

## A Case of Pancreatoblastoma in an Adult

**Dong Hoon Baek, Dong Uk Kim, Guen Am Song**

*Department of Internal Medicine, Pusan National University School of Medicine, Busan, Korea*

**Introduction:** Pancreatoblastoma is a rare neoplasm in adults, which usually affects infants and young children. Adult pancreatoblastomas are large tumors and majority are larger than 8 cm at the time of diagnosis. Metastasis is seen in 26% of adults and usually involves the liver and then the lymph nodes. Metastasis is usually observed in cases where the primary tumor measures more than 10 cm. Pancreatoblastoma is named after its resemblance to fetal pancreatic tissue in the seventh week of life. The presence of squamoid corpuscles with a morular appearance is the most characteristic feature of the tumor. Pancreatoblastomas can have mixed features of both endocrine and exocrine cells; however, acinar differentiation is the most prevalent feature. Its prognosis is better favorable than typical pancreatic cancer.

**Case:** The patient was a 51-year-old woman who complained of dyspepsia and diarrhea for 3 months. She was transferred to Pusan National University Hospital from a local clinic, where computed tomography revealed an intraabdominal mass compressing the stomach. On admission physical examination revealed an epigastric mass that was hard and movable. A biochemical evaluation revealed no significant abnormalities. Serum level of tumor markers (AFP, CA19-9, CA125 and CEA) were within reference limits. Computed tomography and MR pancreas revealed 10cm mass in the pancreatic head. Her preoperative diagnosis was SPEN by abdominal computed tomography and MR pancreas. During laparotomy, the tumor was located in head of pancreas. The lesion was completely resected without the pancreas being damaged. A histopathological analysis revealed a resected mass measuring 9.5×8.7 cm, which was made up predominantly of medium sized round to polygonal cells having finely granular cytoplasm arranged in solid sheet and acinar structure. Squamoid corpuscles with ovoid to elongated cells were also scattered. In the final pathology report following surgery, the diagnosis of pancreatoblastoma was confirmed with histopathological results. After surgery, the patient was treated with concurrent chemoradiotherapy. There is no evidence of recurrence and metastasis for twelve months after surgery.

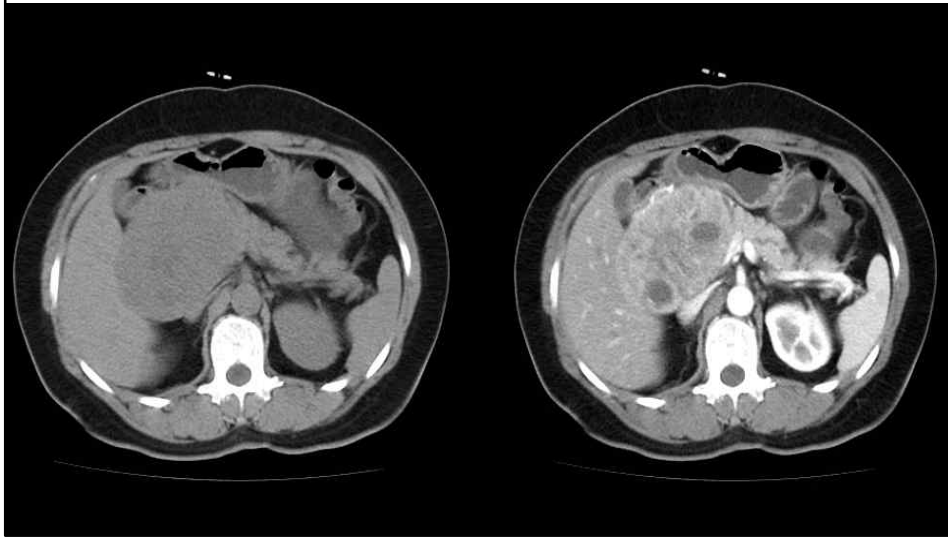
## F/ 51

- C.C) dyspepsia, frequent loose stool for 3 mo
- P.I)
  - visited other hospital
    - EGD : N-S
    - Colonoscopy : N-S
    - CT abdomen : pancreatic mass, 10cm
  - Referred to PNUH

## History

- Past Hx)      DM    HTN    Hepa    Tbc  
                         –        –        –        –
- Social Hx)    Smoking : never  
                         Alcohol : never
- Family Hx)    N/S

## CT abdomen in other hosp.



## Physical examination

- **GA / GC**
  - Acute ill appearance
  - Poor condition
- **HEENT**
  - Not anemic conjunctivae
  - Anicteric sclera
  - dehydrated tongue
  - Not engorged neck v.
  - No CLN, SCLN enlargement
- **Chest**
  - CBS /s r
  - RHB /s m
- **Abdomen**
  - Not distended
  - No tender point
  - **Palpable mass in upper abd.**
- **Extremities**
  - Grossly free
  - No Pitting edema

### Review of system

- Fever/ chilling/ wt loss/ wt gain      -/ -/ -/ -
- General weakness/ Fatigue              -/ -
- Headache/ dizziness/ tinnitus          -/ -/ -
- Cough/ sputum/ dyspnea                -/ -/ -
- Chest pain/ palpitation                  -/ -
- A/ N/ V/ D (**loose stool**) / C          -/ -/ -/ +/-
- Dysuria/ frequency/ dark urine        -/ -/ -

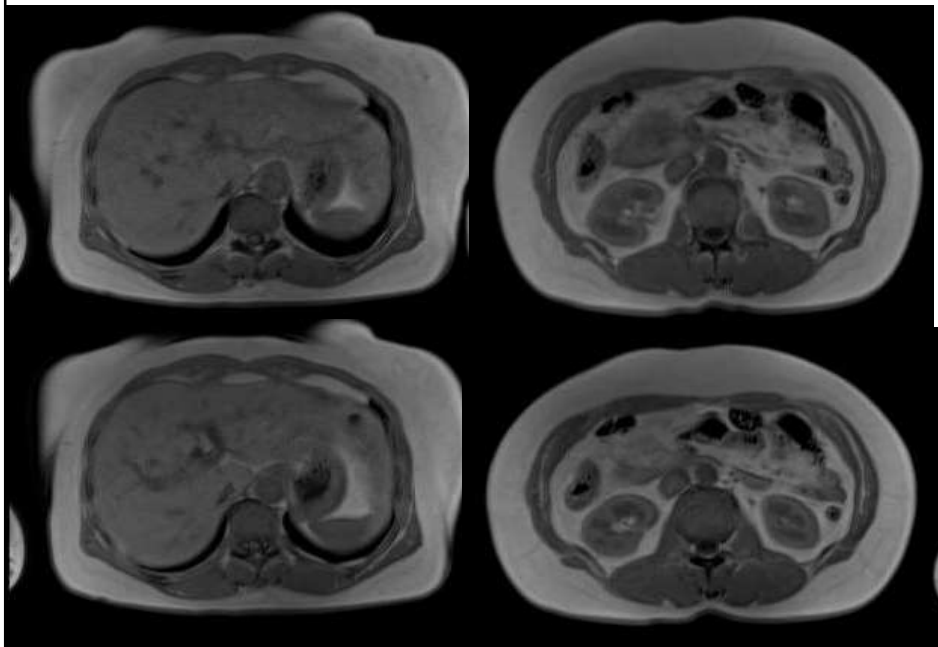
### Initial lab

CBC, electrolyte, APR			LFT		
WBC	8210	/mm <sup>3</sup>	AST / ALT	28 / 24	IU/L
Neutrophil	4950	/mm <sup>3</sup>	ALP / LDH	75 / 211	IU/L
Hb	14.5	g/dL	TB / DB	0.83 / 0.24	mg/dL
PLT	283	10 <sup>3</sup> /mm <sup>3</sup>	TP / Alb	7.50 / 4.9	gm/dL
			BUN / Cr	16.6 / 0.94	gm/dL
Na	143.7	mEq/L	TC / UA	188 / 4.9	gm/dL
K	4.14	mEq/L	Ca / P	10.2 / 4.3	gm/dL
Cl	103.8	mEq/L	GGT	50	IU/L
ESR	5	mm/hr	PT(INR)	1.03	Sec
CRP	0.85	mg/dL	aPTT	30.9	Sec

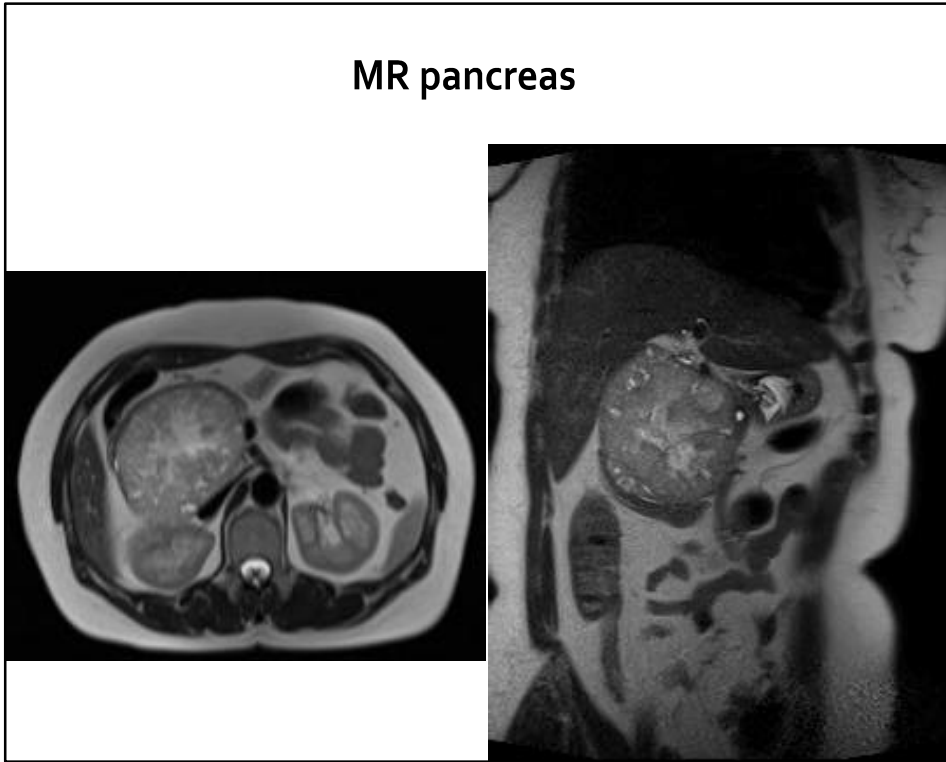
## Initial lab

- Tumor marker
  - CA19-9 6.75 U/mL (0-39)
  - CEA 1.35 ng/mL (0-5)
  - AFP 1.80 IU/mL (0-10)
  - CA125 9.93 U/mL (0-35)
- Amylase 52.7 IU/mL (36-128)
- lipase 38.9 U/mL (22-51)
- HBsAg / Ab- / +
- HCV Ab -

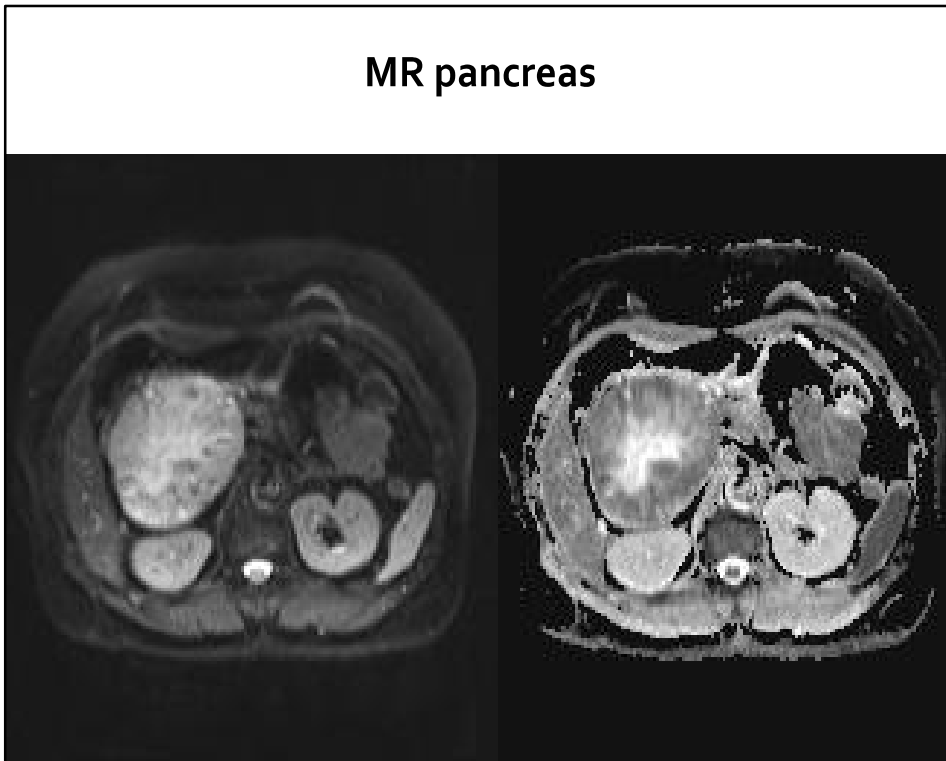
## MR pancreas



MR pancreas



MR pancreas



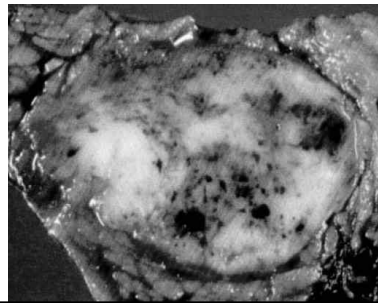
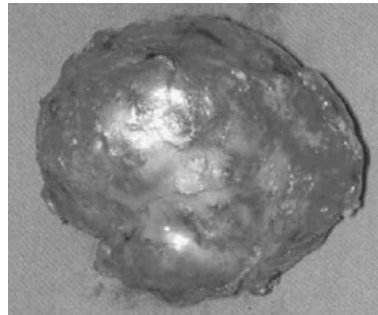


## impression

- SPEN (Solid and papillary epithelial neoplasm of pancreas)

**PPPD**

- PPPD : pylorus-preserving  
pancreatico\_duodenectomy
  
- Gross finding
  - Yellow & pink solid mass
  - 9.5 x 8.7 cm
  - Hemorrhage in mass

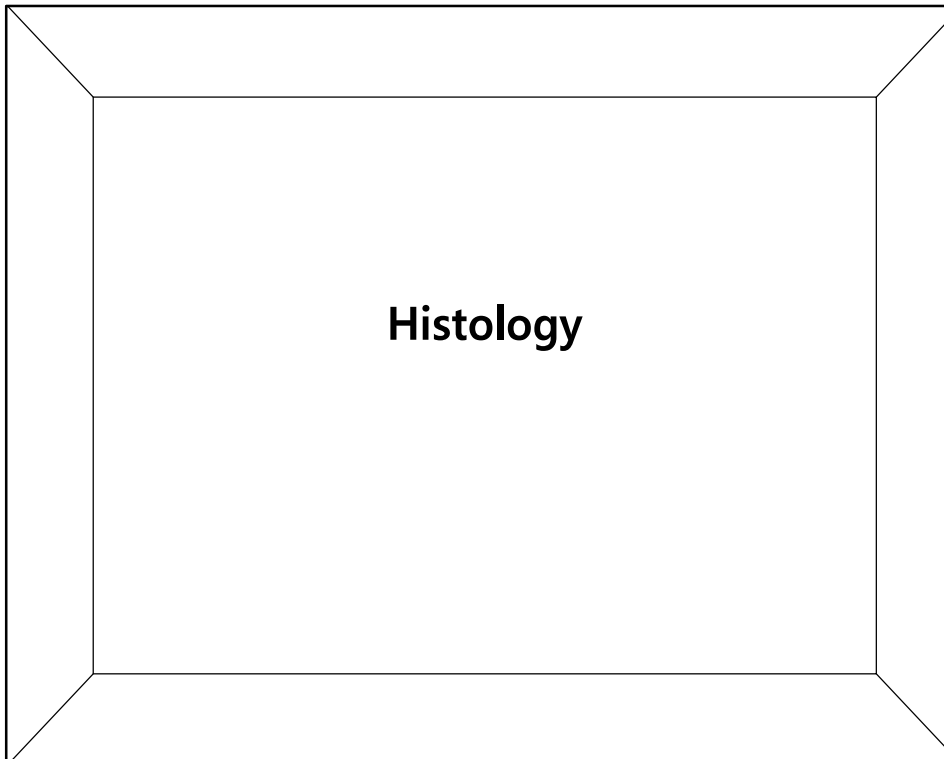


**F/U lab (POD #1)**

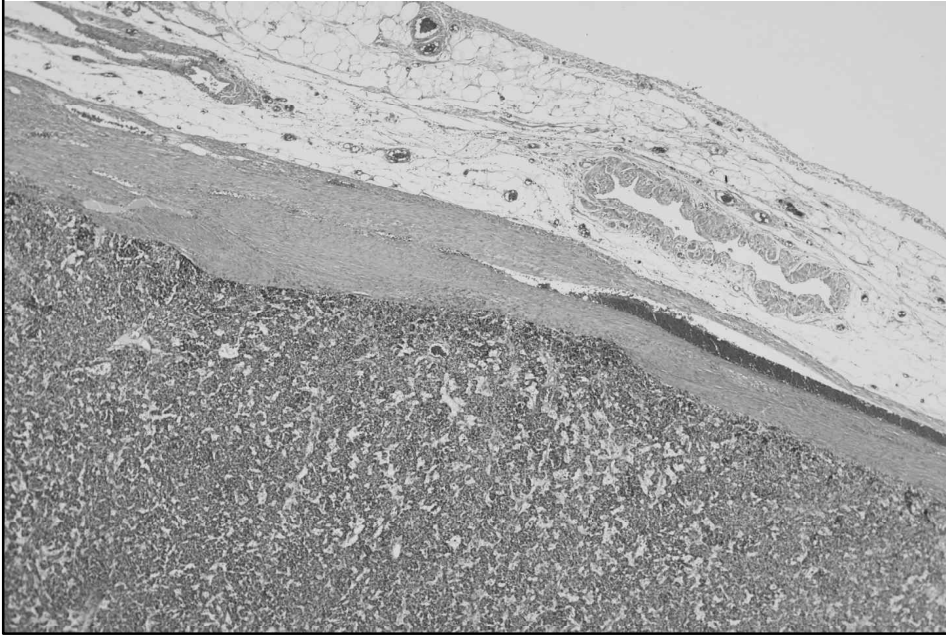
<b>CBC, electrolyte, APR</b>			<b>LFT</b>		
WBC	16760	/mm <sup>3</sup>	AST / ALT	54 / 53	IU/L
Neutrophil	14460	/mm <sup>3</sup>	ALP / LDH	45 / 214	IU/L
Hb	11.7	g/dL	TB / DB	0.74 / 0.76	mg/dL
PLT	197	10 <sup>3</sup> /mm <sup>3</sup>	TP / Alb	5.0 / 3.2	gm/dL
			BUN / Cr	11.1 / 0.56	gm/dL
Na	142.2	mEq/L	TC / UA	130 / 2.6	gm/dL
K	4.14	mEq/L	Ca / P	8.4 / 3.5	gm/dL
Cl	106.2	mEq/L	GGT		IU/L
ESR		mm/hr	Amylase	522.6	IU/L
CRP	11.27	mg/dL	Lipase	248.7	U/L



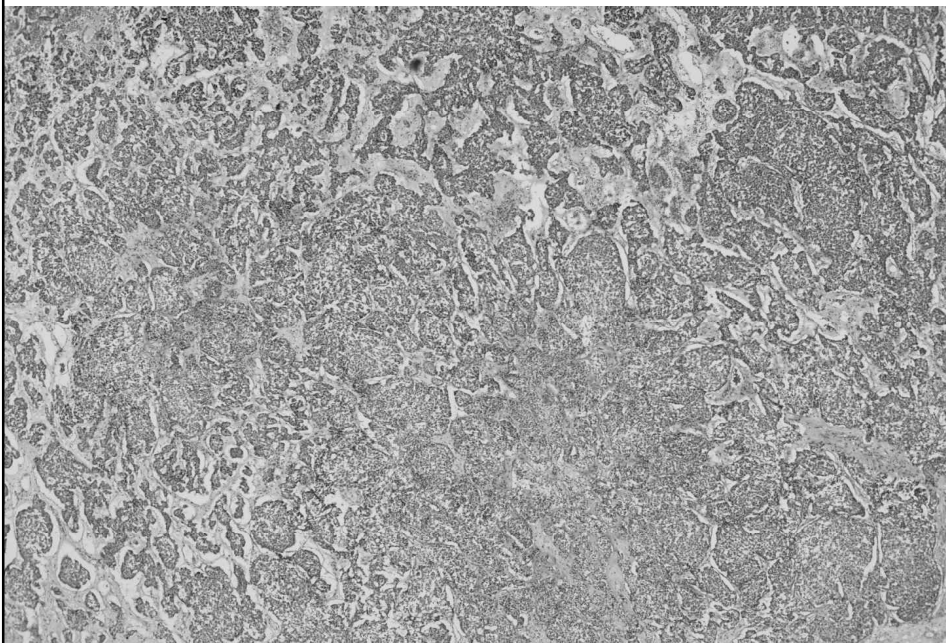
F/U lab (POD #6)					
CBC, electrolyte, APR			LFT		
WBC	10550	/mm <sup>3</sup>	AST / ALT	16 / 28	IU/L
Neutrophil	7880	/mm <sup>3</sup>	ALP / LDH	68 / 236	IU/L
Hb	12.7	g/dL	TB / DB	0.29 / 0.14	mg/dL
PLT	287	10 <sup>3</sup> /mm <sup>3</sup>	TP / Alb	5.7 / 3.7	gm/dL
			BUN / Cr	8.9 / 0.71	gm/dL
Na	142.2	mEq/L	TC / UA	86 / 1.9	gm/dL
K	4.14	mEq/L	Ca / P	9.3 / 38	gm/dL
Cl	106.2	mEq/L	GGT		IU/L
ESR		mm/hr	Amylase	24.8	IU/L
CRP	1.57	mg/dL	Lipase	12.3	U/L



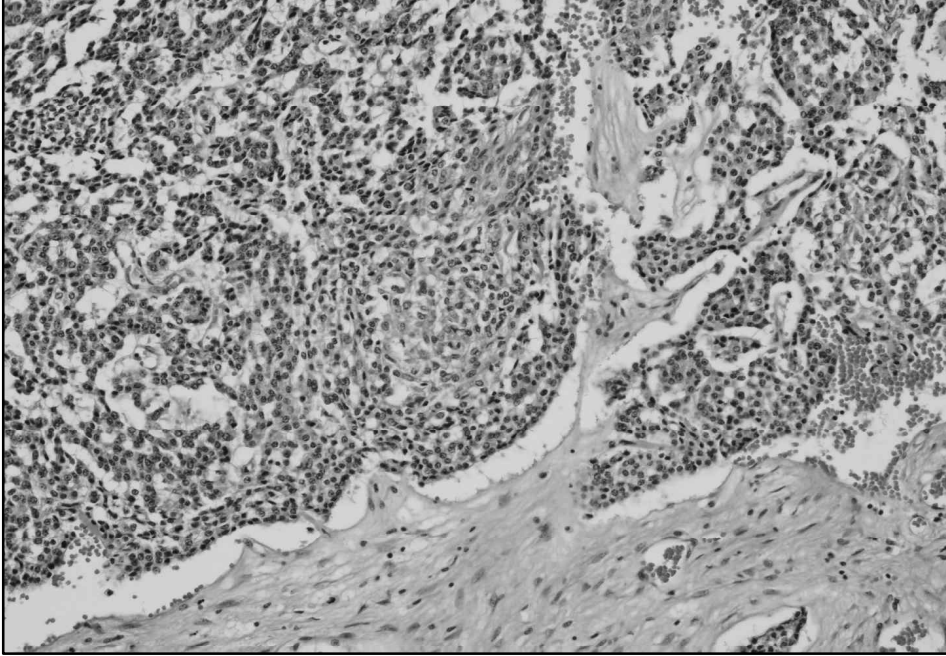
**Histology, LM x40**



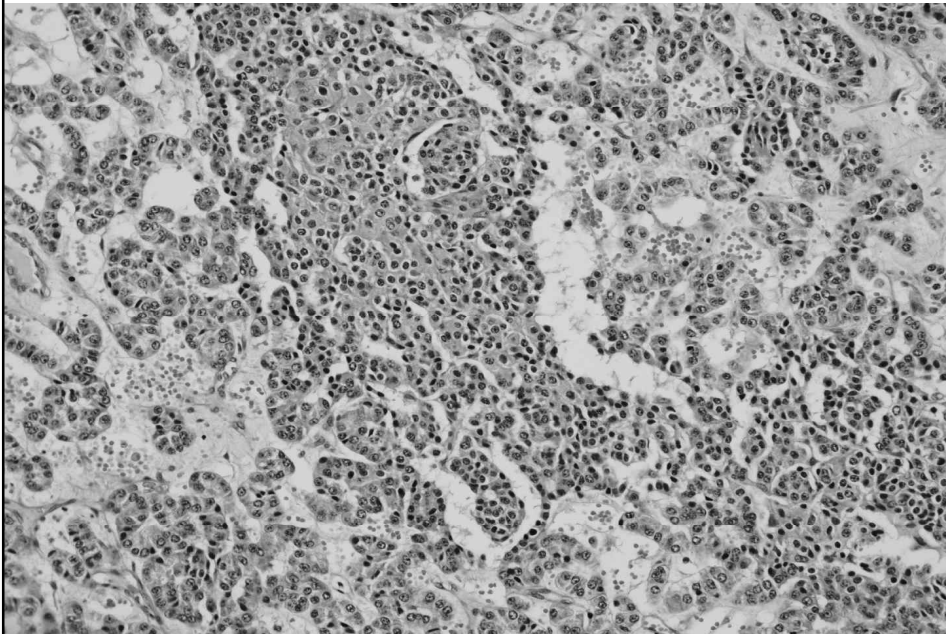
**Histology, LM x40**



**Histology, LM x400**



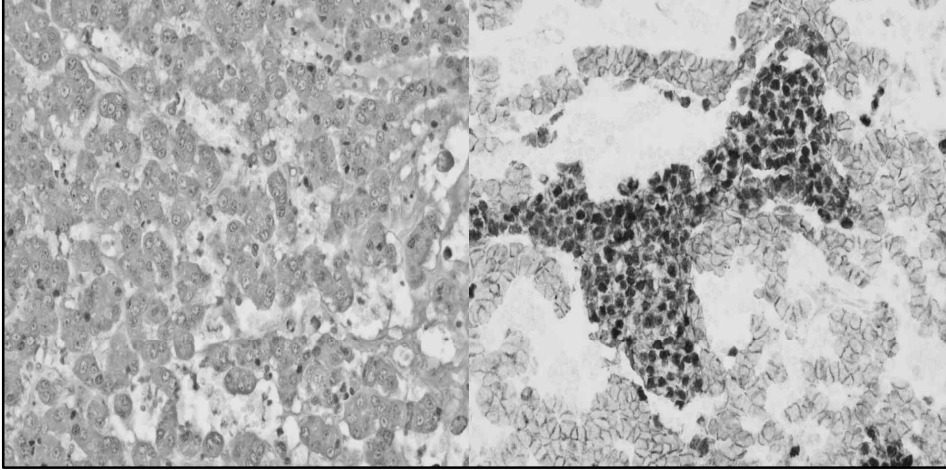
**Histology, LM x400**



## Histology, immuno\_histo\_chemistry

$\alpha$ -1-antitrypsin (+)

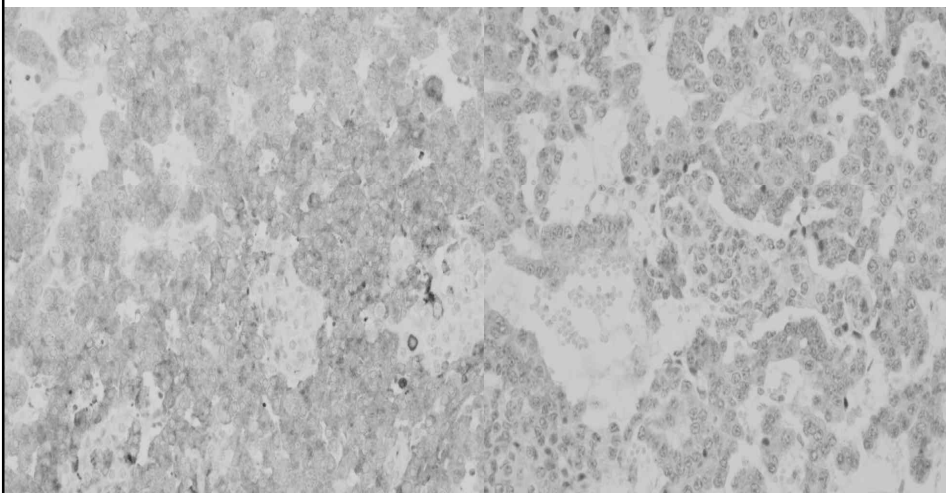
$\beta$ -catenin (+)



## Histology, immuno\_histo\_chemistry

Synaptophysin (+)

Chromogranin A (+)



## Progress

- Dx : Pancreato\_blastoma, (pT2N0M0, LN 0/13)
- CCRT
  - Pt had suffered badly & refused further Tx (except FU exam)
- 2013-10 lastest visit
  - CT abdomen : Recurrence : (-)

Pancreato\_blastoma

## Review



Pancreato\_blastoma

## Pancreato\_blastoma (PB)

- **Extremely rare pancreatic tumor**
  - 0.2-0.5% of pancreatic tumor
  - 0.5% of pancreatic non-endocrine tumor
- **Occur in childhood**
  - PB = “infantile pancreatic carcinoma”
  - Can occur in adult, M > F
    - more aggressive than childhood
    - Px of PB in adult > adult panc. carcinoma

## Sx of PB

- Presented late
  - **Incidental abdominal mass in epigastrium (50%)**
  - Upper abdo. pain (43%)
  - Wt loss (29%)
  - Mechanical obst. of duodenum (in panc. head)
    - vomiting, jaundice, GIB
  - Fatigue, anorexia, splenomegaly

## Dx of PB : tumor marker

- **AFP**
  - ▲ in 68%, ▽ after resection
  - panc. & liver arise from same primitive cells
- CA 19-9, lipase, trypsin, chymotrypsin, chromogranin  
: correlation (-)

## Dx of PB : imaging

- Location : pancreatic head, mainly
- Finding
  - Calcification : not large, like teratoma
  - **Hemorrhagic necrosis : within the tumor**
  - Solid & cystic elements : typically present
  - Metastasis : liver and LN
- pre\_OP Dx : difficult → Histology

## Histology of PB

- Acinar, endocrine, ductal differentiation
    - /c hemorrhage, capsule formation, necrosis
  - Epithelial component
    - separated into distinct lobules by fibrous stroma
- **Squamoid corpuscles**
- : **typical aspect of pancreatoblastoma**

## Immuno\_histo\_chemistry of PB

- Positive
  - **Alpha 1-antitrypsin**
  - glucose 6 phosphatase
  - trypsin, chymotrypsin
- May positive
  - acid phosphatase
  - Esterase
  - entero\_protease
  - **chromogranin, synaptophysin**, neuron-specific enolase
- AFP : positive in solid region
- EM : cytoplasmic neuro\_secretory zymogen granule



## Tx, Px of PB

- Complete resection
  - TOC, may be curative
  - Adjuvant CTx or RTx : controversial
  - Long term FU is necessary (recurrence is common)
- Unresectable : CTx
- Px
  - Good. When resected completely
  - Poor. When metastasis (+) or inoperable