Laparoscopically-Assisted Repair of a Small Bowel Perforation Secondary to Multiple Metastases of Undifferentiated Pleomorphic Sarcoma

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ABSTRACT

We present a very rare case of a laparoscopically-assisted repair of a small bowel perforation secondary to multiple metastases of undifferentiated pleomorphic sarcoma from the posterior mediastinum. A 46-year-old man presented with middle to upper abdominal pain during chemotherapy for lung metastases from undifferentiated pleomorphic sarcoma. Computed tomography revealed intra-abdominal free air, and emergency laparoscopy was performed. Consequently, a perforation was detected in the jejunum, and partial jejunal resection was performed by mini-laparotomy. Pathological examination demonstrated an ulcerated tumor with perforation and four additional tumors in the resected jejunum. Pathological examination also revealed undifferentiated pleomorphic sarcoma in all five tumors. To our knowledge, our case is the first report of a laparoscopically-assisted repair of a small bowel perforation secondary to metastasis of undifferentiated pleomorphic sarcoma. Although the perforation site was unclear preoperatively, laparoscopic observation readily identified the lesion in this patient. Therefore, minimally invasive surgery could be performed successfully with mini-laparotomy following laparoscopic observation. Laparoscopic techniques for the small intestine are viable options, even in acute and uncommon situations, and small bowel perforation secondary to metastasis should be considered in patients with undifferentiated pleomorphic sarcoma and acute abdomen.

Key words laparoscopy; malignant fibrous histiocytoma; sarcoma; small intestine

Undifferentiated pleomorphic sarcoma (UPS) is a relatively common neoplasm that most frequently affects the

soft tissues. Soft tissue sarcoma usually metastasizes to the lungs; tumors arising in the abdominal cavity more commonly metastasize to the liver and peritoneum. We describe an extremely rare case of small intestinal perforation secondary to metastatic UPS.

PATIENT REPORT

A 46-year-old man presented with middle to upper abdominal pain while receiving chemotherapy for lung metastases from posterior mediastinal UPS. He had no significant medical history except for the UPS. He also had no history of previous abdominal surgery. The UPS was resected and diagnosed 2 years earlier, and the right lung recurrence occurred within 2 months after the resection. Therefore, he underwent partial resection of his right upper lobe for the metastatic lesion. Unfortunately, he was diagnosed with newly-arising multiple metastases in both lungs soon after the second resection, and chemotherapy with doxorubicin (60 mg/m²) was the first-line approach. After 4 months, pazopanib (800 mg/day for 2 days) was started as the second-line approach nine days before the initial consultation to us, because of disease progression. On initial consultation, the patient's body temperature was 37.3°C, his heart rate was 92 beats/min, blood pressure was 129/78 mmHg, and SpO₂ was 97% breathing room air. No tumor was detected in the abdomen, but there was tenderness in the epigastric and periumbilical regions with peritoneal signs. His bowel sounds were normal. Laboratory examination revealed that the patient's C-reactive protein level was 9.38 mg/dL and his white blood cell count was 11,300 cells/ μ L. Plain and contrast-enhanced abdominal computed tomography (CT) revealed intra-abdominal free air under the diaphragm, in the hepatic portal region, and around the right paracolic sulcus (Fig. 1). It was impossible to identify the site of perforation; however, the wall of the duodenum was slightly unclear and was a suspected perforation (Fig. 2).

To diagnose and repair the gastrointestinal perforation, we performed emergency laparoscopy under general anesthesia. During the surgery, a 12-mm trocar was inserted through an umbilical incision using an open

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Abbreviations: CT, computed tomography; MFH, malignant fibrous histiocytoma; UPS, undifferentiated pleomorphic sarcoma

Fig. 1. Preoperative CT. White arrow indicates free air under the diaphragm.

Fig. 2. Preoperative CT. White arrow indicates that the wall of the duodenum was slightly unclear, indicating a possible perforation.

technique. A flexible 10-mm laparoscope was inserted into the abdomen via the trocar, and pneumoperitoneum was established. Subsequently, 5-mm and 10-mm trocars were placed in the left and right supraumbilical region, respectively.

There was no evidence of perforation or pus around the stomach, duodenum, or intestinal sections in the pelvic cavity. However, an approximately 10-mm perforation was detected in the jejunum 20 cm distal to the ligament of Treitz (Fig. 3). No tumors were revealed by laparoscopic observation. The umbilical port incision was extended for 4 cm, and a mini-laparotomy was performed. Two more tumors around the perforation were identified, and partial jejunal resection, including all three lesions, with a functional end-to-end anastomosis was performed. The operative time was 94 min, and the amount of intraoperative bleeding was 30 g. The patient was discharged on postoperative day 5 without complications.

Pathological examination demonstrated an ulcerated tumor with perforation and another four tumors in the resected jejunum (Fig. 4). Pathological examination also revealed UPS with bizarre cells in all five tumors (Fig. 5).

Postoperatively, the patient experienced no recurrence of perforation, but he died 2 months after the operation as a result of cancer progression. Autopsy confirmed multiple metastases of UPS (lung, pleura, pericardium, liver, pancreas, peritoneum, mesentery, and paraaortic lymph node).

Fig. 3. The perforation site covered with pus was detected in the jejunum laparoscopically (white arrow).

DISCUSSION

Common subtypes of soft tissue sarcomas include UPS, gastrointestinal stromal tumor, liposarcoma, and leiomyosarcoma.¹ Extremities (43%), the trunk (10%), viscera (19%), retroperitoneum (15%), and head and neck (9%) are the most common primary sites.² Generally, surgical resection with appropriately negative margins is the standard primary treatment for most patients with soft tissue sarcomas, according to the National Comprehensive Cancer Network guidelines.³ For unresectable or metastatic disease, chemotherapy

Fig. 4. The resected jejunum including an ulcerated tumor with perforation and four additional tumors.

with single agents (dacarbazine, doxorubicine, epirubicin, or ifosfamide) or anthracycline-based combination regimens have been widely used. More recently, a number of targeted therapies have shown promising results in patients with certain types of advanced or metastatic soft tissue sarcoma.³

In 2013, the World Health Organization classification of soft tissue tumors was updated. With the advent of advanced diagnostics, the term “malignant fibrous histiocytoma (MFH)” became somewhat antiquated as well as imprecise and was superseded by the term “undifferentiated pleomorphic sarcoma” in this revision.⁴ These tumors have no specific markers for diagnosis and have in common marked cytological and nuclear pleomorphism, often with bizarre tumor giant cells, admixed with spindle cells and often rounded histiocyte-like cells (which may have foamy cytoplasm) in varying proportions.^{5, 6}

MFH occurs principally as a mass on an extremity (lower extremity 49%, upper extremity 19%) or in the abdominal cavity or retroperitoneum (16%) of adults (peak incidence, 61–70 years of age).⁷ The reported local recurrence rate of the tumor is 44%, and the metastatic rate is 42%; furthermore, metastasis is most frequently to the lung (82%) and lymph nodes (32%).⁷

In this report, we presented a case of a laparoscopically-assisted repair of a small bowel perforation secondary to multiple metastases of UPS from the posterior mediastinum. Historically, laparotomy has been the intervention of choice for acute abdomen. However, laparoscopic exploration has emerged as a viable option to identify and treat the source of perforation because of its minimal invasiveness. In particular, perforated peptic ulcer,⁸ acute cholecystitis⁹ and acute appendicitis¹⁰ are

treated laparoscopically. In small bowel emergencies, laparoscopy has been adopted as a first-line option in many elective indications of acute small bowel obstruction, and it is also emerging as a feasible alternative to treat this condition,¹¹ even though open surgery has been the gold standard therapy for adhesive small bowel obstruction for decades. The best approach for small bowel perforation has been evaluated previously,¹² and Sinha et al. described 25 patients with small bowel perforation in whom laparoscopic intracorporeal bowel repair was performed and concluded that laparoscopic intervention was safely and efficaciously extended to such patients, especially regarding the lower rate of wound complications.¹³ Additionally, some case reports described the feasibility and advantages of laparoscopy for small bowel perforation secondary to an ingested foreign body.^{14, 15} These reports suggest that laparoscopic procedures are also beneficial in the treatment of small bowel perforation. Even though, poor laparoscopic view due to such conditions as severe intestinal distension or adhesion can be a limitation of laparoscopy. Conventional laparotomy should be considered for the safety of operation in cases with poor laparoscopic view.

Gastrointestinal perforation is a critical adverse event with molecular-targeted therapies such as bevacizumab¹⁶; however, pazopanib, a small-molecule inhibitor of multiple protein tyrosine kinases that was used in our patient, does not appear to lead to gastrointestinal perforation.¹⁷

Although malignant UPS/MFH is one of the most common soft tissue sarcomas, metastasis to the small bowel has been reported rarely. To the best of our knowledge, there are 12 reported cases to date, including ours, describing small bowel metastasis of UPS/MFH (Table 1). The age range in the published studies was 25–73 years, and 10 patients (83%) were men. The patients had various primary sites of malignancy, namely, the extremities, trunk, and viscera. Symptoms related to gastrointestinal bleeding were seen in four patients (44%), and intussusception was seen in three patients (25%). All procedures were performed by open laparotomy for small bowel metastasis.

Our literature review also revealed that intestinal perforation secondary to metastasis of UPS/MFH is extremely rare, with only one reported patient.¹⁸ We believe that our case is the first report of laparoscopically-assisted repair of a small bowel perforation secondary to UPS metastasis. Although the perforation site was unclear preoperatively, laparoscopy readily identified the lesion in our patient. Therefore, minimally invasive surgery can be performed successfully with mini-laparotomy following laparoscopic observation.

Table 1. Cases of small bowel metastasis in UPS/MFH

No.	Author	Year	Age (years)	Sex	Primary site	Symptoms	Treatment for metastasis
1	Wasserman, T. H.	1974	69	Male	Shoulder	Unknown	None
2	Wasserman, T. H.	1974	78	Male	Unknown	Unknown	None
3	Jun MISONOU	1990	25	Male	Mediastinum	Tarry stool	Resection (OL)
4	Gorman, R. C.	1993	47	Female	Thigh	Intussusception	Resection (OL)
5	Blaauwwekel, E. E.	1997	75	Male	Right foot	Bleeding	Resection (OL)
6	Kanoh, T.	1998	45	Male	Right back	Intussusception	Resection (OL)
7	Tranter, S. E.	2000	73	Female	Left arm	Intussusception	Resection (OL)
8	Tamura, M.	2002	73	Male	Rib	Bleeding	Resection (OL)
9	Agaimy, A.	2007	30	Male	Left atrium	Abdominal pain, Bleeding	Resection (OL)
10	Oguri, Y.	2018	70	Male	Stomach	Abdominal fullness	Resection (OL)
11	Plestina, S.	2019	57	Male	Lung	Perforation	Resection (OL)
12	Our case	2019	46	Male	Mediastinum	Perforation	Resection (laparoscopic)

MFH, malignant fibrous histiocytoma; OL, open laparotomy; UPS, undifferentiated pleomorphic sarcoma.

Fig. 5. Pathological examination showing UPS with bizarre cells (white arrows) in all five tumors. Bar = 100 µm.

Generally, laparoscopic procedures for acute abdomen are limited to severe states related to a primary disease and/or the surgeon's experience, and conversion to open laparotomy is sometimes needed.^{19, 20} However, studies have reported the potential benefits of laparoscopic surgery regarding reduced morbidity, less requirement for analgesics, shorter hospital stay, and quicker recovery compared with open surgery.¹³ As an example, fortunately, our patient had a good outcome following the laparoscopically-assisted procedure, even though his was an extremely rare case of small bowel perforation secondary to UPS metastasis. Laparoscopic techniques for the small intestine are viable options

even in acute and uncommon situations, and small bowel perforation secondary to UPS metastasis should be considered in patients with UPS and acute abdomen.

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The authors declare no conflict of interest.

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