A 3-year-old boy with upper abdominal mass

โรงพยาบาลรามาธิบดี

History:

ผู้ป่วยเด็กชายไทย อายุ 1 ปี มีก้อนในท้องมา 1 ปี

ประวัติปัจจุบัน: 1 yr PTA มารดาสังเกตว่ามีก้อนบริเวณลิ้นปี่ ไม่มีอาการผิดปกติใดๆ ไม่เคยพาไปตรวจที่ไหน;

4 wk PTA กินได้น้อยน้ำหนักลด 3 กก. ใน 1 สัปดาห์ ไม่ปวดท้อง คลำก้อนได้ชัดขึ้น ไป รพ.เอกชน แนะนำให้มา รพ.รามาธิบดี; 1 wk PTA มารดาสังเกตว่าก้อนโตขึ้น แน่นท้องบางครั้ง ไม่มีไข้ ไม่มีตัวเหลืองตาเหลือง ปัสสาวะอุจจาระปกติ

Past history:

เป็นบุตรคนที่ 2/2 คลอดปกติที่ รพ.มหาชัย น้ำหนักแรกเกิด 4,200 กรัม แข็งแรงดีมาตลอด ปฏิเสธโรคประจำตัว

ฉีดวัคซีนครบตามเกณฑ์; ปฏิเสธประวัติเคยได้รับอุบัติเหตุบริเวณช่องท้อง

Family history:

ลุงเป็นมะเร็งถุงน้ำดี; ปู่เป็นมะเร็งต่อมลูกหมาก

Physical examination:

GA: A Thai boy, good consciousness, BW 16 kg Height 100 cm

VS: normal

HEENT: no pallor, no icteric sclera

RS: normal breath sound
CVS: normal S1 S2, no murmur

Abdomen: soft, moderate distension, no tenderness, firm mass size 9x11 cm. at epigastrium area, slightly movable, no splenomegaly, liver just palpable, span 6 cm below RCM

Extremities: no pitting edema, no ecchymosis
Lymnodes: no palpable LN
Neuro signs: grossly intact
Problem lists :

>>Abdominal mass at epigastria area

>>Weight loss

Approach to upper abdominal mass in children:

Stomach	Carcinoma, Leiomyosarcoma, Rhabdomyosarcoma, Myosarcoma Fibrosarcoma
Small bowel	Lymphoma
Omentaum and mesentery	Cysts, Mesenteric fibromatosis, Inflammatory pseudotumor Liposarcoma, Leiomyosarcoma, Fibrosarcoma, Mesothelioma Metastatic tumor
Retroperitoneum	Wilms tumor, Neuroblastoma, Pancreatoblastoma, Lymphoma Rhabdomyosarcoma, Ewing sarcoma, Germ cell neoplasm
Liver	Benign: Adenoma, Hamartoma, Focal nodular hyperplasia Malignant: Hepatoblastoma, HCC, Germ cell tumor, angiosarcoma, Intrahepatic mesenchymal tumor, Rhabdomyosarcoma Vascular: Hemangioendothelioma, Solitary cavernous hemangioma

Basic investigations:

CBC : Hb 10.3 g/dl Hct 32%, WBC 7,260 cell/mm3, PMN 22% L 66% M 10% E 1%, Plt. 465,000 cell/mm3, MCV 70 fL, PTT 26.3 sec, PT 11.1 sec, INR 0.93 TT 10.0 sec

UA: pH 6.0, Pro. Negative, Glu. Negative, Bil. Negative, Uro. Negative, Leu. Negative
WBC 0 – 1, RBC Negative
Electrolytes: Na136 mmol/L, K5.12 mmol/L, Cl 102 mmol/L, HCO3 22 mmol/L
BUN 15 mg/dl, Cr 0.3 mg/dl
LFT: ALP 204 mmol/L, AST 35 mmol/L, ALT 41 mmol/L, GGT 174 mmol/L, ALB 37.4 g/L, Tbil. 0.2 mg/dl, Dbil. 0.1 mg/dl
Hepatitis profiles: HBs Ag negative, Anti HBS positive, Anti HCV negative
AFP 2.34 ng/ml (0-7.02)
Beta- HCG < 1.2
LDH 215 U/L

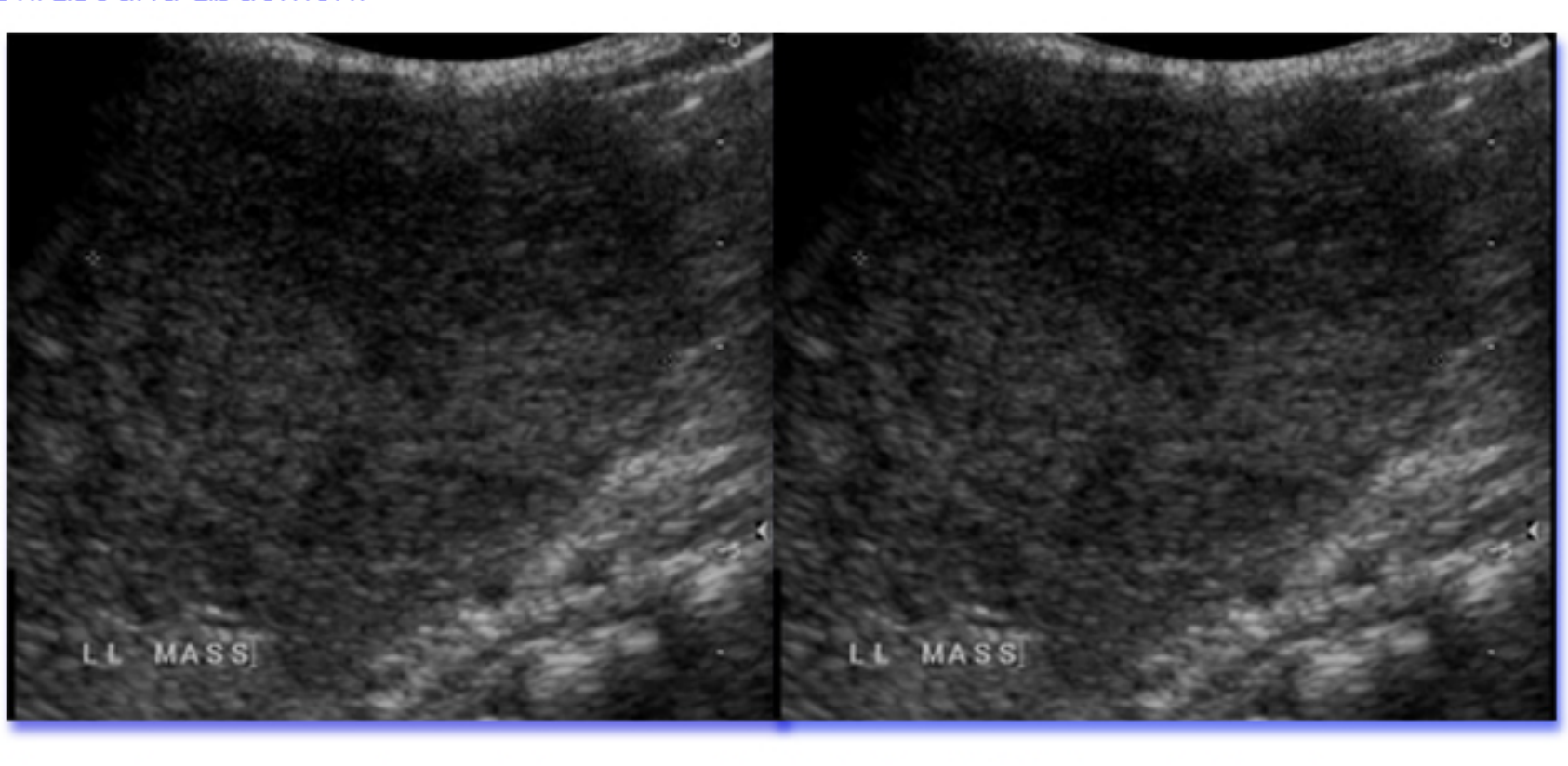
Blood for NSE 80.36 ng/ml(0-15 ng/ml)

Bone marrow biopsy: smear,cytogenetic, pathology

Blood for N-myc amplification

Urine 24 hr for VMA, Cr All result: normal

Ultrasound abdomen:

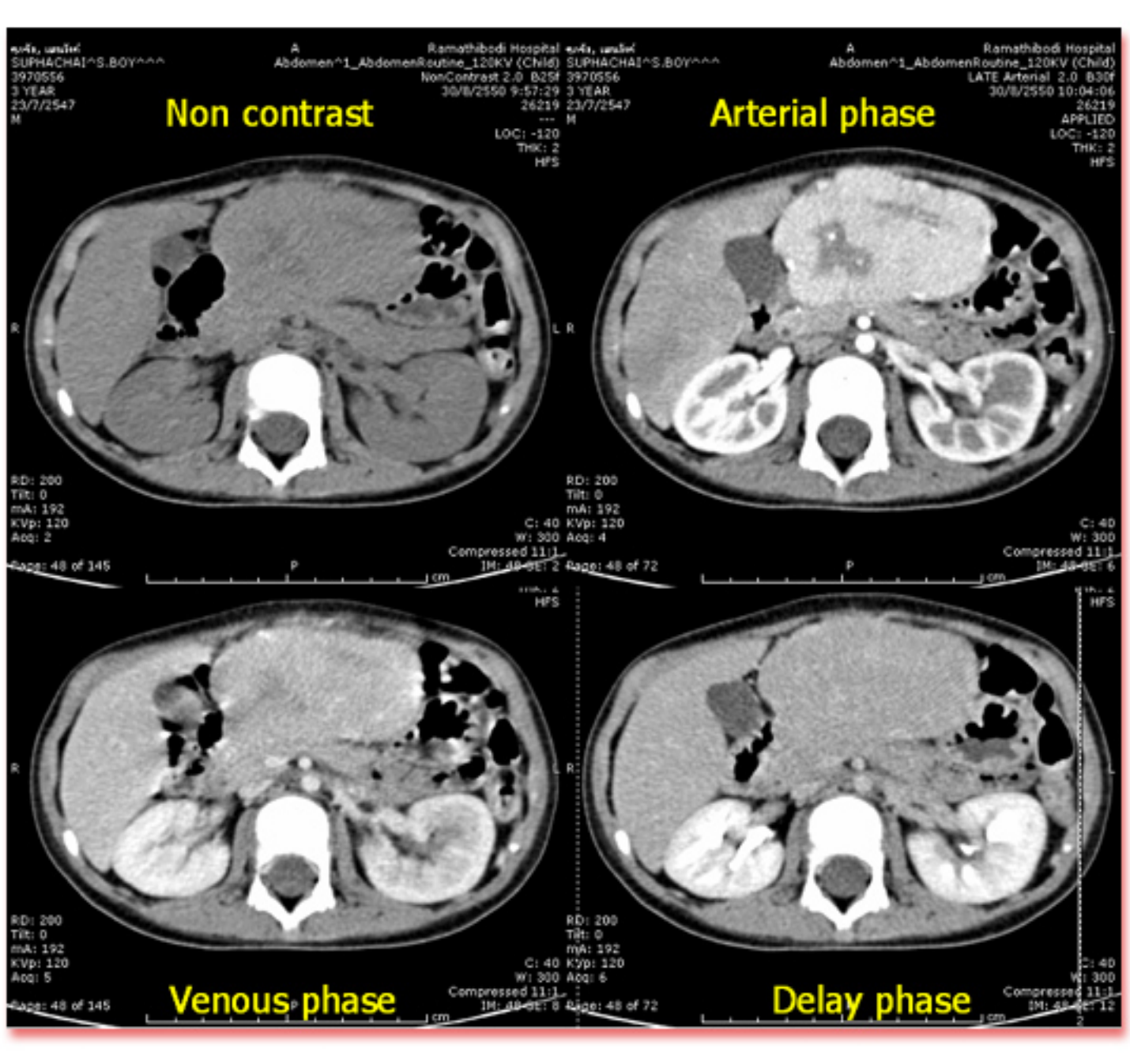


A Large homogenous hypoechoic mass at left lobe liver 5.7x8.3 cm. in diameters
The rest of liver parenchyma appear normal

Differential diagnosis of liver mass by ultrasound:

Hypoechoic mass	Hyperechoic mass
Hepatic adenoma	Hepatic adenoma
Atypical Hemangioma	Hemangioma
Focal nodular hyperplasia	Focal nodular hyperplasia
Metastatic tumor	Metastatic tumor
Lymphoma	Lipoma/angiomyolipoma
Hepatic infarction	Focal fatty change
HCC	Cholangiocarcinoma

Further investigations: CT abdomen:



CT non-contrast: Huge lobulated, exophytic heterogenous arterial enhancing isodense mass without internal calcification involving nearly entire Lt hepatic lobe about 6x8.6x7.2 cm. It contains ill-defined internal non-enhancing area suggestion necrosis/scar.

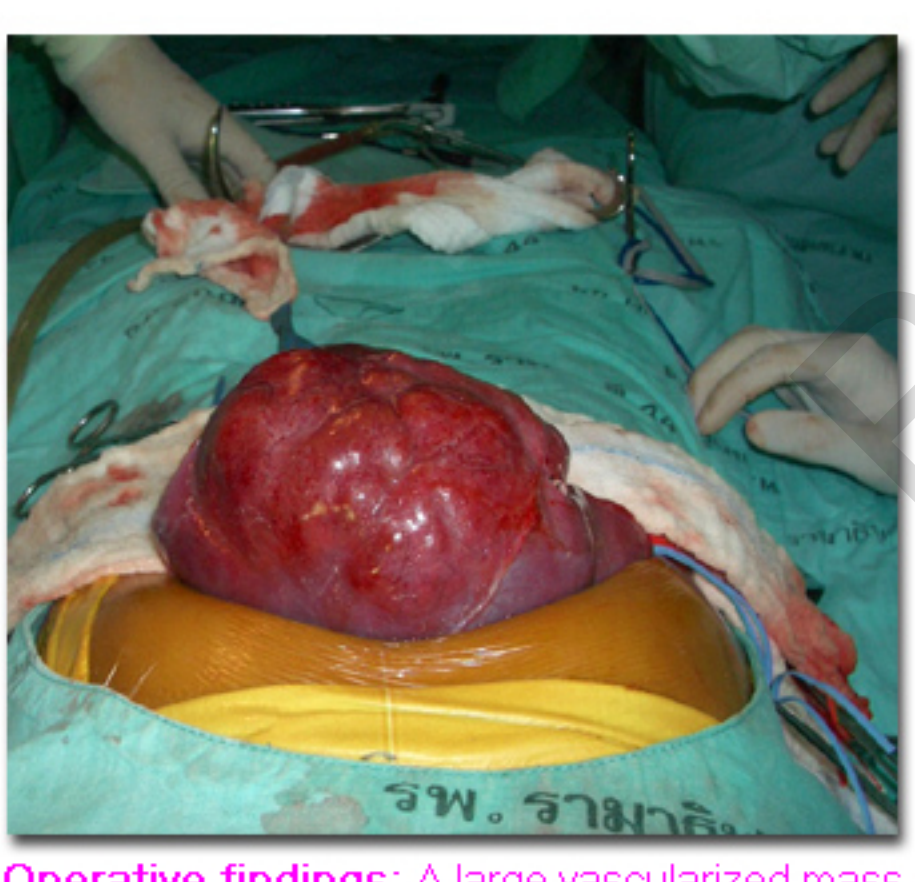
CT arterial and venous phase: Peripheral wedge-shaped hyperdensence lesion and turn to be isodense in portal phase. The mass is supplied by the left

Differential diagnosis of the liver mass based no the result of CT abdomen:

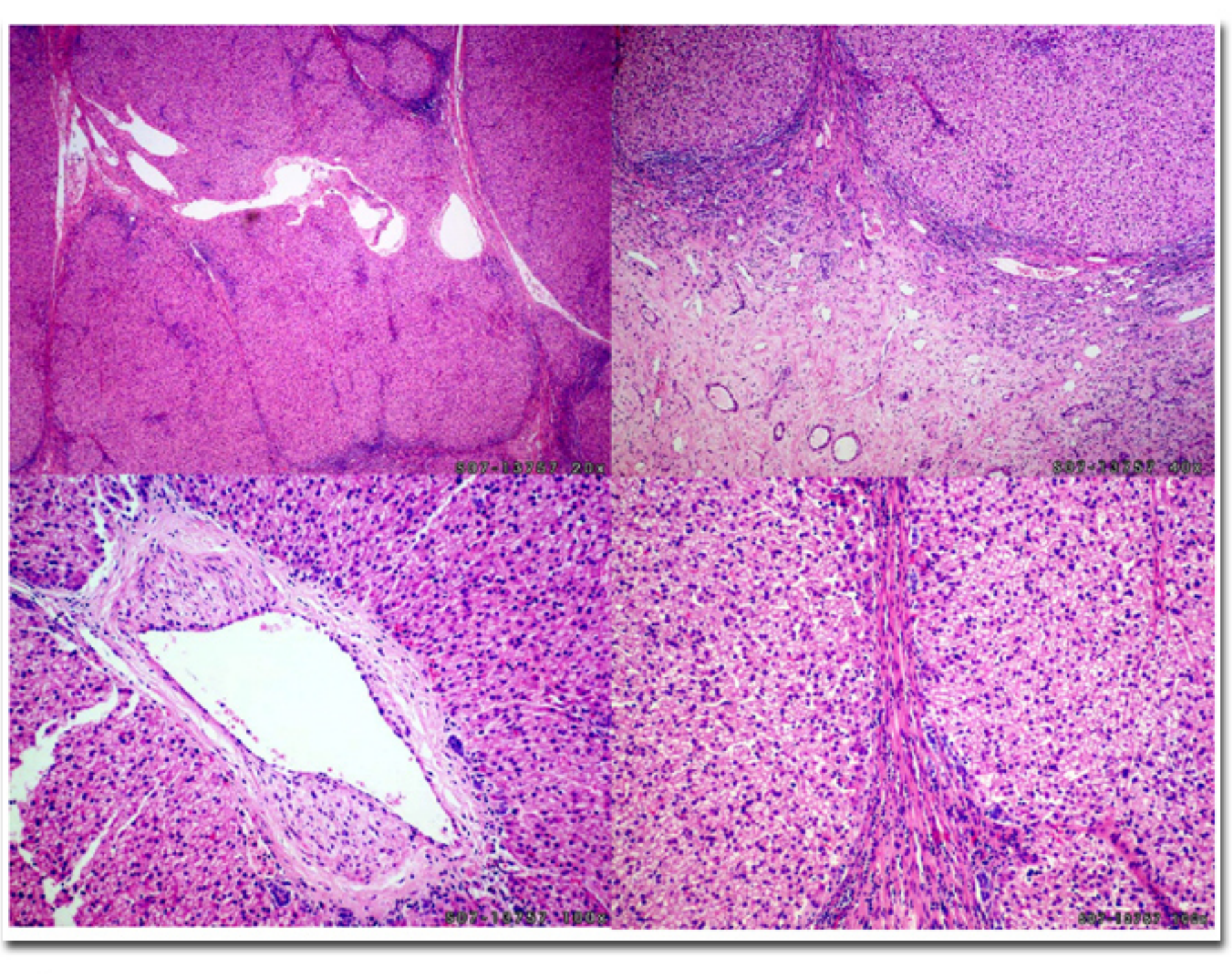
>>Fibronodular hyperplasia

>>Hepatoblastoma
>>Fibrolamellar HCC

Operation



Operative findings: A large vascularized mass in left lobe liver about 10 cm. in diameter, irregular surface. Several hilar node 0.5-1.2 cm. in diameter rubbery consistency. Normal Rt. lobe of liver and gallbladder. Minimal clear peritoneal fluid. Frozen section from core needle biopsy is benign



Pathologic description:

1- Nodular hyperplastic parenchyma.

2 - Nodules: completely or incompletely surrounded by circular or short fibrous septa
3- Hepatic plates may be moderately thickened (two or three cells in thickness) with normal-appearing hepatocytes
4- The central scar:

>>>>Fibrous connective tissue

>>>>Cholangiolar proliferation with surrounding inflammatory infiltrates
>>>>Malformed vessels of various caliber, including tortuous arteries with thickened walls, capillaries, vascular channels of undetermined type, and veins

Pathological diagnosis: Focal nodular hyperplasia

Final diagnosis:

Focal nodular hyperplasia

Focal nodular hyperplasia

Primary liver tumor in children:

Tumor	%
Hepatoblastoma	43
HCC	23
Adenoma	2
Hemangioma, Hemangioendohelioma	13
Mesenchymal hamartoma	6
Sarcoma	6
Focal nodular hyperplasia	2
Other	5

Etiology of the tumor according to age group:

Age group	Malignancy	Benign
Infant/Toddler	Hepatoblastoma (43%) Rhabdoid tumor (<1%) Malignant germ cell (<1%)	Hemangioma/ vascular (14%) Mesenchymal hamartoma (6%) Teratoma(<1%)
School age/Adolescent	Hepatocellular and transitional cell tumors(23%) Sarcomas (7%)	Hepatic adenoma(2%) Focal nodular Hyperplasia(FNH) (2%)

Von Schweinitz D. Management of liver tumors in childhood.Seminar in Pediatric Surgery 2006;15:17–24.

Focal nodular hyperplasia

>Benign epithelial tumor
>Second most common benign liver tumor after hemangioma
>Prevalence 0.9%, M:F = 1:8
>Various names: benign hepatoma, solitary, hyperplastic nodule, focal cirrhosis, cholangiohepatoma, mixed adenoma
>Dx: any age: newborn -> elderly

>In children: common 2-5 yrs of age

Clinical presentation
>Asymptomatic

>Most common symptom is abdominal pain

>>>>abdominal mass
>>>>decreased appetite
>>>>weight loss
>Hepatomegaly
>Normal liver function test
Pathogenesis:
> Vascular malformation & vascular injury -> suggested as the underlying mechanism
>An association with steroids has been denied more recently
Pathology
Mass of a few mm. to 20 cm in diameter; Multiple lesions 20%, Single lesion 80%
Nodular architecture; Central or eccentric scaring (80%); Fibrous septa contain bile duct and malformed vessels; No malignant transformation
Multiple FNH syndrome: The combination of multiple FNH lesions and hemangiomas

Current pathologic classification:

Classic FNH :

>>Abnormal nodular architecture

>>Malformation of vessels

>>Cholangiolar proliferation

Nonclassic FNH :

>>Lack one of the following classic

>>Always show bile ductular proliferation

Diagnosis :
Ultrasound - Not well visualized; A subtle change in echogenicity compared with the surrounding normal liver parenchyma; can be Slightly hypoechoic, isoechoic, or slightly hyperechoic

CT scan - Classical findings are early enhancement mass with central scaring
MRI -
>>>T1: iso- or hypointense
>>>T2 : slightly hyper- or isointense and has a hyperintense central scar
>>>Gadolinium-enhanced
- Arterial phase: intense homogeneous enhancement
- Later phases: enhancement of the central scar
Treatment :
Asymtomatic and diagnosis is certain- follow up with serial US
Symptomatic, progression, unclear diagnosis-
>>>Percutaneous biopsy
>>>Arterial embolization
>>>Liver resection

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