# SPC

Presenter: PGY 甘育安

Supervisor: VS 楊明勳

# GENERAL DATA

- Name: OOO
- Chart no.: OOOOOOO
- Gender: male
- Birth date: OOOO/OO/OO (60 years old)
- Date / time of ER arrival: 2012/10/11, 23:42
- Time of admission: 2012/10/12
- Discharge date: 2012/10/17
- Information source: patient

# CHIEF COMPLAINT

- Epigastric pain and shifted to right lower quadrant abdomen since last evening

# PRESENT ILLNESS

- 60 y/o male, denied any systemic diseases
- Epigastric pain and shifted to right lower quadrant abdomen since last evening
- Denied fever, chillness, shortness of breath, nausea, vomiting, diarrhea or constipation
- Visited our emergency room

# PAST / PERSONAL HISTORY

**Medical history**

- Nil

**Travel history**

- Nil

**Personal history**

- Alcohol: occasional drinking
- Smoke: 2 packs / day for many years
- No known allergy

**Current medication**

- Nil

**Surgical history**

- Nil

**Family history**

- No contribution

# PHYSICAL EXAMINATION

- Height/weight:
  - 160cm/65kg
- Vital signs:
  - T/P/R: 37.0/78/16; BP: 124/78 mmHg
- General appearance:
  - Clear consciousness; acute ill-looking
- Skin:
  - Normal skin turgor, no pigmentations
- HEENT and neck:
  - Eye: anicteric sclera, pink conjunctiva
  - Supple neck; no palpable lymph nodes; no JVE

# PHYSICAL EXAMINATION

- Chest: clear breathing sound, symmetric expansion
- Heart: regular heart beat without murmur
- Abdomen:
  - Mild distended; palpable mass (-); RLQ tenderness (+)
  - Murphy's sign (-); rebound pain (-); muscle guarding (-)
  - McBurney point tenderness (+); hypoactive bowel sound
- Back:
  - No limited ROM; CV angle knocking pain (-)
- Extremities:
  - Freely movable; no pitting edema
  - Symmetric deep tendon reflex and full muscle power

# LAB DATA

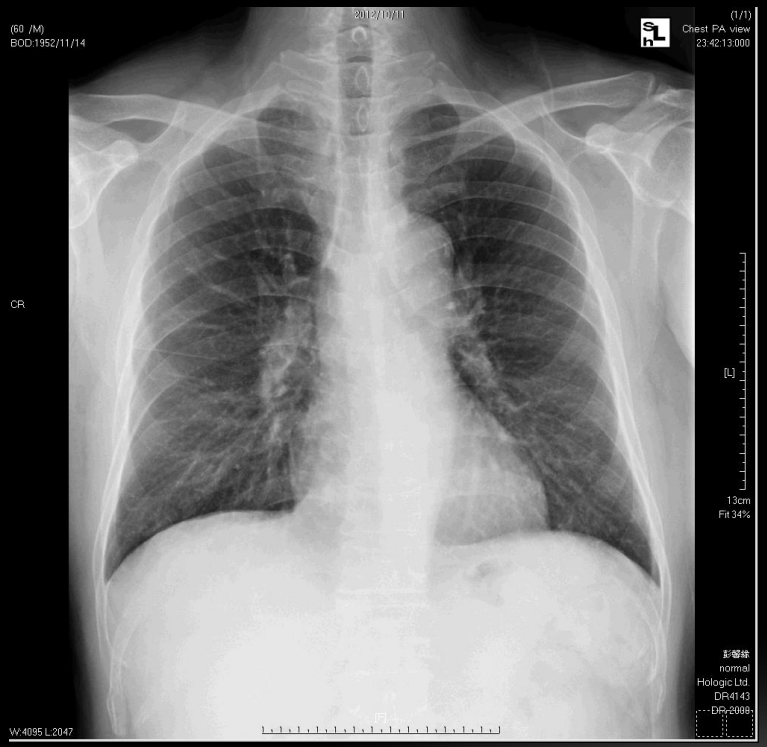
| Blood | |
|---|---|
| RBC | 5,270,000 /uL |
| HGB | 16.9 g/dL |
| HCT | 47.8 % |
| MCV | 90.7 fL |
| MCH | 32.1 pg |
| WBC | 13,000 /uL |
| PLT | 218,000 /uL |
| %Neut | 83.9 % |
| %Lym | 12.1 % |
| PT (INR) | 10.1 sec (0.94) |
| APTT | 28.0 |

| SMA | |
|---|---|
| Glucose AC | 261 mg/dl |
| Creatinine | 0.69 mg/dl |
| ALT | 20 IU/L |

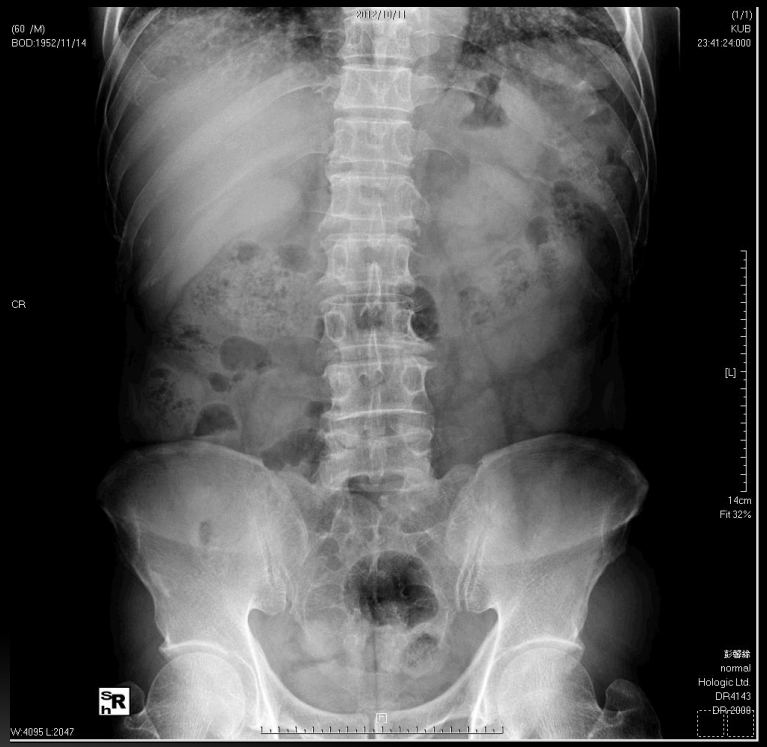
| Urine | | | |
|---|---|---|---|
| Sp. Gr. | 1.031 | Nitrite | - |
| Color | Yellow | OB | - |
| Glucose | 4+ | RBC | 0-2 |
| Bil | - | WBC | 0-2 |
| Prot. | 1+ | Epi. cell | - |
| Ketone | 1+ | Bact. | 0-18 |

# IMAGES

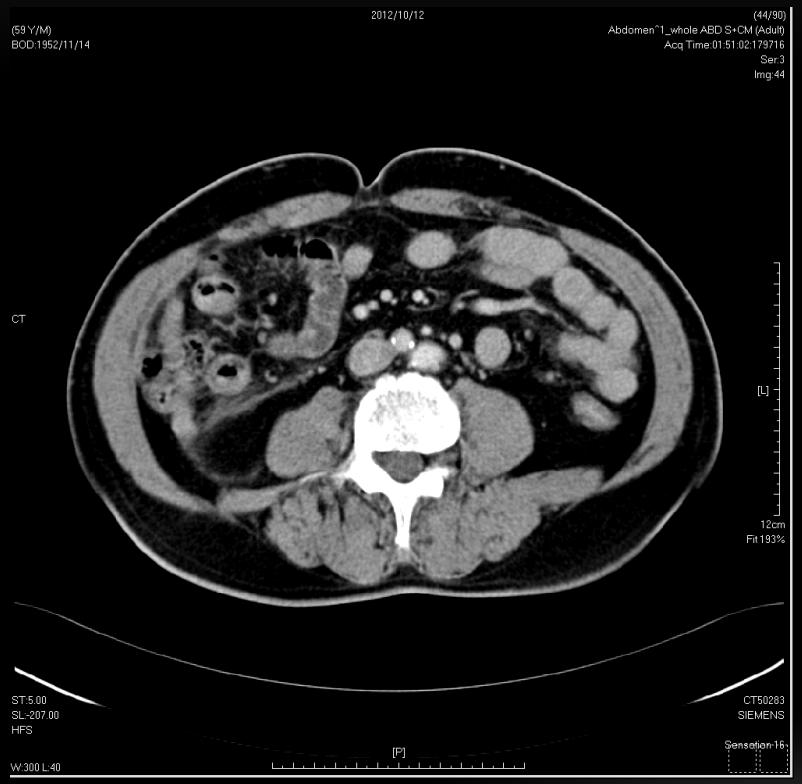
## CXR (2012/10/11)

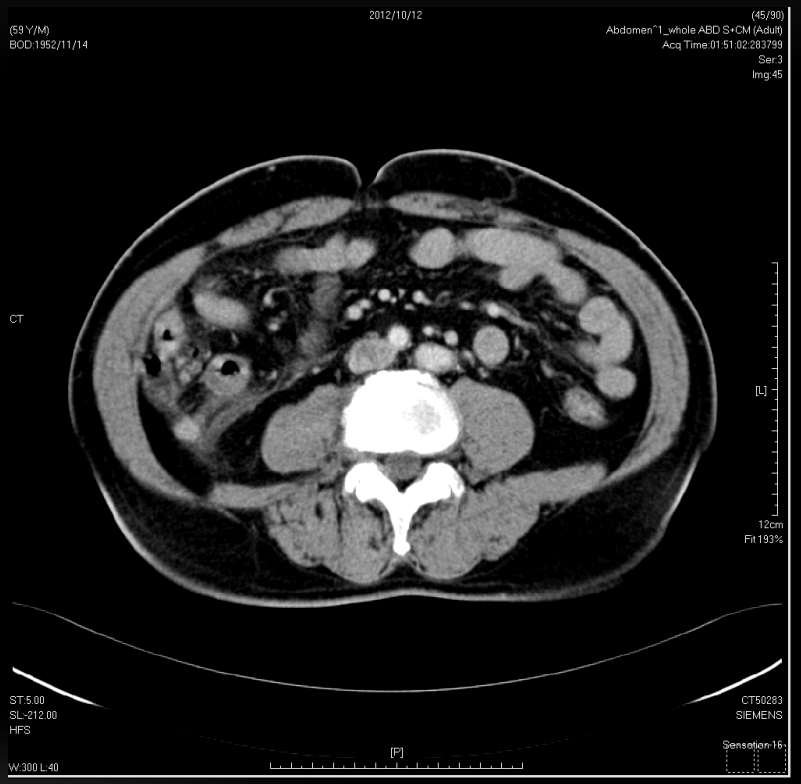



## KUB (2012/10/11)



# IMAGES – ABDOMINAL CT



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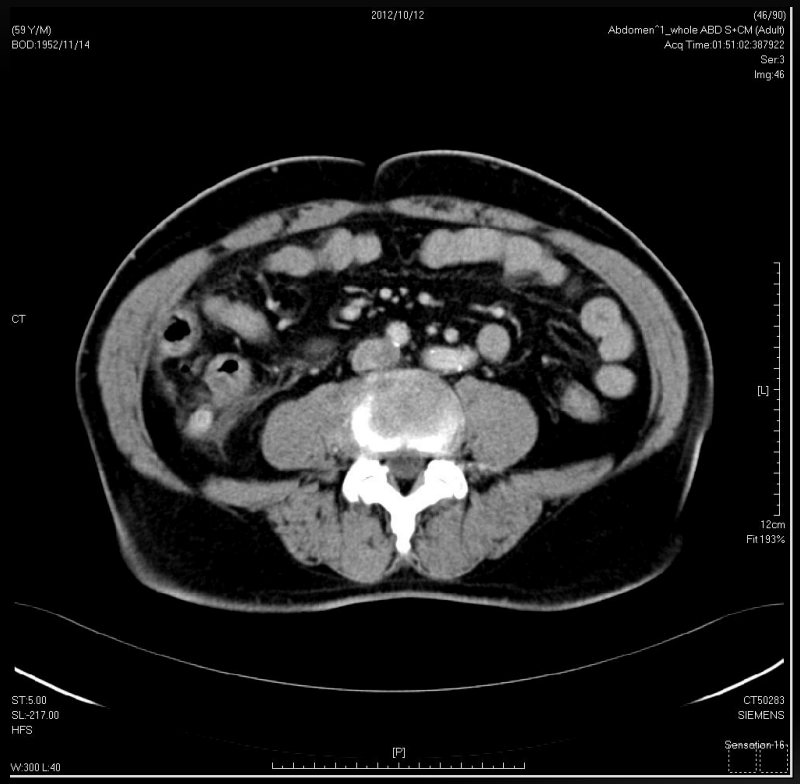


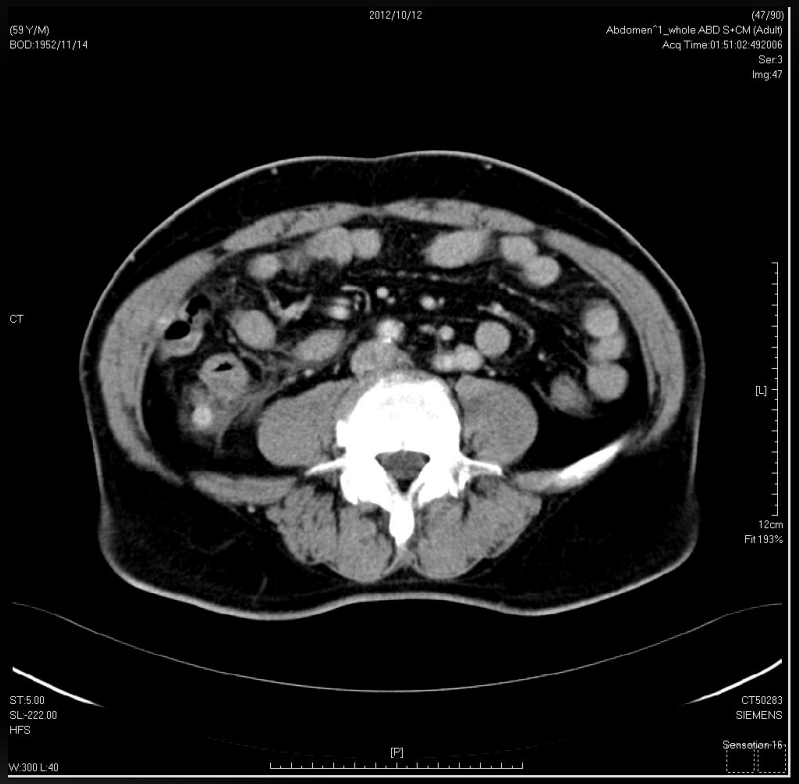

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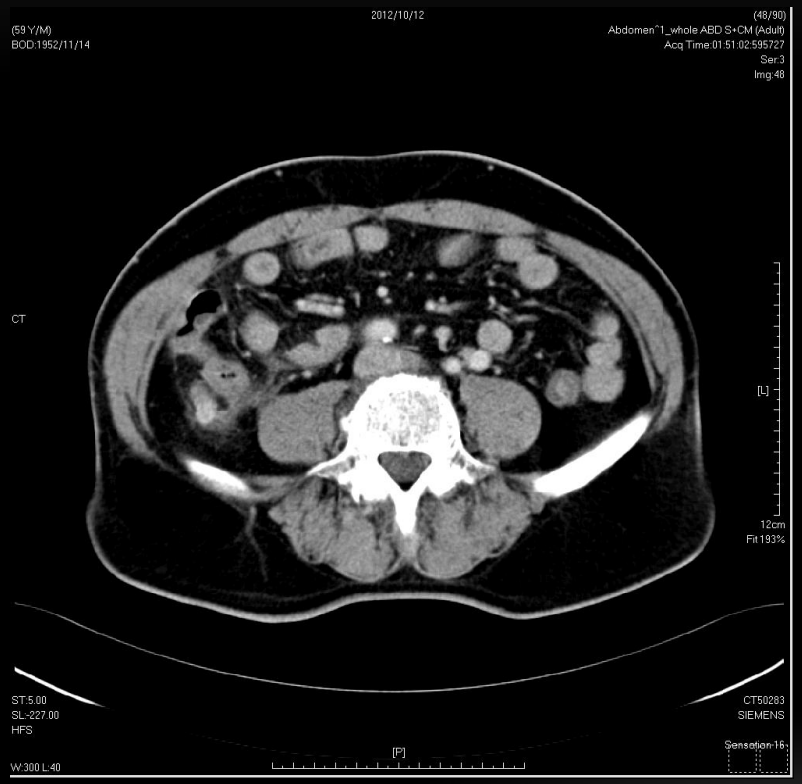


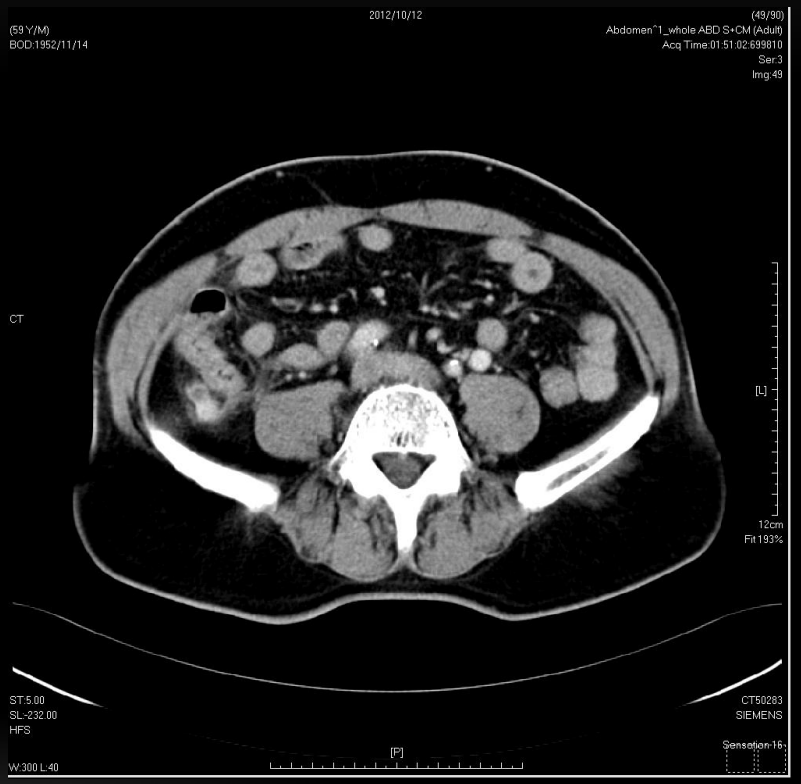

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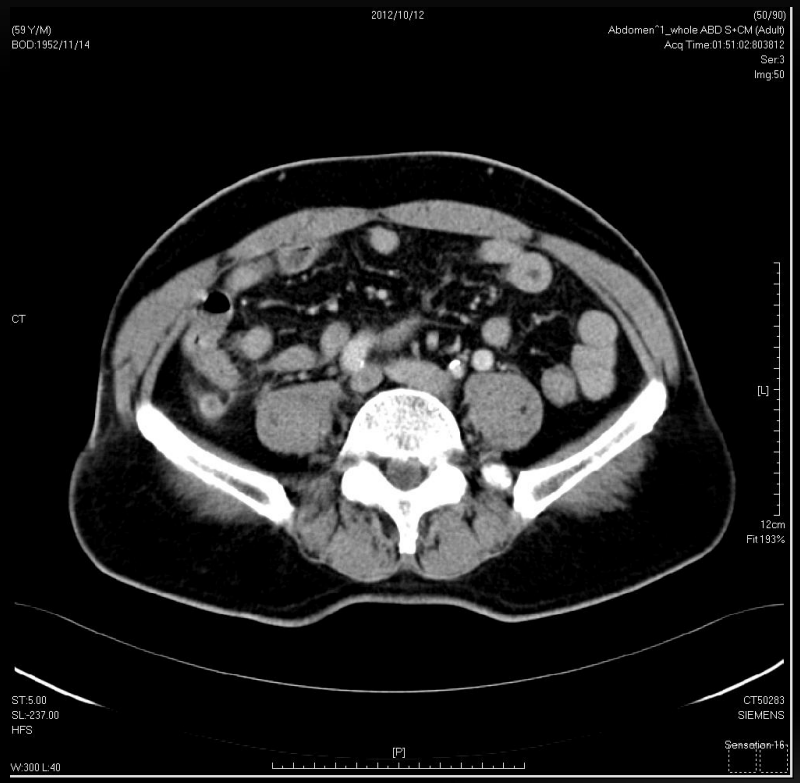


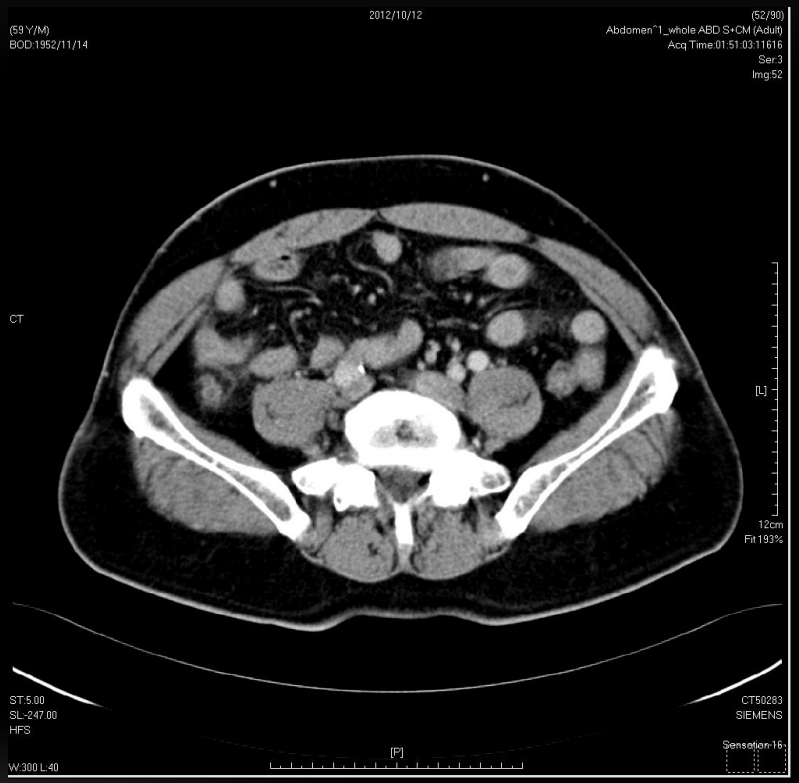

# TENTATIVE DIAGNOSIS

- Acute appendicitis
- Hyperglycemia

# PLAN

- Emperic antibiotics:
  - Cefazolin 1g Q6H IVD
  - Gentamicin 80mg Q12H IVD
  - Metronidazole 500mg Q8H IVD
- Pre-OP evaluations
- Arrange laparoscopic appendectomy

# OPERATION

- Pre-OP diagnosis: acute appendicitis
- Post-OP diagnosis: acute appendicitis
- Procedure: laparoscopic appendectomy
- OP findings:
  - Appendix:
    - No perforation; congested; length in 7 cm
    - Location: pelvic
  - Ascites: clear

# FINAL DIAGNOSIS

- Goblet cell carcinoid of appendix, pT1N0M0, status post laparoscopic appendectomy
- Type 2 diabetes mellitus

# DISCUSSION – GOBLET CELL CARCINOIDS OF THE APPENDIX

*Review Article*

# Goblet Cell Carcinoids of the Appendix

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# Goblet cell carcinoid of appendix: A rare case with literature review

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# INTRODUCTION

- Goblet cell carcinoids (GCCs):
  - An uncommon neoplasm of the vermiform appendix
  - A separate entity from adenocarcinoma and carcinoid tumors
  - Mixed tumors with partly neuroendocrine differentiation and partly goblet cell type morphology
  - Different names:
    - Adenocarcinoids, goblet cell tumors, mucinous adenocarcinoids and crypt cell carcinoma

# INTRODUCTION

- GCC almost exclusively occur in the appendix but may occasionally be found in other parts of the gastrointestinal tract
- Extra-appendiceal locations of GCC and found that true primary extra-appendiceal GCC is extremely rare

# EPIDEMIOLOGY

- Occur in 0.3%–0.9% of appendectomies
- Extremely rare (Incidence in 1973-2001: 0.05/100,000 per year )
- McCusker et al., who made a population-based study from the SEER database, 1973–1998:
  - Most often seen in:
    - Patients in their fifties or sixties
    - Caucasian population

"Primary malignant neoplasms of the appendix: a population-based study from the surveillance, epidemiology and end-results program, 1973–1998," Cancer, vol. 94, 2002.

# EPIDEMIOLOGY

- Jiang et al.
  - A possible connection between GCC and schistosomiasis
  - Appendiceal schistosomiasis
    -> Increased proliferation and neuroendocrine differentiation of mucosal pluripotent crypt cells
    -> Development of GCC

Y. Jiang et al. “Schistosomiasis may contribute to goblet cell carcinoid of the appendix,” Journal of Parasitology, vol. 98, 2012.

# CLINICAL PRESENTATION

- Up to 60% of the patients present with signs and symptoms of acute appendicitis due to luminal obstruction
- The tumor cells **proliferate sparsely**
  - Do not form nodules
  - Diffuse thickening, fibrous proliferation, and contraction of the appendiceal lumen

# CLINICAL PRESENTATION

- Other manifestations:
  - Asymptomatic patients
  - Intussusception
  - A palpable mass
  - Gastrointestinal bleeding
  - Chronic intermittent lower abdominal pain
  - Secondary genitourinary complications

# CLINICAL PRESENTATION

- In cases with disseminated disease:
  - Abdominal pain associated with an abdominal mass and weight loss
  - More prevalent in women
  - Often involved ovaries and peritoneum
  - Compared to metastases of intestinal carcinoid tumors and adenocarcinomas, metastases to the lungs and liver are rare

# DIAGNOSIS

- Histology:
  - **Scattered positivity** for chromogranin A and synaptophysin
    - Classic appendix carcinoids: **homogeneous** staining for both chromogranin A and synaptophysin
  - Positivity for CK20 and CEA
  - Ki-67: proliferation index
    - Jiang et al.: a mean Ki-67 index with 5 ± 3% with significantly higher levels than typical appendix carcinoid tumors

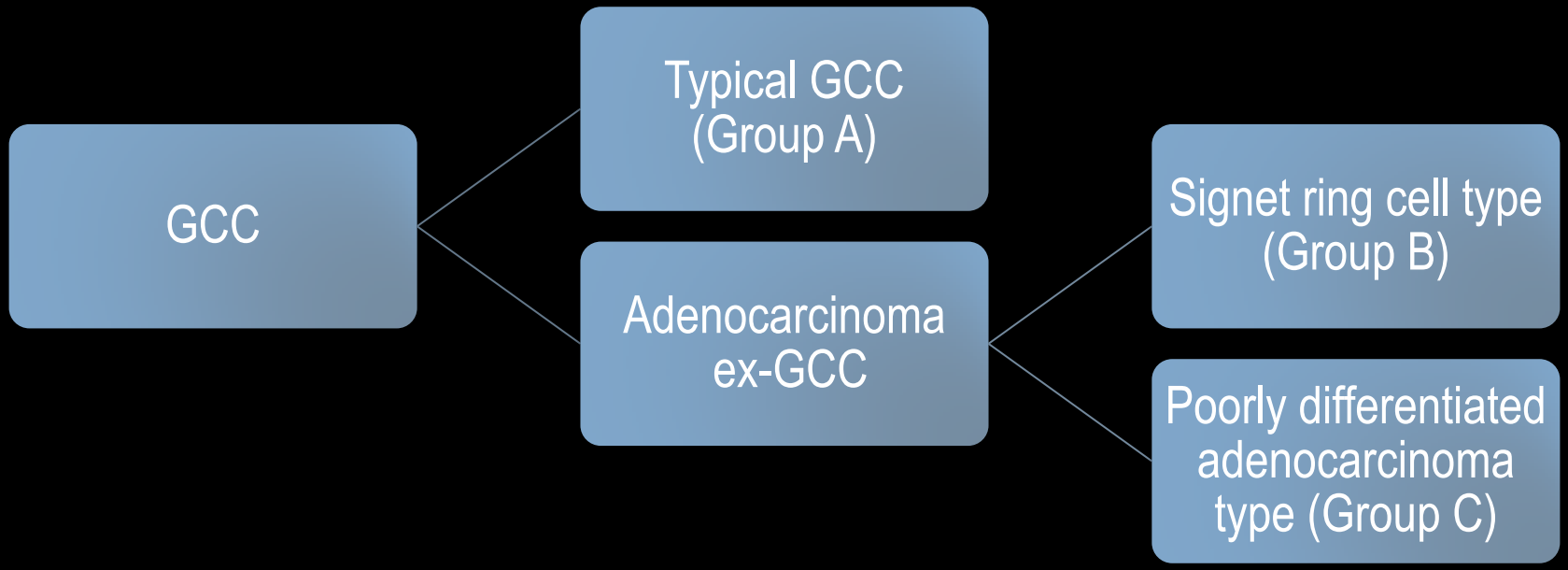
# DIAGNOSIS

- Biochemistry:
  - Plasma chromogranin A (CgA):
    - Usually negative
    - No specific neuroendocrine markers have been observed
  - Urinary 5-hydroxyindoleacetic acid (5-HIAA) level:
    - Mostly within normal limits
  - CEA, CA-19-9, and CA-125:
    - May be elevated in patients with disseminated disease

# DIAGNOSIS

- Imaging:
  - No specific diagnostic studies of imaging focused on GCC
  - Somatostatin receptor scintigraphy (SRI) or Gallium-DOTANOC-PET scans:
    - Usually not useful; the presence of somatostatin receptors on goblet cells in general is sparse or lacking
  - FDG-PET:
    - May be useful in patients with high ki67 index
  - CT / MRI:
    - Low sensitivity for local spread of the disease
    - To rule out metastasis to the lymph nodes and liver

# GRADING / STAGING




L. H. Tang et al., "Pathologic classification and clinical behavior of the spectrum of goblet cell carcinoid tumors of the appendix," American Journal of Surgical Pathology, vol. 32, 2008.

# GRADING / STAGING

- TNM 7th edition staging (appendix carcinoma):

T1 Submucosa

T2 Muscularis propria

T3 Subserosa, non-peritonealize periappendiceal tissues

T4a Perforates visceral peritoneum/Mucinous peritoneal tumour within right lower quadrant

T4b Other organs or structures

N1 ≤ 3 regional

N2 > 3 regional

M1a Intraperitoneal metastasis beyond right lower quadrant

M1b Non-peritoneal metastasis

Like colon, based on depth; includes goblet cell carcinoid

# GRADING / STAGING

| | | |
|---|---|---|
| Stage 0 | Tis | N0 |
| Stage I | T1, T2 | N0 |
| Stage II | T3, T4 | N0 |
| Stage IIA | T3 | N0 |
| Stage IIB | T4a | N0 |
| Stage IIC | T4b | N0 |

Basic categories unchanged

Subdivisions expanded

Changes from TNM 6

| | | | |
|---|---|---|---|
| Stage III | Any T | N1-2 | |
| Stage IIIA | T1, T2 | N1 | |
| | T1 | N2a | |
| Stage IIIB | T3, T4a | N1 | |
| | T2-T3 | N2a | |
| | T1-T2 | N2b | |
| Stage IIIC | T4a | N2a | |
| | T3-T4a | N2b | |
| | T4b | N1-2 | |
| Stage IV | Any T | Any N | M1 |
| Stage IVA | Any T | Any N | M1a |
| Stage IVB | Any T | Any N | M1b |

# TREATMENT

- Localized stage I tumors:
  - Appendectomy alone
- In higher stages:
  - Right hemicolectomy
    - For nodal sampling (increased risk for local lymph node metastases)
  - A prophylactic removal of the ovaries:
    - High incidence of metastases to the ovaries
- With peritoneal carcinomatosis:
  - Cytoreductive surgery with intraperitoneal chemotherapy (HIPEC)

# TREATMENT

- Post-OP follow up:
  - Imaging: CT or MRI is recommended
  - Biochemistry:
    - CgA determination is not recommended.
    - CEA, CA-125, and CA-19-9 are suggested as tumor markers

# TREATMENT

- When disseminated at time of diagnosis:
  - Debulking surgery is recommended when possible followed by adjuvant chemotherapy with regimens similar to colorectal adenocarcinoma
  - There are case reports of regimens using streptozotocin and 5FU or platin-based therapies in combination with etoposide
  - More aggressive combinations: FOLFOX/FOLFIRI

# PROGNOSIS

- McCusker et al. (227 patients):
  - GCCs are associated with an 80% 5-year survival rate and 65% 10-year survival rate
  - Overall biologic behavior of GCC in their series was intermediate between that of adenocarcinomas and carcinoid tumors:
    - Age at diagnosis
    - Extent of disease spread at diagnosis
    - Number of cases with lymph node involvement
  - Most of the GCC are still localized at time of diagnosis

# PROGNOSIS

- Pham et al. (57 patients):
  - A 45% 5-year survival rate
  - Found a tendency for GCC to occur more frequently in women and simultaneously that half of the female patients had metastasis to the ovaries at time of initial presentation

# PROGNOSIS

- Tang et al. (63 patients):
  - Separated the tumors into 3 groups according to histology
  - A 5-year survival rate that decreased from 100% to 0%, from group A to group C
  - Low Ki67 index + Early stage
    - Similar to the classic appendiceal neuroendocrine tumors
  - High Ki67 index + disseminated disease
    - Similar to gastrointestinal adenocarcinomas

# THANK YOU!