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Rectal GIST mimicking an ovarian mass; a case report

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ABSTRACT

Introduction. Gastrointestinal stromal tumors (GISTs) are the most frequent mesenchymal neoplasms of the gastrointestinal (GI) tract. However, rectal GISTs are rare among these tumors. *Case Presentation*. A 60-year-old Caucasian woman had surgery for an ovarian mass with carcinomatosis. The patient underwent exploratory laparoscopy, which found a large amount of ascites, epiploic and appendicular carcinomatosis. The Hudson operation was performed to completely and en bloc remove the recto-sigmoid junction, the uterus, and adnexa. The appendix and the carcinomatosis were also removed with no residual tumor. After that, she was sent to the oncology department to start Imatinib therapy. *Conclusion*. Rectal GIST is a rare entity with varied clinical manifestations and a high risk of recurrence, which may complicate the diagnosis in women with an abdominopelvic tumor mass. Consequently, even if GIST can mimic gynecological tumors, such as ovarian tumor, the surgeon must be aware of this condition and take it into account in establishing the differential diagnosis, especially since surgical treatment is essential in localized rectal GIST.

Category: Case Presentation

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Introduction

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract, with an incidence of 10–15 per million per year [1-3]. Most of them originate from the stomach (50–60%), followed by the small intestine (20–30%), the rectum (5%), and the colon 1% [4-6].

Rectal gist can mimic an ovarian mass, which in some cases can lead to a differential diagnosis dilemma for the physician. Our aim is to discuss from this case report the differential diagnosis when faced with a pelvic mass in the post-menopausal woman.

Case presentation

We report the case of a 60-year-old Caucasian woman with no past medical history who was referred to our department for abdominal pain associated with a left abdominopelvic mass. Her blood pressure was 13/7 mm/Hg, and she had a normal heart rate. The body temperature was 37^0 Celsius. On physical examination, a 15 cm left- abdominal mass was found associated with a mass of the Douglas pouch with reduced mobility.

Ultrasonography showed a 16.5 cm large solid pelvic mass with a cystic component extended to the abdominal cavity, with abundant ascites. Thoraco-abdomino-pelvic computed tomography (CT) showed a 17 cm large solid mass of probable ovarian origin, with cystic component adherent to the posterior surface of the uterus and peritoneal carcinomatosis. No other sign of malignant disease was found on CT (Figures 1-3).

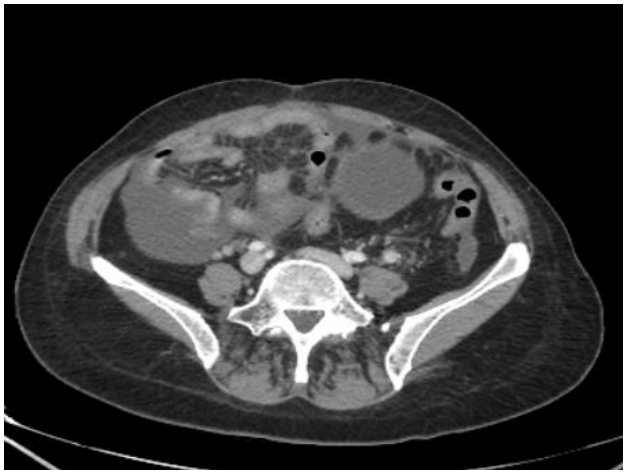

Figure 1. CT scan axial section

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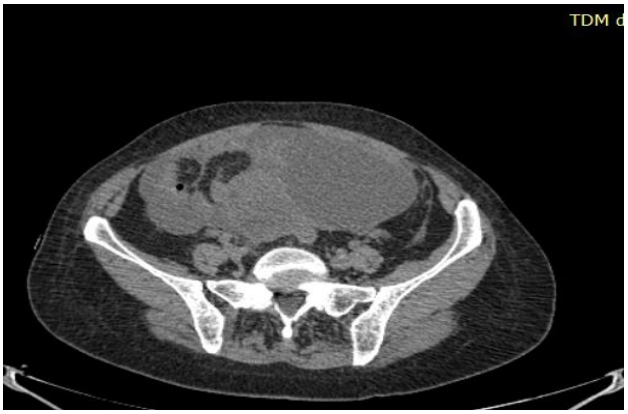

Figure 2. Ovarian solid mass with cystic component on CT scan axial section

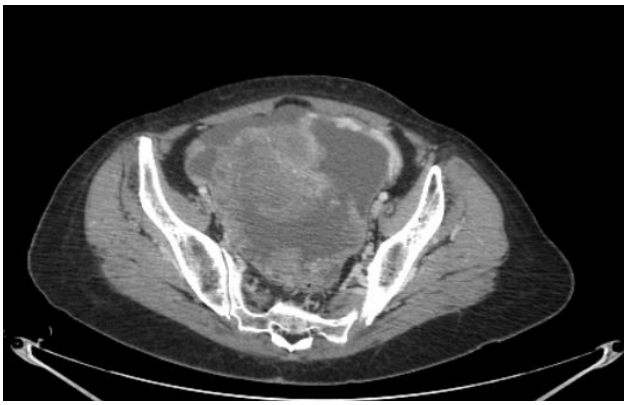

Figure 3. Ovarian mass adherent to the posterior surface of the uterus on CT scan axial section

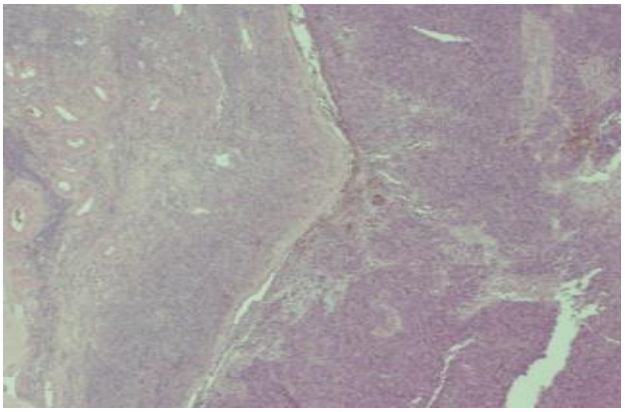

Figure 4. The tumor invaded the ovary (x 100)

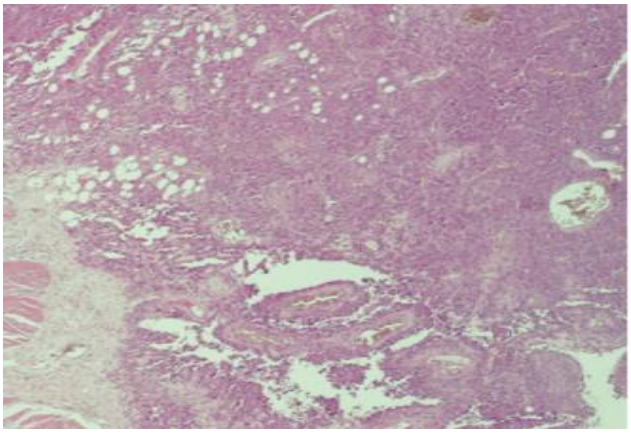

Figure 5. Tumor centered in the rectal subserosa (x 100)

Laboratory findings showed an increase in CA-125 (776,80 U/mL) and normal CA-19.9 and CEA.

The multidisciplinary team decided to start with upfront surgery. The patient had exploratory laparoscopy, which found a-3L- ascites, epiploic and appendicular carcinomatosis. The pelvis was completely filled by a 17 cm mass apparently arising from the ovaries. The mass was adherent to the sigmoid colon, the ovaries, the posterior wall of the uterus, and the peritoneum covering the bladder. The mass was already ruptured and clogged by the ileum and the omentum. Therefore, we decided to carry on a midline laparotomy. Hudson surgery was undertaken to remove completely and on bloc the recto-sigmoid junction, the uterus, and adnexa. The appendix and the carcinomatosis were also removed with no residual tumor. Surgery took three hours and twenty minutes and the patient was transfused with four red blood cell bags. The immediate follow up was without complications.

After the first postoperative day, the patient was mobilized. On the third day, she returned to normal bowel function and received a clear liquid diet. The next day she started a regular diet. She was finally discharged home after five days. She was admitted after ten days for stercoral peritonitis by anastomotic leakage. She had a Hartman procedure and was discharged after five days.

Histopathology was concordant with a 17cm- high risk-colorectal GIST invading the myometrium, the cervix, the peritoneum covering the bladder, and adnexa. Immunohistochemical staining was positive for CD34 and CD117(C-KIT) (Figures 4-7).

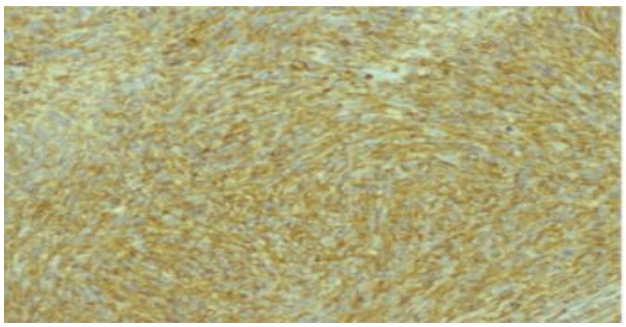

Figure 6. CD34 immunostaining (x200)

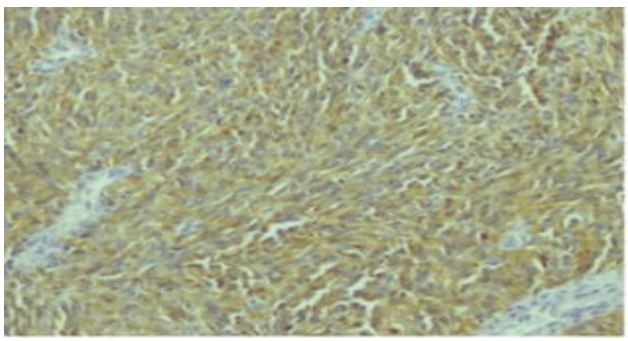

Figure 7. CKIT immunostaining (x 200)

Adjuvant treatment by Imatinib was planned with three months follow-up, the patient being well and disease-free. The reverse Hartman will be scheduled in 6 months.

Discussion

GISTs are rare tumors and may present a diagnostic dilemma in women with abdominopelvic mass. It is typically present in adults over 50 years. They could present a pelvic mass and lead to misdiagnosis with gynecologic tumors [7-9].

GISTs arise from the interstitial cells of Cajal, which can be found in the myenteric and submucosal plexus of the GI tract [10-12].

The most common presentations of GISTs include gastrointestinal bleeding followed by abdominal pain and bowel obstruction. Other presenting symptoms are abdominal distention, palpable mass, and features of perforation. Abdominal pain and/or GI bleeding are seen more often in large GISTs owing to the vascular nature of the tumor [13-15].

In addition to the nonspecific clinical presentation, no characteristic biochemical markers or typical imaging features of GIST have been demonstrated. Thus, a contrast-enhanced CT scan should be performed to determine the extent of the tumor and the presence or absence of metastatic disease, as well as the possibility of complete resection. Diagnosis is established based on histopathology and immunophenotyping. CD117 (C-kit) and DOG-1 have the most diagnostic value with high sensitivity (94%) [16]. In our case, the diagnosis was confirmed through C-Kit.

Pelvic mass in a post-menopausal woman is a common disorder that represents a spectrum of conditions with many causes. A mass may be gynecologic, digestive, or ureteral. Concerning gynecological masses, they are commonly ovarian but may also arise from the uterus or fallopian tube. The main etiologies have been summarized in Table 1 [17-19].

Table 1. The main etiologies of pelvic mass in a post-menopausal woman

gynecologic		**ureteral**	**digestive**	**others**
benign	malignant			
*Mature teratoma *Ovarian torsion *Serous/mucinous cystadenoma *Endometrioma *Hydrosalpinx *Leiomyoma *Tubo ovarian abscess	*Borderline tumor *Epithelial carcinoma *Ovarian germ cell tumor *Sex cord tumor *Endometrial carcinoma *Fallopian tube carcinoma	*Bladder diverticulum *Diverticular abscess *Pelvic kidney	*Gastrointestinal carcinoma *Krukenberg tumor *Sigmoidite	*aneurysm *Retroperitoneal sarcoma

Clinical history and examination are crucial to determine the possible causes. Pain is a common symptom. Obstructive symptoms such as urinary, bowel dysfunction, or even lower limb venous thromboembolism are frequent symptoms. Uterine masses can be associated with symptoms of vaginal bleeding or dyspareunia and dysmenorrhea. Clinical abdominal examination may reveal a palpable mass or demonstrate ascites in the cases of ovarian cancer.

The standard treatment of localized GIST is surgical with negative margins without lymph node resection. During surgery, it is important to guarantee complete tumor resection and avoid tumor rupture.

The surgical treatment of rectal GISTs can be grouped into two categories: local resection and radical excision. The former includes trans-anal endoscopic microsurgery (TEM) and trans-anal minimally invasive surgery (TAMIS), while the latter includes low anterior resection and abdominoperineal resection (APR), and pelvic exenteration [20-22].

Large and aggressive rectal GISTs with extensive extra-rectal growth— invading adjacent structures, such as the prostate, vagina, or sacrum, usually require a more extensive resection [23,24].

The most significant factor related to outcome is complete resection, and this can be accomplished in 40-60% of all GIST patients [25,26]. Rectal GISTs have a high rate of local recurrence regardless of the surgical procedure. That's why adjuvant medical therapy has gained importance in the management of colorectal GIST. Beyond minimizing disease recurrence, adjuvant therapy can also be useful in case of intraoperative tumor rupture and incomplete tumor removal.

Imatinib, a specific tyrosine kinase receptor inhibitor (TKI), is the only available and approved therapy as the first-line treatment for advanced GIST [27-29].

The colorectal GISTs usually metastasize to the liver, peritoneum, and sometimes to the lung and bone. Imatinib is the first-line treatment for advanced disease for patients with sensitive mutations to TKI, which should be continued for an unlimited period of time according to the ESMO [30,31].

The prognosis of the tumor depends on the age at presentation, anatomic location, size, immuno-histo-chemistry, molecular genetics, and mitotic activity [32,33].

Recurrence of GIST is a common complication despite full resection with negative margins. That's why regular follow-up examinations and investigations are compulsory. The NCCN guidelines suggest physical

examination and abdominopelvic CT scan every 3 to 6 months in the first 3 to 5 years after surgery and annually thereafter [34-36].

Conclusions

Rectal GIST is a rare tumor with malignant potential and a high risk of recurrence, which poses a diagnostic dilemma in women with abdominopelvic mass. As it may mimic gynecologic tumors like an ovarian tumor, the surgeon should be aware of this condition and consider it in the differential diagnosis.

Authors' contribution

MG, OK, SR: data collection, drafted the manuscript

MS, RD: review of the literature

RC, KR: drafted the manuscript

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Conflict of interest disclosure

There are no known conflicts of interest in the publication of this article. The manuscript was read and approved by all authors.

Compliance with ethical standards

Any aspect of the work covered in this manuscript has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

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