Case Report

A ruptured jejunal gastrointestinal stromal tumor with hemoperitoneum mimicking ovarian carcinoma

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Abstract: Background: Gastrointestinal stromal tumor (GIST) of the small bowel is a rare malignancy and accounts for only 0.1-3% of all gastrointestinal neoplasms. GISTs may mimic gynecologic tumors. The authors present an unusual case of ruptured jejunal GIST with hemoperitoneum mimicking ovarian carcinoma. Case presentation: A 57-year-old postmenopausal woman presented with progressive, generalized low abdominal pain for 3 months. Computed tomography (CT) imaging depicted a 9.6 cm-sized, complex and solid mass on the left pelvic cavity and a 3.7 cm-sized heterogeneous, enhancing mass on the right adnexa, suspicious for ovarian cancer accompanied by hemoperitoneum. There was no active bleeding. Laboratory examination revealed a low level of serum hemoglobin (6.7 g/dL), and a raised serum level of CA-125 (107.0 U/mL). Based on CT imaging findings and an elevated serum level of CA-125, bilateral ovarian cancer was suspected. The patient underwent exploratory laparotomy, and frozen section of the excised mass indicated malignancy originating from the small bowel jejunum. Consequently, small bowel segmental resection with mesenteric resection was done, and a debulking operation including hysterectomy, BSO, BPLD, omentectomy, and excision of multiple metastatic masses in the peritoneum was performed. Subsequent histopathologic examination confirmed the final diagnosis of high risk GISTs of the primary small bowel jejunum. The patient’s postoperative course was uneventful, and adjuvant Imatinib was administered. Conclusion: The authors report an unusual case of ruptured jejunal GIST with hemoperitoneum mimicking ovarian carcinoma. Therefore, GIST, in addition to ovarian cancer, should be considered in patients with an increased serum level of CA-125 and an abdominopelvic mass.

Keywords: Gastrointestinal stromal tumors (GISTs), small bowel and mesentery, jejunum

Background

Gastrointestinal stromal tumors (GISTs) is a rare tumor and it represents between 0.1 and 3% of all newly diagnosed gastrointestinal neoplasms but is the most common mesenchymal tumor of the gastrointestinal tract [1, 2]. Data from the surveillance, epidemiology, and end results registry from the U.S. National Cancer Institute found an age-adjusted yearly incidence of 6.8 per million [3, 4]. In Korea, GIST occurs in 10-20 people per year per 1 million people, and about 20-30% of the total GISTs are known to show a clinical progression to malignancy. Considering that Korea is composed of about 50 million people, it is estimated that 500-1000 new GIST patients are being reported annually, and the number of patients with malignant progression would be 100-300 people per year [5].

It mostly occurs in patients in the sixth decade of life and can arise at any site of the GI tract from the esophagus to the rectum. It also may present with GI hemorrhage, hemoperitoneum, bowel obstruction, or abdominal distension [6].

The mainstays of the treatment options are surgical resection and adjuvant therapy with tyrosine kinase inhibitors (imatinib), and this has led to dramatic improvements in survival of patients with GIST [7, 8].

Here, we report an unusual case of ruptured jejunal GISTs with hemoperitoneum mimicking ovarian carcinoma and describe its clinicopathologic findings in a postmenopausal woman.
Case presentation

A 57-year-old postmenopausal woman presented with progressive, generalized low abdominal pain of 3 months. She had several prior surgeries; she had undergone subtotal thyroidectomy due to benign thyroid nodule, open salpingectomy due to ectopic pregnancy, appendectomy and right breast mass excision due to a borderline tumor. Her family history included liver cancer (parental) and hypertension (sibling). On physical examination upon her admission, her vital signs were stable. An abdominal examination revealed tenderness with rebound tenderness. No abdominal mass was palpated. A digital rectal exam was normal.

On contrast enhanced CT scan, the 9.6 cm-sized mass was detected in the left pelvic cavity. This mass presented a heterogeneous density, representing hemorrhagic change. The mass slightly abutted the adjacent small bowel, but ovarian malignancy was suspected based on the location of the mass. Moreover, another 3.7 cm-sized heterogeneous enhancing mass was also detected in the right adnexa. We regarded the mass as bilateral ovarian cancer. In addition to the hemoperitoneum, sentinel clot sign, indicating the anatomic sites of hemorrhage, was suspected around the huge mass on pre-contrast CT images. There was no active bleeding (Figure 1). Hemoperitoneum was suspected to be due to left ovarian carcinoma rupture.

Laboratory examination revealed an increased serum level of CA-125 (107.0 U/mL). Other than the low Hb (6.7 g/dL), there were no unusual findings. Based on CT imaging findings and the elevated CA-125 level, the patient was suspected to have bilateral ovarian cancer.

The patient underwent exploratory laparotomy under suspicion of rupture of ovarian cancer with hemoperitoneum. Bloody ascites with a volume of 2-3 L was aspirated, and cytology was done. Abdominal wall adhesiolysis and multiple omental and peritoneal wall biopsies were done. The bleeding focus was the abdominal wall mass from the small bowel. The small bowel main mass was suspected to be small bowel sarcoma or small bowel GIST. The tumor was a 10-15 cm sized, friable mass (Figure 2). Small bowel segmental resection with mesenteric resection was done. Frozen section result...
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was spindle cell sarcoma from the small bowel. Adhesiolysis, metastatic site and retroperitoneal dissection were done. To obtain oncologic certainty, hysterectomy with pelvic node dissection, BSO, and infracolic omentectomy were done.

Microscopically, the tumor was composed of moderately cytologically atypical spindle cells with increased mitotic activity (20/50 high-power fields). Omentum and small bowel showed gastrointestinal stromal tumor, with high risk of malignant potential. Uterus, both adnexa and peritoneal wall biopsy had no specific findings. Pelvic and para-aortic lymph nodes were free from tumor cells, and peritoneal washings were negative for malignant cells. Immunohistochemical staining showed tumor cells were positive for C-kit (CD117), DOG1, CD34, and SMA (Figure 3).

The patient’s postoperative care was uneventful. Imatinib chemotherapy was administered and serum CA-125 decreased to normal at 1 month after surgery.

Discussion

GISTs originate from the interstitial cells of Cajal, and can arise at any site of the GI tract from the esophagus to the rectum; particularly, jejunal GISTs comprise 10% of all GISTs [9]. Since its symptoms are rather unclear, it is difficult to diagnose jejunal GIST preoperatively, so a high index of suspicion is needed for diagnosis of GISTs. GIST mostly has an endophytic pattern of growth [10]. GIST may present as an abdominal emergency, including GI hemorrhage, perforation, and obstruction. Especially, perforation of the mass is more common in GISTs of the small bowel compared to those of other anatomic sites [6].

Although small bowel GIST is a rare tumor, suspecting GIST with appropriate imaging, such as CT enterography, can be helpful in diagnosis process prior to surgery. CT enterography has been increasingly used to investigate when small bowel bleeding is suspected [11]. In the previous case study, there were reports of hemorrhagic potential due to the ulceration of the
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mucosa, and the incidence of bleeding was higher in duodenal GIST and small bowel GIST [6].

Although the diagnostic procedure may include several examinations, such as barium examination, capsule endoscopy, endoscopic ultrasonography, CT, MRI, and FDG-PET, none of them can establish the correct diagnosis [10]. In our case, we performed the CT of abdomen & pelvis instead of CT enterography because there was no active bleeding. The typical CT feature of GIST as a heterogeneously enhancing, well-defined mass. On our contrast-enhanced CT scan, a 9.6 cm size heterogeneously enhancing mass was detected, in the left pelvic cavity. There was also a sentinel clot, resulting in hemoperitoneum, around the huge mass in the left pelvic cavity. Moreover, another 3.7 cm-sized heterogeneous enhancing mass was also detected in the right adnexa. Based on CT image findings and elevated CA-125 level, ovarian malignancy with hemorrhagic change needed to be ruled out. There was no active bleeding, so the patient underwent exploratory laparotomy under suspicion of ovarian cancer.

The final diagnosis of GIST was made by histopathologic findings. GIST tumors have variable expression of markers, including C-KIT (CD117), CD34, CMA, S-100 and desmin, but are almost always positive for C-KIT (CD117) and DOG1 [6]. In our case, the mass was positive for C-kit (CD117), DOG1, CD34 and SMA.

GISTs may be categorized as very low, low, intermediate and high-risk tumors by the Fletcher et al. prognostic classification [6]. In our case, tumor size was > 10 cm and mitotic count was > 5/50 HPF. Thus, our case was defined as high risk.

In our case, an incorrect initial diagnosis of ovarian cancer was made because ovarian cancer is more common in postmenopausal women, and CT findings with the tumor locations of left pelvic cavity and right adnexa suggested bilateral ovarian cancer. The unusually huge, multiseptated cystic mass occupying the entire abdomen and accompanying hemoperitoneum was initially misdiagnosed as ovarian cancer based on such CT imaging findings and elevated CA-125.

Conclusion

In summary, we present an unusual case of ruptured GIST of the jejunum, which mimicked ovarian malignancy based on anatomic location, CT imaging findings, and serum level of CA-125, in a postmenopausal woman.

Small bowel GIST is a rare tumor with spontaneous perforation and life-threatening hemorrhage. Our case suggests that gynecologists should consider GIST in the differential diagnosis of patients presenting with an abdominopelvic mass.

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Disclosure of conflict of interest

None.

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