Recurrent Dermatofibrosarcoma Protuberans with Metastases to the Lungs and Spinal Canal: A Case Report

Zhijun Li[1], Hongwei Li[2], Zheng Pan[1], Xiaodong Niu[2], Yong Huang[2]*

[1]*Department of Imaging, The 81st Group Army Hospital of PLA, China*

[2]*Department of Pathology, The 81st Group Army Hospital of PLA, China*

***Correspondence:** Yong Huang, Department of Pathology, The 81st Group Army Hospitalof PLA, China

pathxhy@163.com

Radiology Case. 2026 February; 20(2):1-6 :: DOI: 10.3941/jrcr.5969

AUTHORS' CONTRIBUTIONS
Each author contributed to the idea of the paper, the research of the case, and the editing of the case report.

ACKNOWLEGMENTS
The authors are very grateful to the patients for the providing their reports.

DISCLOSURES
No disclosures for all authors

CONSENT
No consent due to no patient identifiers used.

HUMAN AND ANIMAL RIGHTS
No human or animal experiments were conducted.

CONFLICT OF INTEREST
No conflicts of interest to declare.

ETHICAL APPROVAL
The study was approved by the hospital audit committee prior to commencement of the study.

ABSTRACT

Background: Dermatofibrosarcoma protuberans (DFSP) has a high local recurrence rate and strong invasiveness after surgical resection, and metastasis rarely occurs. In particular, epidural metastasis within the spinal canal is extremely rare. This article explores the imaging features, pathological characteristics, differential diagnosis, and metastatic mechanisms of DFSP lung and spinal canal metastases through this case.

Case presentation: An elderly male had a recurrence of the surgical site on his back after initial surgery for dermatofibrosarcoma protuberans on the back at another hospital and chemotherapy with anlotinib taken orally, and developed pulmonary and epidural metastases within the spinal canal.

Conclusion: Dermatofibrosarcoma protuberans is a rare low - grade malignant mesenchymal tumor originating from the skin fibrous tissue. It is crucial to clarify the scope, depth of DFSP and the local infiltration of the tumor through MRI examination before surgery to reduce the risk of postoperative recurrence and metastasis. If epidural metastasis within the spinal canal occurs, it should be differentiated from lymphoma, leukemia, hemangiopericytoma, infectious lesions and malignant solitary fibrous tumor.

CASE REPORT

CASE REPORT

DFSP is a slow-growing, borderline/low-grade mesenchymal tumor primarily affecting the trunk, limbs, and head-neck regions [1]. Despite low metastatic potential, local recurrence rates are high due to its infiltrative growth. Fibrosarcomatous transformation increases metastatic risk and worsens prognosis. We present a rare case of DFSP metastasizing to the lungs and spinal epidural space, emphasizing imaging-pathology correlation to guide surgical management and reduce recurrence.

CASE PRESENTATION

An elderly male presented with recurrent DFSP on the back. Initial presentation included a painless, immobile, bean-sized mass that progressively enlarged. Previous resection and

anlotinib chemotherapy failed to prevent recurrence. Physical examination revealed a 25×10×8cm firm, non-tender mass with a 15cm surgical scar and localized ulceration. Spinal mobility was restricted. Imaging Findings: MRI (T4-T12 level): T1WI (1a): Isointense mass with patchy hyperintensity and linear hypointense septa. T2WI (1b): Hyperintense mass with irregular isointense/hyperintense areas and septa. Contrast (1d): Rim and patchy enhancement with fat/skin/fascia "tail signs." Spinal metastasis (T7-T8) (1b, 1c, 1d): Epidural nodular mass compressing the dural sac and spinal cord, involving vertebral bone. CT: Recurrent back mass, bilateral pulmonary nodules (1e), and T7-T8 vertebral destruction (1f). Surgical Findings: Wide excision (5cm margins) of the back mass and resection of T7-T9 spinous processes/laminae exposed a gray-white epidural tumor compressing neural structures and invading T7-T8 vertebrae. Microscopy features: Skin mass: Storiform-patterned spindle cells with focal fibrosarcomatous transformation and increased mitoses (>4/10 HPF). Diagnosis: DFSP with fibrosarcomatous transformation. Spinal mass: Spindle cell sarcoma with hemangiopericytoma-like areas. Immunohistochemistry: CD34 was positive, but STAT6, SMA and SSTR2 were negative.

DISCUSSION

DFSP was classified into the category of fibroblast/myofibroblast tumors in the 2020 version of soft tissue tumor classification released by the World Health Organization (WHO) [2]. To reduce tumor recurrence rates, selecting appropriate preoperative imaging for surgical planning is critical. MRI is the optimal imaging modality for soft tissue tumors, clearly delineating the extent of DFSP, its relationship with the skin, subcutaneous tissue, and deep muscles, and assessing tumor size, boundary morphology, and local invasion. MRI typically shows a protuberant cutaneous mass with T1-weighted signal intensity (T1WI) iso- or slightly hypointense to muscle within subcutaneous fat. On T2-weighted sequences (T2WI), signal varies with tumor composition: hyperintensity correlates with myxoid degeneration or necrosis, while iso-/hypointensity reflects dense collagen fibers and sparse spindle cells. Concurrent hyperintensity suggests spindle cell-rich areas. Small DFSP lesions exhibit homogeneous enhancement, whereas larger tumors often show heterogeneous enhancement due to hemorrhage; cystic changes or calcifications are rare. The "double hypointense sign" (non-enhancing linear strands on T1WI/T2WI) may result from stromal fibroblasts or dense connective tissue aggregation, potentially a specific imaging feature of DFSP. The presence of the 'three-tail sign' in DFSP: skin tail sign (infiltration into adjacent skin, often accompanied by skin thickening), fat tail sign (moisture reaching the fat layer), and fascia tail sign (infiltration into the fascia layer), indicates a higher malignancy degree of the tumor [3]. Incomplete resection increases recurrence risk, and repeated recurrences damage peritumoral vasculature/lymphatics, shortening the recurrence-metastasis interval and elevating distant metastasis risk. Metastatic DFSP typically involves distant sites (e.g., lungs, liver) [4], consistent with this case. However, spinal epidural metastasis with vertebral involvement is unreported. MRI revealed no direct intraspinal invasion; metastasis likely occurred via postoperative recurrence and spread through spinal capillary/venous/lymphatic networks.

Intraspinal metastatic DFSP is rare. While classic DFSP morphology is characteristic, biopsy-based diagnosis remains challenging. Immunohistochemical findings are supportive but nonspecific. Most DFSP (80%-100%) express CD34 and vimentin, while factor XIIIa, desmin, SMA, S100, and keratin are negative. Differential diagnoses include: Adult-type fibrosarcoma: Lacks storiform architecture and subcutaneous infiltration; CD34-negative. Solitary fibrous tumor: STAT6 nuclear positivity with NAB2-STAT6 fusion. Synovial sarcoma: SS18-SSX fusion and CK/EMA expression. Intraspinal metastatic DFSP presents as a hypervascular extramedullary epidural mass, requiring differentiation from lymphoma [5], leukemia [6], hemangiopericytoma [7], tuberculous or infectious abscesses [8], and malignant solitary fibrous tumor [9,10]. A history of primary tumor recurrence is key. CD34 loss/weakness does not exclude DFSP; FISH testing is recommended (Figure 1-3).

Metastatic mechanisms of DFSP include: FS-DFSP was an aggressive subtype with 15%-20% metastasis risk (lung, bone, lymph nodes). Genetic drivers, such as PDGFB-COL1A1 fusion (t(17;22)) in 85%-95% cases activates PDGFR signaling; TP53 mutations and CDKN2A deletions disrupt cell cycle control. Tumor microenvironment plays some roles, for example, ECM remodeling via HIF-1/TGF-β/VEGF pathways creating "fibrotic channels" for tumor migration. Immune evasion may impair immune recognition, facilitating metastasis.

TEACHING POINT

DFSP is a rare low-grade malignant mesenchymal tumor originating from the skin's fibrous tissue, characterized by high local recurrence rates and strong invasiveness. Extramedullary spinal metastasis is extremely rare and needs to be differentiated from other malignant tumors such as solitary fibrous tumor.

QUESTIONS

Question 1: Which high-grade transformation is the most common pathological change in dermatofibrosarcoma protuberans ?

A. Fibrosarcomatous change (applies)
B. Pleomorphic sarcoma
C. Myxofibrosarcoma-like areas
D. Rhabdomyosarcomatous differentiation
E. Osteosarcomatous or chondrosarcomatous differentiation

Explanation: [Fibrosarcomatous change is the most frequent]. Architectural Pattern Dense, uniform storiform (cartwheel) pattern Long, intersecting fascicles in a herringbone pattern.Cellularity moderately was markedly hypercellular. Nuclear features were monomorphic, small, hyperchromatic nuclei with minimal pleomorphism. Pleomorphic, vesicular nuclei with prominent nucleoli were frequently seen. Pleomorphic sarcoma change is characterized by severe pleomorphism and a storiform pattern, but with high-grade

nuclear features. Feature of myxofibrosarcoma-like had areas of a myxoid stroma with long, curvilinear vessels and pleomorphic cells. Rhabdomyosarcomatous differentiation was seen the emergence of cells with rhabdomyoblastic features, which may express myogenic markers such as desmin and myogenin. Osteosarcomatous or chondrosarcomatous differentiation was characteristic by the presence of malignant osteoid production or chondroid matrix.

Question 2: Which are the common spindle cell tumors of the spine, with a focus on those in the differential diagnosis of dermatofibrosarcoma protuberans?

A. Fibroblastic variant meningioma (applies)
B. Schwannoma
C. Solitary fibrous tumor
D. Malignant peripheral nerve sheath tumor
E. Synovial sarcoma

Explanation: Meningioma is the most common spindle cell tumor in the spinal canal. Fibroblastic variant meningioma is composed of intersecting fascicles of spindled cells with elongated nuclei, often with whorls and psammoma bodies. Immunohistochemically, positive for EMA (Epithelial Membrane Antigen) and Vimentin. Often positive for SSTR2A (Somatostatin Receptor 2A). Negative for CD34 in the fibroblastic type, which is a key distinction from DFSP.

Question 3: Which is the preferred imaging method for dermatofibrosarcoma protuberans (DFSP) ?

A. Radiographic plain film
B. Ultrasound
C. CT scan
D. Magnetic Resonance Imaging (MRI) (applies)
E. Scintigraphy**Explanation:** Although MRI cannot replace pathological diagnosis, it has extremely high soft tissue resolution and can clearly display characteristic features of DFSP, such as the fat tail sign, skin tail sign, and fascial tail sign. [It accurately assesses the tumor's extent, its relationship with surrounding fascia and muscles, precisely delineates boundaries, shows infiltration depth, guides surgical planning, and serves as a baseline for postoperative follow-up and monitoring recurrence]. Therefore, it is the preferred imaging method before and after surgery.

Question 4: Which is the main value of CT scanning in the diagnosis of DFSP ?

A. Show characteristic calcifications within the tumor.
B. To estimate the presence of metastases in the lungs (applies)
C. As the preferred local assessment method before surgery
D. To differentiate between benign and malignant tumors
E. Assess the patient's liver and kidney function (excluding surgical contraindications).

Explanation: DFSP rarely calcifies, and the soft tissue resolution of CT is inferior to MRI, so it is not the preferred method for local assessment. [Its main value lies in performing chest CT scans to rule out distant metastasis and bone metastasis (although the rate of metastasis is low, it still needs to be evaluated)].

Question 5: Which is the most common site of distant metastasis for DFSP ?

A. Lung (applies)
B. Bones
C. Eidural space within the spinal canal
D. Liver
E. Lymphatic metastasis

Explanation: DFSP is a low-grade malignant soft tissue sarcoma, characterized by an extremely high local recurrence rate and a very low distant metastasis rate (<5%), primarily through hematogenous spread. Lymphatic metastasis is extremely rare, [and the lungs, with their rich capillary bed, become the most common target organ].

REFERENCES

[1] Bichakjian CK, Olencki T, Alam M, et al. Dermatofibrosarcoma protuberans, version 1.2014. J Natl Compr Canc Netw. 2014; 12(6): 863-868. PMID: 24925197.

[2] WHO classification of soft tissue and bone tumours. (5th edn). Lyon: IRAC Press, 2020: 100-103.

[3] Bergin P, Rezaei S, Lau Q, Coucher J. Dermatofibrosarcoma protuberans, magnetic resonance imaging and pathological correlation. Australas Radiol. 2007; 51 Spec No: B64-B66. PMID: 17875163.

[4] Zaraa, Cherif E, Ferchichi L, et al. Dermatofibrosarcoma protuberans. About 18 cases. Tunis Med. 2005; 83(10): 622-626. PMID: 16370214.

[5] Barz M, Aftahy K, Janssen I, et al. Spinal Manifestation of Malignant Primary (PLB) and Secondary Bone Lymphoma (SLB). Curr Oncol. 2021; 28(5): 3891-3899. PMID: 34677250.

[6] Berti PP, Pirillo V, Rizzo P, Ria A, Mannelli F, Broger M. Collision tumor between a spinal schwannoma and chronic lymphocytic leukemia/small lymphocytic lymphoma: case report and review of the literature. Br J Neurosurg. 2023; 37(5): 1266-1268. PMID: 33249871.

[7] Giordan E, Marton E, Wennberg AM, Guerriero A, Canova G. A review of solitary fibrous tumor/hemangiopericytoma tumor and a comparison of risk factors for recurrence, metastases, and death among patients with spinal and intracranial tumors. Neurosurg Rev. 2021; 44(3): 1299-1312. PMID: 32556679.

[8] Garg RK, Malhotra HS, Gupta R. Spinal cord involvement in tuberculous meningitis. Spinal Cord. 2015; 53(9): 649-657. PMID: 25896347.

[9] Yang C, Xu Y, Liu X. Spinal Intramedullary Solitary Fibrous Tumor: A Rare and Challenging Diagnosis. Cancer Manag Res. 2019; 11: 10321-10326. PMID: 31849521.

[10] Chen Y, Xu Z, Liu M, Xu H. Recurrent solitary fibrous tumor of the spinal cord: A case report and literature review. Clin Neuropathol. 2020; 39(2): 86-91. PMID: 31670648.

FIGURES

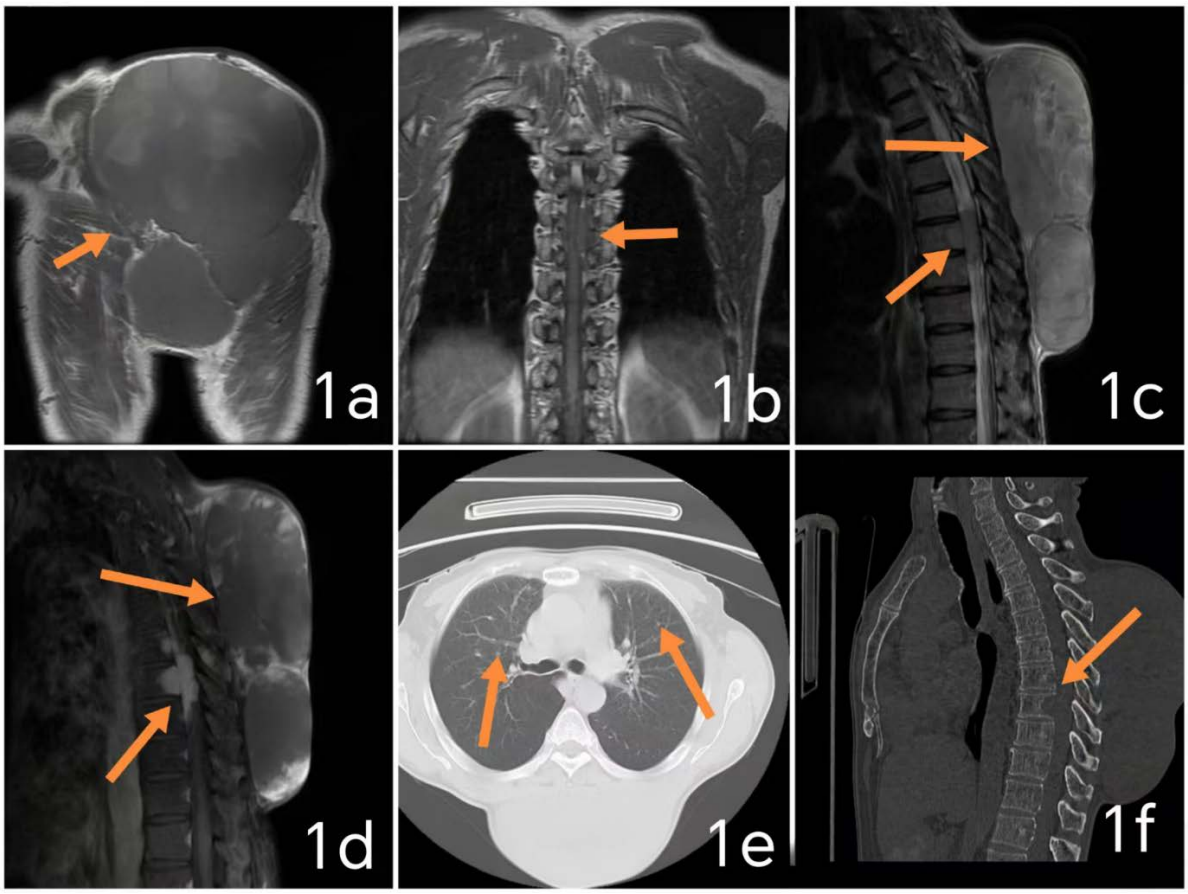

Figure 1: (1) In Figure 1a(arrowhead), on T1WI, the dorsal skin mass shows mostly isosignal, with multiple slightly hyperintense signals within the lesion, and linear hypointense septations can be seen locally. In Figure 1c(long arrowhead), on T2WI, the lesion shows mostly hyperintense signal, with isointense and slightly hyperintense signals within the lesion, and linear hypointense septations can be seen locally. Fat tail sign, skin tail sign, and fascia tail sign can be observed. In Figure 1d (long arrowhead), on enhanced scanning, multiple patchy areas of significant enhancement of varying sizes can be seen at the edge of the dorsal mass and within the lesion. (2) At the level of the T7 - T8 vertebral bodies, there are strip - shaped and nodular masses in the epidural space of the spinal canal. In Figure B (arrowhead), on T1WI, it shows isosignal, and in Figure 1c(short arrow), on T2WI, it shows slightly hyperintense signal. The mass invades the posterior edges of the adjacent T7 and T8 vertebral bodies. In Figure 1d (short arrow), on enhanced scanning, the mass shows significant enhancement, compressing the dural sac and spinal cord. Diagnosis: Subcutaneous soft - tissue mass on the back, consistent with the manifestations of dermatofibrosarcoma protuberans; mass in the epidural space of the spinal canal at the T7 - T8 level, involving the posterior edges of the T7 and T8 vertebral bodies, considered as metastatic tumor. (2) CT diagnosis: Subcutaneous soft - tissue mass on the back, consistent with the manifestations of postoperative recurrence of dermatofibrosarcoma protuberans; nodules in both lungs (Figure 1e) (arrowhead), and bone destruction of the T7 and T8 vertebral bodies (Figure 1f) (arrowhead), considered as metastases.

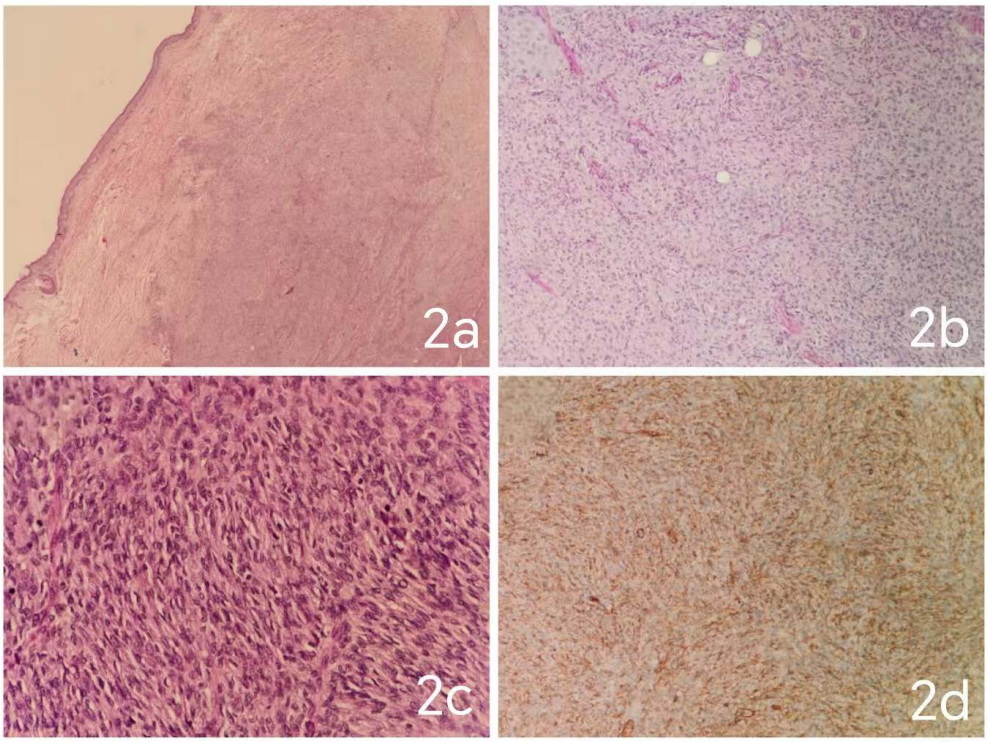

Figure 2: Skin Tumor: **2a**: Tumor in the superficial dermis and subcutaneous tissue, densely arranged (HE×40); **2b**: The tumor invades the subcutaneous adipose tissue (HE×100); **2c**: In focal areas, the tumor is densely arranged in bundles, and mitotic figures are easily visible (HE×200); **2d**: Immunohistochemistry shows that tumor cells are diffusely positive for CD34 (IHC×200).

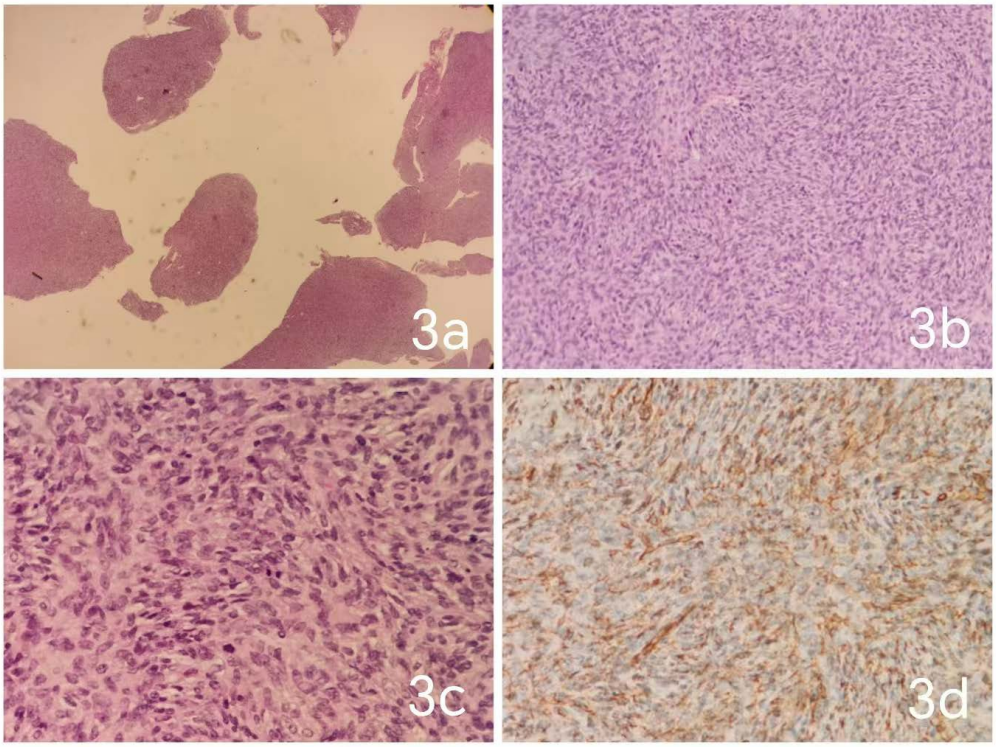

Figure 3: Intraspinal mass: **3a**: Fragments of tumor tissue under low power microscopy, with dense arrangement of tumor tissue (HE×20); **3b**: The tumor shows a cartwheel-like arrangement (HE×100); **3c**: The tumor cells are densely arranged, and mitotic figures can be seen (HE×200); **3d**: Immunohistochemistry shows diffuse positive CD34 in tumor cells (IHC×200).

KEYWORDS

Dermatofibrosarcoma protuberans; Spinal canal metastasis; Imaging features; Pathological diagnosis; Differential diagnosis

ABBREVIATIONS

DFSP = Dermato Fibro Sarcoma Protuberans
MRI = Magnetic Resonance Imaging
CT = Computed Tomography
FS = Fibro Sarcomatous
T1WI = T1-Weighted Imaging
T2WI = T2-Weighted Imaging
WHO = World Health Organization
FISH = Fluorescence In Situ Hybridization