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Undifferentiated Pleomorphic Sarcoma Presenting as a Gluteal Abscess: A Diagnostic Pitfall with a Fulminant Clinical Course

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Abstract

Introduction: Undifferentiated pleomorphic sarcoma (UPS) is a high-grade soft-tissue sarcoma characterized by aggressive biological behavior, with high rates of local recurrence and early metastatic spread. It typically presents as a deep, painless, progressively enlarging mass; however, acute inflammatory or abscess-like presentations are rare and may delay diagnosis.

Presentation of Case: We report the case of a 32-year-old man presenting with fever and a presumed right gluteal abscess. Computed tomography revealed a large fluid-density lesion with peripheral enhancement. Surgical drainage yielded atypical hematic-mucoid material. Histopathological and immunohistochemical evaluation confirmed grade 3 UPS. Staging studies excluded distant metastases. Despite urgent referral for multidisciplinary oncologic management, the clinical course was fulminant, and the patient died one month after diagnosis due to septic complications.

Discussion: UPS represents a significant proportion of adult soft-tissue sarcomas, and although uncommon, pseudo-infectious or abscess-like presentations have been documented and pose a diagnostic challenge. Tumor size, depth, and histologic grade remain key prognostic determinants. Complete surgical resection with negative margins is the cornerstone of treatment, often complemented by multimodal therapy depending on individual risk factors.

Conclusion: Deep soft-tissue collections with atypical intraoperative characteristics should raise suspicion for an underlying malignancy. Early tissue sampling and prompt referral to specialized sarcoma centers are essential to optimize outcomes.

Keywords: Undifferentiated pleomorphic sarcoma; Soft tissue sarcoma; Gluteal mass; Diagnostic pitfall; Abscess like presentation

Introduction

Undifferentiated pleomorphic sarcoma (UPS), previously termed malignant fibrous histiocytoma, is an aggressive malignant mesenchymal neoplasm characterized by marked cellular pleomorphism and absence of specific lineage differentiation [1]. It is currently classified by the World Health Organization within the group of undifferentiated/unclassified soft-tissue sarcomas, reflecting its heterogeneous morphology and poorly defined histogenesis [1]. Soft-tissue sarcomas account for approximately 1% of adult malignancies, and UPS represents one of the most frequent high-grade subtypes in later adulthood [1,6]. Although most commonly arising in the deep soft tissues of the extremities, UPS may develop in virtually any anatomical region, including the trunk, retroperitoneum, mediastinum, and head and neck [2,3].

Clinically, UPS typically presents as a slowly enlarging, painless mass, a pattern that often contributes to delayed medical evaluation [1]. Systemic inflammatory symptoms, rapid enlargement, or abscess-like features are distinctly uncommon and may lead to

initial misdiagnosis as infectious or inflammatory disease [1,3]. Such atypical presentations have been described only in isolated reports but are clinically relevant, as diagnostic delay in high-grade sarcomas may adversely affect outcomes due to their early metastatic potential [1,2].

Case Presentation

A 32-year-old man with a history of morbid obesity and active smoking presented to the emergency department with a 5-day history of fever, weakness, and progressive right gluteal discomfort. He denied recent trauma, intramuscular injections, immunosuppression, or prior radiotherapy factors that may predispose to soft-tissue infections or secondary sarcomas [4–6]. On examination, he was febrile (38.6 °C) and tachycardic, with a deep, fluctuant mass approximately 10 cm in diameter in the right gluteal region. The overlying skin appeared intact, without erythema or drainage, a finding that can occur in deep soft-tissue sarcomas that mimic abscesses or inflammatory collections [1,7].

Laboratory testing revealed marked systemic inflammation, including leukocytosis of 28,000/μL and elevated C-reactive protein of 13 mg/dL. These abnormalities, although suggestive of infection, have also been described in sarcomas presenting with necrosis or rapid growth, occasionally leading to misdiagnosis as an infectious process [1,7]. Contrast-enhanced computed tomography of the pelvis demonstrated a large hypodense lesion with peripheral enhancement in the right gluteal compartment, accompanied by ipsilateral iliac lymphadenopathy (Figure 1). These findings were initially interpreted as compatible with a complicated abscess. Similar radiologic misinterpretations have been reported in the literature, as UPS may present with cystic or necrotic areas that resemble fluid collections [1,2,7].

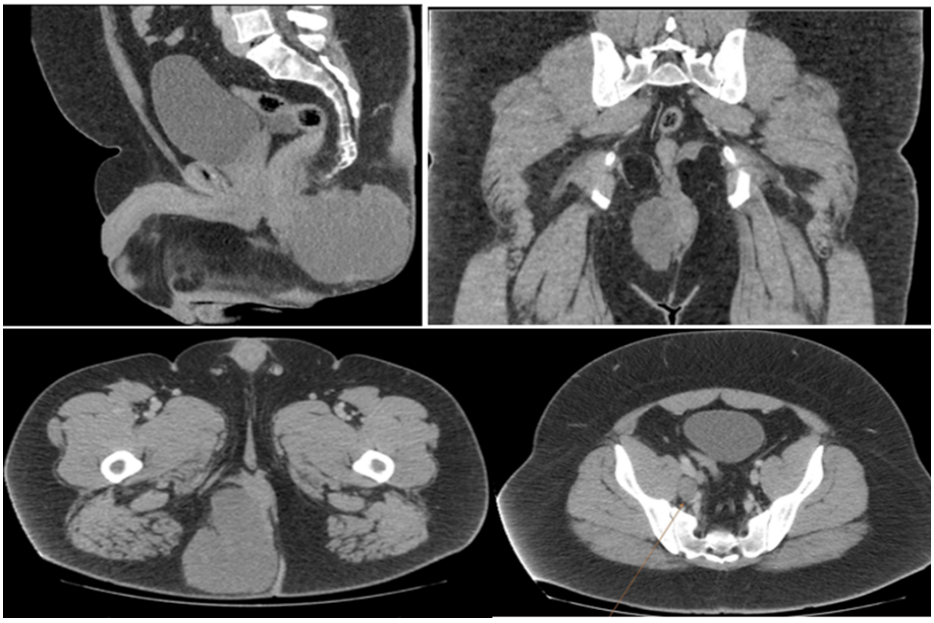

Figure 1: Contrast enhanced computed tomography of the pelvis: sagital, coronal and axial slices of large hypodense lesion with peripheral enhancement in the right gluteal compartment, accompanied by ipsilateral iliac lymphadenopathy marked with a orange arrow.

Given the presumed diagnosis of a deep gluteal abscess, urgent surgical drainage was performed. Intraoperatively, however, no purulent material was encountered. Instead, the cavity contained hematic–mucoid tissue with friable solid components. This discrepancy between preoperative imaging and intraoperative findings has been described in atypical presentations of UPS and should raise suspicion for an underlying malignancy [1,7]. Samples were obtained for microbiological and histopathological evaluation. All cultures remained negative.

Microscopic examination revealed a highly cellular spindle-cell neoplasm arranged in a storiform-pleomorphic pattern, with marked nuclear atypia, 34 mitoses per 10 high-power fields, and <50% tumor necrosis. These features corresponded to FNCLCC grade 3, consistent with the aggressive biological behavior characteristic of UPS [4–6,8]. Immunohistochemistry demonstrated positivity for smooth muscle actin, while S-100, desmin, CD34, BCL-2, HMB-45, and Melan-A were negative (Figure 2). This immunoprofile, although nonspecific, is typical of UPS, which is a diagnosis of exclusion [5] requiring the absence of lineage-specific markers [4-6,9].

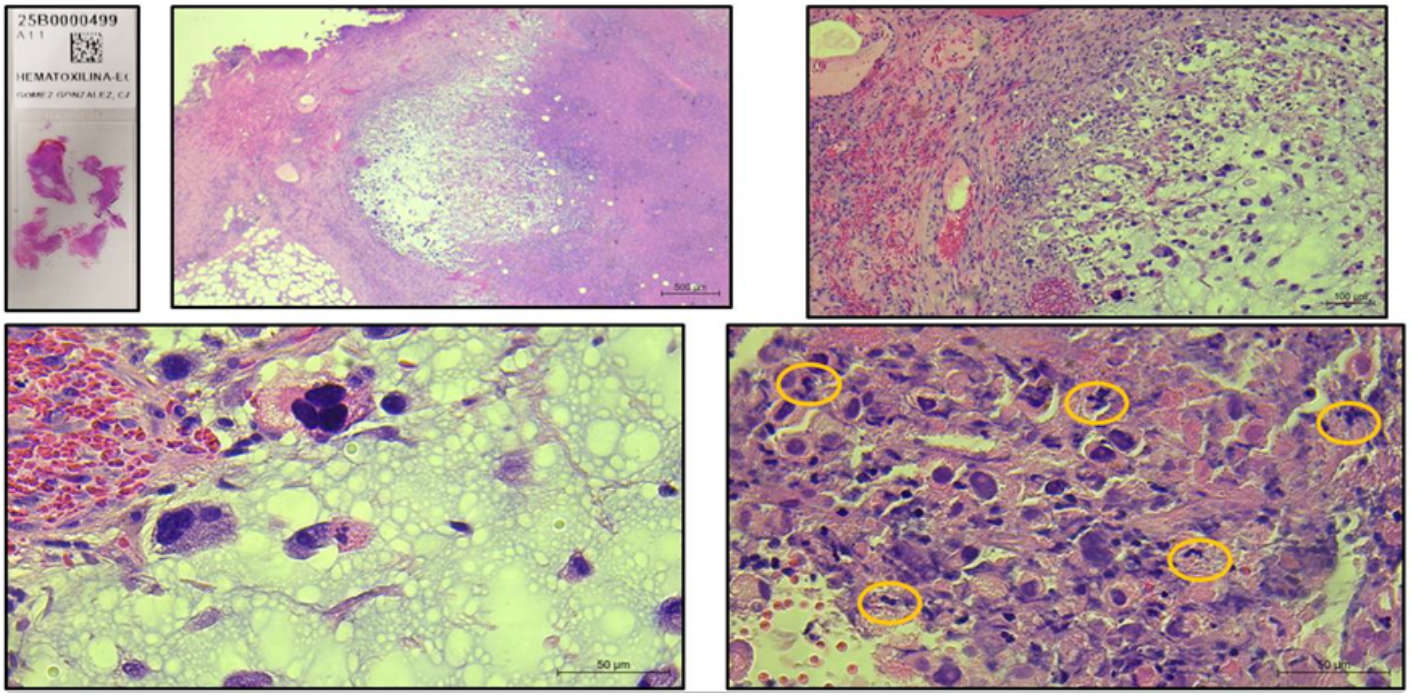

Figure 2: Histopathology: The tumor is composed of cells lacking clear tissue differentiation. The nuclei are markedly pleomorphic, with occasional multinucleated forms and frequent mitotic figures (ovoid in appearance). The cytoplasm is abundant, eosinophilic, and often shows xanthomatous changes with microvacuolization. Immunohistochemical analysis demonstrates diffuse positivity for vimentin and focal positivity for smooth muscle actin (AML). No expression was observed for S100, HMB45, SOX10, CK AE1/AE3, EMA, SALL4, CD34, CD45, CD68, desmin, or MyoD1.

Comprehensive staging was performed, including thoracoabdominal computed tomography, positron emission tomography-CT, and pelvic magnetic resonance imaging. No distant metastases were identified. Although UPS frequently metastasizes early most commonly to the lungs localized disease at presentation has been reported in up to 50% of cases [2,5,10]. The patient was referred to a multidisciplinary sarcoma team for definitive oncologic management, in accordance with current recommendations emphasizing centralized care in specialized centers [5,10].

Despite timely referral, the patient's clinical condition deteriorated rapidly. Within days, he developed worsening systemic inflammation, progressing to septic shock and acute liver failure (Figure 3). He died one month after diagnosis. Fulminant clinical decline has been described in high-grade UPS, particularly in cases with extensive necrosis or rapid tumor progression, although death from septic complications is uncommon [1,7,11].

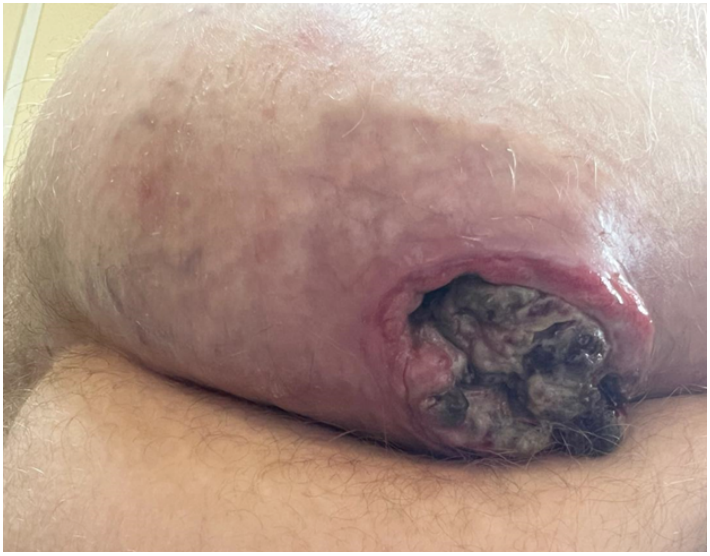

Figure 3: Extensive gluteal lesion with a necrotic appearance and slow, unfavorable evolution, located in the area previously subjected to drainage for a presumed perianal abscess. The wound shows irregular, raised borders with central areas of necrosis. The surface is friable, with foci of hemorrhage and persistent serohematic exudate.

Case Discussion

Undifferentiated pleomorphic sarcoma (UPS) is a high-grade malignant mesenchymal neoplasm characterized by marked cellular pleomorphism and absence of a definable line of differentiation, making it a diagnosis of exclusion [5] supported by immunohistochemistry [4-6]. Historically termed malignant fibrous histiocytoma, UPS was reclassified following advances in molecular pathology demonstrating that many tumors previously grouped under this label represent distinct sarcoma subtypes. Despite its rarity, UPS remains one of the most common high-grade soft-tissue sarcomas in adults, typically arising in the sixth and seventh decades of life, although cases in younger individuals have been documented [1,7].

The clinical presentation of UPS is classically that of a gradually enlarging, painless deep-tissue mass, most frequently located in the extremities, particularly the thigh [4,6]. However, atypical presentations may occur and represent a significant diagnostic challenge. The abscess-like presentation observed in this patient is uncommon but has been described in the literature. Tumor necrosis, intralesional hemorrhage, and the associated inflammatory response may produce imaging appearances indistinguishable from infection, including fluid-density lesions with peripheral enhancement and surrounding inflammatory changes [1,7].

Radiologically, UPS often demonstrates heterogeneous enhancement, central necrosis, and variable hemorrhage. These features can mimic infectious collections, especially in deep anatomical compartments such as the gluteal region. Although lymph node involvement is uncommon in UPS, reactive lymphadenopathy may occur in the setting of tumor necrosis or inflammation. Early biopsy preferably core-needle biopsy performed in a sarcoma center is recommended to avoid inappropriate surgical approaches that may compromise future oncologic management.

Histopathological evaluation remains the cornerstone of diagnosis. UPS is defined by a storiform-pleomorphic architecture, marked nuclear atypia, brisk mitotic activity, and variable necrosis [4-6]. Immunohistochemistry is essential [6] to exclude other spindle-cell neoplasms. The immunoprofile in this case positivity for smooth muscle actin with absence of S-100, desmin, CD34, BCL-2, HMB-45, and Melan-A is consistent with UPS, which typically lacks lineage-specific markers and is therefore a diagnosis of exclusion [5,4-6].

Prognosis in UPS is strongly influenced by tumor size, depth, and histological grade. High-grade tumors larger than 10 cm are associated with significantly worse outcomes, including increased risk of local recurrence and distant metastasis [5,10]. Data from institutional series consistently identify tumor size >5 cm, deep location, and FNCLCC grade 3 as independent predictors of poor survival. Although the lungs are the most common site of metastasis, aggressive local behavior and rapid progression may occur even in the absence of detectable metastatic disease, as observed in this patient. Fulminant deterioration has been described in high-grade sarcomas with extensive necrosis or rapid proliferative activity, although death from septic complications is uncommon [1,7].

The mainstay of curative treatment for localized UPS is wide surgical excision with negative margins [2,9,10]. Achieving adequate margins is critical, as positive or close margins are associated with significantly higher rates of local recurrence [5,10]. Adjuvant radiotherapy improves local control in selected cases, particularly for high-grade or large tumors, or when margins are close or positive. Chemotherapy, typically doxorubicin-based regimens, may be considered in high-risk disease, although its impact on overall survival remains uncertain and is the subject of ongoing investigation. Emerging systemic therapies [7,14], including immunotherapy and targeted agents, are under study, with early evidence suggesting potential benefit in selected patients based on tumor immunogenicity and microenvironmental features.

The rapid clinical decline in this patient, despite the absence of detectable metastases at staging, underscores the aggressive nature

of UPS and the importance of early diagnosis and specialized multidisciplinary management. In this case, the combination of systemic inflammation, tumor-associated necrosis, and possible occult microvascular invasion may have contributed to the severe systemic response culminating in septic shock and acute liver failure. This fulminant course highlights the need for heightened clinical suspicion when evaluating deep soft-tissue collections with atypical features.

This case reinforces several key principles. First, deep soft-tissue masses with abscess-like imaging characteristics should not be assumed to be infectious without careful evaluation, particularly when risk factors for sarcoma are present or when intraoperative findings are inconsistent with infection. Second, early biopsy is essential when the clinical picture is atypical. Third, management of suspected sarcomas should occur in specialized centers with expertise in multidisciplinary care, as recommended by international guidelines. Finally, atypical presentations of UPS, although rare, carry significant risk for diagnostic delay and adverse outcomes, emphasizing the importance of maintaining a broad differential diagnosis in patients presenting with deep soft-tissue collections.

Conclusion

Undifferentiated pleomorphic sarcoma can rarely present with clinical and radiological features mimicking a deep soft-tissue abscess, creating a significant diagnostic pitfall. Surgeons should maintain a high index of suspicion when intraoperative findings are atypical or cultures are negative, and routine histopathological evaluation of drained material is imperative. Given the aggressive nature and rapid progression of this malignancy, early diagnosis and prompt referral to specialized sarcoma centers are essential to optimize patient outcomes.

Declarations

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