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## Undifferentiated Pleomorphic Sarcoma: Indolent, Tail-like Recurrence of a High-grade Tumor

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### Abstract

Recurrence of a soft tissue sarcoma typically manifests as a round or oval mass at imaging, and recurrent high-grade soft tissue sarcomas generally enlarge relatively rapidly. We present a case of high-grade undifferentiated pleomorphic sarcoma in the calf of a 48-year-old male that recurred as a thin, curvilinear “tail” of enhancing tissue at magnetic resonance imaging (MRI), with extremely indolent growth over a 7-year period. The unusual imaging finding of a slowly enlarging “tail” should not be dismissed as postoperative changes, even for a high-grade soft tissue sarcoma.

### Keywords

Undifferentiated pleomorphic sarcoma; Malignant fibrous histiocytoma; Soft tissue sarcoma; Postoperative change; Magnetic resonance imaging

### Introduction

Undifferentiated pleomorphic sarcoma (UPS) (previously termed malignant fibrous histiocytoma) [1], often presents as a palpable mass in an extremity. At CT or MRI, UPS generally manifests as a rounded or oval mass that heterogeneously enhances and contains areas of necrosis. UPS, as well as a related tumor, myxofibrosarcoma (MFS), may have an infiltrative border with tail-like extensions at imaging [2]. Such extensions of tumor are believed to account for the high propensity for these tumors to recur locally. Recurrence generally occurs within the first two years after surgery, again usually appearing as a round

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or oval mass in or near the surgical bed [2, 3]; however, a minority of cases of UPS and MFS recur only as tail-like thickening along fascia [2].

We present an unusual case of recurrent high-grade UPS that grew extremely slowly in a tail-like configuration, to draw attention to the fact that, rarely, such findings do represent recurrent tumor and should not be misinterpreted as “evolving postoperative changes.”

## Case Report

A 48-year-old male presented to an outside facility for evaluation of a palpable mass in the proximal anterior left calf that had been present for one month. A dermatologist unsuccessfully attempted to aspirate the mass. The patient then underwent MRI of the calf, demonstrating a  $3.0 \times 1.8$  cm lobulated solid mass in the subcutaneous tissues, extending deep to the fascia and contacting the tibial cortex, with a small enhancing tail of tumor extending along the fascia anteriorly (Fig. 1). Subsequent biopsy and resection were performed by a general surgeon elsewhere. Histopathologic examination of the outside slides from resected specimen showed a spindle and pleomorphic sarcoma with storiform growth, diffusely infiltrating the subcutaneous tissue and fascial planes, consistent with UPS. Surgical margins were positive for tumor, and amputation was recommended.

The patient then presented to our institution, and wide re-resection of the tumor and the surgical bed was performed 27 days after the initial resection. Multiple foci of UPS were present, the largest measuring 2.1 cm in diameter, with microscopic foci surrounding the biopsy cavity. UPS infiltrated dermis, deep dermis, and skeletal muscle. The surgical margins were positive at the lateral and inferior margins. The patient then underwent a course of adjuvant radiation treatment to the tumor bed (total dose, 63 Gy). The re-resection specimen showed a high grade spindle and pleomorphic sarcoma with a diffuse growth pattern similar to the prior submitted material (Fig. 2). The patient had an uneventful postoperative course, and was followed with serial contrast-enhanced MRI at various facilities, at approximately 6–10-month intervals. The postoperative MRIs showed small amounts of linear and reticular signal changes in the subcutaneous tissues in and near the surgical bed, presumed by the interpreting radiologists to represent postoperative changes; most of these decreased over time. The radiologist interpreting the MRI obtained 7.3 years after resection noted that a curvilinear, tail-like band of T2-hyperintense, enhancing tissue measuring a few millimeters thick was located along the muscular fascia in the subcutaneous tissues in and beyond the operative bed. Upon additional directed review, it became clear that this tissue had been enlarging extremely slowly, both in its extension circumferentially along the fascia and in overall thickness (initially 0.1 cm thick, increasing in size to 0.4 cm at 7 years) — a fact that became evident only by comparing the most recent scan to the most remote postoperative scan (Fig. 3 a–d). A tail of tumor had been present in this region on the initial MRI (Fig. 1b), but appeared less prominent on the next subsequent scan.

At this time, the possibility of recurrence was suggested to the surgeon, who performed a biopsy that confirmed recurrent UPS (Fig. 4), similar in appearance to the original tumor. The patient underwent resection of the recurrent UPS, without wound closure. Histopathologic examination revealed two positive margins. Two subsequent re-excisions

were performed, resulting in negative margins. Temporary occlusive dressings were applied following the two resections until negative histologic margins were confirmed on permanent histopathologic examination. A reconstruction of the left calf wound then was performed with a left anterolateral thigh free-flap, and split-thickness skin graft.

The patient has not had any metastatic disease, and remains disease-free 15 months after the most recent resection. He notes some swelling of his left foot and ankle, but is otherwise asymptomatic in his left lower extremity.

## Discussion

UPS is one of the most common soft tissue sarcomas in adults, showing a slight male predilection and an average age of 59 years at presentation, manifesting as a palpable, painless, slowly enlarging mass within an extremity [4]. UPS may demonstrate aggressive regional infiltration at presentation, with tumor spreading from the dominant mass in a “tail-like” fashion. Such tumor extension is often underestimated on preoperative MRI, which is thought to demonstrate only macroscopic tumor. These tails of tumor are believed to account for the high propensity of these tumors to recur following treatment [2]. A wider surgical margin than is typically determined from clinical examination or imaging is required to minimize the risk of local recurrence. Given these facts, surgeons at our institution currently recommend leaving the tumor bed open and applying a temporary dressing or wound vacuum until results of histopathologic examination indicate that the surgical margins are negative; once confirmed, the surgeon performs final wound closure.

Microscopic residual tumor may remain in the surgical bed after initial soft tissue sarcoma resection, often along vessels or nerves that contacted the tumor [5]. Adjuvant radiation treatment has been reported to decrease, but not entirely eliminate, the risk of soft tissue sarcoma recurrence, in part by destroying these residual tumor cells. In the case presented, a small, tail-like area of enhancement was present in the initial surgical bed, and it is not possible to be certain that it was entirely resected, given the presence of postoperative changes in that region on first post-resection MRI (not shown). Even on the MR image obtained 3.4 years after re-resection (Fig. 3a), the enhancing tail seems less prominent than on the initial preoperative image (Fig. 1b), suggesting that the original tail had been resected. Also, any local recurrence of sarcoma is presumed to be due to growth of viable tumor cells that were not excised at surgery or killed with chemotherapy or radiation therapy. However, this point would be difficult to prove, making the distinction between “recurrent tumor” and “slowly growing residual tumor” necessarily somewhat arbitrary.

Sarcoma recurrences, which are more common in high-grade tumors than in low-grade ones, may have adverse implications for limb preservation and overall patient survival [6,7]. The majority of soft tissue sarcoma recurrences occur in the first two years after the initial surgical resection, though tumor may recur at any point during a patient’s lifetime [3]. Local recurrences of sarcomas typically manifest as an ovoid or round mass at imaging, and enlarge relatively rapidly [8]; however, Yoo et al [2] recently reported that a minority of UPS and MFS recurrences grow only as plaque-like tails of enhancing tissue, without associated discrete masses. We concur with their assessment that focal, thickened fascial enhancement

in a tail-like configuration is consistent with tumor involvement. Detailed time courses of tail-like recurrences of UPS were not specified in that report, however.

After initial surgery for soft tissue sarcoma, patients are typically followed with serial MRIs, which optimally should be performed with use of an intravenous gadolinium contrast agent, to assist in detection of subclinical recurrence [9]. Contrast material is particularly important in evaluation for recurrent UPS or MFS at MRI, as the tails of such tumor enhance, facilitating their detection and distinction from post-treatment edema [2,10].

After resection of a primary UPS or MFS, surveillance MRI that demonstrates curvilinear, T2-hyperintense, enhancing tissue located along fascial planes in or adjacent to the operative bed should strongly suggest the presence of recurrent tumor, rather than benign postoperative changes. Furthermore, postoperative changes should decrease or remain unchanged in size and extent over time; any enlargement should be considered at least suspicious for recurrent tumor. The case presented shows that enlargement of a tail of recurrent UPS can be extremely slow, despite the high-grade histology of the tumor. To appreciate such minimal growth, it is essential to meticulously compare current MRI findings with those on the most remote postoperative scans. Most radiology studies are displayed on picture archiving and communication systems (PACS), which usually are configured to automatically display the current study and the most recent prior study of the same type. Although time consuming, extra effort should be expended to also compare to the most remote studies to allow the radiologist to more promptly diagnose an unusual, slow-growing recurrence. We can find no additional reported case of such indolent growth in the literature, nor can we explain the discrepancy between the indolent biologic behavior and high-grade histology.

This case emphasizes that even subtle changes in postoperative MRI findings in soft tissue sarcomas that have a known tail-like growth pattern (predominantly UPS and MFS) should not be ignored or dismissed without meticulous comparison to remote prior examinations. Also, it is important to recognize that even a high-grade UPS rarely may exhibit extremely slow growth of a tail as the sole manifestation of recurrence, with only minimal enlargement over a period of multiple years. Thus, any enlargement of enhancing, tail-like tissue should be considered suspicious for recurrent UPS, and biopsy recommended.

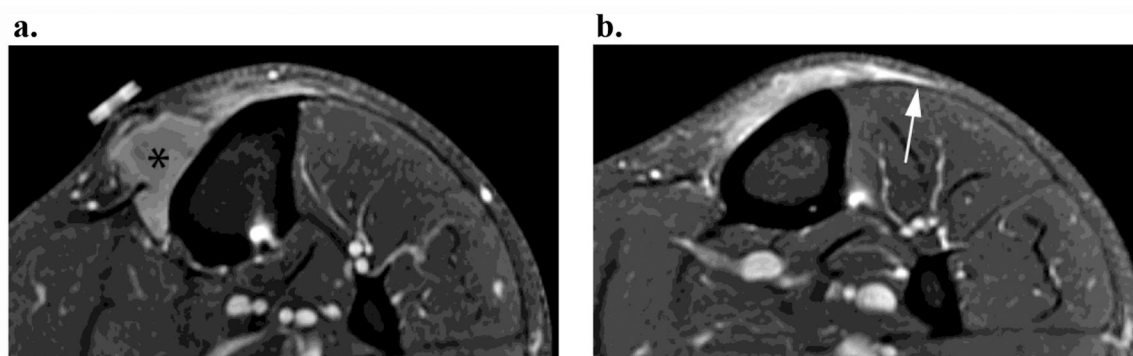
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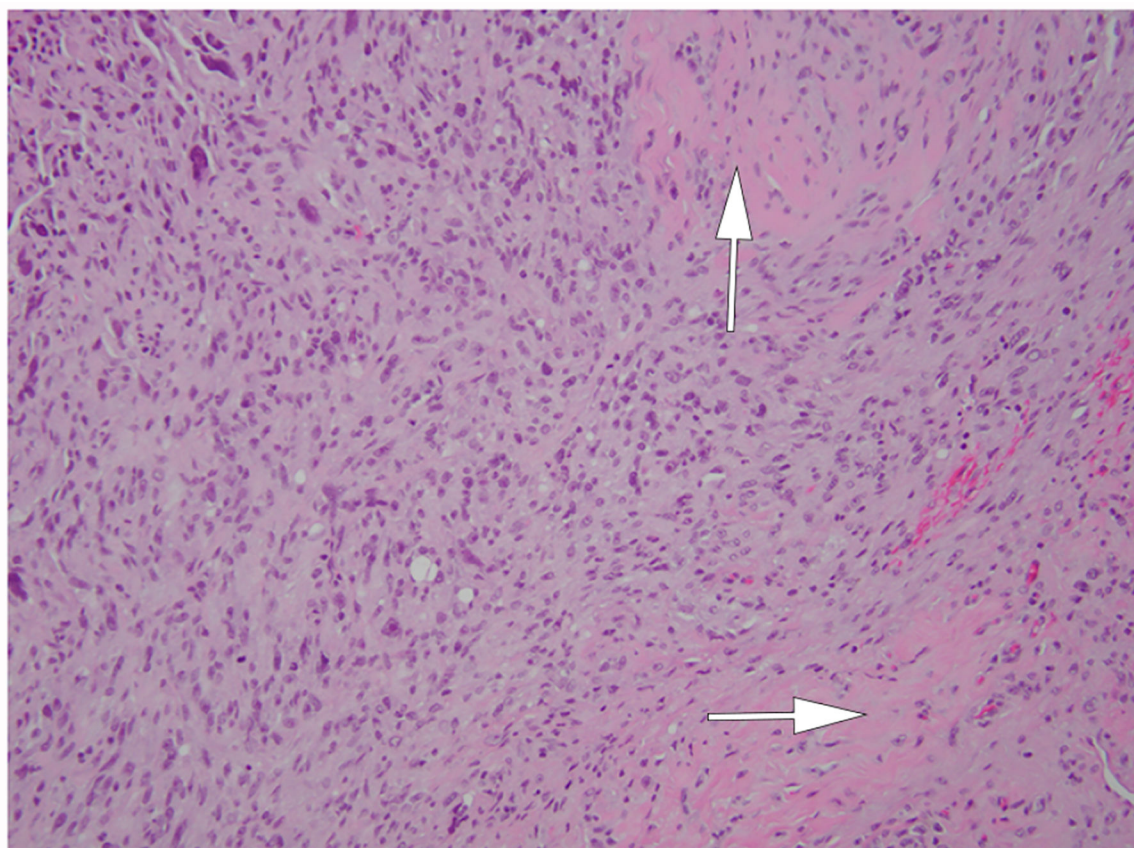
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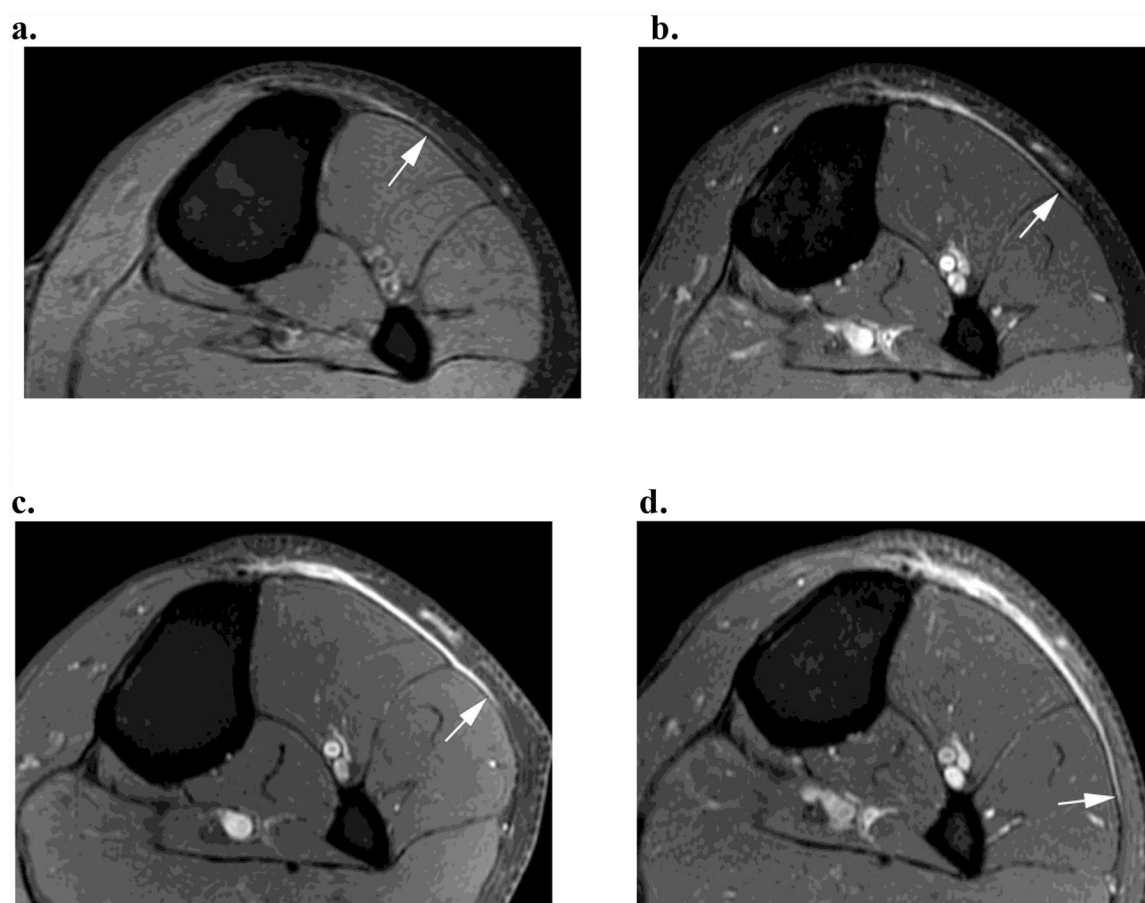


**Fig. 1.**  
**a-b** Preoperative axial fat-suppressed T1-weighted MR images obtained after administration of gadolinium contrast material show enhancement of the original mass (**a**, \*) in subcutaneous tissues, extending deep to contact the tibia, and a “tail” (**b**, arrow) extending laterally from the mass.



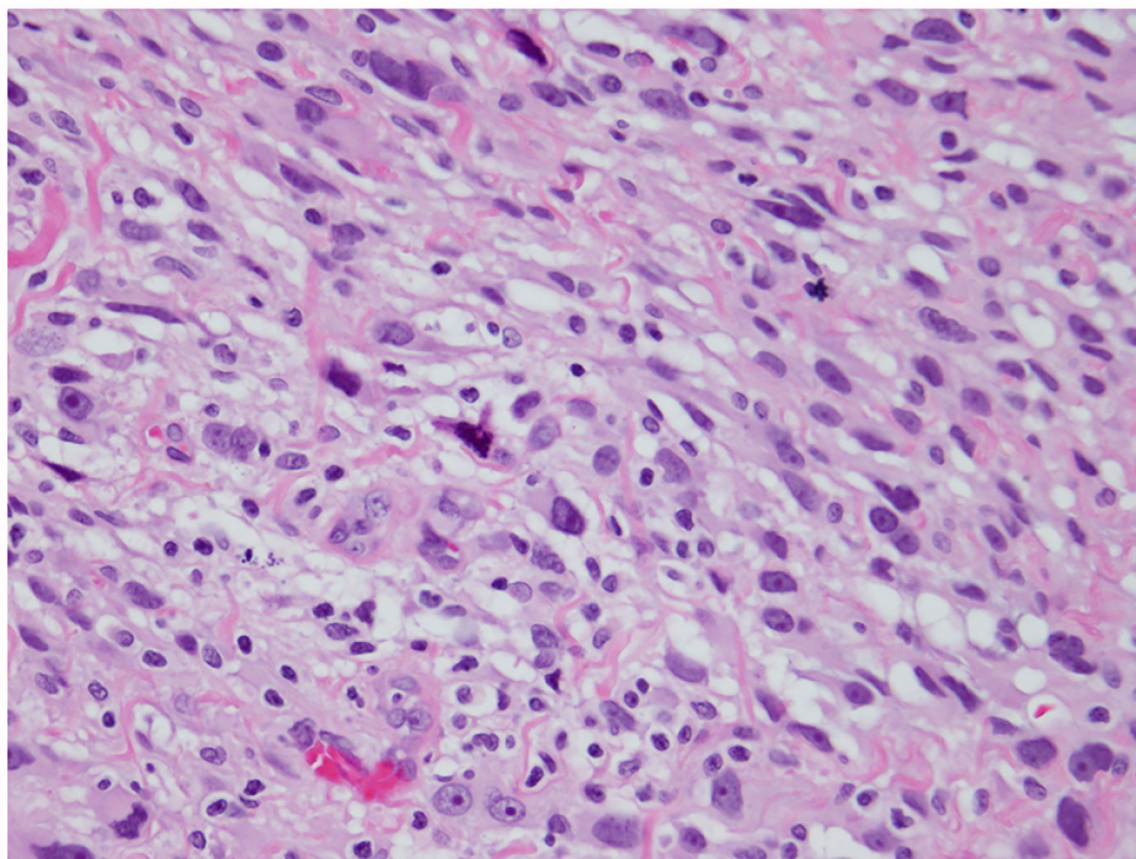


**Fig. 2.** Pathology from original re-resection. Photomicrograph shows spindle and pleomorphic cells, infiltrating in a diffuse manner within fascia (arrows). (x 200X)



**Fig. 3.**  
**a-d** Postoperative axial fat-suppressed T1-weighted MR images obtained after administration of gadolinium contrast material show enhancement and gradual enlargement of the tail-like recurrent tumor (arrow) at **a** 3.4 years, **b** 5.0 years, **c** 6.6 years, and **d** 7.3 years after initial resection.





**Fig. 4.**  
Pathology of recurrent tumor. Photomicrograph shows spindle and pleomorphic tumor cells similar to those at prior resection. (X 400X)