

## Patient Information

- Name: 李X光
- Age: 58 y/o
- Gender: male
- Marital status: married
- Hospitalization: 96-01-02

## Chief Complaint

- Sudden onset low abdominal cramping pain in the afternoon

## Present Illness

- Persistent low abdominal fullness without association with meals and RLQ tenderness since 6 days ago.
- Mildly easy fatigue and dizziness in this week
- Sudden onset of severe low abdominal cramping pain, especially RLQ
- Non radiating, accompanied by fever with chills and nausea, without vomiting this afternoon

## Past History

- Duodenal ulcer 29 years ago

## Physical Examination

- Low abdominal tenderness, especially RLQ.
- Diffuse rebounding tenderness(+),
- Murphy's sign(-).

## Lab Data

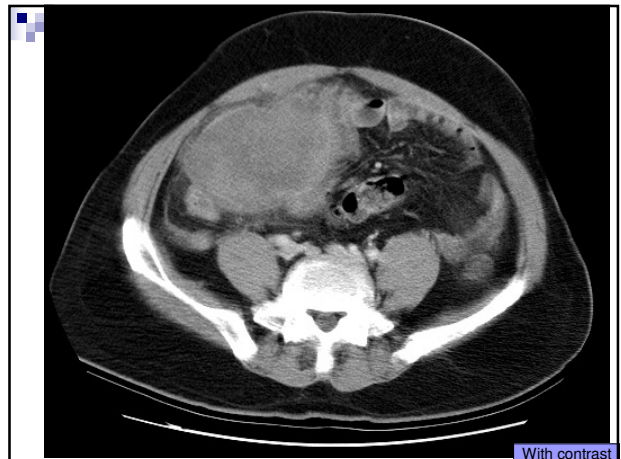
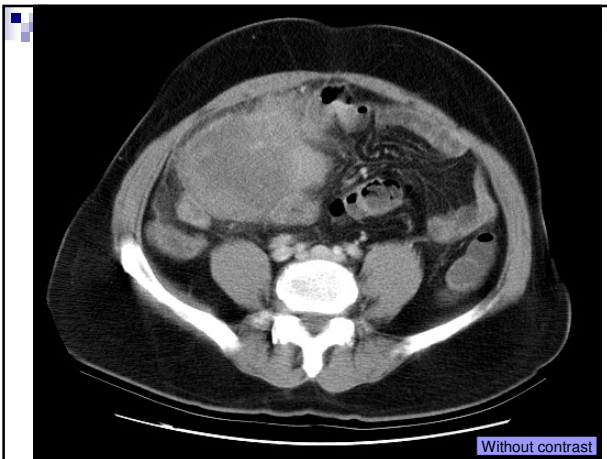
- |                                  |                                 |
|----------------------------------|---------------------------------|
| ■ 血液 (2007-1-02)                 | ■ 生化取樣 (2007-1-02)              |
| ■ WBC [4.0-11.0 x10.e3/uL] 14.89 | ■ Glucose(血液)[80-140 mg/dl] 153 |
| ■ RBC [4.2-6.1 x10.e6/uL] 3.99   | ■ CRP (血液)[0.0-0.8 mg/dl] 10.00 |
| ■ HGB [12-18 g/dL] 8.8           |                                 |
| ■ HCT [37-52 %] 28.5             |                                 |
| ■ MCV [80-99 fL] 71.4            | ■ 尿液檢驗 (2007-1-02)              |
| ■ NEUT [40-74 %] 82.8            | ■ Negative finding              |
| ■ %LYM [19-48 %] 14.0            |                                 |

## Imaging study-1

- Chest x-ray: negative finding
- KUB:
  - Nonspecific gaseous pattern of bowel.
  - Fecal material distension of abdominal-pelvis

## Imaging study-2

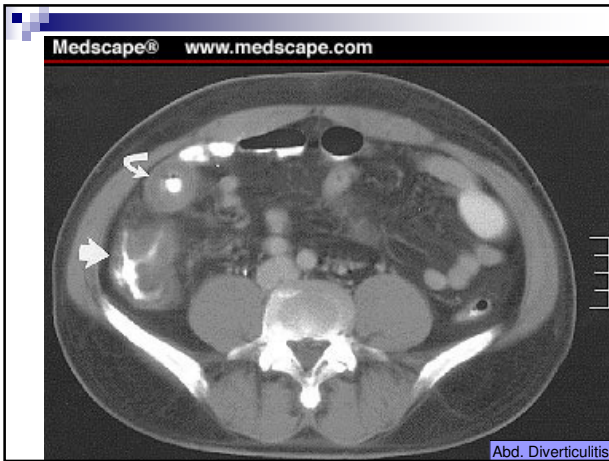
- CT
  - An irregular soft tissue mass, 9.5x7 cm at the right lower abdomen, inhomogeneous enhancement
  - The mass is suspected arising from the terminal ileum.
  - Right lower abdominal mass, r/o small bowel tumor, abscess, diverticulitis



## Different diagnosis

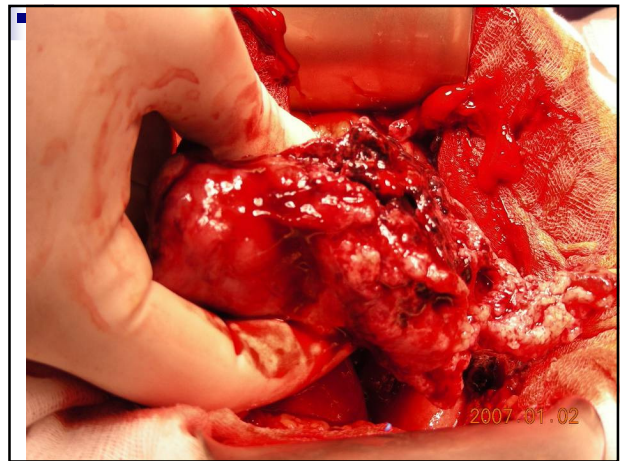
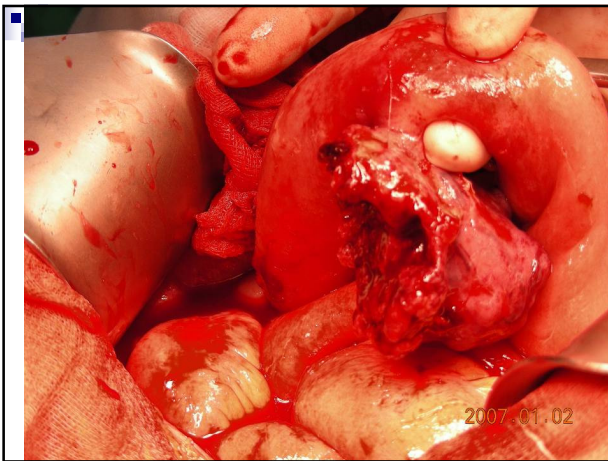
- Small bowel tumor
- Abscess: small bubbles or large collection of air, fluid-fluid level
- Diverticulitis: an exophytic pouch with air-filling or gas, small collection of fluid





## Operation

- Pre-op dx: Peritonitis
- OP method: Excision of intraperitoneal tumor by segmental resection of intestine & Partial omentectomy
- Post-op dx: Intestine tumor , R/O malignant GIST with rupture and hemoperitoneum
- Op finding :
  - One 10 X10 X 10 cm fragile exophytic hypervascular tumor arising from jejunum , 40 cm below ligament of Trietz with rupture.
  - Psuedotumor formation surrounding by omentum , R't side colon & intestine
  - About 200 cc blood in abdominal cavity on opening peritoneum



## Pathologic Report-1

- Intestine, small, jejunum, segmental resection, gastrointestinal stromal tumor (GIST), high risk with ruptured
- Soft tissue, intra-abdomen, excision, tumor involvement
  - one excised mass with attached small bowel 7.2 cm\*7.1 cm
  - 3 separate soft tissue fragments and 1 membranous soft tissue, in fresh status

## Pathologic Report-2

- Microscopically, ruptured high risk GIST
  - **High risk** GIST according to the risk categories of Fletcher CDM et al (**size>5 cm and mitotic figures> 5 / 50HPF**)
- The immunohistochemical study diffusely strongly positive of **CD117** and **CD34** and negative of desmin and S-100 protein.
- The both cut ends of small intestine are free of tumor.

## Discussion

## Clinical Presentation-1

- Frequently diagnosed incidentally with nonspecific symptoms during endoscopy or surgery or radiologic studies
- Manifestation: GI disease or an emergent condition such hemorrhage or obstruction
- Up to 75% of GIST found < 5 cm
- >5 cm more likely to be symptomatic → a palpable abdominal mass or swelling, abdominal pain, nausea, vomiting, anorexia, and early satiety
- 60% to 70% stomach, 20% to 30% small intestine and less than 10% in the esophagus, colon, and rectum

## Clinical Presentation-2

- GIST metastases : intra-abdominal, either with metastases to the liver, omentum, or peritoneal cavity
- Lymphatic metastasis is rare
  - most lesions thought to be nodal metastases by imaging studies simply represent metastatic deposits of tumor nodules in the omentum or peritoneum rather than true lymphatic spread
- Intestine GIST: worse prognosis
  - Predominant site : jejunum, followed by ileum and duodenum
  - The large lesions may be highly vascularized

## Lab

- No specific lab test

## Image study

- CT: important to diagnosis and staging
  - Small GIST(<5cm) homogeneous density
  - Intermediate GIST(5~10cm) irregular shape, heterogeneous density
  - Large GIST(>10cm) : irregular shape, heterogeneous density, locally aggressive behavior, distant metastasis

## Treatment

- **Primary, localized**
  - Surgery: definitive treatment
  - A clinical trial: Pre-op imatinib with resectable or potential resectable
- **Malignant or metastasis, recurrent**
  - Conventional C/T: 0~4% benefit
  - Intraperitoneal C/T: useless
  - Surgery: not significant role, due to liver or peritoneal metastasis
  - Target therapy: imatinib → blocking KIT receptor (Tyrosine kinase) inducing apoptosis, optimal dose undetermined

## Prognosis

- Primary, localized
  - 40% with complete surgical resection 5-year survival rate 54%, high risk for recurrence and ultimately prove to be a life-threatening disease.
  - large tumor size (>10cm) the only factor that reduced disease-specific survival
  - Size <5cm with complete resection → good prognosis
  - 10% disease free with long term follow-up

1	400mg	Major response, durable for >2yr.
40 (36 GIST)	400 to 1000mg	Partial remissions in 19/36 (53%) GIST with additional minor responses in 6/36 (17%). Total clinical benefit rate = 70% No responses in non-GIST.
147	400 or 600mg	Partial remissions in 97/147 (66%) with additional minor responses and durable stable disease in 25/147 (17%). Total clinical benefit rate = 83%, no differences in doses.
51 (27 GIST)	800mg	Complete remissions in 4%, partial remissions in 67%, with additional minor responses and durable stable disease in 18%. Total clinical benefit rate = 89%.
346	400 vs. 600mg	Complete remissions in 5%, partial remissions in 45%, with additional minor responses and durable stable disease in 32%. Total clinical benefit rate = 82%, no differences in doses.
746	400 vs. 800mg	Complete remissions in 2%, partial remissions in 46%, with additional minor responses and durable stable disease in 26%. Total clinical benefit rate = 74%, no differences in doses.

## Reference:

- Robbins: pathologic basis of disease 6th edition
- Feldman: Sleisenger & Fordtran's Gastrointestinal and Liver Disease, 8th ed
- Armstrong: Diagnostic imaging