

## Acute perforation of Gastrointestinal stromal tumor

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**Section:** Abdominal imaging

**Area of Interest:** Abdomen

**Imaging Technique:** CT

**Special Focus:** Abscess Neoplasia Case Type: Clinical Cases

**Authors:** Michaela Kubincova<sup>1,2</sup>, Kris Mertens<sup>2,3</sup>, Kristof De Cuyper<sup>2</sup>, Leen Mortier<sup>4</sup>, Luc Verheyen<sup>5</sup>, Filip Vanhoenacker<sup>2,6,7</sup>

**Patient:** 32 years, female

### Clinical History:

A 32-year-old female presented at the emergency department with fever and progressive abdominal pain in the left iliac and periumbilical region lasting 3 days. Laboratory examination showed elevated inflammatory markers C-reactive protein and leucocytosis. The patient is followed for ovarian cysts every year and has no history of previous surgery.

### Imaging Findings:

Contrast-enhanced computed tomography (CECT) with rectal contrast was done to rule out diverticulitis. CECT demonstrated a pelvic mass with central air posteriorly to abdominal wall surrounded by thickened and enhancing peritoneal reflections and mesenteric fat stranding (Figure 1 a, b, c). This mass is complicated by a fistula formation superiorly towards the bowel (Figure 2 a, b). In addition, free intraperitoneal air was seen in the left hypochondrium (Figure 3 a, b).

Laparotomy revealed that the mass originated from the antimesenteric side of the jejunum distal to the ligament of Treitz, descending into the pelvis covered by greater omentum.

Partial enterectomy and mass resection was done. Histopathological examination revealed spindle cell type of gastrointestinal stromal tumour (GIST). Molecular analysis showed expression of tyrosine kinase growth factor receptor, also called c-Kit proving the sensitivity to a tyrosine kinase inhibitor. The patient was treated with an adjuvant molecular targeted therapy with Imatinib.

### Discussion:

GIST's are mesenchymal tumours arising from the interstitial cells of Cajal accounting for 1-3% of all gastrointestinal tumours. They express a KIT protein (90%). Sporadic form occurs in adults. Inherited forms are associated with inherited syndromes. There are 3 histological types: spindle cell (70%), epithelioid (20%), mixed (10%). They commonly arise from the stomach (50-60%), small intestine (30-35%) and rarely colon, oesophagus, peritoneum, omentum and mesentery [1,2].

Clinical manifestations are nonspecific. Complications of GISTs are haemorrhage, rupture, bowel perforation or obstruction. They may metastasize to the liver, peritoneum, omentum, mesentery and rarely lymph nodes, lungs or bones [1].

Ultrasound (US) is often used as the initial examination but the lesion's origin may be unclear. US demonstrates a homogeneous or heterogeneous solid/cystic mass. Endoscopic ultrasound may diagnose small submucosal GISTs [1,3].

Barium examination depicts intraluminal or submucosal mass with smooth filling defect and obtuse marginal angles but is rarely performed nowadays [1,3,4].

CT imaging is the method of choice for detection, diagnosis and staging of GIST's providing evaluation of tumor size, organ invasion, distant metastasis and peritoneal seeding. [1,3,5].

Small tumours (<5 cm) are solid, well-defined, mostly intraluminal and homogeneously enhancing. Larger lesions are heterogeneous due to haemorrhage, necrosis or cystic degeneration. They are either well- or ill-defined and heterogeneously enhancing with exophytic or 'dumbbell-like' appearance. Cavitory lesions may contain air, air-fluid levels or contrast resulting from mucosal ulceration and fistulous communication with the bowel. The "*Toricelli-Bernoulli*" crescentic necrosis sign consists of deep ulceration with air-fluid level [1,2,4].

Additional Magnetic Resonance Imaging (MRI) may be used to assess the origin and extent of the disease and internal structure. Diffusion Weighted Imaging (DWI) helps to for characterize the cellularity and internal tumoral components [1,5]. MRI shows heterogeneous mass due to haemorrhage, necrosis or cystic degeneration. Signal intensity of the haemorrhage depends on its age. The solid portions are low on T1- and high on T2- weighted images and enhance on post-contrast images. [1,4].

Positron emission tomography is used for staging, treatment monitoring and to differentiate viable, malignant or recurrent tumours. GISTs show uptake of fluorodeoxyglucose and photopenic necrotic areas [1,3,4].

Histologic examination and molecular analysis for KIT-protein expression are mandatory for final diagnosis and defining treatment regime [1].

GIST's are treated by resection with an overall 5-year survival rate of 40-65%. Adjuvant immunotherapy is used in KIT-positive, inoperable, recurrent and metastatic disease. Treatment-related complications are haemorrhage, pneumatosis, perforation and fistulisation [1].

Written informed consent for publication has been obtained.

**Differential Diagnosis List:** Complicated gastrointestinal stromal tumour, Inflamed Meckel's diverticulum, Infected urachal cyst, Gastrointestinal lymphoma, Gastrointestinal leiomyosarcoma, Small bowel adenocarcinoma

**Final Diagnosis:** Complicated gastrointestinal stromal tumour

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## Figure 1

a



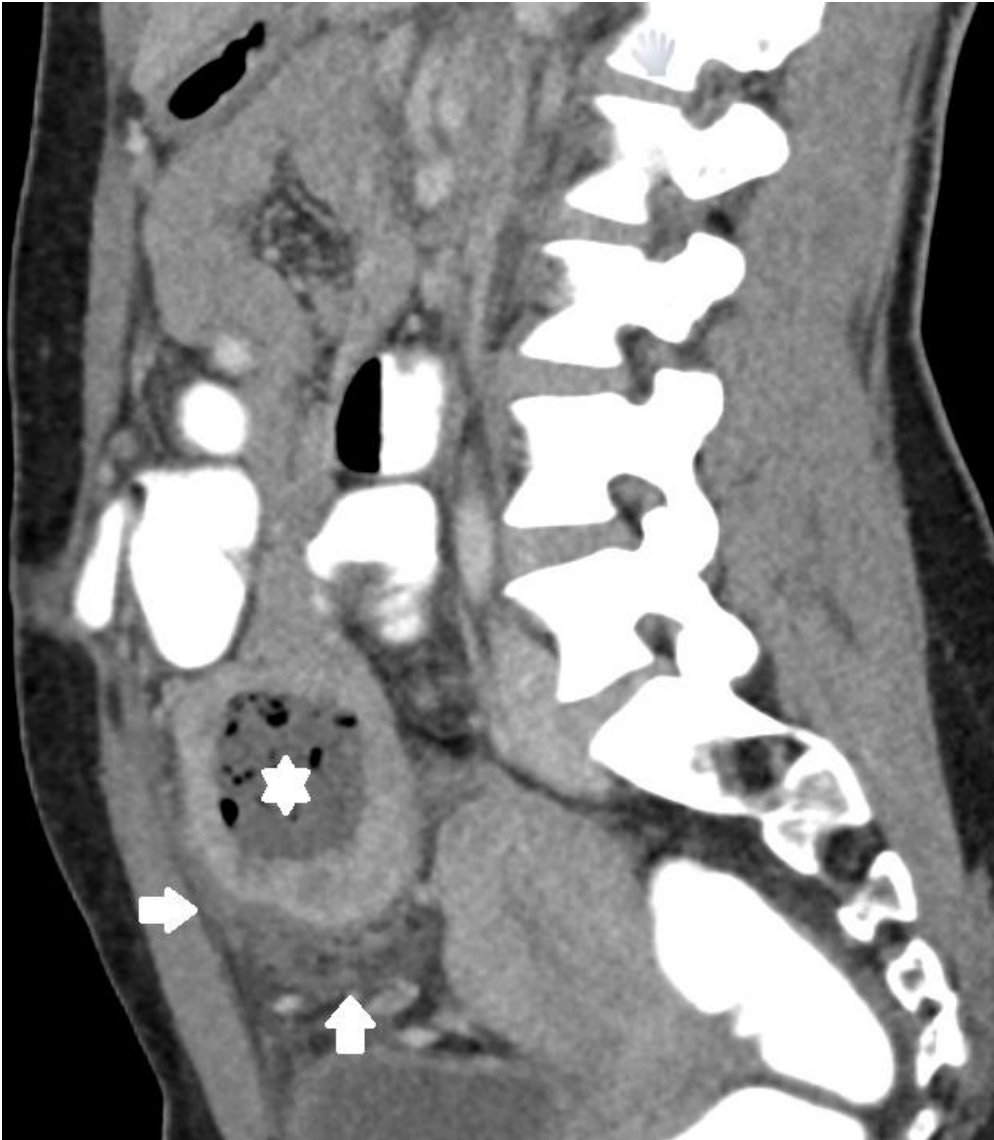
**Description:** Axial (a), coronal (b) and sagittal (c) images. There is a well-defined extraluminal intraperitoneal mass with thickened enhancing wall and low attenuating central necrotic region containing air in the pelvic region inferior to the antimesenteric side of the proximal jejunum side (asterisk in a/b/c). There is surrounding mesenteric fat stranding with thickening and enhancing of the peritoneal reflections in keeping with metastatic spread (white arrows in a/b/c) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium

b



**Description:** Axial (a), coronal (b) and sagittal (c) images. There is a well-defined extraluminal intraperitoneal mass with thickened enhancing wall and low attenuating central necrotic region containing air in the pelvic region inferior to the antimesenteric side of the proximal jejunum side (asterisk in a/b/c). There is surrounding mesenteric fat stranding with thickening and enhancing of the peritoneal reflections in keeping with metastatic spread (white arrows in a/b/c) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium

c



**Description:** Axial (a), coronal (b) and sagittal (c) images. There is a well-defined extraluminal intraperitoneal mass with thickened enhancing wall and low attenuating central necrotic region containing air in the pelvic region inferior to the antimesenteric side of the proximal jejunum side (asterisk in a/b/c). There is surrounding mesenteric fat stranding with thickening and enhancing of the peritoneal reflections in keeping with metastatic spread (white arrows in a/b/c) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium

**Figure 2**

a



**Description:** Coronal (a) and sagittal (b) images. Note a fistula containing air (white arrowhead in a/b). There is also a small non-incarcerated umbilical hernia containing colon transversum on the sagittal images (b, black arrowhead) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium

b

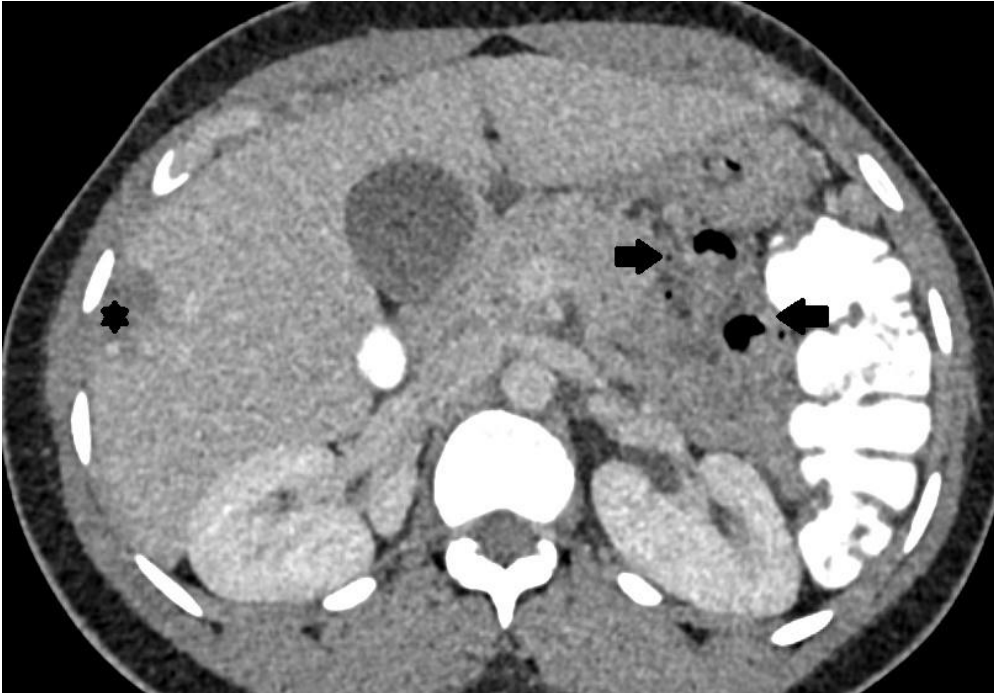


**Description:** Coronal (a) and sagittal (b) images. Note a fistula containing air (white arrowhead in a/b). There is also a small non-incarcerated umbilical hernia containing colon transversum on the sagittal images (b, black arrowhead) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium

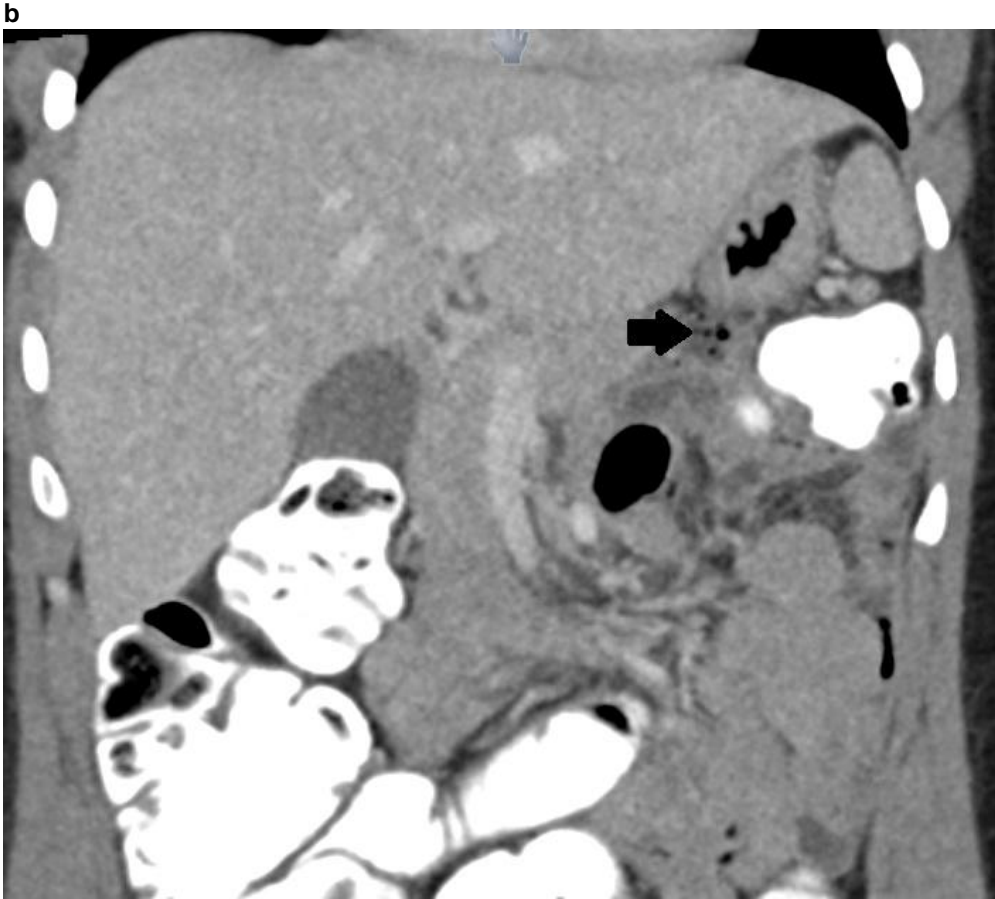


### Figure 3

a



**Description:** Axial (a) and coronal (b) images. There is an extraluminal intraperitoneal air - pneumoperitoneum in the left hypochondrium indicative of tumor rupture (black arrows in a/b). Also note an incidental liver hemangioma in segment V on the axial images (black asterisk in a) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium



**Description:** Axial (a) and coronal (b) images. There is an extraluminal intraperitoneal air - pneumoperitoneum in the left hypochondrium indicative of tumor rupture (black arrows in a/b). Also note an incidental liver hemangioma in segment V on the axial images (black asterisk in a) **Origin:** © F.M. Vanhoenacker, AZ Sint-Maarten, Mechelen, Belgium