Case Presentation

中國醫藥大學附設醫院　兒童醫學中心
R3 林佐臨／ VS 吳淑芬 ／VS陳安琪
2013/ 12/ 07

General Data

- ID no.: 13498955
- Name: 徐O傑
- Gender: male
- Age: 13 years
- Admission Date: 2012/ 9/ 23

Chief Complaint

- Abdominal pain for 1 week

Underlying Disease

- Smith-Lemli-Opitz syndrome
- Mental retardation
- Type I DM since 7 years old
  - Control with SC Novorapid/NPH 20~25U/day (~0.5~0.6 U/kg/day)
  - Usual sugar AC: 200~300 mg/dL
  - HbA1C: 9.1% (2011/12) => 9.0% (2012/4) => 7.9% (2012/7)

Present Illness

- Severe epigastric pain on and off for 1 week
- Post-pradinal vomiting 4 times a day
- No stool passage in recent 2 days
- Tarry stool for 3 months

Physical Examination

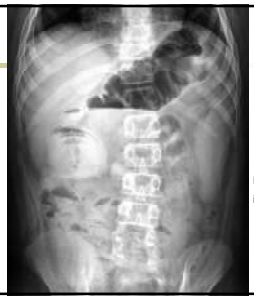
- General appearance: **chronically ill, weak, pale-looking**
- Height: 133cm (<3th percentile)
- Weight: 38Kg (15-50th percentile)
- Conscious level: alert, responsive
- Vital signs：BT36.5℃,PR:**138/min**, RR:24/min
- BP: 123 / 72 mmHg
- HEENT: **sunken eye**, conjunctiva: **pale**
- Neck: supple, no palpable mass

Physical Examination

- Chest: symmetrical expansion, clear breathing sound
- Heart: regular heart beat, no murmur
- Abdomen:
  - Auscultation: normoactive bowel sound
  - Percussion: tympanic
  - Palpation: distended, **severe epigastric tenderness(+), rebounding pain (+/-)**, no muscle guarding, no hepatomegaly, no splenomegaly
- Extremities: warm, freely movable, no deformity, no pitting edema
- Skin: **acanthosis nigra over the whole body, dry skin turgor**

Lab Data (in admission)

- WBC= 6130, DC: N/L/M= 78.8%/ 16.0%/ 4.7%
- **RBC= 2.53 * x10^6/ul, Hb= 5.1, HCT= 18.7%**
- **RDW= 18.1%, MCV= 73.9 fl, MCH= 20.2 pg, MCHC= 27.3 g/dL** (microcytic hypochromic anemia)
- Reticulocyte= 1.81%
- **Blood smear: RBC hypochromia 3+**
- Platelet= 229K
- **Glucose= 211**
- Other biochemistry data normal



KUB
in admission

Initial Treatment

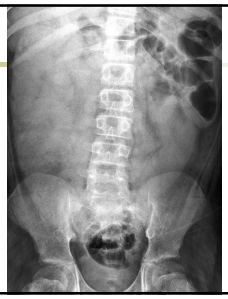
- PRBC 1U transfusion
- IV Ranitidine
- IV Metoclopramide
- Glycerin enema

Hospital Course

- Upper GI panendoscope:
  - Esophagus: a esophageal ulcer with blood clot about 0.3cm
  - Stomach: several hemorrhagic spots with erosion over fundus, body to antral area
  - H1 gastric ulcer,sized about 0.3 cm, over high body area
- Discharged 3 days later since symptoms improved

Hospital Course

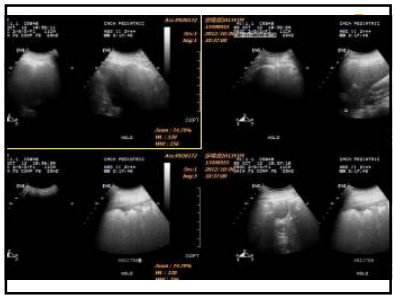
- Admitted again 1 week later
- Vomiting several times, no coffee ground vomius
- Diffused abdominal pain
- No stool passage for 3-4 ；days

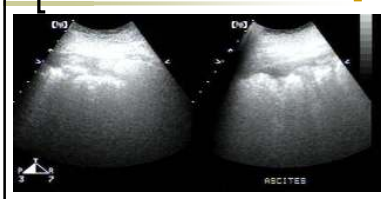


KUB
In 2nd admission

Hospital Course

- Still no stool passage after 4 days of conservative treatment
- Severe bowel distention
- Abdominal echo: ascites (+)



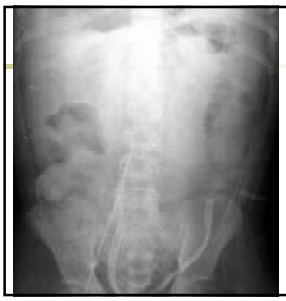


Hospital Course

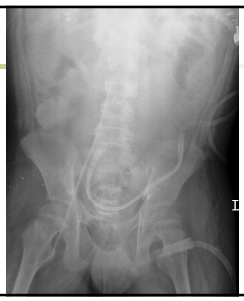
- Consult pediatric surgeon and laparotomy was done
- Severe distended small bowel from the ligament of Treitz to the ileocecal valve
- Much fecoloma retention in the A-T colon
- The D-S colon was collpase
- The appendix was congestive
- No true obstruction site was identified

Operation

- Tried to remove the fecoloma on table but failed
- Decompressed the bowel retrogradely but failed due to thick material in the stomach.
- Opened the terminal ileum and made an ileostomy
- Appendectomy



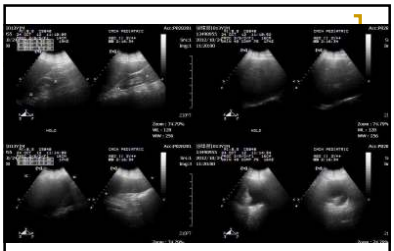
Post OP



Post OP day 3

Hospital Course

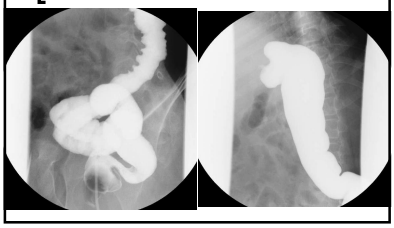
- Transferred to PICU after operation
- Wound pus formation after OP => MRSA => Vancomycin => Teicoplanin
- Transfer to ward after infection has controlled
- Still poor feeding (could not tolerate 300cc/day and vomiting) after almost 2 months after OP => arrange lower series

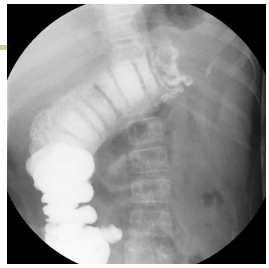


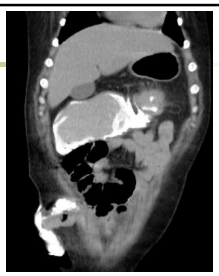
Abdominal echo, 2 weeks after OP

Lower GI Series + Abdominal CT

- Contrast medium was inflated from anus, and **it could not pass through T-colon**
- Contrast medium was inflated though the ileostomy, **and obstruction at T-colon is noted**
- CT findings:
  - Bowel wall thickening with narrowing lumen of T-colon
  - Enlarged paraaortic and mesentery lymph nodes







Hospital Course

- Consult pediatric surgeon again
- Extended right hemicolectomy + stoma takedown + ileocolostomy + enterolysis

OP finding

- Severe peritoneal adhesion with bowel loops
- Stony harddish yellowish tumor mass about 8 x 7 cm in size located at T-colon near splenic flexure
- Multiple mesenteric root LNs enlargement, including the mesoileum
- Luminal narrowing by external compression of tumor
- Diffuse scattering mucosal ulceration and blackish spots in the proximal side of obstruction.
- The mucosa of the distal colon and terminal ileum were not involved

Pathology

- Large intestine, transverse colon, extended right hemicolectomy, **moderately differentiated intestinal type adenocarcinoma** (tumor size about 4.0-cm in maximal diameter) invading through pericolic soft tissue and penetrating to the serosal surface, revealing moderate stromal desmoplasia with evident fibroplasia, moderate mixed acute and chronic inflammatory tumor-host reaction, presence of focal lymphovascular permeation, but no definite perineural invasion identified.
- Serosa, outmost pericolic soft tissue, ascending colon, extended right hemicolectomy, penetrated through by moderately differentiated intestinal type adenocarcinoma of transverse colon (section A5).

Pathology

- Surgical margin, proximal and distal, ileum and colon, extended right hemicolectomy, free of tumor invasion or glandular dysplasia.
- Large intestine, ascending colon, proximal from main tumor, extended right hemicolectomy, two tubular adenomas with low- to focally high-grade glandular dysplasia, evident intramucosal hemorrhage, and free of invasive malignancy.
- Large intestine, ascending colon, non-tumor part, extended right hemicolectomy, mucosal and submucosal congestion with multifocal hemorrhage and frequent small lymphocytic aggregation in lamina propria, overlying mild mucosal erosion, submucosal edema, and free of tumor invasion or glandular dysplasia.

Pathology

- Small intestine, ileum, extended right hemicolectomy, one ileostoma, revealing submucosal and peri-ileal fibrosis, vascular congestion, hemorrhage, focal inflamed granulation tissue formation, and free of tumor involvement or glandular dysplasia.
- Lymph node, regional, lymphadenectomy, metastatic moderately differentiated intestinal type adenocarcinoma (1/20, intranodal lesion less than 0.1 mm in diameter) without extranodal extension.
- Omentum, omentectomy, no specific pathologic change, and free of tumor
- Lymph node, mesenteric root and mesoileum, lymphadenectomy, metastatic moderately differentiated intestinal type adenocarcinoma (1/2, intranodal lesion about 0.7 cm in diameter) with evident extranodal soft tissue extension.

Pathology

- Immunohistochemistry study for primary tumor cells: CK7 (negative), CK20 (positive, diffuse strongly membranous staining), CDX2 (positive, diffuse moderately nuclear stain), c/w the immunoprofiles of **moderately differentiated intestinal type adenocarcinoma of colonic primary**
- **EGFR** study for nodal metastatic tumor cells: **positive** (moderately basolateral membranous staining in 80% tumor cells).
- The KRAS gene mutation test: already arranged at section A5 and will be performed as soon as possible.
- pTNM: T4aN1bM0, stage IIIB

Final Diagnosis

- Colon adenocarcinoma, wild type KRAS
- Staging: cT3N2M0, pT4aN1bM0
- Bowel obstruction due to mass effect

Thank you for your attention.