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CASE REPORT

# Dermatofibrosarcoma protuberans with contiguous infiltration of the underlying bone

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**Abstract**

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade, cutaneous neoplasm with pronounced tendency for local recurrence. A case of DFSP that showed direct infiltration into the underlying bone marrow is described. To the best of our knowledge, such direct bony involvement by dermatofibrosarcoma has not been reported in the English literature to date. The role of imaging is also discussed for planning adequate initial treatment, which will result in a lower recurrence rate and improved clinical outcome.

**Keywords:** *Dermatofibrosarcoma protuberance; bone infiltration; recurrence; imaging.*

## Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, low-grade cutaneous malignancy that originates from the dermis[1]. It can occur in any part of the body, but has predilection for the trunk, head and neck region, and the proximal extremities [2]. The tumor begins as a small bump on the skin surface and then grows in the dermis to form a larger mass that swells and bulges outwards, hence the name 'protuberans'. Distant metastasis of DFSP is extremely rare; however, it is a locally aggressive tumor and shows marked tendency to recur even after wide local excision.[3,4] We emphasize the role of imaging in the preoperative evaluation of these tumors so that more accurate treatment plans can be instituted, leading to decreased recurrence rates.

## Case report

A 20-year-old male presented to our hospital with a progressively increasing swelling on the right leg for the previous 9 months. It was painless and non-tender to start with, but showed rapid growth over the last few weeks with onset of pain that was continuous and mild to moderate in intensity. There was no history of any fever, loss of weight or any trauma to the local site. Complete blood cell counts and biochemical laboratory parameters were within normal limits. The physical examination revealed an irregular, protuberant swelling with a large indurated, non-ulcerated plaque on the right leg. The swelling appeared to be fixed to the underlying bone. No regional lymphadenopathy was noted.

Radiography of the right leg revealed a rounded soft tissue mass approximately $5 \times 3$ cm in size along the anteromedial aspect of the right tibia with associated cortical erosions in the tibia (Fig. 1A,B). No soft tissue calcification or periosteal reaction was noted. Magnetic resonance imaging (MRI) was performed to further characterize the mass and delineate the extent of bone marrow involvement. The bone marrow of proximal metadiaphysis of the right tibia over a segment of approximately 7 cm revealed an abnormal hypointense signal on T1-weighted images and hyperintense signal on inversion

recovery and T2-weighted images (Fig. 2A,B). There was evidence of cortical breach along the anterior aspect of the tibia with a large associated soft tissue mass that was infiltrating the subcutaneous tissues and bulging anteriorly. The soft tissue was predominantly hypointense on T1-weighted images and hyperintense on T2-weighted images depicting florid enhancement after contrast administration. The underlying bone marrow also showed heterogenous post-contrast enhancement (Fig. 3A,B). These findings were suggestive of an unresectable malignant sarcomatous tumor.

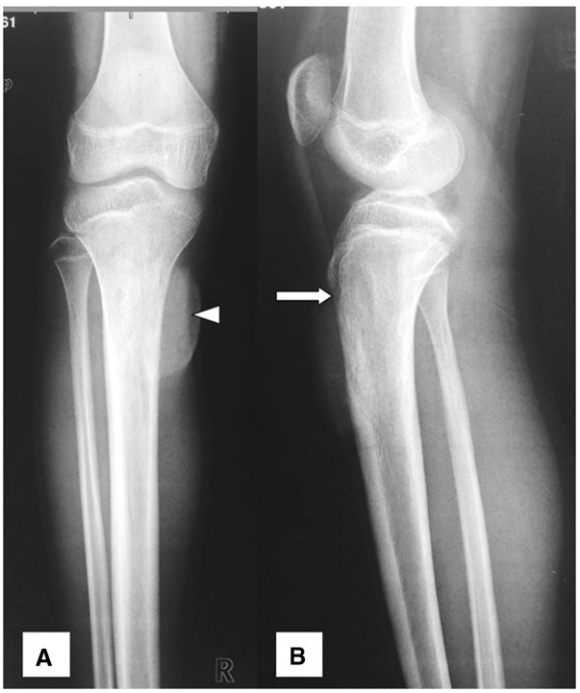


*Figure 1* **Radiographs of the right leg, anteroposterior (A) and lateral projection (B) showing a well-defined soft tissue mass along the anteromedial aspect of the right tibia (arrowhead, A) with suggestion of cortical breach (arrow, B).**

The patient underwent core-needle biopsy under local anaesthesia from swelling on the right leg which showed highly cellular smears with clusters of spindle-shaped cells in a fibromyxoid background. The cells showed moderate pleomorphism and storiform pattern in places (Fig. 4A). These features were suggestive of DFSP. The stains for CD34 were positive on immunohistochemistry (Fig. 4B). The patient is scheduled for neoadjuvant therapy with imatinib mesylate to be followed by surgical excision of the tumor.

## Discussion

Dermatofibrosarcoma protuberans is a rare, painless, monoclonal, cutaneous soft tissue sarcoma of unknown cause that was first described by Taylor in 1890[5]. It constitutes approximately 1% of all soft tissue sarcomas, shows slight male preponderance and generally presents in the third and fourth decades of life[6,7]. Its clinical manifestations are non-specific. It often masquerades as a benign, indolent tumor on the trunk and proximal extremities. The surface of the tumor is characterized by irregular protuberant swellings, and a hard indurated plaque of irregular outline forms the base. It is a slow-growing neoplasm; however in the later stages, it may show rapid

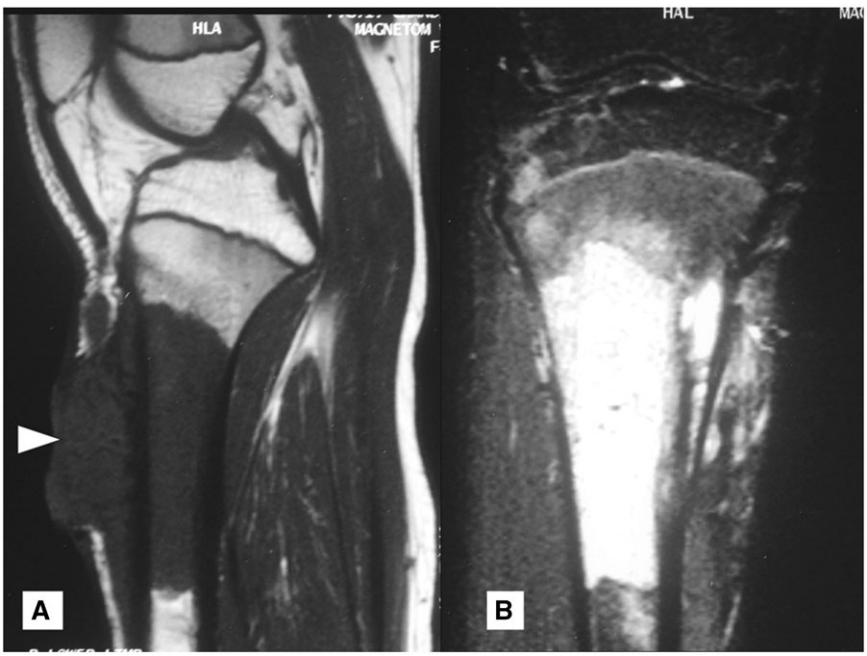


*Figure 2* **MRI sagittal T1-weighted (A) and fat suppressed coronal T2-weighted images (B) demonstrating abnormal marrow signal of the proximal metadiaphysis of the right tibia with sharp zone of transition. Also note the large soft tissue seen anteriorly (arrowhead, A).**

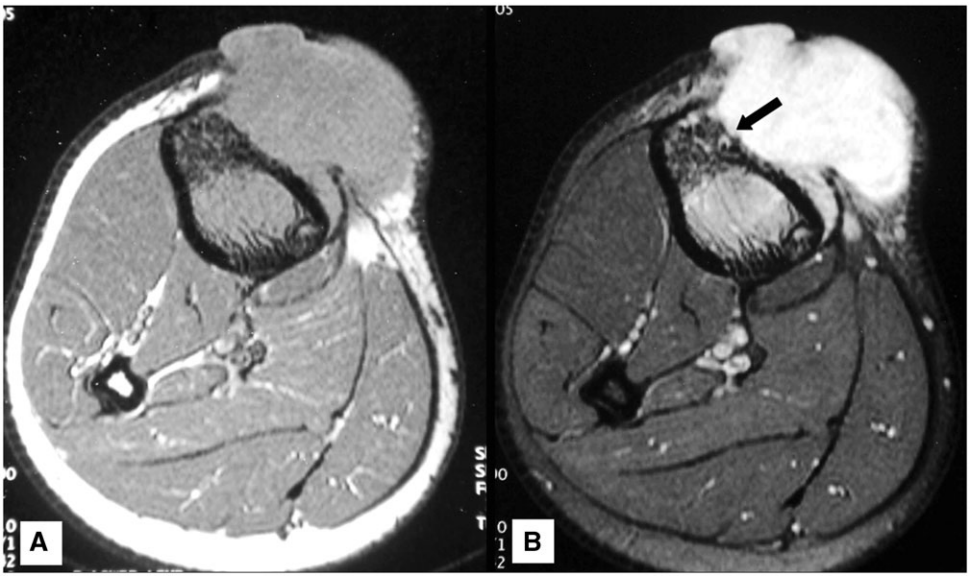


***Figure 3*** **MRI axial T1-weighted pre-contrast (A) and post-contrast (B) images depicting large soft tissue bulging anteromedial to the right tibia and showing florid enhancement after contrast administration. Mild heterogeneous enhancement is also noted in the underlying bone marrow. The soft tissue is seen to infiltrate the subcutaneous planes associated with cortical breach in the tibia anteriorly (arrow, B).**

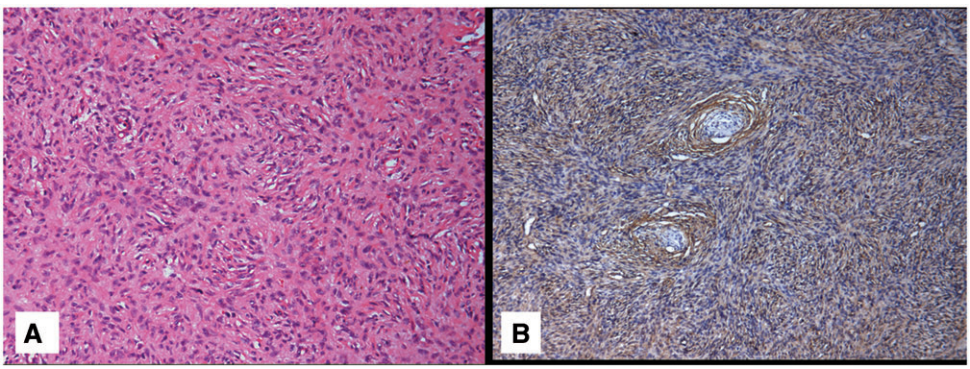


***Figure 4*** **Biopsy smear showing highly cellular tumor with fibroblastic spindle cells producing storiform pattern (A) and immunohistochemical stain for CD34 (B) showing strong membranous positivity around the tumor cells.**

growth and the lesion may become painful, ulcerated and can discharge.

Surgery with wide local excision or Mohs micrographic surgery remains the treatment of choice in these patients[8]. Radiation therapy, hitherto with a limited role, is recently being used as an adjunct to surgical excision, primarily in cases in which the tumor is unresectable, there are positive resection margins on histology and when the surgery is going to leave major cosmetic or functional deficits. Chemotherapy has rarely been used in the treatment of DFSP. Recently, a very promising neoadjuvant molecular therapy with imatinib mesylate, a potent and selective inhibitor of several protein-tyrosine kinases, is advocated in adult patients with unresectable, recurrent or metastatic DFSP[9].

DFSP is a locally invasive fibrohistiocytic tumor with a recurrence rate of up to 25% even after wide local excision with a safety margin of 3–5 cm[4]. This is attributed to the fact that DFSP extends far beyond the assessed clinical margins, spreading locally in the dermis, subcutaneous tissues and muscles. Our index case illustrates that it can extend even deeper to directly involve the bone with cortical erosions and bone marrow infiltration. To the best of our knowledge such deep invasion of the tumor with infiltration into the underlying bone has not been described in the literature to date except in one case of DFSP of the scalp where the calvarium was also infiltrated[10]. This could partly be due to the fact that imaging is rarely a part of the preoperative workup in these patients.

DFSP is a low-grade malignancy with only a few sporadic cases of distant metastases[11в13]. The diagnosis of DFSP generally comes from core-needle or incisional biopsy. Histologically, it consists of uniform but atypical spindle cells placed in a fibrotic stroma in the dermis and subcutaneous tissues exhibiting characteristic 'cartwheel' or 'storiform' appearance[4]. Immunohistochemistry studies have shown that DFSP stains positive for CD34 and negative for factor XIIIa, which are very useful markers to differentiate DFSP from dermatofibroma.[9] The poor prognostic indicators of DFSP include advanced age, high mitotic index, increased cellularity, DNA aneuploidy and fibrosarcomatous changes within the tumor[8]. Multiple local recurrences can also predispose to distant metastasis, thus highlighting the significance of adequate initial treatment.

The extent of the tumor and the degree of invasion of underlying tissues is usually assessed by physical examination and not much importance has been given to the role of imaging except to rule out metastasis. Mendenhall et al.[5] and McArthur[9] discussed the role of MRI in determining deep tumor invasion. Because DFSP is a locally invasive tumor with a high rate of post-operative recurrence, we advocate that imaging (preferably MRI) should be done in all such cases especially those involving the extremities. MRI will ascertain the exact depth of invasion by showing the tumor margins, and will guide the clinician to institute an adequate initial treatment by choosing an appropriate surgical plan with or without adjunctive chemo-/radiotherapy. This in turn will lead to a reduced rate of local recurrence and improved clinical outcome. However, the cost/benefit ratio needs to be further substantiated by larger prospective studies.

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