



Rare Presentation of Small Bowel Undifferentiated Pleomorphic Sarcoma with Perforation and Obstruction

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Abstract

Undifferentiated pleomorphic sarcoma (UPS) of the small intestine is extremely rare, with few cases including both obstruction and perforation. We discuss a case of small bowel UPS with acute perforation and luminal obstruction, focusing on diagnostic problems, surgical therapy, and pathological findings. Small intestine UPS is an uncommon tumor, and vague symptoms may cause delays in diagnosis. Imaging may indicate a malignant small bowel tumor; however, it cannot distinguish UPS from gastrointestinal stromal tumor, lymphoma, or adenocarcinoma. The simultaneous occurrence of perforation and blockage is unusual and presents management concerns. Complete surgical resection with negative margins is the primary treatment option; the role of adjuvant chemotherapy or radiotherapy is unknown because to a lack of data. The prognosis is generally poor, with a high recurrence and metastasis rate. This case report emphasizes the importance of being aware that undifferentiated pleomorphic sarcoma, albeit rare in the small intestine, can present tragically with perforation and obstruction. Prompt surgical intervention and a comprehensive pathologic examination are required. Additional case reports and studies are required to advise appropriate care and better understand the behavior of such tumors in the gastrointestinal tract.

Subject Areas

Urology

Keywords

Gastrointestinal Stromal Tumour, Gastrointestinal Motility, Interstitial Cell of Cajal, Tyrosine Kinase Inhibitor, Immunohistochemistry

1. Introduction

Sarcomas are malignant tumors arising from mesenchymal cells, representing a rare and heterogeneous group that accounts for approximately 1% of adult malignancies [1]. Among these, undifferentiated pleomorphic sarcoma (UPS)—formerly known as malignant fibrous histiocytoma—is a high-grade soft tissue sarcoma comprising about 5% to 10% of adult soft tissue sarcomas [2]. Histologically, UPS is characterized by marked cellular pleomorphism, atypical mitotic figures, and regions of necrosis.

UPS most commonly arises in the extremities and retroperitoneum. Primary involvement of the small bowel is exceedingly rare, with only around 21 cases reported in the English-language literature to date [3]. Imaging characteristics—such as CT and MRI findings—are generally nonspecific and may mimic other small bowel neoplasms, including gastrointestinal stromal tumors (GIST), lymphoma, or adenocarcinoma [3].

Due to its rarity, evidence-based guidelines for management remain limited, and treatment typically involves surgical resection with or without adjuvant therapy. We report a rare case of small bowel UPS in a young adult male who presented with features of intestinal obstruction, complicated by perforation.

2. Case Report

A 33 years old gentleman with no underlying illnesses presented to our center with the inability to pass stool for 3 days, associated with vomiting for 2 days and abdominal pain for 1 day. Physical examination revealed a palpable bowel loop. An abdominal X-ray showed dilated small bowel loops. Contrast-enhanced CT demonstrated dilatation of the proximal jejunum with a transition point in the lower mid-abdomen, likely at the mid-ileum. At this level, a short segment of enhancing wall thickening caused luminal narrowing, with a small bowel feces sign seen proximally. Distal small bowel loops appeared collapsed.

The patient subsequently underwent exploratory laparotomy. Intraoperatively, a perforated small bowel tumor measuring approximately 4 cm × 4 cm was noted 200 cm distal to the duodenojejunal (DJ) flexure. No feculent material was found in the peritoneal cavity. A 20-cm segmental resection of the small bowel was performed, and the resected ends were fashioned into a double-barrel stoma after decompression. The decision to avoid primary anastomosis was made in view of the recent perforation, potential microscopic contamination, and the patient's intraoperative hemodynamic instability, all of which increased the risk of anastomotic leakage. The remaining bowel appeared viable.

Histopathological examination revealed diffuse infiltration of tumor cells within the bowel wall. The tumor extended into the mucosa but spared the glandular epithelium. The bowel mucosa showed no evidence of dysplasia. The tumor ulcerated the bowel mucosa and perforated the serosal layer. The tumor cells were large, polygonal, and epithelioid, with moderate-to-marked nuclear pleomorphism, vesicular nuclei, and prominent macronucleoli. Atypical mitoses, apoptotic bodies, and

tumor necrosis were readily identified. The cytoplasm was abundant, with focal clearing or vacuolation. The bowel resection margins were free of tumor, but the tumor had perforated beyond the peritonealized margin. Immunohistochemistry demonstrated tumor cell positivity for INI-1, CD163, and CD68. The following markers were negative: CK7, CK20, panCK, EMA, CAM5.2, S100, CD34, SMA, Desmin, Myogenin, LCA, CD3, CD20, CD21, CD138, CD117, DOG-1, SALL4, CD30, HcG, HMB45, SOX10, TTF1, CDX2, p40, Synaptophysin, Chromogranin, MPO, CD43, and BRAF V600E. The Ki-67 proliferation index was high (>80%). The final diagnosis was consistent with an undifferentiated pleomorphic sarcoma, FNCLCC Grade 3.

Postoperatively, the stoma functioned well. Despite counselling, the patient declined further oncological management and was discharged against medical advice. One month later, his family reported that he had passed away. Written informed consent have been taken from the patient for the purpose of publication (See **Figures 1-6**).



Figure 1. Plain abdominal radiograph at presentation showing multiple dilated small bowel loops with air-fluid levels.

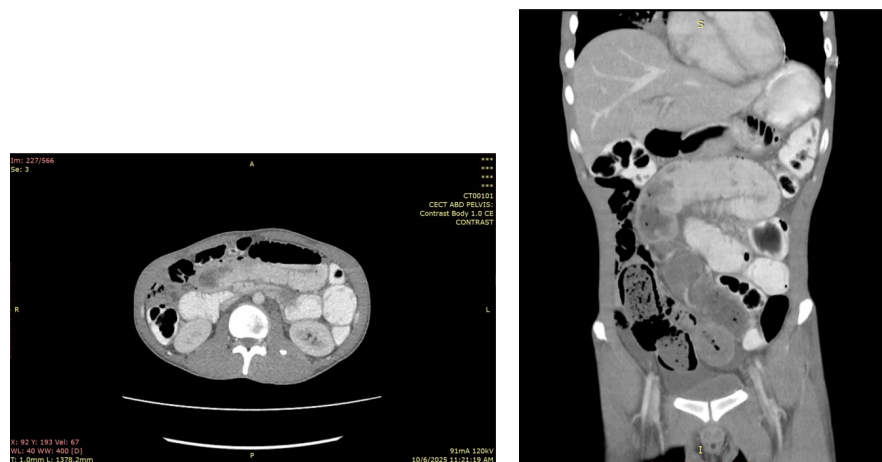


Figure 2. Axial and coronal contrast-enhanced CT of the abdomen and pelvis showing a heterogeneously enhancing intraluminal soft tissue mass in the small bowel with proximal bowel dilatation, consistent with small bowel obstruction.



Figure 3. Resected segment of small bowel showing a circumferential ulceroinfiltrative mass with surface necrosis and hemorrhage. Proximal stump is tagged with a long suture and distal stump with a short suture for orientation.



Figure 4. Macroscopically, there is a diffuse thickening of the small bowel wall with whitish, firm cut surface and friable mucosa. Arrow show bulging over the mucosa with adjacent normal small bowel mucosa.

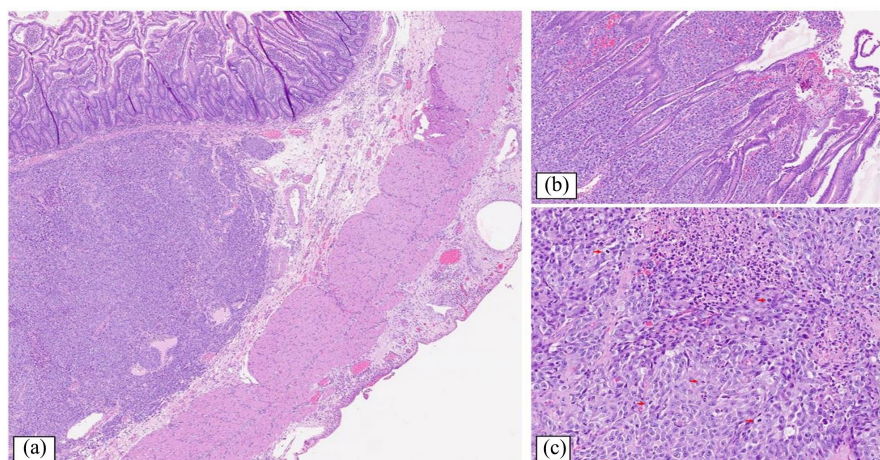


Figure 5. Microscopically, the tumour is seen within submucosa region (a) and extending up to the mucosa (b) in between colonic glands. The tumour shows diffuse spindle to epithelioid cells with enlarged nuclei, ample cytoplasm and brisk mitosis (red arrows show mitosis).

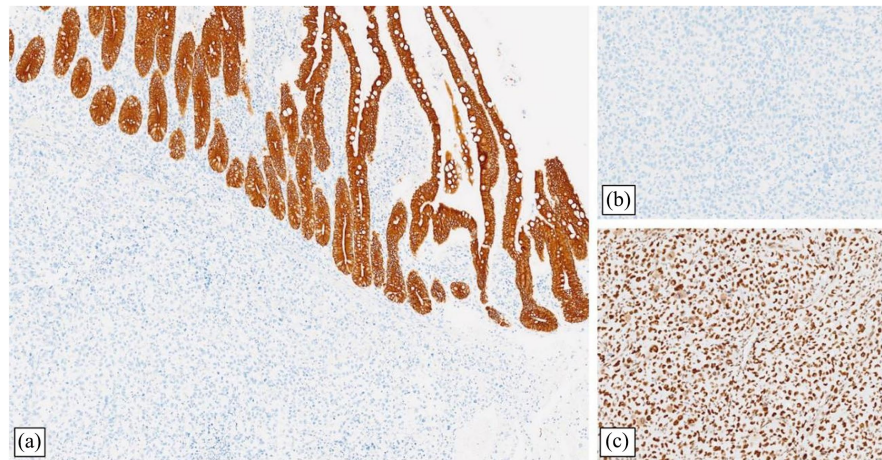


Figure 6. Undifferentiated pleomorphic sarcoma is a diagnosis of exclusion. Tumour cells are negative for pan-CK (a), SALL4 (b) and intact INI-1 expression (c).

3. Discussion

Undifferentiated pleomorphic sarcoma (UPS) of the small bowel is an exceptionally rare and aggressive malignancy, with only a handful of cases reported in the literature. The etiology of UPS remains incompletely understood. While the majority of cases occur sporadically without identifiable predisposing factors, some evidence suggests potential associations with prior radiation exposure or chronic tissue injury. However, these links remain inconsistent across reported cases [4].

UPS typically follows an indolent clinical course, frequently remaining asymptomatic for prolonged periods [5]. Many cases are detected incidentally on imaging or when the tumor reaches a sufficient size to cause local symptoms. The most common presenting features relate to mass effect, including localized pain or neurological deficits from compression of nearby structures [6]. Unlike many malignancies, systemic symptoms are uncommon and, when present, typically indicate advanced metastatic disease [7].

Gastrointestinal stromal tumors (GISTs) and adenocarcinomas often cause bleeding or perforation. This is typically due to mucosal ulceration or transmural invasion [8]. Intestinal lymphomas can also perforate, but this results from rapid tumor necrosis. In contrast, UPS rarely presents with these acute symptoms. When UPS perforation occurs, it stems from secondary ischemic necrosis rather than mucosal invasion. This key pathological mechanism helps differentiate UPS from other aggressive small bowel malignancies during acute presentations [9].

Routine laboratory studies (CBC, metabolic panel, coagulation studies), while nondiagnostic, are essential for treatment planning. Elevated lactate dehydrogenase (LDH) levels, though nonspecific, may signal aggressive disease [10]. CT imaging of UPS typically reveals a well-demarcated soft tissue mass that may appear homogeneous or contain hypodense areas corresponding to necrosis/hemorrhage [11]. MRI may provide additional soft tissue characterization, particularly in assessing local invasion [12].

The diagnosis of undifferentiated pleomorphic sarcoma (UPS) is one of exclusion. Microscopically, UPS demonstrates three hallmark features: 1) marked cellular heterogeneity, 2) significant nuclear and cytoplasmic pleomorphism, and 3) disorganized architectural patterns. While immunohistochemical staining may aid in diagnosis by excluding other malignancies, UPS lacks a definitive immunoprofile. No consistent immunohistochemical marker exists to facilitate further subclassification, reflecting the tumor's undifferentiated nature.

In this case of a small bowel tumor, the differential diagnosis included gastrointestinal stromal tumor (GIST)—the most common small bowel soft tissue lesion [13]—as well as other soft tissue sarcomas. The patient's young age raised the possibility of a metastatic germ cell tumor. Morphologically, the epithelioid appearance of the tumor cells suggested histiocytic sarcoma as another key differential. Immunohistochemistry was crucial in the diagnostic workup. GIST was excluded by negative staining for CD117, CD34 and DOG-1. Additional exclusions included epithelial tumors (negative for panCK, EMA, CAM5.2), lymphoma (negative for LCA, CD20, CD3, CD30), germ cell tumors (negative for SALL4), and neuroendocrine tumors (negative for synaptophysin and chromogranin). Other intra-abdominal soft tissue tumors, such as clear cell sarcoma, extrarenal rhabdoid tumor and histiocytic sarcoma were also ruled out through appropriate immunohistochemical analysis [14]. The interpretation of specific markers was critical. The retained nuclear expression of INI-1 helped exclude malignancies characterized by its loss, such as malignant rhabdoid tumor and epithelioid sarcoma. Furthermore, the focal and non-specific positivity for CD163 and CD68 was insufficient to support a diagnosis of histiocytic sarcoma. The comprehensive exclusion of these entities ultimately supports the diagnosis of UPS. This diagnosis is consistent with the tumor's aggressive clinical behavior, as evidenced by the patient's rapid demise.

The primary treatment for UPS involves complete surgical resection with negative margins, ideally performed at the earliest feasible opportunity. [15] For extremity UPS, adjuvant radiotherapy has demonstrated established efficacy in local disease control. However, its role in bowel-associated UPS remains uncertain due to limited evidence and potential toxicity concerns in abdominal locations [16]. Systemic therapy with doxorubicin and ifosfamide is typically reserved for advanced disease, including unresectable primary tumors or metastatic cases [17]. In the refractory setting, therapeutic alternatives such as the gemcitabine-docetaxel regimen or trabectedin may be employed [18]. Furthermore, immune checkpoint inhibitors (e.g., pembrolizumab, nivolumab) have demonstrated preliminary efficacy in select cases of advanced UPS, although their application remains confined to investigative protocols [19].

Following curative resection, vigilant surveillance is critical given the high risk of recurrence. Current soft-tissue sarcoma guidelines recommend clinical evaluation and cross-sectional imaging (CT or MRI) every 3 - 4 months for the first 2 - 3 years, every 6 months until year 5, and annually thereafter, with chest imaging

at each visit to monitor for pulmonary metastases [20] [21]. The administration of adjuvant systemic therapy is warranted in patients with high-risk features (e.g., >5 cm, high-grade, or positive margins) to reduce the likelihood of relapse [22], and lifelong follow-up is mandated by the documented occurrence of late recurrences [21].

4. Conclusion

Undifferentiated pleomorphic sarcoma (UPS) of the small bowel is an exceptionally rare and aggressive malignancy, with limited cases described in the literature. Its nonspecific clinical and radiologic features pose significant diagnostic challenges, often leading to delayed recognition until complications such as obstruction, bleeding, or perforation occur. Histopathological evaluation with comprehensive immunohistochemistry remains essential to establish a definitive diagnosis by excluding other mimickers, particularly gastrointestinal stromal tumors (GIST). Surgical resection with negative margins continues to be the cornerstone of management. This case underscores the importance of maintaining UPS in the differential diagnosis of small bowel tumors presenting with acute abdomen and highlights the need for further research to define optimal adjuvant treatment strategies and improve prognostic outcomes.

Conflicts of Interest

The authors declare no conflicts of interest.

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