
CASE REPORT

Radiological Findings of a Large Dermatofibrosarcoma Protuberans with Pathological Correlation

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare, low- to intermediate-grade soft tissue sarcoma that is fibrohistiocytic in nature and originates from the dermis. Dermatofibrosarcoma protuberans is a locally aggressive tumour that invades deep subcutaneous tissues and has high recurrence rates. Radiological investigation is important to narrow down the differential diagnoses and to evaluate the local extent, particularly the level of infiltration of the tumour. Tumour infiltration is important for preoperative planning, especially for large tumours, as clinical examination alone may not be completely accurate and local recurrence is common, particularly for tumours with inadequately resected margins. Magnetic resonance imaging, with its superior soft tissue contrast, is particularly helpful. This report is of a patient with a large dermatofibrosarcoma protuberans on the right side of the anterior abdominal wall, and describes the radiological appearance and histopathological correlation.

Key Words: Dermatofibrosarcoma; Magnetic resonance imaging; Radionuclide imaging; Tomography, X-ray computed; Ultrasonography

中文摘要

巨大隆突性皮膚纖維肉瘤的放射學表現及其與病理學的相關性

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隆突性皮膚纖維肉瘤是一種罕見的低到中度惡性軟組織肉瘤，性質屬纖維組織細胞，起源於真皮層。它是一種侵入深層皮下組織的局部侵襲性腫瘤，復發率高。放射學檢查對縮小鑑別診斷範圍，評估腫瘤的局部範圍，尤其是浸潤程度很重要。腫瘤浸潤度對術前計劃相當重要，尤其對大體積腫瘤而言；單靠臨床評估可能不完全準確，同時局部復發很常見，特別對切除邊緣不完全的腫瘤。磁共振成像軟組織對比度高，特別有助於評估。本文報告一宗右腹前壁巨大隆突性皮膚纖維肉瘤的病例，並描述其放射性影像，以及其組織病理學相關性。

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), a tumour described by Hoffman in 1925,¹ is a rare slow-growing soft tissue tumour originating from the dermis. DFSP comprises about 2.0% of all soft tissue sarcomas.^{2,3} Diagnosis of small and superficial lesions may be made by clinical appearance. However, radiological investigation may be necessary to evaluate the complete extent of larger tumours, as local recurrence is common, especially for tumours with inadequately resected margins. DFSP is usually staged according to the American Musculoskeletal Tumor Society staging system, based on histological grade, local extension, and presence of metastasis.⁴ DFSP can also be staged in accordance with the 7th American Joint Committee on Cancer Staging for sarcoma based on tumour size and depth, nodal involvement, distant metastasis, and histological grade.⁵

This report presents the radiological findings of a patient with a large soft tissue mass over the anterior abdominal wall. Subsequently, the patient was referred to a surgeon for excision, and histopathology confirmed the tumour to be DFSP. As the diagnosis of DFSP can be made clinically, the imaging appearance is underreported in the literature. This report discusses the combined imaging spectrum of ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), and scintigraphy correlating with the pathological findings of DFSP, thus reiterating the importance of imaging, especially for large tumours.

CASE REPORT

A 45-year-old man presented in July 2011 with a mass on the right side of the anterior abdominal wall that had slowly, but progressively, increased in size over the previous 10 years. He had no history of weight loss or bone pain. Clinical examination revealed a non-tender firm oval mass measuring approximately 7 x 6 cm over the right hypochondriac and epigastric regions, with palpable dermal induration and adhered overlying skin (Figure 1). The mass was mobile over the anterior abdominal wall muscles.

Using a linear array transducer, ultrasonographic evaluation of the mass showed a large oval tumour with well-defined margins in the subcutaneous fat layer. There was a predominantly hypoechoic echo pattern with internal septations and some heterogeneity (Figure 2), and low blood flow in the periphery of the mass



Figure 1. A large flesh-coloured nodular mass over the right side of the anterior abdominal wall.

on colour Doppler imaging. No calcification was seen within the mass.

CT revealed a well-defined lobulated soft tissue mass (mean Hounsfield units, 32), measuring 3.6 cm x 5.1 cm x 5.2 cm in the subcutaneous tissue on the right side of the anterior abdominal wall. Posteriorly, the lesion was superficial to the rectus muscle, and intervening fat planes in the right rectus muscle were lost in a few places (Figure 3a). Two satellite nodules measuring 1.0 x 2.5 cm and 2.0 x 1.3 cm were noted adjacent to the mass. The overlying skin was stretched over the mass and was not separately identifiable. No calcification was noted within the mass. On post-contrast images, the tumour showed moderate inhomogeneous enhancement, with poorly enhancing intervening areas. Sagittal maximal intensity projection (MIP) images delineated the vascularity of the mass very well (Figure 3b). No significant lymphadenopathy was noted.

MRI revealed a well-delimited lobulated subcutaneous mass of altered signal intensity over the right anterior abdominal wall. The mass appeared hypointense to fat and isointense to skeletal muscles on T1-weighted images (Figure 4a). On T2-weighted images, the mass appeared hypointense to fat and hyperintense to muscle. Short tau inversion recovery images showed the mass to be hyperintense (Figure 4b). Diffusion-weighted imaging showed restricted diffusion.

After injecting ^{99m}technetium-methylisobutyl isonitrite

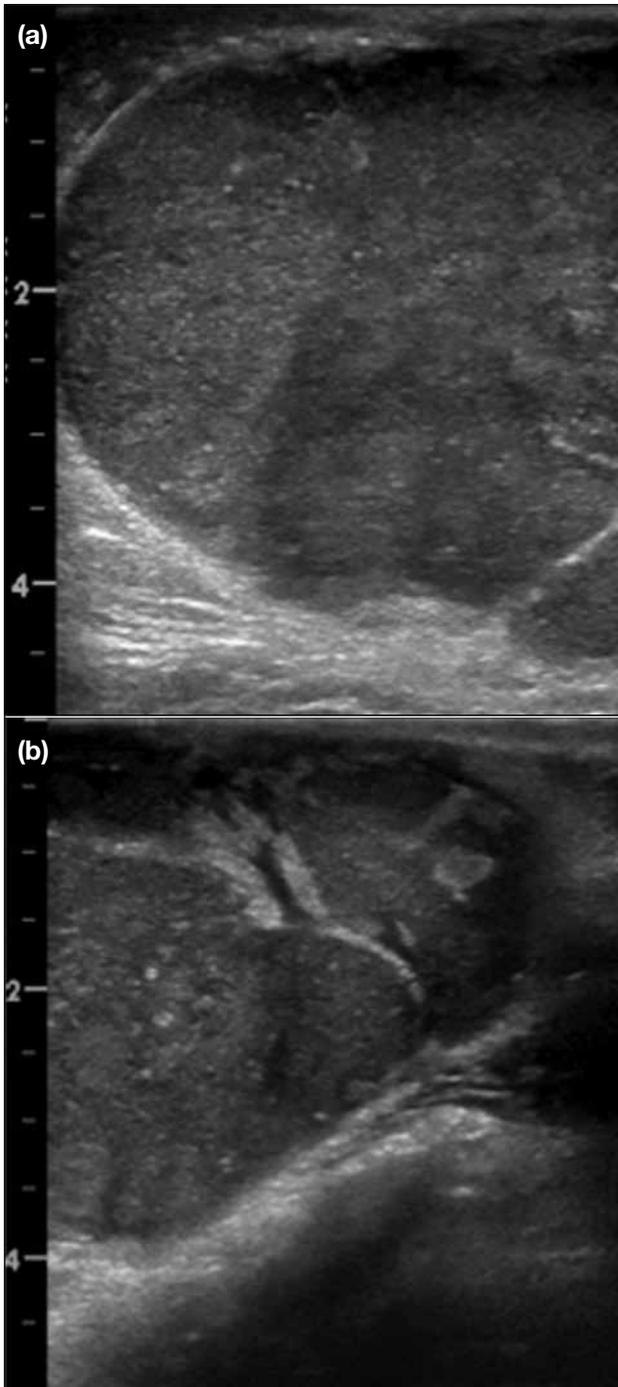


Figure 2. B-mode ultrasonographic images of dermatofibrosarcoma protuberans show (a) a well-defined oval heterogenous, but hypoechoic, mass in the subcutaneous fat layer of the anterior abdominal wall and (b) an internal septation within the mass.

15 mCi intravenously, dynamic scintigraphic images of the thorax and upper abdomen were acquired for 1 minute, followed by static images at 5 minutes, 30 minutes, and 2 hours. Physiological uptake was seen in the heart, liver, spleen, gall bladder, intestines, and

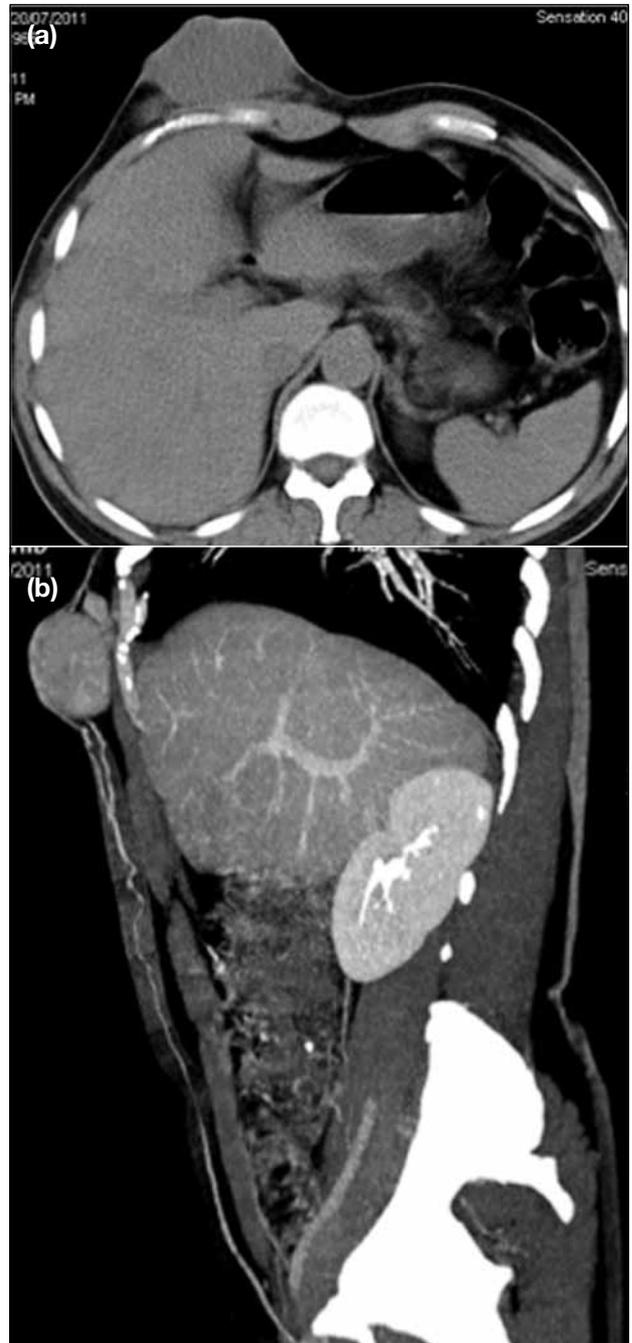


Figure 3. Computed tomography images of dermatofibrosarcoma protuberans: (a) an axial unenhanced view showing a well-circumscribed subcutaneous mass with intermediate density on the right side of the anterior abdominal wall and a satellite nodule, and (b) a sagittal maximum intensity projection view showing the vascularity of the mass.

kidneys. The initial perfusion phase showed increased tracer accumulation in the upper abdomen anterior to the liver corresponding to the site of the lesion and suggestive of increased vascularity. The blood pool phase showed an abnormal tracer accumulation at the

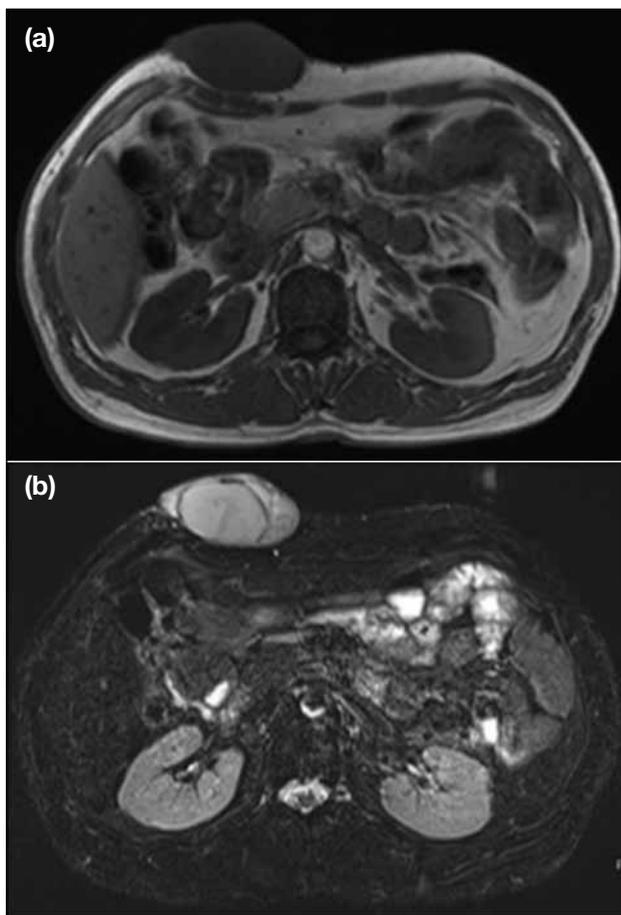


Figure 4. Magnetic resonance images of the abdomen: (a) an axial T1-weighted image demonstrating a well-defined nodular mass in the subcutaneous adipose tissue of the anterior abdominal wall with intermediate signal intensity that was hypointense to fat and isointense to muscle, and (b) an axial short tau inversion recovery image showing the high signal mass that was hyperintense to fat and skeletal muscle.

same site. However, no significant tracer accumulation was seen on delayed images. The tumour was surgically removed using a wide excision of 2-cm margins. No adjuvant chemo- or radio-therapy was given.

The cut surface of the excised surgical specimen revealed a grey white bosselated tumour measuring 5.0 cm x 4.5 cm x 3.0 cm. The tumour was well circumscribed and extended up to the deep resected margin. Haematoxylin and eosin stained sections revealed densely packed spindle shaped cells arranged in a storiform pattern (Figures 5 and 6). The tumour was infiltrating into the underlying subcutaneous fat, but was 1 mm away from the deep resected margin. A diagnosis of DFSP was confirmed.

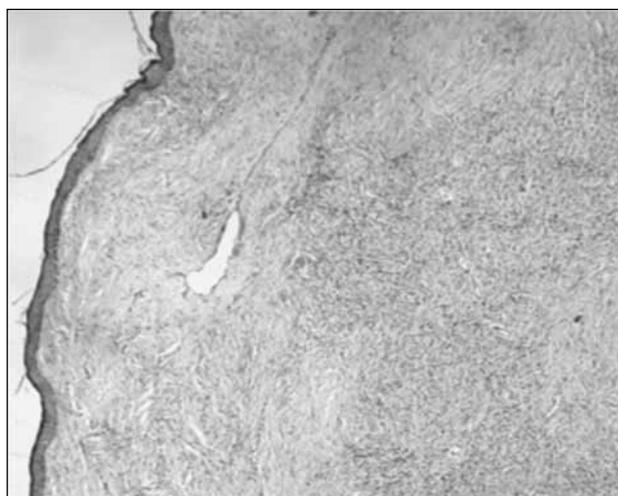


Figure 5. A photomicrograph shows spindle-shaped tumour cells occupying the dermis. A thin grenz zone separates the tumour from the epidermis (H&E; original magnification, x 40).

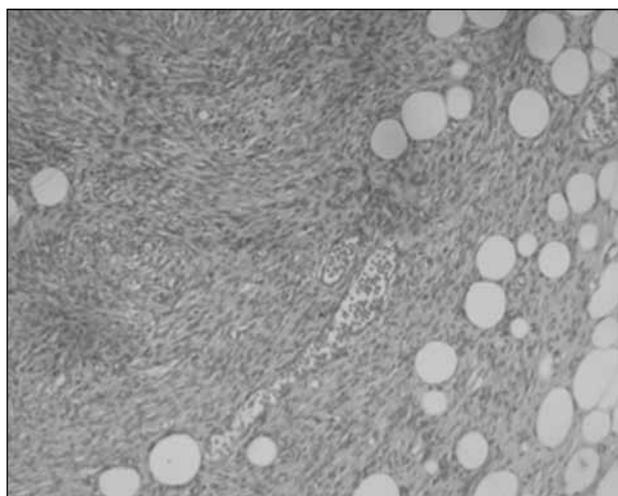


Figure 6. A high-power photomicrograph highlighting the typical storiform pattern shows the tumour infiltrating into the underlying subcutaneous fat (H&E; original magnification, x 100).

DISCUSSION

DFSP is an uncommon tumour with low-to-intermediate malignant potential. This report is of a rare DFSP on the anterior abdominal wall and the radiological and histopathological correlation. Among the radiological investigations, ultrasonography is the most easily accessible and cost-effective modality, and details the composition and marginal infiltration of the tumour in a non-invasive manner.⁶ According to the literature, if an oval mass with lobulated margins and mixed to hypoechogenicity is encountered in the subcutaneous tissue, the possibility of DFSP should be considered.^{6,7}

On CT, earlier reports have identified DFSP as an unmineralised well-defined nodular soft tissue mass with tissue attenuation equal to or higher than skeletal muscle and moderate enhancement after intravenous contrast administration.⁸ CT helps to decide the line of incision, thus avoiding inadequate excision. The newer post-processing techniques such as volume-rendered images and MIP images can delineate the vascularity of the mass, as was the case for this patient.

MRI, with its high soft tissue resolution and multiplanar capability, is the superior imaging modality for such patients. MRI facilitates the surgical planning and assessment of depth of tumour extension and involvement of critical structures.⁹ MRI also helps to establish the diagnosis of difficult and atypical lesions such as large-sized tumours, those with a suspected deeper component, recurrent tumours, those at critical anatomic locations, and re-excision of tumours with positive surgical margins. The MRI appearance is non-specific, showing mostly low signal intensity on T1-weighted images, and high signal intensity on T2-weighted images. MRI also shows moderate-patchy enhancement on post-gadolinium scans.⁸⁻¹¹

Scintigraphic findings have shown abnormal tracer accumulation at the site of the mass in the blood pool phase.¹²

Histologically, the tumour shows densely packed spindle-shaped cells arranged in a characteristic storiform pattern on a background of fibrous stroma and infiltration of surrounding subcutaneous fat.¹³⁻¹⁵

Radiological evaluation confirms the clinical diagnosis and shows the extent of the tumour. Such evaluation provides the surgeon with the extent and location of this locally aggressive entity, which helps with surgical planning, and improves the overall care of the patient.

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