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Case Report

Primary Retroperitoneal Undifferentiated Pleomorphic Sarcoma Arising from the Psoas Muscle with Suspected Contralateral Involvement: A Rare Case Report

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Abstract

Undifferentiated pleomorphic sarcoma (UPS) is an aggressive soft-tissue malignancy, and primary involvement of the psoas muscle is exceptionally rare. We describe a 66-year-old man with a remote history of a treated left psoas abscess who presented with progressive left lower-limb swelling and extensive iliofemoral deep vein thrombosis. Contrast-enhanced CT demonstrated a 14.2 × 11.4 × 11.0 cm heterogeneous left iliopsoas mass with close vascular relationships and a separate right psoas lesion. MRI demonstrated heterogeneous T2 signal, necrotic changes, and restricted diffusion in viable tumor components. Image-guided biopsy revealed high-grade UPS after immunohistochemical exclusion of other lines of differentiation. The patient underwent neoadjuvant chemoradiotherapy, resection of the left-sided tumor with negative margins, and postoperative chemotherapy. The right psoas lesion decreased in size following systemic treatment but was not histologically confirmed as metastatic. This case illustrates the diagnostic challenge posed by concomitant venous thrombosis, the value of determining the tumor epicenter, and the essential role of histopathology in distinguishing rare retroperitoneal neoplasms.

Keywords: *Undifferentiated Pleomorphic Sarcoma; Psoas Muscle; Retroperitoneal Neoplasm; Iliofemoral Deep Vein Thrombosis; Computed Tomography; Magnetic Resonance Imaging*

Introduction

Undifferentiated pleomorphic sarcoma (UPS), formerly known as malignant fibrous histiocytoma, is a rare high-grade soft-tissue sarcoma characterized by marked cellular pleomorphism and the absence of a specific line of differentiation. The UPS commonly arises in the extremities, particularly the lower limbs, retroperitoneal involvement is uncommon, and primary origin within the psoas muscle is exceptionally rare [1,2].

Because of its deep location and nonspecific clinical manifestations, psoas UPS may remain clinically silent until it reaches a considerable size or causes symptoms related to compression or invasion of adjacent structures. Presentation with extensive deep venous thrombosis is particularly unusual and may represent a diagnostic challenge, especially in patients with a previous history of psoas infection. We report a rare case of primary UPS of the left psoas muscle in a 66-year-old man presenting with extensive iliofemoral deep venous thrombosis, with associated vascular involvement and a contralateral psoas lesion suspicious for secondary muscular involvement. This case emphasizes the pivotal role of multimodality imaging in diagnosis, assessment of tumor extent, and therapeutic planning [1,3].

Case Presentation

A 66-year-old man with a medical history of hypertension and type 2 diabetes mellitus, and a history of chronic cigarette smoking, presented to the emergency department with progressive swelling, erythema, warmth, and partial functional impairment of the left lower limb. Two years earlier, he had been treated conservatively with antibiotics for a left psoas abscess, with complete clinical resolution.

On physical examination, the left lower limb was warm, swollen, and tender to palpation, with pain during passive mobilization. Enlarged left inguinal lymph nodes were palpable. Laboratory testing showed C-reactive protein of 39 mg/L, leukocytosis, thrombocytosis, and anemia (hemoglobin 10 g/dL). Despite these inflammatory abnormalities, there was no convincing clinical infectious syndrome at presentation.

Given the clinical suspicion of acute deep venous thrombosis (DVT), an urgent Doppler ultrasound examination was performed. It demonstrated extensive thrombosis involving the left common femoral vein and extending into the external iliac vein. Multiple enlarged left inguinal lymph nodes were also identified. In addition, a large heterogeneous abdominal mass was incidentally detected but could not be adequately characterized by ultrasonography because of its deep retroperitoneal location.

In view of these findings, contrast-enhanced thoracoabdominal computed tomography was immediately performed for further characterization of the retroperitoneal mass and assessment of its local extension (Figure 1). It revealed a large, heterogeneous, solid mass centered on the left iliopsoas muscle, measuring approximately 14.2 × 11.4 × 11.0 cm. The lesion extended inferiorly toward the iliac and inguinal regions, with close contact with the left iliac bone and hip joint, without definite osseous destruction. Medially, the mass displaced the adjacent pelvic structures, involved the left external iliac artery and was closely related to the left iliac vascular axis, in the setting of the previously demonstrated extensive iliofemoral venous thrombosis. Multiple enlarged left inguinal lymph nodes were also present, measuring up to approximately 2.9 cm.

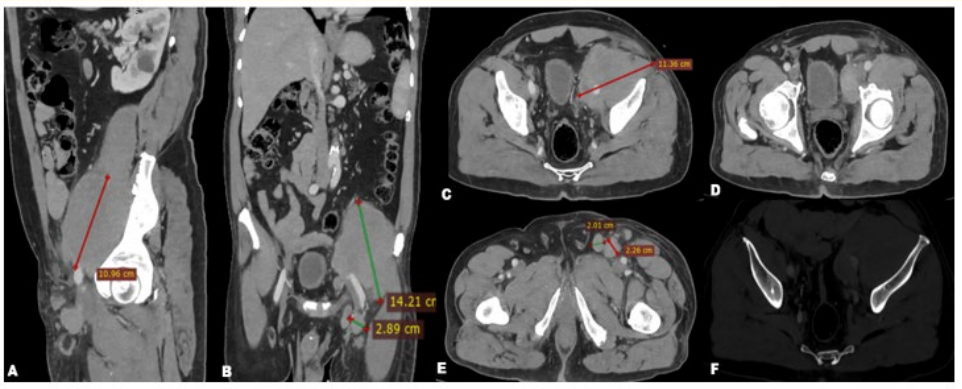


Figure 1: *Contrast-enhanced thoraco-abdomino-pelvic CT images. (A, B) Sagittal and coronal reformatted images demonstrate a large heterogeneous mass centered on the left iliopsoas muscle, measuring approximately 14.2 × 11.4 × 11.0 cm, with inferior extension toward the iliac and inguinal regions. (C, D) Axial images show the relationship of the tumor with adjacent pelvic structures and involvement of the left external iliac vascular axis. (E) Multiple enlarged left inguinal lymph nodes are identified. (F) A smaller contralateral intramuscular lesion involving the right psoas muscle is seen, suspicious for secondary muscular involvement. No other distant metastatic lesions were identified on the thoraco-abdomino-pelvic staging CT.*

Given the aggressive features identified on CT, pelvic MRI was subsequently performed for further tissue characterization and assessment of locoregional extension (Figure 2). MRI confirmed a large, well-defined but heterogeneous solid mass centered on the left iliopsoas muscle, extending from the iliac region to the proximal thigh. The lesion demonstrated heterogeneous intermediate-to-high signal intensity on T2-weighted images, with internal areas of necrosis. Diffusion-weighted imaging showed high signal intensity with corresponding low signal on the ADC map, consistent with restricted diffusion within the viable solid components. The mass exerted a marked mass effect on the adjacent pelvic structures and showed close relationships with the left iliac vascular axis. No definite osseous invasion was identified. These MRI features further supported the diagnosis of an aggressive soft-tissue neoplasm.

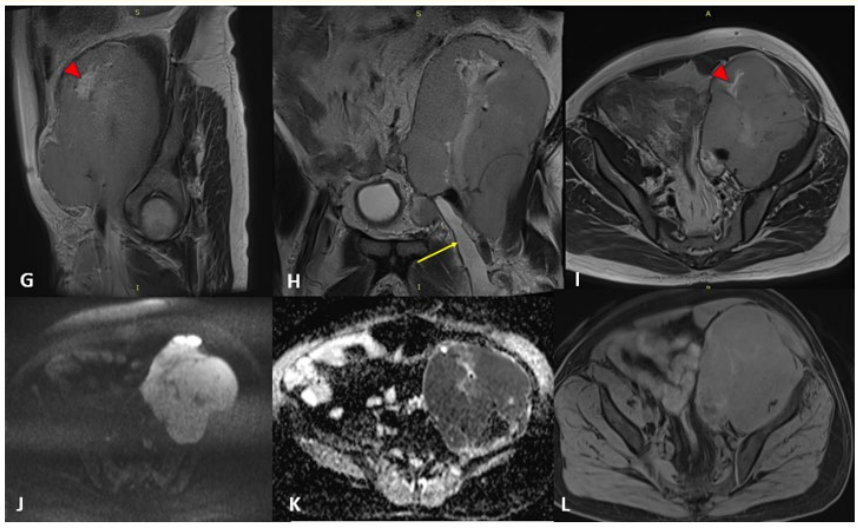


Figure 2: *Magnetic resonance imaging features of a left iliopsoas undifferentiated pleomorphic sarcoma. (G-I): Multiplanar T1-weighted MR images demonstrating a large, well-defined but heterogeneous soft-tissue mass centered in the left iliopsoas region, with marked displacement of adjacent anatomical structures. Small intratumoral areas of altered signal intensity are visible (Red arrowheads). The coronal image (H) also demonstrates the close relationship of the mass to the adjacent femoral vascular structures (Yellow arrow). (J, K): Diffusion-weighted imaging and corresponding apparent diffusion coefficient (ADC) map demonstrating areas of restricted diffusion within the solid tumor components. (L): Axial fat-suppressed T1-weighted image showing the heterogeneous internal architecture of the mass.*

MRI also demonstrated a second, smaller, well-defined intramuscular mass involving the contralateral right psoas muscle. The lesion showed heterogeneous T2 signal intensity and restricted diffusion, with high signal on diffusion-weighted imaging and corresponding low signal on the ADC map. Given its similar imaging characteristics to the dominant left iliopsoas tumor, this contralateral lesion was considered highly suspicious for secondary muscular involvement.

An image-guided core needle biopsy of the dominant left iliopsoas mass was performed. Histopathological examination revealed a high-grade spindle cell sarcoma composed of markedly pleomorphic cells with pronounced nuclear atypia, frequent atypical mitotic figures, and extensive tumor necrosis. Immunohistochemical analysis showed diffuse vimentin expression, whereas cytokeratins, S100, desmin, smooth muscle actin (SMA), MDM2, and CDK4 were negative. In the absence of a specific line of differentiation, the morphological and immunohistochemical findings supported the diagnosis of high-grade undifferentiated pleomorphic sarcoma (UPS), formerly referred to as malignant fibrous histiocytoma.

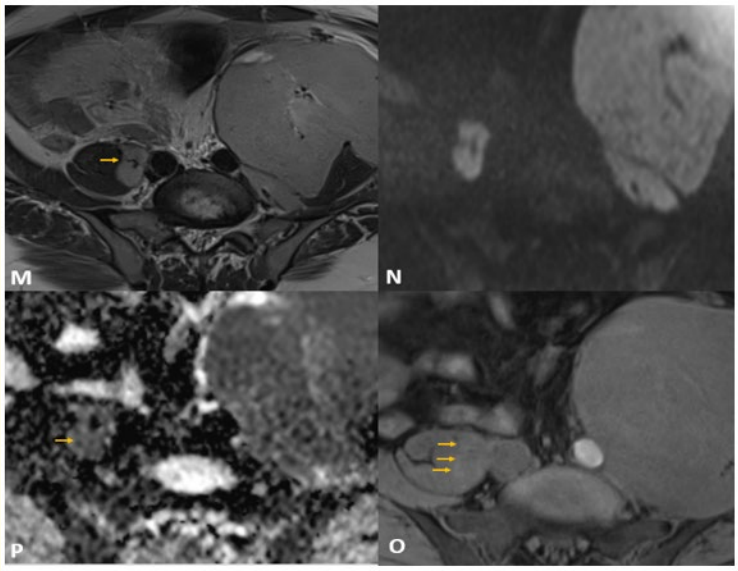


Figure 3: *Contralateral right psoas lesion. (M) Axial T2-weighted image demonstrates a well-defined intramuscular mass within the right psoas muscle (arrow), distinct from the dominant left iliopsoas tumor. (N, P) Diffusion-weighted imaging and corresponding ADC map demonstrate restricted diffusion within the lesion. (O) Axial image further delineates the right psoas mass (arrows). Given its imaging similarity to the primary left-sided tumor, the lesion was considered suspicious for secondary muscular involvement.*

Following multidisciplinary review, the patient received neoadjuvant chemoradiotherapy including doxorubicin and ifosfamide. Subsequent imaging demonstrated a decrease in size of the contralateral right psoas lesion. The dominant left iliopsoas tumor was then surgically resected with negative margins. Only the left-sided tumor underwent resection; the right psoas lesion was neither biopsied nor removed.

The patient subsequently received six cycles of adjuvant doxorubicin- and ifosfamide-based chemotherapy. Postoperatively, the experienced asthenia followed by gradual partial clinical improvement. Long-term oncological follow-up data were unavailable; therefore, durable disease control and the metastatic nature of the right psoas lesion could not be established.

Discussion

Undifferentiated pleomorphic sarcoma (UPS), formerly classified as malignant fibrous histiocytoma, is a high-grade mesenchymal malignancy defined by marked pleomorphism without a specific line of differentiation. It predominantly occurs in older adults and more often involves the extremities and trunk. Primary retroperitoneal UPS is uncommon, while origin within the psoas muscle is exceptionally rare and described mainly in isolated reports. Its deep location permits substantial growth before symptoms develop. The older malignant fibrous histiocytoma literature must be interpreted cautiously because many retroperitoneal tumors were subsequently reclassified, particularly as dedifferentiated liposarcoma [1-3].

UPS is an aggressive malignancy associated with local recurrence and distant metastatic spread, most commonly to the lungs. In a retrospective cohort of 266 patients with UPS, five- and ten-year overall survival rates were 60% and 48%, respectively; large tumor size and metastatic disease at presentation were adverse prognostic factors [4]. These outcomes reflect UPS across anatomical sites and cannot be directly extrapolated to the exceptionally rare psoas location. In our patient, the separate right psoas lesion was suspicious for

secondary involvement, particularly because it decreased after systemic therapy. However, without tissue confirmation, it should not be designated a proven metastasis.

Clinical manifestations of psoas UPS are nonspecific and may include lumbar or abdominal pain, a palpable mass, and symptoms from compression of adjacent structures. In our case, extensive left iliofemoral thrombosis dominated the initial presentation, with limb swelling and pain. Elevated inflammatory markers were nonspecific and did not establish infection, despite a remote history of psoas abscess [1,3].

Imaging plays a central role in the detection of retroperitoneal UPS, morphological characterization, locoregional staging, assessment of vascular relationships, image-guided biopsy, and treatment planning. Ultrasonography may reveal a deep heterogeneous solid mass but is limited by its retroperitoneal location. Contrast-enhanced CT is the principal initial staging modality and typically shows a large, heterogeneous soft-tissue mass with irregular enhancement and low-attenuation areas related to necrosis, hemorrhage, or cystic change. MRI further defines the muscular epicenter and relationships to adjacent structures: viable tumor commonly exhibits low-to-intermediate T1 signal, heterogeneous intermediate-to-high T2 signal, irregular enhancement, and sometimes restricted diffusion. None of these appearances is specific for the UPS histological subtype [3,5].

In our case, CT identified a 14.2 × 11.4 × 11.0 cm heterogeneous left iliopsoas mass with necrotic components and close relationships to the iliac vessels. MRI demonstrated heterogeneous T2 signal and restricted diffusion in solid regions. The large size, muscular epicenter, and aggressive morphology supported a high-grade soft-tissue malignancy, but did not establish UPS before biopsy [3,5].

Dedifferentiated liposarcoma is an important alternative diagnosis because it is common in the retroperitoneum and may appear as a heterogeneous nonlipomatous mass. Identification of an adjacent macroscopic fatty component favors this diagnosis. No macroscopic fat was described in our patient, although entirely nonlipomatous dedifferentiated tumors remain possible; negative MDM2 and CDK4 immunostaining provided further support against this diagnosis. Molecular testing for MDM2 amplification may be required when doubt persists [2,3,5].

Retroperitoneal leiomyosarcoma may also appear as a large heterogeneous necrotic mass. Some tumors arise from major venous walls and demonstrate intraluminal and extraluminal growth. In our patient, extensive iliofemoral thrombosis made it initially difficult to determine whether venous disease was secondary to the adjacent mass or reflected a primary vascular neoplasm. Careful assessment of the tumor epicenter favored the left iliopsoas muscle rather than the venous wall, making vascular leiomyosarcoma less likely. This did not exclude soft-tissue leiomyosarcoma on imaging alone; absent desmin and smooth muscle actin expression argued against smooth muscle differentiation [3,5].

Retroperitoneal lymphoma may produce a large soft-tissue mass with diffusion restriction, but more often shows relatively homogeneous enhancement, associated lymphadenopathy, and vessel encasement without major luminal compromise. In our patient, extensive internal heterogeneity and necrosis favored a high-grade sarcoma, although the inguinal lymphadenopathy and restricted diffusion meant lymphoma could not be excluded radiologically [3,5].

A recurrent psoas abscess was also considered because the patient had been treated for a left psoas abscess two years previously. Abscesses typically show a fluid-containing center, peripheral rim enhancement, surrounding inflammatory changes, and possible restricted diffusion of pus. Here, the absence of a convincing clinical infectious syndrome and the predominantly solid, irregular tumor morphology favored neoplasia. Nevertheless, inflammatory laboratory abnormalities and imaging overlap with necrotic tumors made tissue sampling necessary [3,5].

The radiological overlap among these entities makes histopathological examination indispensable. UPS is a diagnosis of exclusion requiring morphology and immunohistochemical assessment for other differentiated sarcoma subtypes and mimics. In our patient, biopsy demonstrated a high-grade pleomorphic spindle cell sarcoma expressing vimentin but not cytokeratins, S100, desmin, smooth muscle actin, MDM2, or CDK4, supporting UPS [2,6].

Management requires specialist multidisciplinary evaluation. Complete resection with negative margins is the cornerstone of potentially curative therapy for localized retroperitoneal sarcoma; the roles of perioperative radiotherapy and chemotherapy depend on histology, grade, resectability, and disease extent and are not universally established. In our patient, neoadjuvant doxorubicin/ifosfamide-based chemoradiotherapy was followed by negative-margin resection of the left-sided tumor and six postoperative chemotherapy cycles. The right psoas lesion decreased in size after systemic therapy but was not sampled. Although partial clinical recovery occurred, long-term oncological outcome was unavailable [7].

Conclusion

Primary UPS arising within the psoas muscle is exceptionally rare and can mimic other retroperitoneal malignancies or a recurrent abscess. Extensive iliofemoral thrombosis may obscure the lesion's origin; identification of the muscular tumor epicenter and multimodal imaging are essential for characterization and local staging. Imaging cannot reliably establish the UPS subtype, which requires histopathological and immunohistochemical confirmation. Multidisciplinary treatment and careful surveillance are crucial, especially when a contralateral lesion remains suspicious but unconfirmed.

Patient Consent for Publication

Written informed consent for publication of this case report and the accompanying clinical images: [To be confirmed and completed by the authors before submission].

Acknowledgments

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