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# General Data

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- ◆ ge: 63-year-old
- ◆ Sex: male
- ◆ Date of admission: 2004-03-01

# Chief Complaint

- ◆ Accident gastric mass found by PES in health check-up at 敏盛 hospital on 2004/02/04

# Present Illness - 1

- ◆ This 63 y/o man denied any systemic disease before. He received health check-up at 敏盛 hospital on 2004/02/04 and gastric mass found by PES was told. Before this, he complained post-prandial fullness for one year. Nausea, vomiting, body weight loss, or coffee ground was denied.

## Present Illness - 2

- ◆ PES → 2004/02/16
- ◆ Upper GI series → 2004/02/16
- ◆ EUS → 2004/03/01
- ◆ Abdominal and pelvic CT → 2004/03/01
- ◆ Operation → 2004/03/02



# Personal History



- ◆ Smoking : 2 PPD for 20+ years
- ◆ Drinking : social
- ◆ Allergy : denied
- ◆ Denied specific past or family history

# Lab Data

WBC = 7180 /uL

RBC = 4960000 /uL

HGB = 16.0 g/dL

Platelet = 208000 uL

Blood type = O(+)

GOT = 26 IU/L

GPT = 23 IU/L

Stool OB = (+/-)

# PES Findings

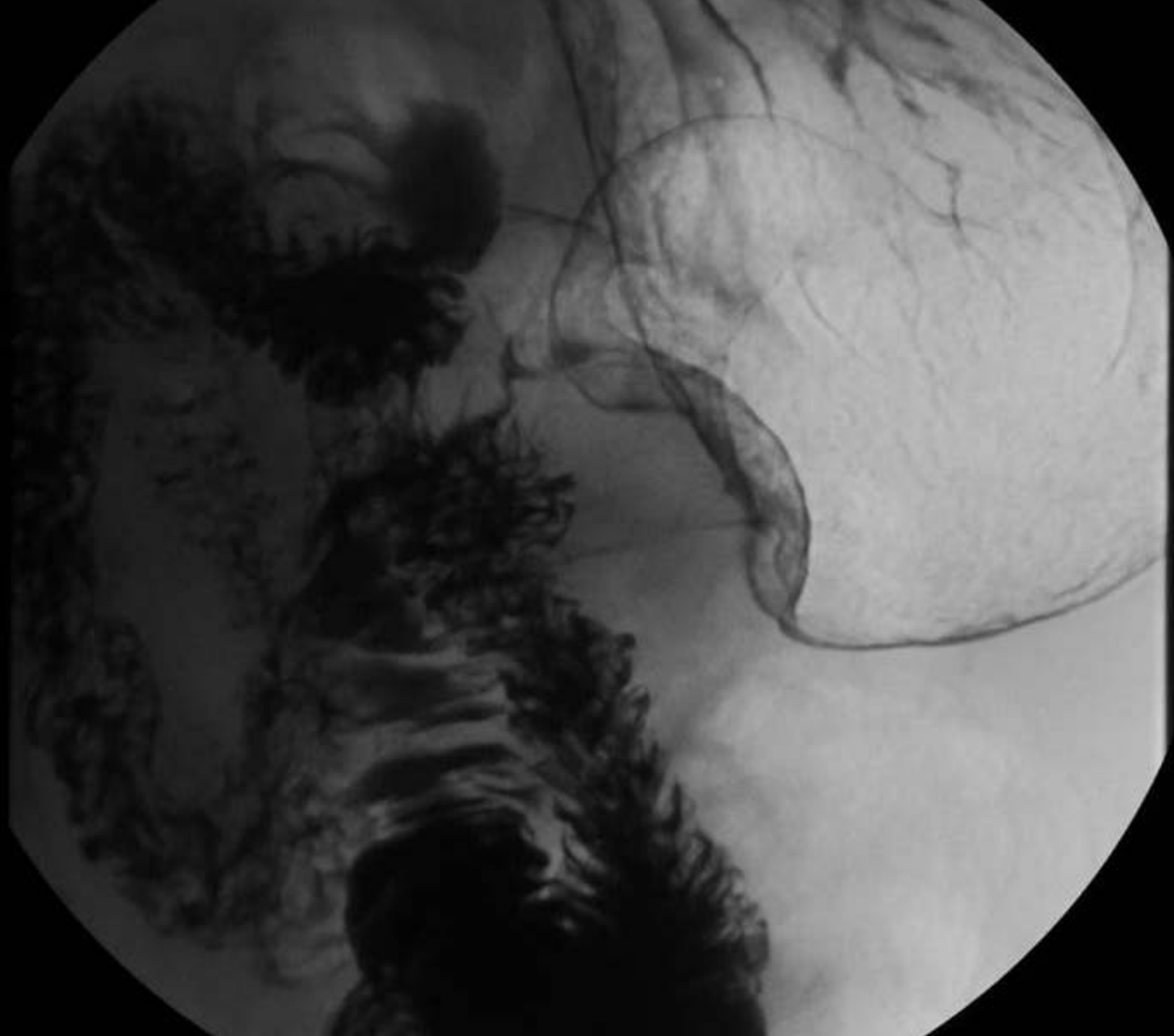


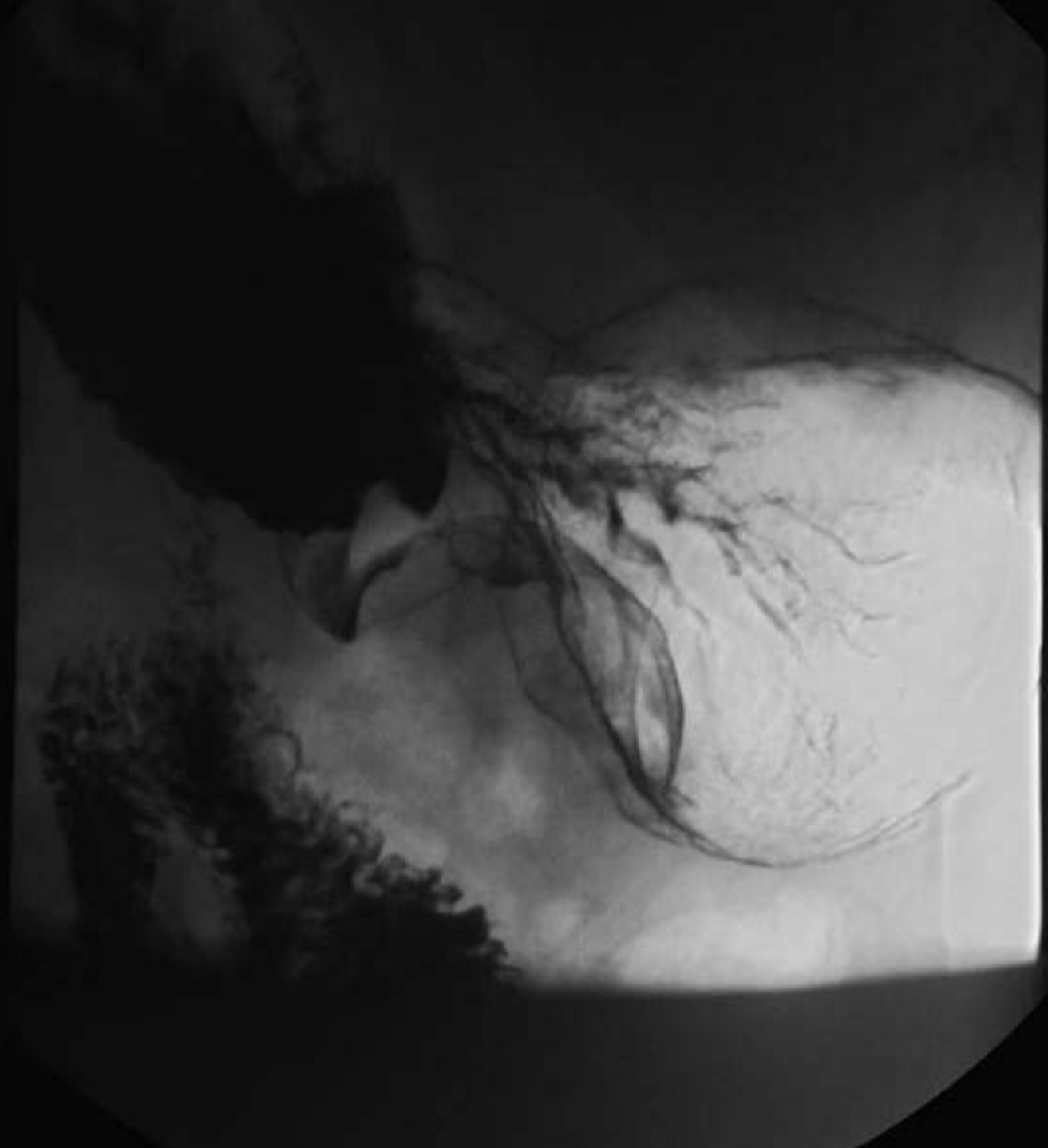
- ◆ A round, well-defined, protruding submucosal mass about 2-3 cm in diameter
- ◆ Biopsy → chronic gastritis

# Image Findings - UGI

1. One single, smooth, regular, and round filling defect (3.2 cm x 3.5 cm in dimension) with acute angle at greater curvature side of posterior wall of gastric antrum, forming the "double contour" appearance, no relationship to position changing
2. No central ulceration nor orifice was observed
3. No duodenal curve dilatation
4. The mucosa and gastric fold were relatively intact







# Image Findings – EUS

- ◆ A mixed echoic tumor (about 26 mm in diameter) with some internal hypoechoic density that arised form 4th layer of gastric wall

- ◆ Impression :  
Intramural submucosal tumor  
→ leiomyoma or leimyosarcoma  
from muscularis propria  
→ benign lesion is more favored (small size, smooth margin)



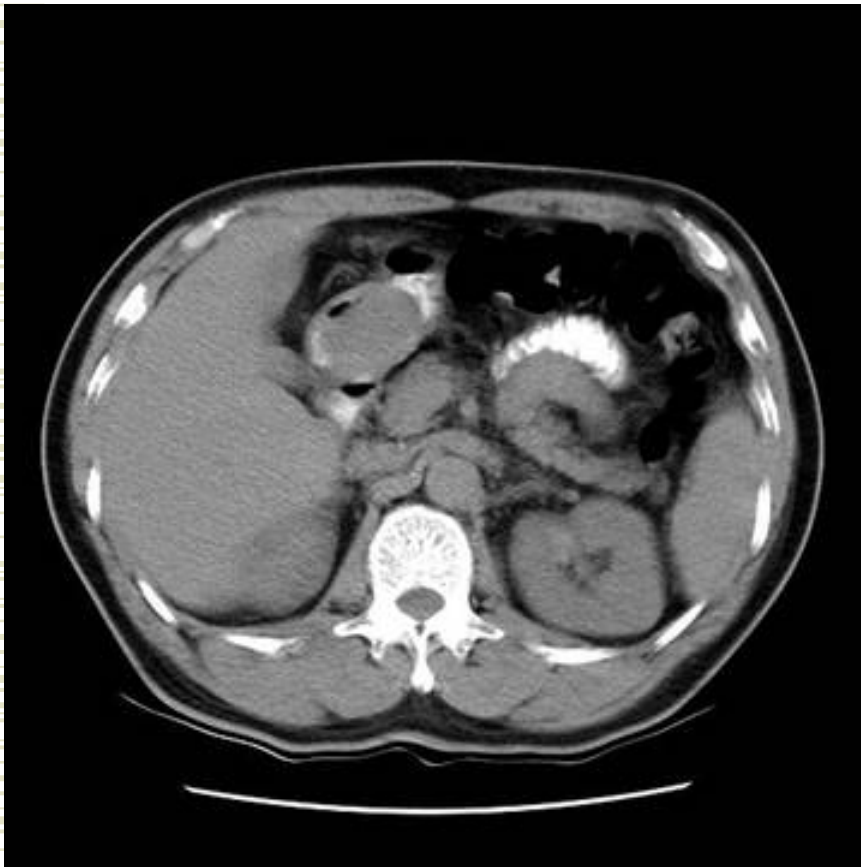
# Image Findings – Abdominal CT Scout View

Bowel gas in A-colon and T-colon → Suspect due to EUS



# Image Findings – Abdominal CT with and without contrast

1. A well-defined, oval shape, regular and intact margin, homogeneous soft tissue density, heterogenous moderately-enhanced mass (2-3 cuts, about 2.6 cm x 3.8 cm in dimension) at posterior wall of gastric antrum region
2. No calcification nor hemorrhage present in pre-CM films
3. The fatty planes between this mass and pancreatic head is preserved
4. No perigastric lymphnodes enlargement
5. No definite abnormal enhanced, space-occupying lesions within the liver, spleen, pancreas and both kidneys.



Pre-CM

Post-CM



# Differential Diagnosis - 1

1. Gastrointestinal stromal tumor
2. Carcinoid tumor (sunburst pattern in CT scan)
3. Ectopic pancreas (central orifice)
4. Inflammatory Fibroid Polyp (uncommon submucosal lesion)
5. Gastric carcinoma (irregular, malignant ulcer, LAP, and mucosal lesions are not likely)
6. Pancreatic head tumor or other external compression (extramural lesions are not likely)

# Differential Diagnosis - 2

- ◆ Other submucosal tumors

1. Leiomyoma → 4-5 cm average size
2. Leiomyosarcoma → 12 cm, ulceration, central necrosis
3. Lipoma or Liposarcoma → pedunculated, compressible, pliability, fat density in CT scan
4. Fibromas → dense & intricate bundles
5. Hemangioma → phleboliths
6. Neurogenic tumors



# OP Findings

- ◆ OP method : Antrectomy + Truncal vagotomy
- ◆ Gastric submucosal tumor at antrum with 5 cm in diameter, smooth surface, movable and elastic nature; no ulceration, no pedunculation

# Pathological Findings

- ◆ Grossly : A protruding tumor 4x3x2 cm covered by intact mucosa and located at submucosal layer with focal myxoid change. No necrosis nor hemorrhage is found.
- ◆ Microscopically : A picture of GIST composed mainly spindle cells arranged in interlacing fascicles in the vascular stroma. The tumor is limited to submucosal area.  
All the 27 dissected out lymph nodes and the surgical margins are free from the tumor involvement



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# Discussion

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# Gastrointestinal Stromal Tumors (GISTs)

- ◆ A kind of spindle cell tumor overwhelming majority of benign submucosal gastric neoplasms
- ◆ Related to the muscle-like nerve cells which are called the Interstitial Cells of Cajal, which coordinate the automatic movements of the GI tract

# Definition of GISTs

- ◆ They are defined as c-KIT (CD117, stem cell factor receptor, a tyrosine kinase)-positive mesenchymal spindle cell or epithelioid neoplasms primary in the GI tract, omentum, and mesentery



# Statistics of GISTs

- ◆ 5,000 ~ 10,000 people each year develop GISTs in USA
- ◆ Men and women are equally affected
- ◆ Most often diagnosed in people 50 years of age or older



# Risk Factors

- ◆ With a history of neurofibromatosis (NF1)
- ◆ Familial urticaria pigmentosa (a skin disorder)
- ◆ There are rare instances of familial GISTs

# Anatomy of GISTs

- ◆ GISTs are most common in the stomach (60-70%), followed by small intestine (20-25%), colon and rectum (5%), and esophagus (<5%)
- ◆ Constituting less than 3% of all GI malignant neoplasm. 20% of small-bowel malignant neoplasms; 1-2% of gastric malignancies; and less than 1% of malignancies involving the esophagus, colon, and rectum





# Symptoms of GISTs



- ◆ Abdominal discomfort or pain
- ◆ Vomiting
- ◆ Blood in stools or vomit
- ◆ Fatigue due to anemia

# Imaging Findings – GI series

- ◆ Barium-enhanced images demonstrate predominantly intramural masses with potential exophytic components
- ◆ The tumor margins usually are smooth. En face, the intraluminal surfaces often have well-defined margins
- ◆ Intramural but extramucosal → overlying mucosa can be intact  
In the stomach → preserved area gastric pattern over the tumor mass
- ◆ Overlying mucosal ulcerations are often present → more common in malignant GISTs → bull's eye or target-lesion appearance

# Imaging Findings – CT scan

- ◆ Smaller GISTs appear as smooth, sharply defined intramural masses with homogenous attenuation
- ◆ Contrast enhancement may be rimlike or uniform
- ◆ Larger GISTs with necrosis appear as heterogeneous masses with enhancing borders of variable thickness and irregular central areas of fluid, air, or oral contrast attenuation that reflect necrosis
- ◆ CT is also sensitive for the detection of metastatic liver, peritoneal, lung, and bone lesions

# Imaging Findings - MRI

- ◆ GISTs appear as sharply delineated, heterogeneous masses with cystic and necrotic areas
- ◆ The masses tend to be isointense relative to skeletal muscle on T1-weighted images and hyperintense on T2-weighted images
- ◆ Signal intensity voids are present if gas is present within areas of necrotic tumor

# Imaging Findings - Sonography

- ◆ Endoscopic ultrasonography can be valuable in the evaluation of GISTs
- ◆ The tumors appear as hypoechoic masses that are contiguous with the fourth hypoechoic layer of the GI wall, which corresponds to the muscularis propria
- ◆ Characteristics associated with malignancy include tumor size greater than 4 cm, an irregular extraluminal border, echogenic foci, and cystic spaces

# Prognostic Factors

Factor	Risk of Malignancy	
	Low	High
Most useful		
Tumour size	<5 cm	>5 cm
Mitoses	<5 per 50 HPF	>5 per 50 HPF
Less useful		
Cellularity	Low	High
Necrosis	Absent/minimal	Present
Growth pattern	No infiltration	May invade adjacent tissue
<i>c-kit</i> mutations	Absent	Frequently present

HPF = High-power field.

# Treatment

- ◆ The only treatment for GIST had been surgery
- ◆ The majority of GISTs (~95%) are highly resistant to radiation and systemic therapy
- ◆ Unresectable or malignant GISTs → palliative surgery or chemotherapy has been the only therapeutic option

# Prognosis

- ◆ About 10-30% of GISTs have malignant behavior
- ◆ A benign GIST cannot be conclusively diagnosed
- ◆ GISTs rarely spread to regional lymph nodes (<10%)
- ◆ Distant metastases most commonly involve liver (50-65%) and peritoneum (21-43%); only 10% occur in the lungs or bones



# Mortality & Morbidity

- ◆ Survival rates with malignant GISTs are 69% at 1 year, 38-44% at 3 years, and 29-35% at 5 years.
- ◆ Median disease-specific survival was 60 months with primary disease, 19 months with metastatic disease, and 12 months with local recurrence
- ◆ Recurrence is typical, and the rate has been reported to be as high as 90% at long-term follow-up