

## GIST with Intravascular Large B-cell Lymphoma

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Intravascular large B-cell lymphoma (IVLBCL)는 diffuse large B cell Lymphoma (DLBL)의 매우 드문 형태로 악성 림프구가 혈관 내로 성장하는 것을 말하며, 대부분 치명적인 경우가 많다. 한편 Gastrointestinal stromal tumor (GIST)는 위장관을 침범하는 기질성 또는 간질성 종양이다. 비록 GIST는 위장관을 침범하는 비상피성 양성종양 중에는 가장 흔하다고 알려져 있으나, 간질성 종양은 일차성 소화기계 종양 중에서는 낮은 비율을 보인다.

본 증례는 내원 2주 전 시작된 혈변과 혈뇨를 주소로 내원한 66세 여자에서 IVLBCL과 GIST가 동시에 발생한 경우이다. 10년 전부터 고혈압 진단 받은 환자로 복부 검진 상 특이소견 없었으며, 혈액검사 상 Hb 12.0 g/dL (normal, 12-16 g/dL)으로 정상이었다. 혈뇨를 주소로 비뇨기과 내원하여 시행한 복부 CT상 십이지장 3<sup>rd</sup> portion의 조영 증강 되는 mass 관찰되어 소화기내과 전과되었으며, 내시경 상 GIST에 합당한 소견 관찰되었고 조직검사에서도 부합하는 소견 관찰되었다. PET-CT 상 십이지장 병변의 Hypermetabolism 증가되어 있었으나 조직검사에서는 Low risk potential으로 판단되었다(mitosis count : 3/50 HPF, spindle cell). 점막침범 소견은 관찰되지 않았으나, 점막 하 혈관구조 내에 림프종이 의심되는 비정형 세포가 관찰되었다. 면역조직화학검사 상 혈관구조를 제외한 부분에서 GIST에서 정상적으로 양성소견을 보일 수 있는 C-kit, SMA, D2-40의 양성 소견 보였다. 한편 IVLBCL에서 관찰될 수 있는 CD20이 혈관구조 내에서 관찰되었다. 골수 검사 상 골수침범 소견은 관찰되지 않았고 IVLBCL에 준하여 R-CHOP 3cycles 후 시행한 PET-CT 상 Complete remission 소견 관찰되었다. R-CHOP 6cycles까지 진행 후 시행한 PET-CT에서는 sigmoid colon에서 tumoral uptake 관찰되었고 3개월 후 F/U 하기로 예정된 상태이다.

**66 / Female**

**Chief Complaints**

- Epigastric discomfort for 4 weeks

**Present Illness**

- 5 days before admission, she was examined at a local hospital for the symptom and abdomen CT showed a large mass in the 3<sup>rd</sup> portion of the duodenum.

**66 / Female**

**Past History**

- Hypertension on medication for 10 years

**Family History**

- No specific history

**Systemic Review**

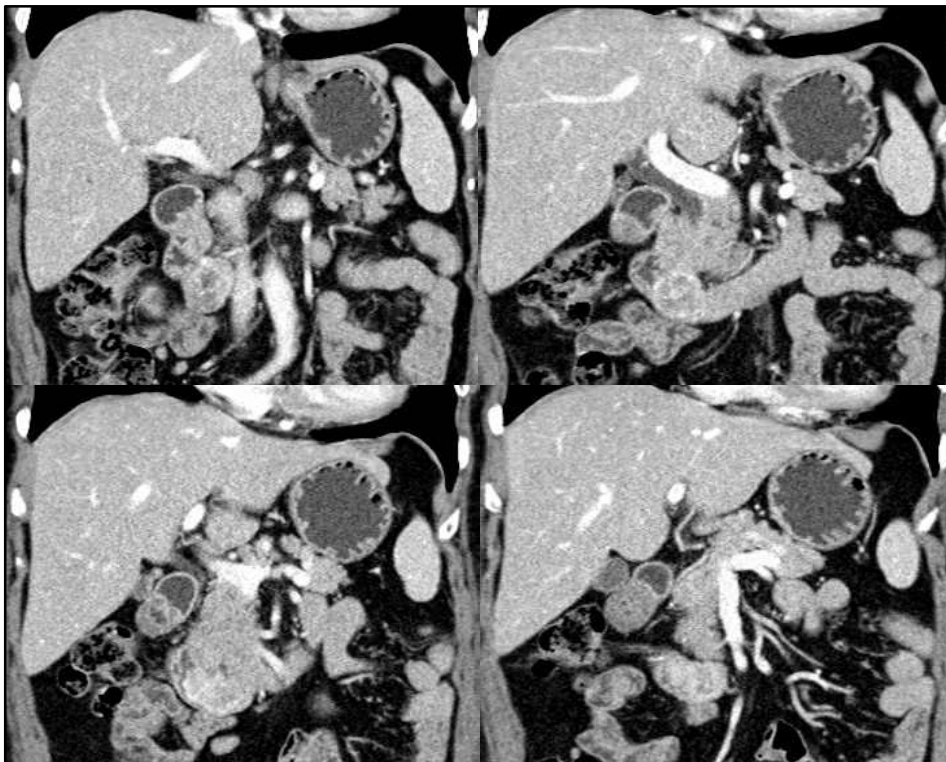
- Nothing to comment

**Physical Examination**

- No abnormal finding

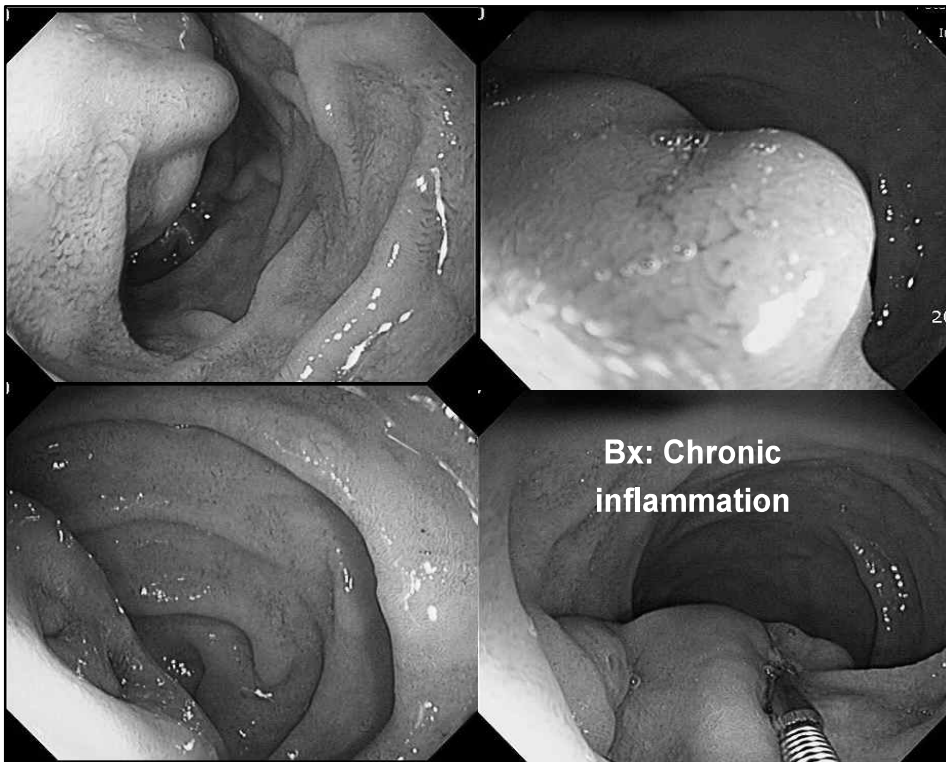
**66 / Female****Laboratory Findings**

WBC	6.29×10 <sup>3</sup> /μL	Protein	6.9 g/dL
ANC	3.46×10 <sup>3</sup> /μL	Albumin	4.3 g/dL
Hemoglobin	12.0 g/dL	Bilirubin	0.4 mg//dL
Platelet	214×10 <sup>3</sup> /μL	AST	20 U/L
CRP	0.39 mg/dL	ALT	16 U/L
<b>Glucose</b>	<b>128 mg/dL</b>	ALP	48 U/L
BUN	9.8 mg/dL	Cholesterol	144 mg/dL
Creatinine	0.6 mg/dL	CEA	0.8 ng/mL
<b>LDH</b>	<b>233 IU/L</b>	CA 19-9	7.1 ng/mL
Calcium	9.4 mg/dL		
Phosphorus	3.0 mg/dL		



**What Kind of Tests would be Needed  
for Diagnosis ?**

Esophagogastroduodenoscopy (EGD)



## What Kind of Tests would be Needed for Diagnosis ?

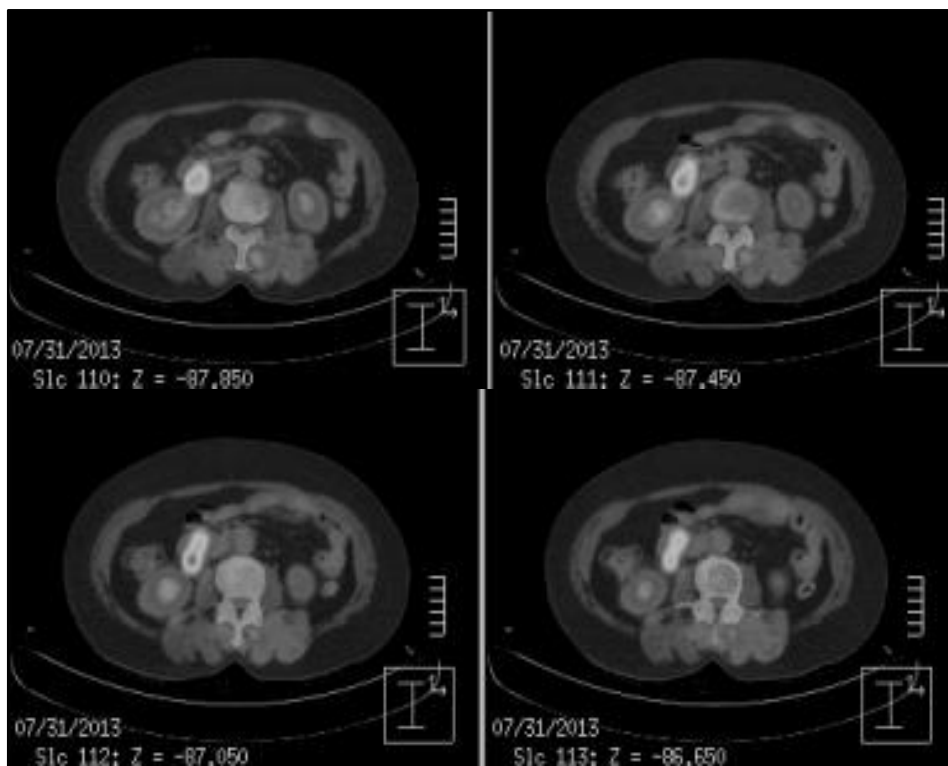
Esophagogastroduodenoscopy (EGD)

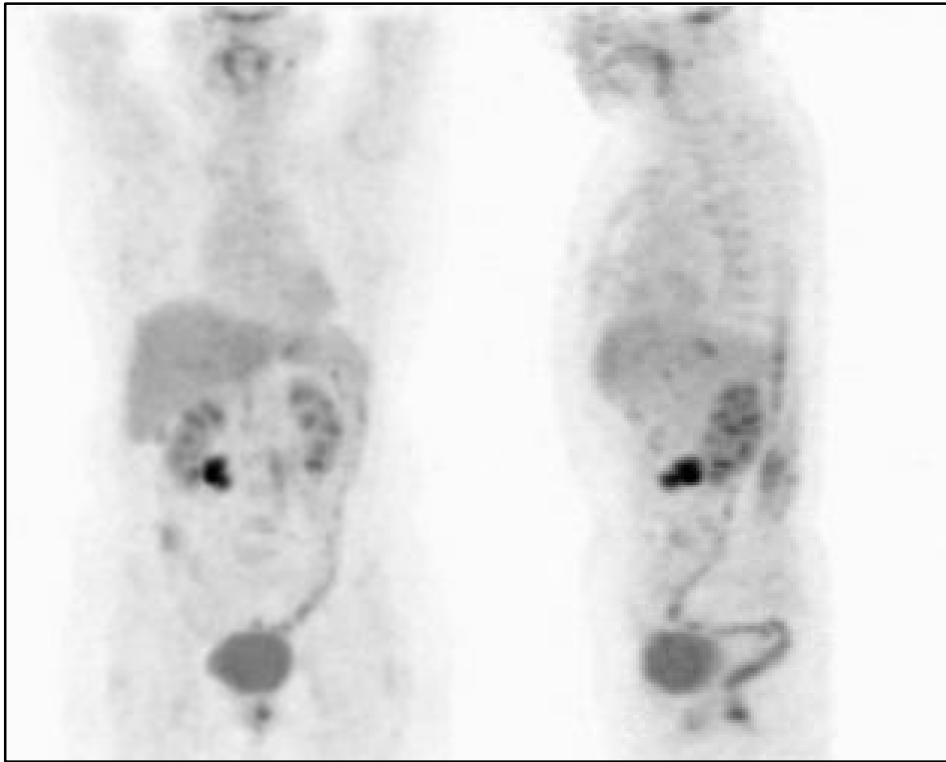
Endoscopic ultrasonography (EUS)

- Radial

- Linear for fine needle aspiration (FNA)

Positron emission tomography (PET)





**What would be the most Probable Diagnosis ?**

Duodenal adenocarcinoma

Neurogenic tumor

Brunner's gland hamartoma

**Gastrointestinal stromal tumor (GIST)**

## What is Your Opinion for Management ?

Regular follow-up

EGD

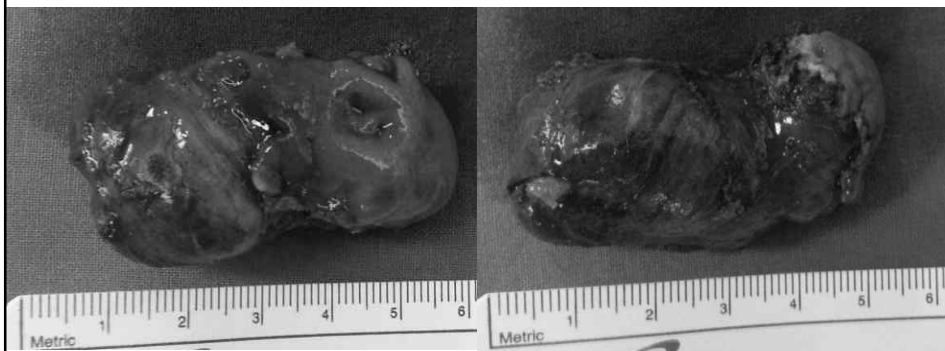
CT

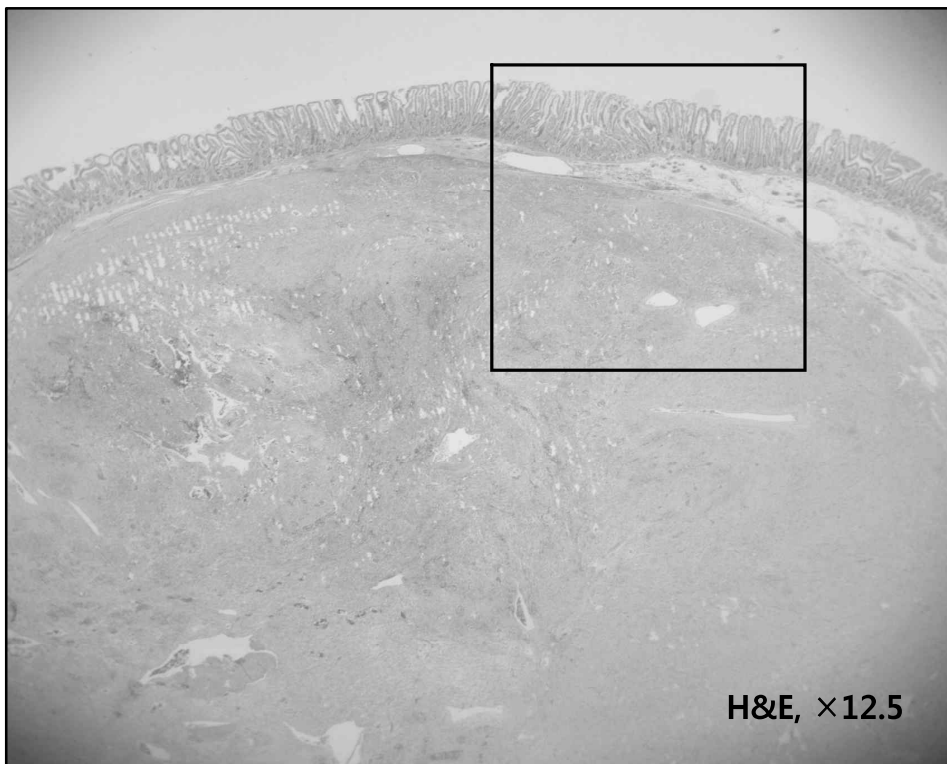
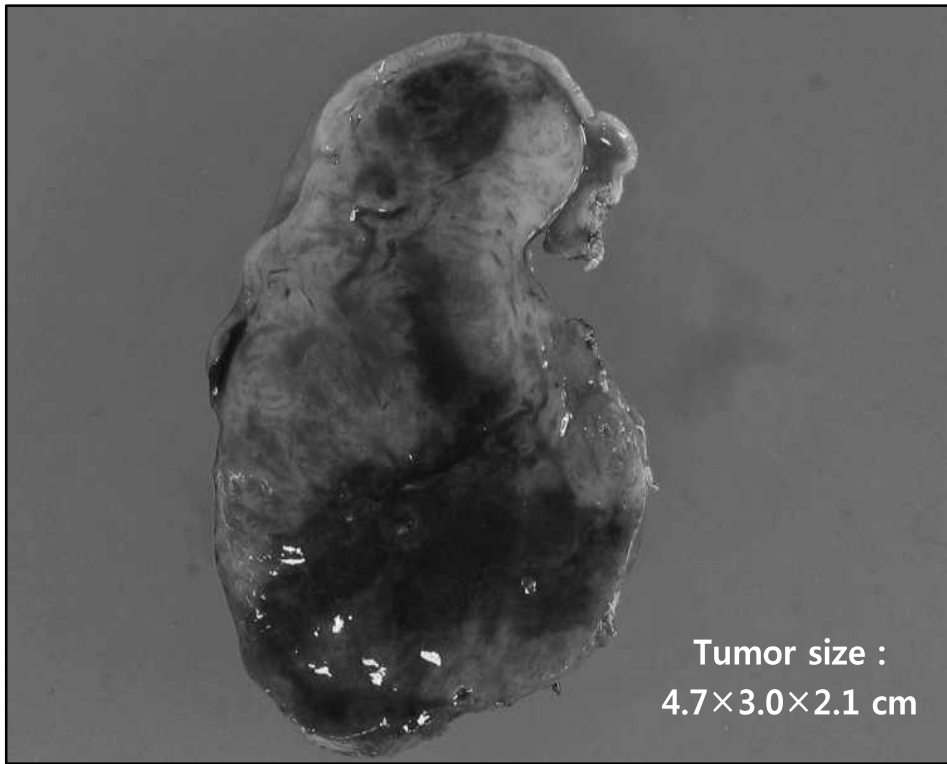
Radial EUS

EUS-FNA

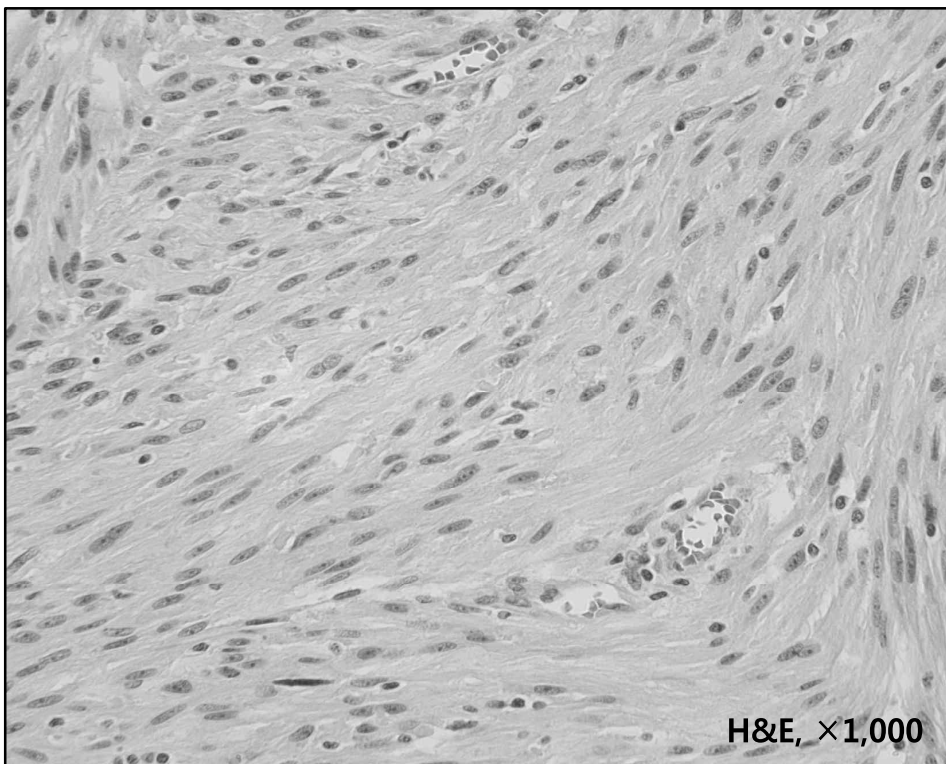
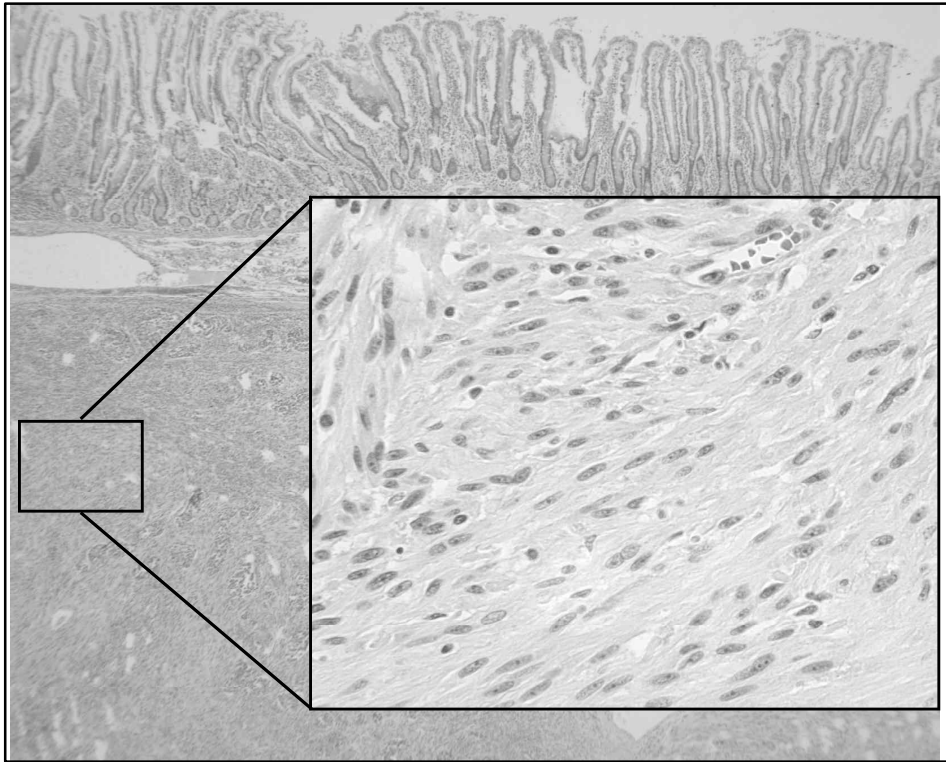
**Surgical resection**

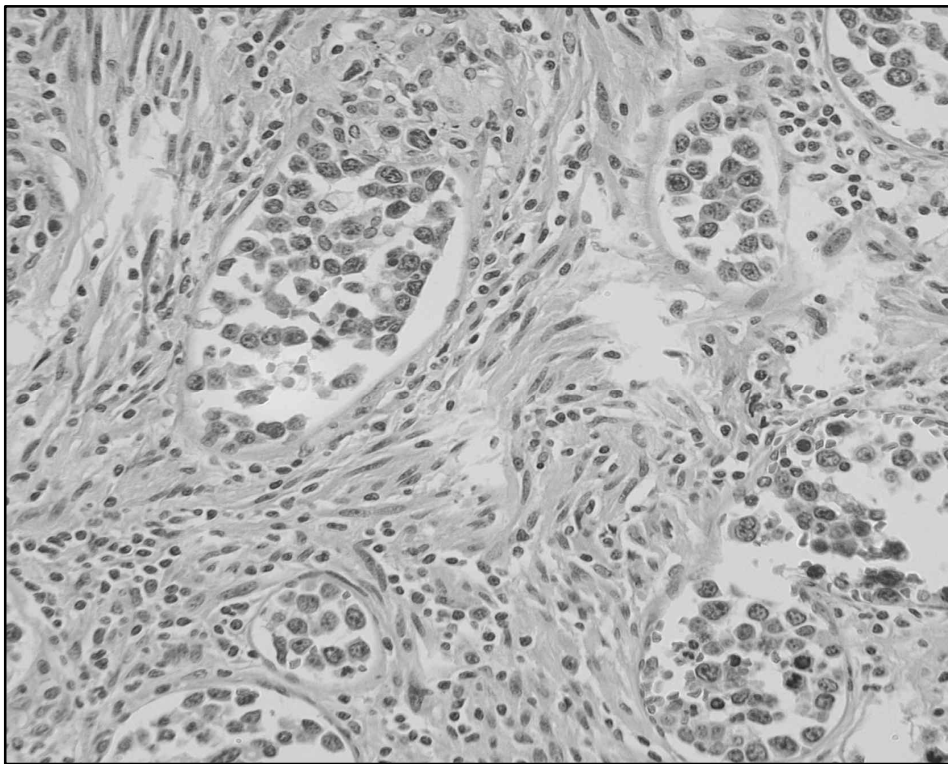
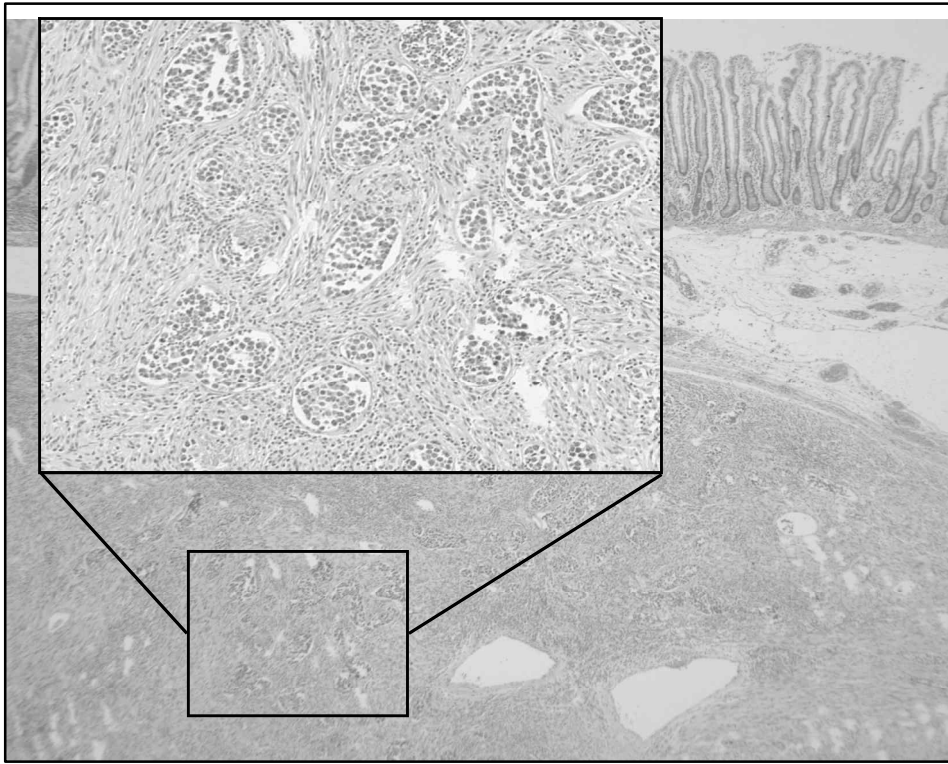
## Duodenal Wedge Resection with G-J Stomy

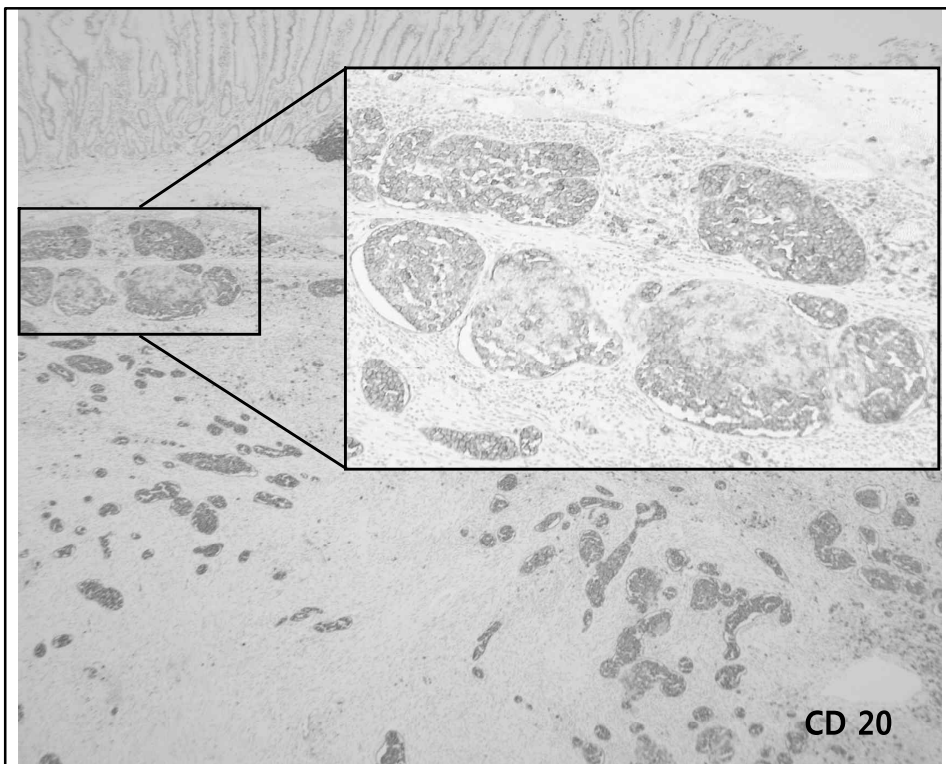
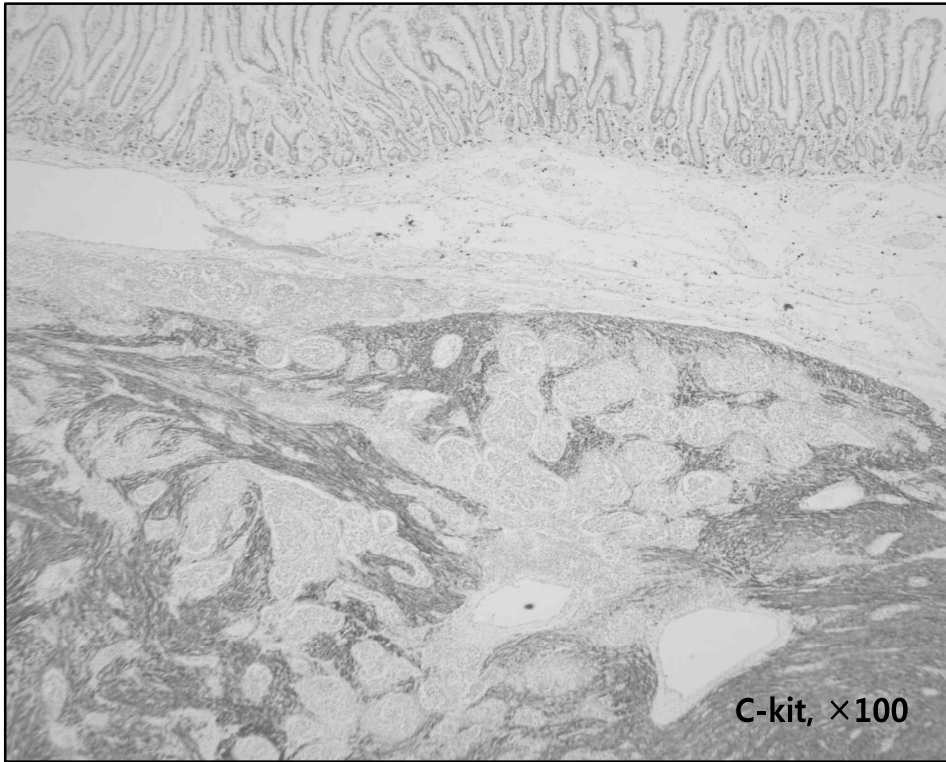


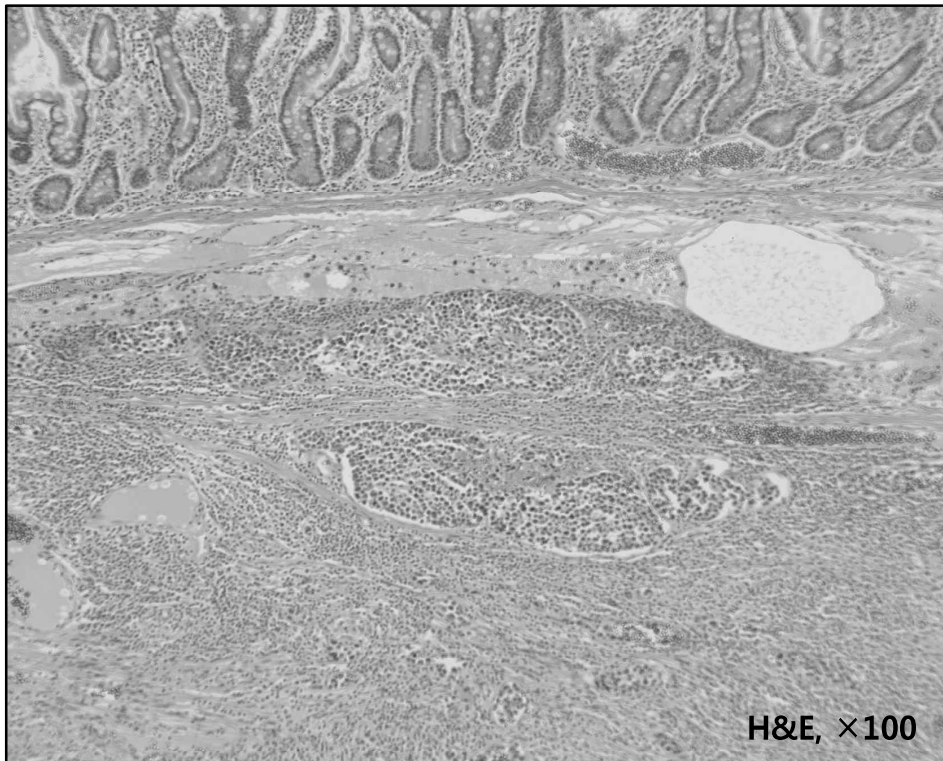












### Final Pathology

1. **GIST** of low risk of malignant potential by NIH (2002) and low risk of progressive disease by Miettinen (2006) :
  - 1) Tumor size : 4.7 x 3 x 2.1 cm
  - 2) Mitosis : 3/50 HPF (high powered fields)
  - 3) Histological type : spindle
  - 4) Necrosis : absent
  - 5) Cellularity : intermediate
  - 6) Cellular atypia : mild to moderate
  - 7) Invasion into mucosa: absent
  - 8) Resection margin (RM) involvement: abutting to RM *combined with*
  
2. **Intravascular large B-cell lymphoma**

## Post-operative Course

- Bone marrow: no involvement
- 6 cycles of CHOP chemotherapy
- In *complete remission*

## IVLBCL (Intravascular Large B Cell Lymphoma)

: Rare subtype of large cell lymphoma with proliferation of **lymphoma cells within the lumina of small blood vessels without an obvious extravascular tumor mass**

- **Extremely unusual** for the **GI system** to be the primary diagnostic site
- **Median age at diagnosis:** sixth to seventh decades
- **Symptoms:** constitutional B symptoms (fever, night sweat, weight loss)
  - ✓ Western countries: CNS (39%), skin (39%) > BM, spleen, liver
  - ✓ Asian countries: BM (75%), spleen (67%), liver (55%) > CNS, skin

## Disease Review

- **Immunohistochemical features:** CD20 (96%), MUM1/IRF4 (95%), Bcl-2 (91%), CD19 (85%), immunoglobulin k light chain (71%), CD5 (38%), Bcl-6 (26%)
- **Diagnosis:** biopsy of involvement area (skin, lung, brain, liver, BM), CD20
- **Treatment**
  - Systemic therapy: (R-)CHOP
  - CNS directed therapy (30-40% of ILBL)