical staining is complete. DFSP stains positive for CD34 in 84 % to 100 % and to Vimentin. It stains negative for other stains, such as S-100, HMB-45, SMA, and Desmin. In short, the classical pathognomonic staining pattern for DFSP is a positive stain for CD34 and a negative one for factor XIIIa [3,15,25].

Evidence-based therapeutic approach is primarily consistent of local lesion surgical excision with a recommended negative margin of 2 to 3 cm, in addition to a three-dimensional surgical excision plain involving subcutaneous tissue, fascia, and skin [14,21].

When discussing local recurrence rates of DFSP, the rate subsides when increasing the perimeter of the excised negative margins [14]. We must be aware of the variety of factors which are closely intercalated with raising neoplastic recurrence rates of DFSP. These include the high mitotic rates, size, cellularity, histological variants, and lesion location [26,27].

Studies have clarified, that in cases where the surgeon excised negative margins of 5 cm, the recurrence rate dwindled to smaller than 5 % [21].

The vast local recurrence rates can be owed to the DFSP's expansive characteristics. It projects itself in several axis and thus can potentially involve deeper organs. As a result, surgical resection -even when aggressive- cannot completely remove residual neoplastic lesions [3,28].

Late phase local recurrence can take place in approximately 24 to 90 % of cases [13].

Most recurrences are revealed within the first three years postoperatively, with an estimation of 50 % of them happening in the first year of postoperative excision. Unfortunately, relevant new articles are showing that some DFSP lesions are recurring even 5 years postoperatively. These results highlight the necessity of clinical monitoring of affected patients for a considerable timeframe with a 6-month interval between each patient clinical assessment postoperatively [3,13,25].

Meticulous local resection of DFSP yields astoundingly positive prognoses for patients. It was estimated that the resulting 5-year tumor specific patient survival rate of said patients reached 99 % [29].

Strict postoperative surveillance regimens of a period between 6 and 12 months are highly advised due to DFSP's extreme tendencies to recur [30].

#### 4. Conclusion

DFSP is a rare malignancy and poorly documented in the literature, especially DFSP which originates from an inguinal hernial repair scar -as was our patient's-The clinical manifestations are vague and non-pathognomonic. This demands attention and time investment in research and documentation endeavors to mark its epidemiological features and clinical characteristics. It can easily masquerade as a different pathology, including dissimilar neoplasia. Misdiagnoses can be

a consequence, and that will lead to the implementation of improper management. Therefore, we're required to carefully study and preoperatively consider this form of neoplasia in a surgical patient. In addition, taking well-informed interventional decisions will result in saving lives, avoid wasting resources, and spare time for medical providers and patients.

Establishing an accurate diagnosis will eliminate the morbidity and mortality related to this highly recurring neoplasia.

Documentation will aid in conducting statistical studies and setting-up preoperative diagnostic modalities, innovative surgical techniques, and surveillance protocols for patients.

#### Abbreviations

DFSP	Dermatofibrosarcoma Protuberans
IV	Intravenous
CT	Computed Tomography
IHC	Immunohistochemistry
SMA	Smooth Muscle Actin
MRI	Magnetic Resonance Imaging

#### Ethical approval

This study is exempt from ethical approval in our institution.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the Data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

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#### Author contribution

OA: Conceptualization, resources, who wrote, original drafted, edited, visualized, validated, and literature reviewed the manuscript.

FA, JS: Supervision, project administration, resources, and review of the manuscript.

LA, DI: Supervision and review of the manuscript.

MA: General Surgery Consultant who performed and supervised the operation. Supervision and review of the manuscript.

OA: The corresponding author who submitted the paper for

publication.

All authors read and approved the final manuscript.

#### Declaration of competing interest

None.

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#### References

- [1] J. Darier, M. Ferrad, Dermatofibromes progressifs et récidivants ou fibrosarcomes de la peau, *Ann. Dermatol. Syphiligr.* 5 (1924) 545–562.
- [2] K. Eguzo, B. Camazine, D. Milner, Giant dermatofibrosarcoma protuberans of the face and scalp: a case report, *Int. J. Dermatol.* 53 (2014) 767–772, <https://doi.org/10.1111/j.1365-4632.2012.05639.x>.
- [3] S. Bhamri, A. Desai, J.Q. Del Rosso, N. Mobini, Dermatofibrosarcoma protuberans: a case report and review of the literature, PMID, *J. Clin. Aesthet. Dermatol.* 1 (2008) 34–36, <https://www.ncbi.nlm.nih.gov/pubmed/21103308>.
- [4] G. McArthur, Dermatofibrosarcoma protuberans: recent clinical progress, *Ann. Surg. Oncol.* 14 (2007) 2876–2886, <https://doi.org/10.1245/s10434-007-9480-y>.
- [5] D. Lemm, L.O. Mügge, T. Mentzel, K. Höffken, Current treatment options in dermatofibrosarcoma protuberans, *J. Cancer Res. Clin. Oncol.* 135 (2009) 653–665, <https://doi.org/10.1007/s00432-009-0550-3>.
- [6] V.D. Criscione, M.A. Weinstock, Descriptive epidemiology of dermatofibrosarcoma protuberans in the United States, 1973 to 2002, *J. Am. Acad. Dermatol.* 56 (2007) 968–973, <https://doi.org/10.1016/j.jaad.2006.09.006>.
- [7] S.A. Felek, M. Ibas, S. Dursun, et al., Dermatofibrosarcoma protuberans of the neck: a brief review of the literature, *Indian J. Otolaryngol. Head Neck Surg.* 71 (Suppl 1) (2019) 369–372, <https://doi.org/10.1007/s12070-018-1314-7>.
- [8] A.E. Acosta, C.S. Velez, Dermatofibrosarcoma protuberans, *Curr. Treat. Options Oncol.* 18 (9) (2017) 56, <https://doi.org/10.1007/s11864-017-0498-5>.
- [9] K.L. Kreicher, D.E. Kurlander, H.R. Gittleman, et al., Incidence and survival of primary dermatofibrosarcoma protuberans in the United States, *Dermatol. Surg.* 42 (Suppl 1) (2016) S24–S31, <https://doi.org/10.1097/dss.0000000000000300>.
- [10] Riaz A. Agha, Thomas Franchi, Catrin Sohrabi, Ginimol Mathew, Ahmed Kerwan, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, *International Journal of Surgery*. ISSN: 1743-9191 84 (2020) 226–230, <https://doi.org/10.1016/j.ijssu.2020.10.034>.
- [11] H.M. Gloster, Jr dermatofibrosarcoma protuberans, *J. Am. Acad. Dermatol.* 35 (pt 1) (1996) 355–374, [https://doi.org/10.1016/s0190-9622\(96\)90597-6](https://doi.org/10.1016/s0190-9622(96)90597-6).
- [12] Y.J. Hong, Y.W. Choi, K.B. Myung, H.Y. Choi, A case of myxoid dermatofibrosarcoma protuberans, *Ann. Dermatol.* 23 (2011) 379–381, <https://doi.org/10.5021/2Fad.2011.23.3.379>.
- [13] P. Rutkowski, A. Wozniak, T. Switaj, Advances in molecular characterization and targeted therapy in dermatofibrosarcoma protuberans, *Sarcoma* 2011 (2011), 959132, <https://doi.org/10.1155/2011/959132>.
- [14] D.M. Dragoumis, L.A. Katsohi, I.K. Amlianitis, A.P. Tsiftoglou, Late local recurrence of dermatofibrosarcoma protuberans in the skin of female breast, *World J. Surg. Oncol.* 8 (2010) 48, <https://doi.org/10.1186/2F1477-7819-8-48>.
- [15] O.G. Pérez, R. Righetti, A. Woscoff, H. Amante, Case for diagnosis Dermatofibrosarcoma protuberans 85 (2010) 245–247, <https://doi.org/10.1590/s0365-05962010000200021>.
- [16] G.T. Pack, E.J. Tabah, Dermato-fibrosarcoma protuberans: a report of 39 cases, *AMA Arch. Surg.* 62 (1951) 391–411, <https://doi.org/10.1001/archsurg.1951.01250030397008>.
- [17] B.R. Burkhardt, E.H. Soule, R.K. Winkelmann, J.C. Ivins, Dermatofibrosarcoma protuberans. Study of fifty-six cases, *Am. J. Surg.* 111 (1966) 638–644, [https://doi.org/10.1016/0002-9610\(66\)90031-6](https://doi.org/10.1016/0002-9610(66)90031-6).
- [18] C. Reddy, P. Hayward, P. Thompson, A. Kan, Dermatofibrosarcoma protuberans in children, *J. Plast. Reconstr. Aesthet. Surg.* 62 (2009) 819–823, <https://doi.org/10.1016/j.jbjs.2007.11.009>.
- [19] A. Stivala, G.A. Lombardo, G. Pompili, M.S. Tarico, F. Fraggetta, R.E. Perrotta, Dermatofibrosarcoma protuberans: our experience of 59 cases, *Oncol. Lett.* 4 (2012) 1047–1055, <https://doi.org/10.3892/2Fol.2012.887>.
- [20] W.B. Bowne, C.R. Antonescu, D.H.Y. Leung, et al., Dermatofibrosarcoma protuberans. A clinicopathologic analysis of patients treated and followed at a single institution, *Cancer* 88 (2000) 2711–2720. PMID: 10870053.
- [21] C.K. Chang, I.A. Jacobs, G.I. Salti, Outcomes of surgery for dermatofibrosarcoma protuberans, *Eur. J. Surg. Oncol.* 30 (3) (2004) 341–345, <https://doi.org/10.1016/j.ejso.2003.12.005>.
- [22] P.Y. Shih, C.H. Chen, T.T. Kuo, C.Y. Yang, Y.H. Huang, C.H. Yang, Deep dermatofibrosarcoma protuberans: a pitfall in the ultrasonographic diagnosis of lipoma-like subcutaneous lesions, *Dermatol. Sin.* 28 (2010) 32–35, [https://doi.org/10.1016/S1027-8117\(10\)60005-5](https://doi.org/10.1016/S1027-8117(10)60005-5).
- [23] P. Inampudi, J.A. Jacobson, D.P. Fessell, R.C. Carlos, S.V. Patel, L.O. Delaney-Sathy, Soft-tissue lipomas: accuracy of sonography in diagnosis with pathologic correlation, *Radiology* 233 (2004) 763–767, <https://doi.org/10.1148/radiol.2333031410>.
- [24] W.C. Torreggiani, K. Al-Ismaïl, P.L. Munk, S. Nicolaou, J.X. O'Connell, M. A. Knowling, Dermatofibrosarcoma protuberans: MR imaging features, *AJR Am. J. Roentgenol.* 178 (2002) 989–993, <https://doi.org/10.2214/ajr.178.4.1780989>.
- [25] S. Labonte, W. Hanna, B. Bandarchi-Chamkhaleh, A study of CD117 expression in dermatofibrosarcoma protuberans and cellular dermatofibroma, *J. Cutan. Pathol.* 34 (2007) 857–860, <https://doi.org/10.1111/j.1600-0560.2007.00731.x>.
- [26] D. DuBay, V. Cimmino, L. Lowe, T.M. Johnson, V.K. Sondak, Low recurrence rate after surgery for dermatofibrosarcoma protuberans: a multidisciplinary approach from a single institution, *Cancer* 100 (2004) 1008–1016, <https://doi.org/10.1002/cncr.20051>.
- [27] J.R. Goldblum, R.J. Tuthill, CD34 and factor-XIIIa immunoreactivity in dermatofibrosarcoma protuberans and dermatofibroma, *Am. J. Dermatopathol.* 19 (1997) 147–153, <https://doi.org/10.1097/0000372-199704000-00008>.
- [28] D. Ratner, C.O. Thomas, T.M. Johnson, V.K. Sondak, T.A. Hamilton, B.R. Nelson, N. A. Swanson, C. Garcia, R.E. Clark, D.J. Grande, Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans. Results of a multiinstitutional series with an analysis of the extent of microscopic spread, *J. Am. Acad. Dermatol.* 37 (600–613) (1997), [https://doi.org/10.1016/s0190-9622\(97\)70179-8](https://doi.org/10.1016/s0190-9622(97)70179-8).
- [29] P. Rouhani, et al., Cutaneous soft tissue sarcoma incidence patterns in the U.S.: an analysis of 12,114 cases, *Cancer* 113 (3) (2008) 616–627, <https://doi.org/10.1002/cncr.23571>.
- [30] S.J. Miller, et al., Dermatofibrosarcoma protuberans, *J. Natl. Compr. Cancer Netw.* 10 (3) (2012) 312–318, <https://doi.org/10.6004/jnccn.2012.0032>.