Basic Data

Name :林O晰

Sex : 女　　Age : 9

Residence : 台北市

Chief complaint

Fever off and on and liver mass for 1+ month

Present Illness

- Underlying Dx : right adrenal tumor s/p total excision on 2000-9-19

- fever on and off with chills since March 2006

- Hung liver tumor was found. She had recipient surgical (opened and closed) intervention on last week at 新光 hospital.

Past History

- Adrenal cortical adenoma s/p total excision on 2000 at 新光醫院

  ※ pathology : Adrenal cortical adenoma with malignant potential, nuclear pleomorphism with capsular invasion

Personal History

- Drug Allergy : NKA
- Food Allergy : NKA

95/04/22

Lab data : (4/22)

WBC : 12680 / ul

RBC : 3.70 x 10^6 / ul

HGB : 9.0 g/dl

HCT : 29.7 %

MCV : 80.3 fL

MCHC : 30.3 g/dl

RDW : 14.9%

GOT : 16 IU / L

GPT : 23 IU/ L

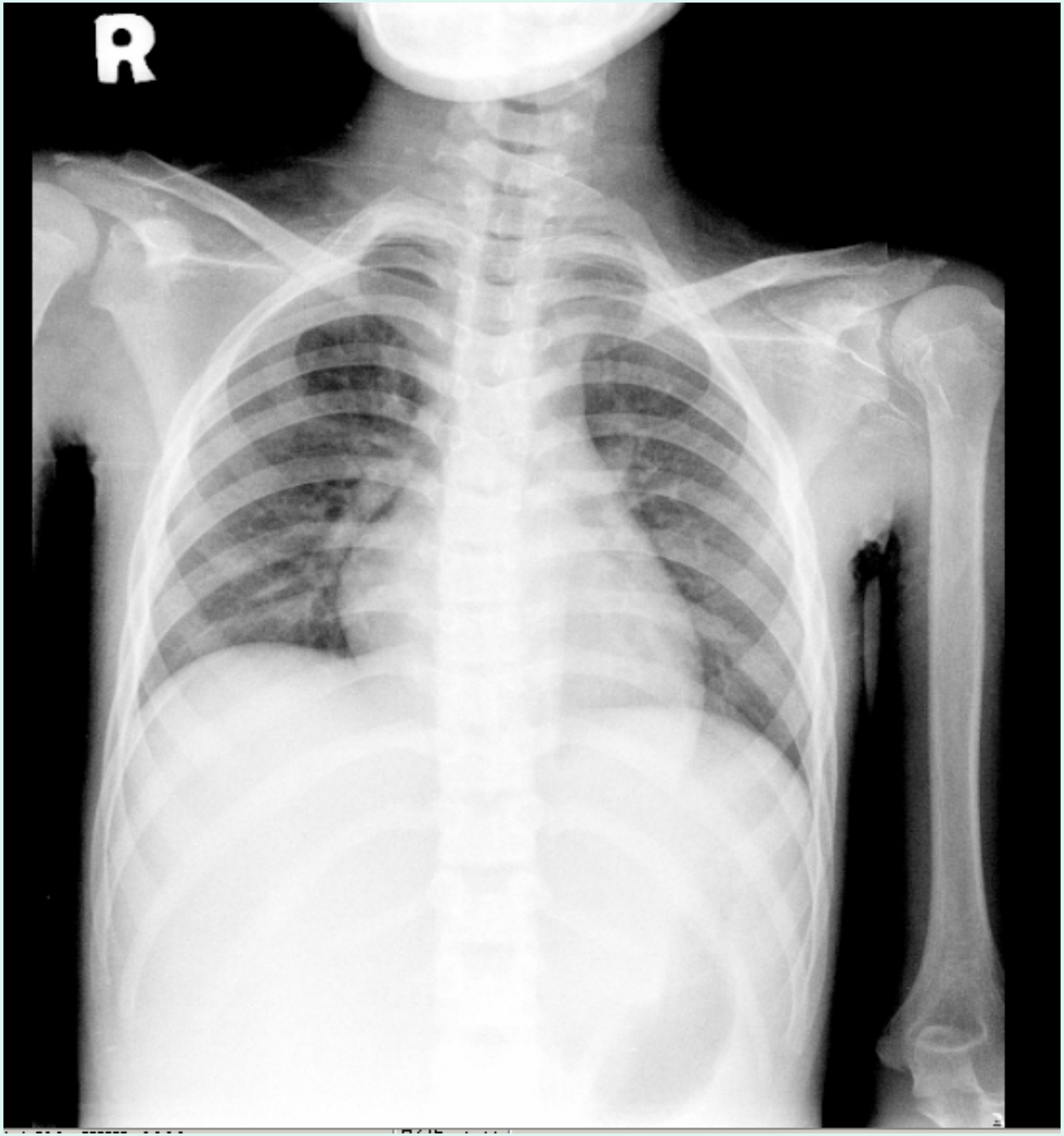
Bilirubin D : 0

Bilirubin T : 0.4

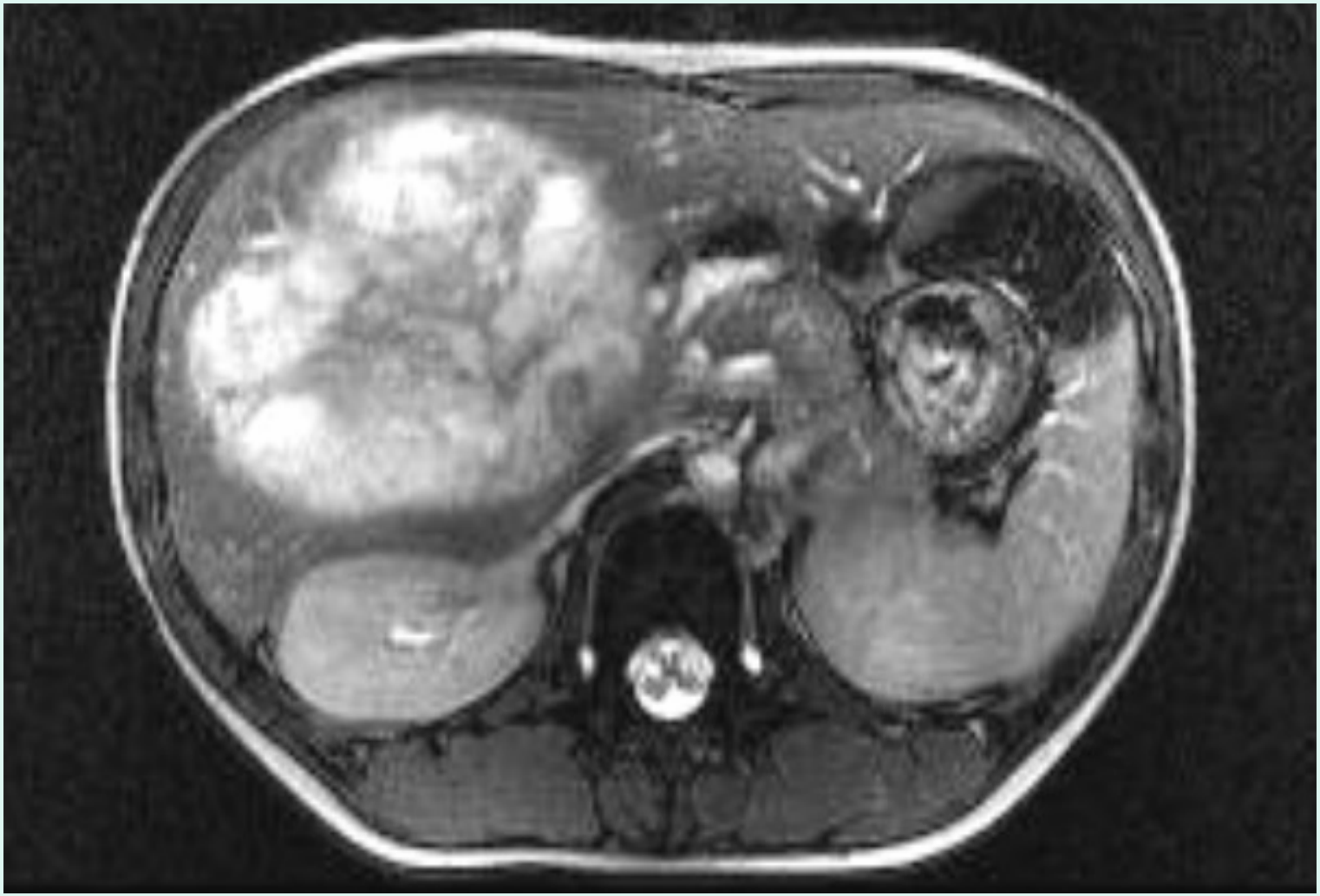
MCH : 24.3 pg

CXR

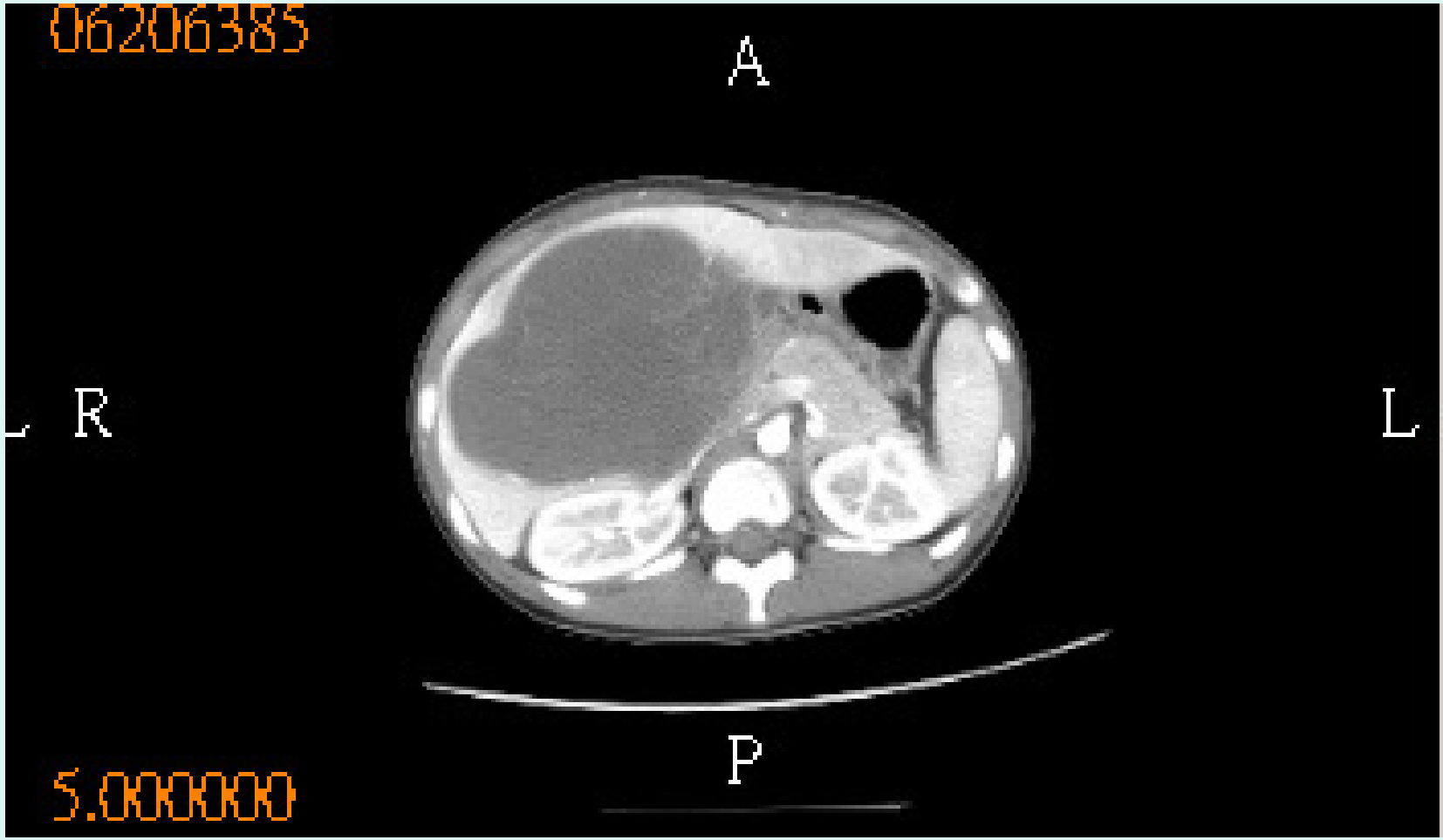
(950422)

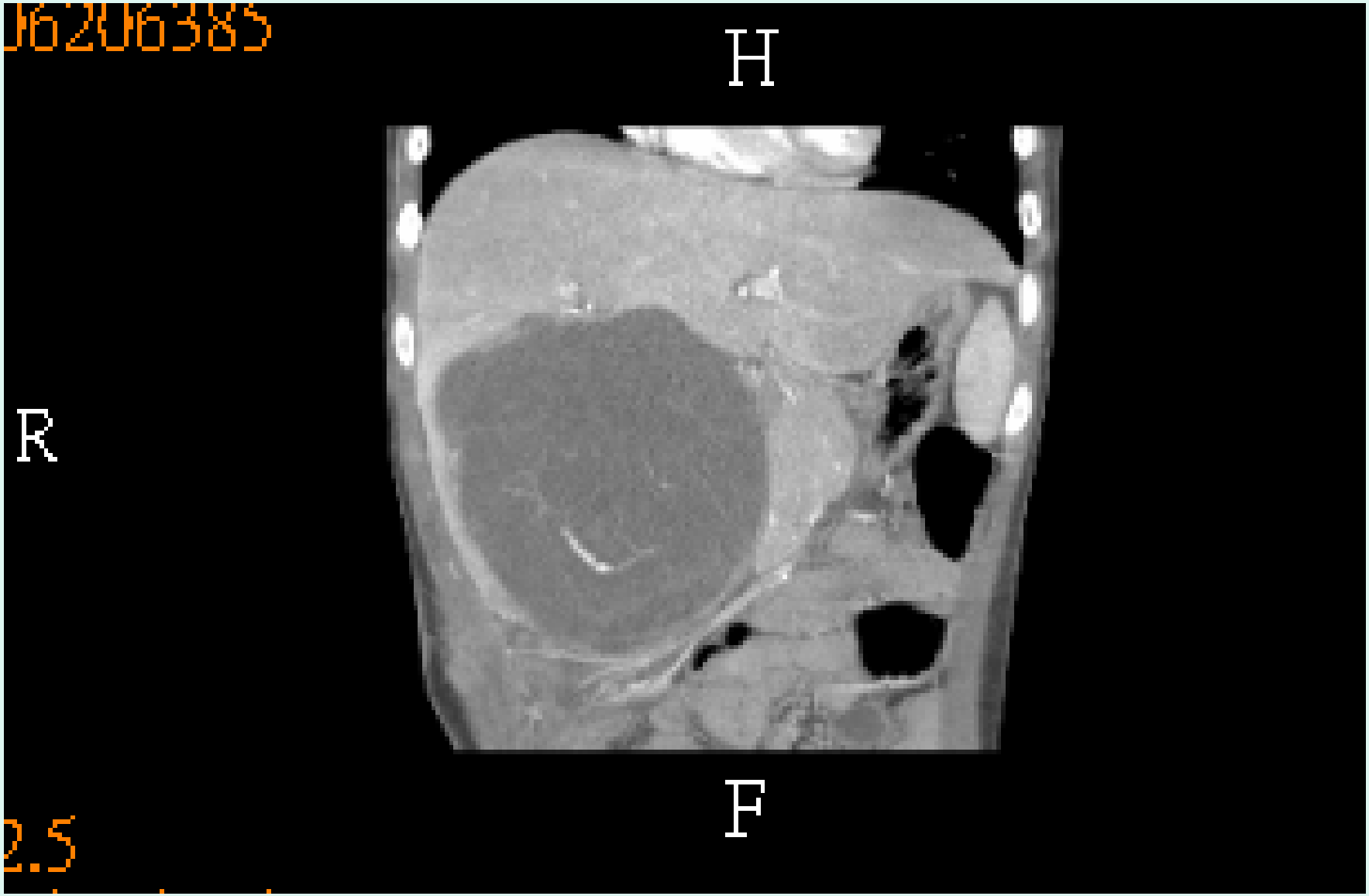


Abdominal MRI (新光)



VCT-Abdomen CT (940422)




H
R
F

Imagine Finding

1. There is huge size heterogenous hypodense mass with some internal enhanced density,measuring about 12x10.2cm in largest dimension,occupy at Seg IVb,V and VI of liver with remarkable adjacent compressing effect.

2. There is obvious compression of IVC (from T10-T12 level) ,gall bladder ,portal vein, pancreas and extrahepatic biliary system, duodenum and bowel structure

3. There is suggest early occlusion ,only severe adhesion of IVC by tumor mass, at L1-2 levels

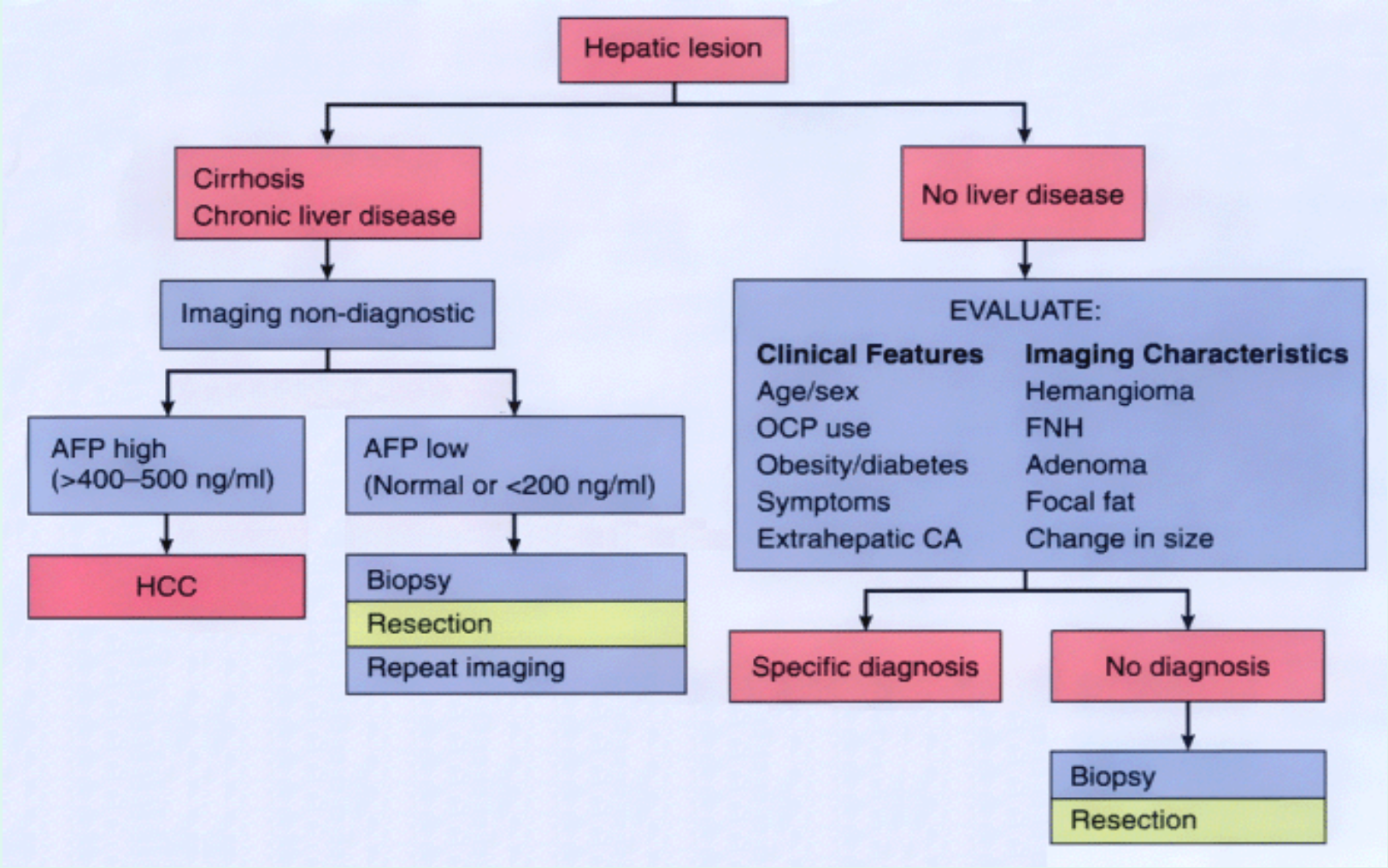
4. There is no definite >1cm LN enlargement in the paraaortic region and pelvic side walls(include inguinal)

5. There is no evidence of focal peritoneal infiltration could be identified.

Imagine impression

- Liver mass about 12X10.2 cm
  - r/o adrenal tumor metastasis
  - r/o HCC
  - r/o Hepatoblastoma

Liver tumor


Hepatic lesion
Cirrhosis
Chronic liver disease
No liver disease
Imaging non-diagnostic
EVALUATE:
Clinical Features
Age/sex
OCP use
Obesity/diabetes
Symptoms
Extrahepatic CA
Imaging Characteristics
Hemangioma
FNH
Adenoma
Focal fat
Change in size
AFP high
(>400–500 ng/ml)
AFP low
(Normal or <200 ng/ml)
HCC
Biopsy
Resection
Repeat imaging
Specific diagnosis
No diagnosis
Biopsy
Resection

HCC

- Children who are affected with biliary atresia, infantile cholestasis, glycogen storage diseases, and a wide array of cirrhotic diseases of the liver are predisposed to developing HCC.

- Many children also experience localized pain, nausea, vomiting, and weight loss. Nearly 25% of patients present with jaundice

- Liver biopsy is the most important procedure to consider when HCC is suspected.

Imagine diagnosis

- Initial staging evaluation should include, but is not limited to, chest, abdomen, and pelvic CT scans.

- If surgical resection is anticipated, use MRI and magnetic resonance angiography (MRA) of the liver to best determine tumor margins and vasculature

- Unenhanced CT typically reveals an iso-hypodense mass. central areas of necrosis may be seen.

- In the hepatic-arterial phase, lesions typically are hyperdense (relative to hepatic parenchyma) as a result of hepatic-arterial supply. Larger tumors may have necrotic central regions that typically are hypodense during this imaging phase.

- In the portal-venous phase, small lesions may be isodense or hypodense and difficult to see, since the remainder of the liver increases in attenuation. Larger lesions with necrotic regions remain hypodense.

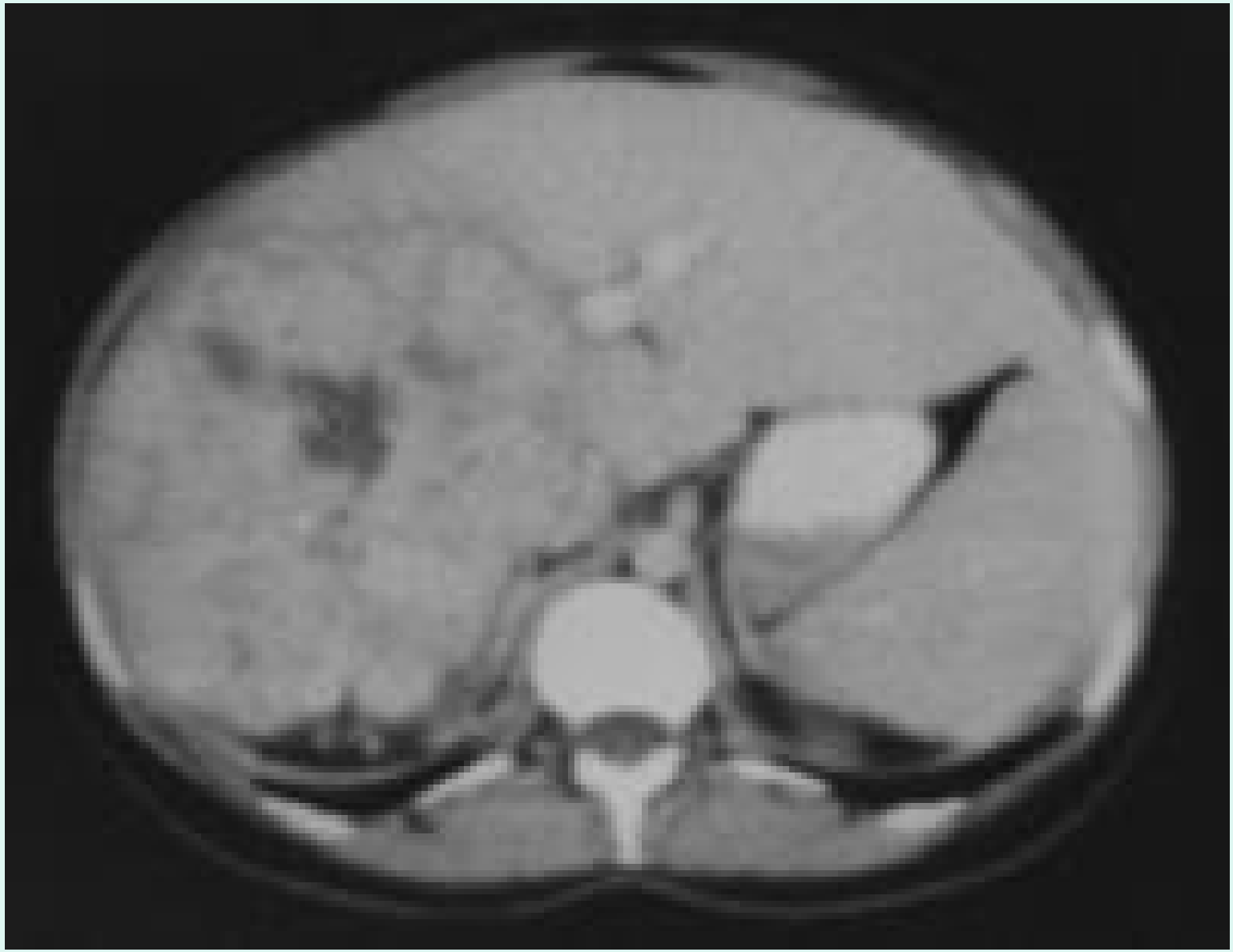
Fibrolamellar HCC

- A histologic variant of hepatocellular carcinoma ,It is associated with cirrhosis in less than 10% of patients and typically presents in a background of normal liver function and histologic architecture.

- Reported series indicate a patient age range of 5-69 years with a mean age of 23 years at the time of initial diagnosis.

Imagine diagnosis - FLC

- Abdominal CT is the preferred imaging method for the diagnosis, staging, and follow-up
- On nonenhanced scans, the primary fibrolamellar tumor typically appears as a large, solitary, hypoattenuating mass with well-circumscribed and lobulated margins.
- During the arterial-enhancing phase, the tumor is heterogeneously enhancing and becomes generally hyperattenuating with respect to the relatively less strongly enhancing surrounding liver.



Hepatoblastoma

- Hepatoblastoma usually affects children younger than 3 years, and the median age at diagnosis is 1 year.

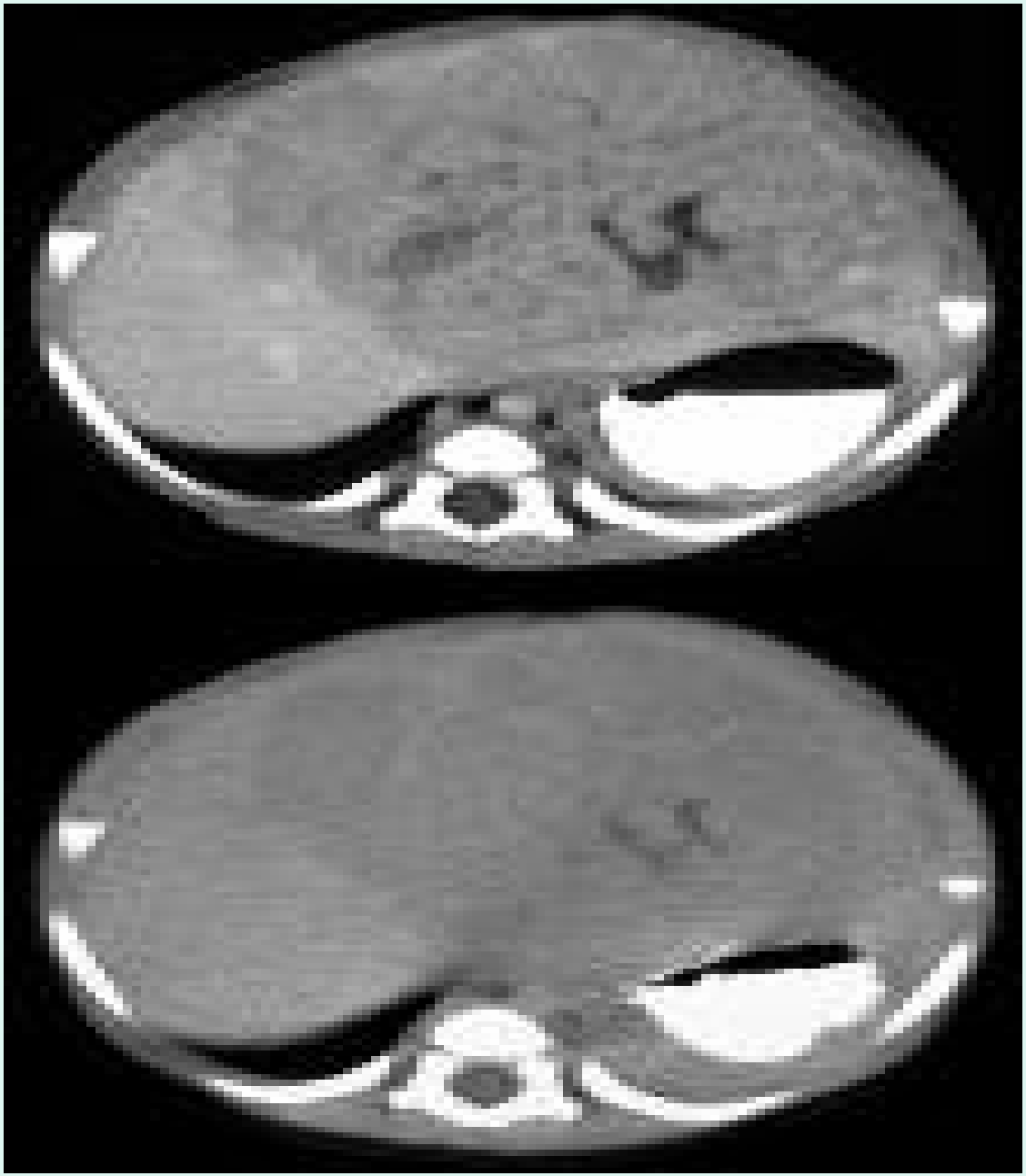
- Patients with hepatoblastoma usually are asymptomatic at diagnosis. Disease is advanced at diagnosis in approximately 40% of patients, and 20% have pulmonary metastases.

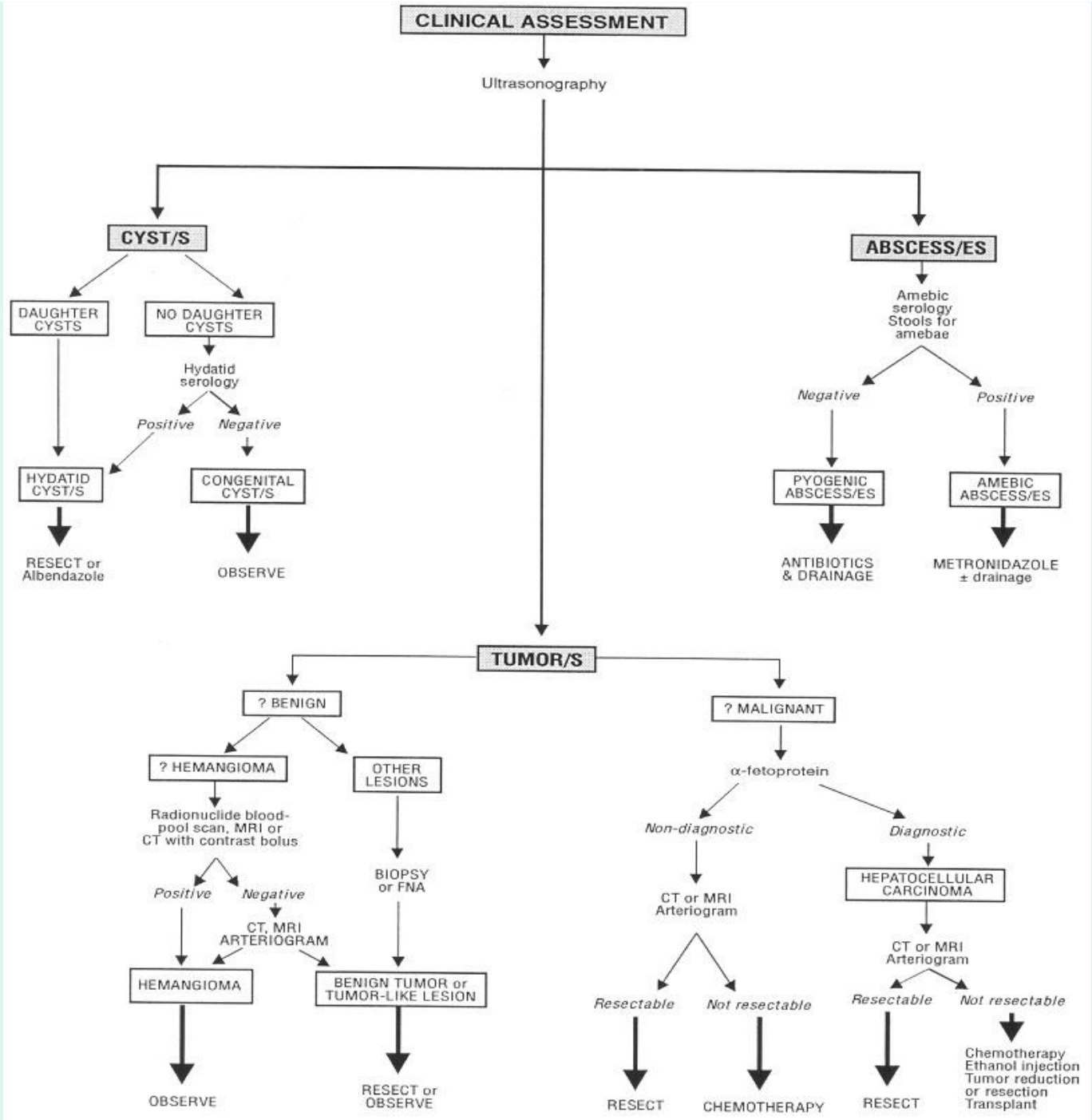
- AFP is found in high concentrations in fetal serum and in children with hepatoblastoma, hepatocellular carcinoma, germ cell tumors, or teratocarcinoma.

Imagine diagnosis - hepatoblastoma

- Prior to contrast administration, an epithelial-type tumor appears as a homogeneous hypodense mass, while a mixed mesenchymal-epithelial tumor demonstrates a more heterogeneous appearance. Calcifications may be present

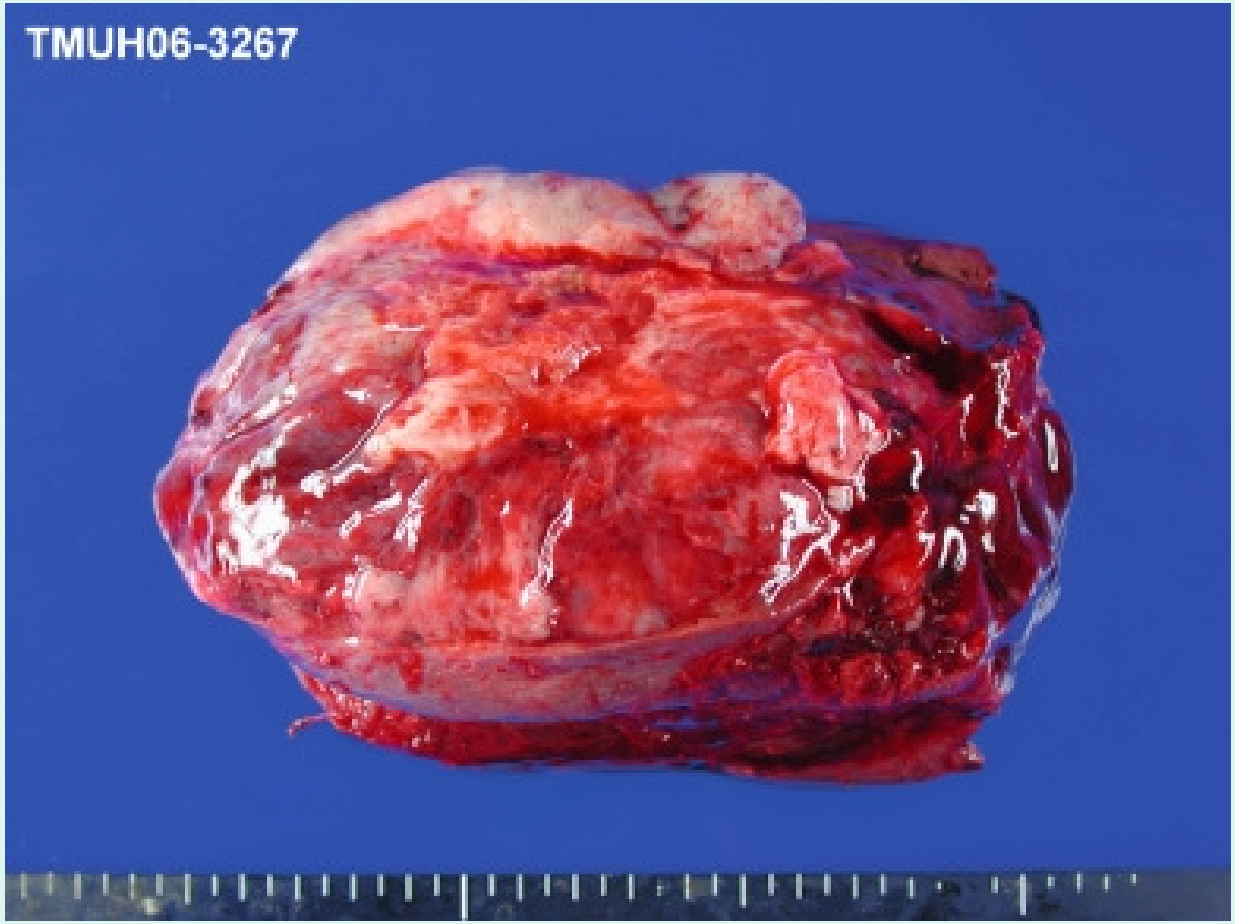
- The enhancement pattern typically is inhomogeneous, and a peripheral rim of enhancement may be observed if imaging is performed during the early arterial phase.

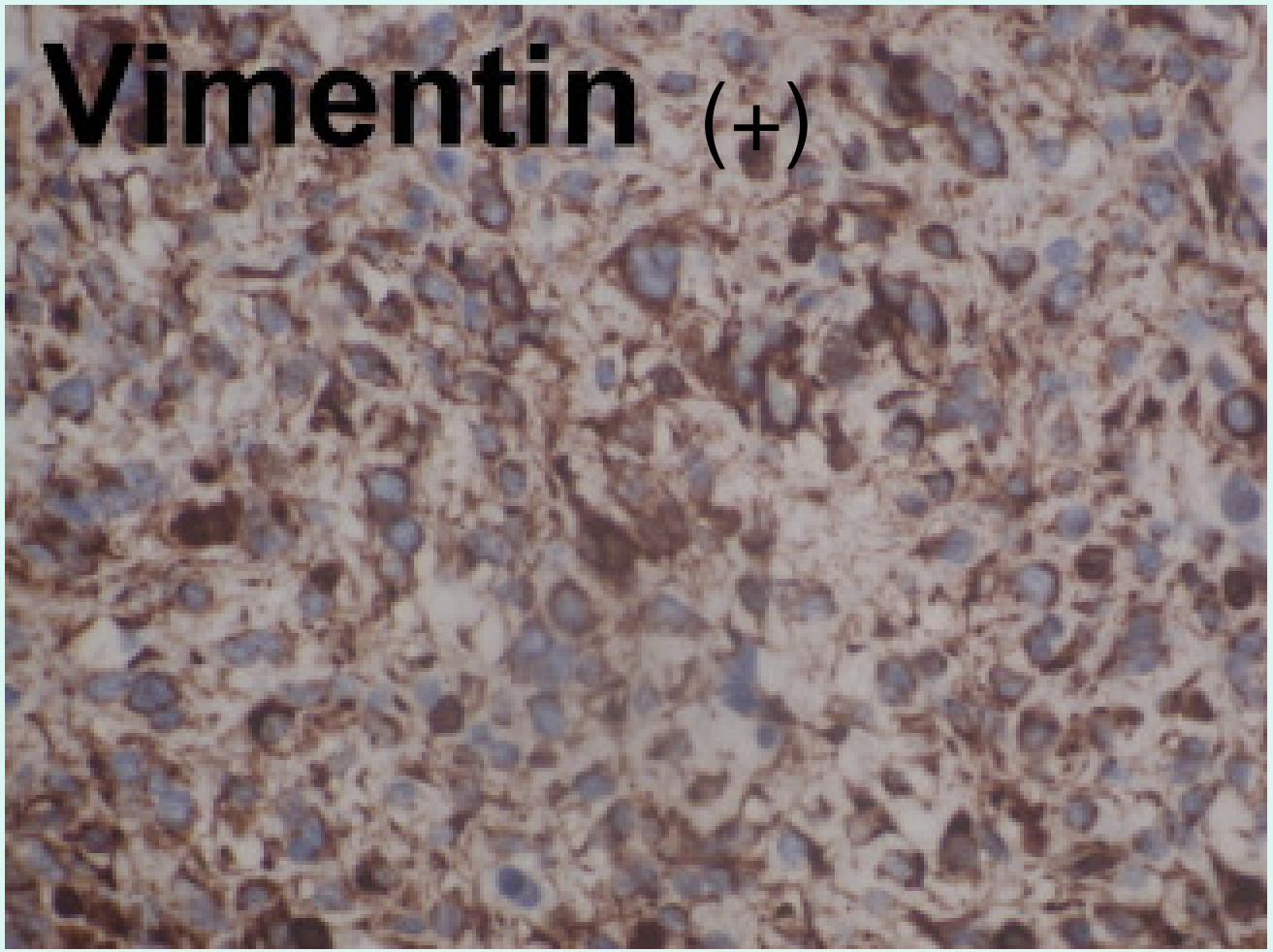


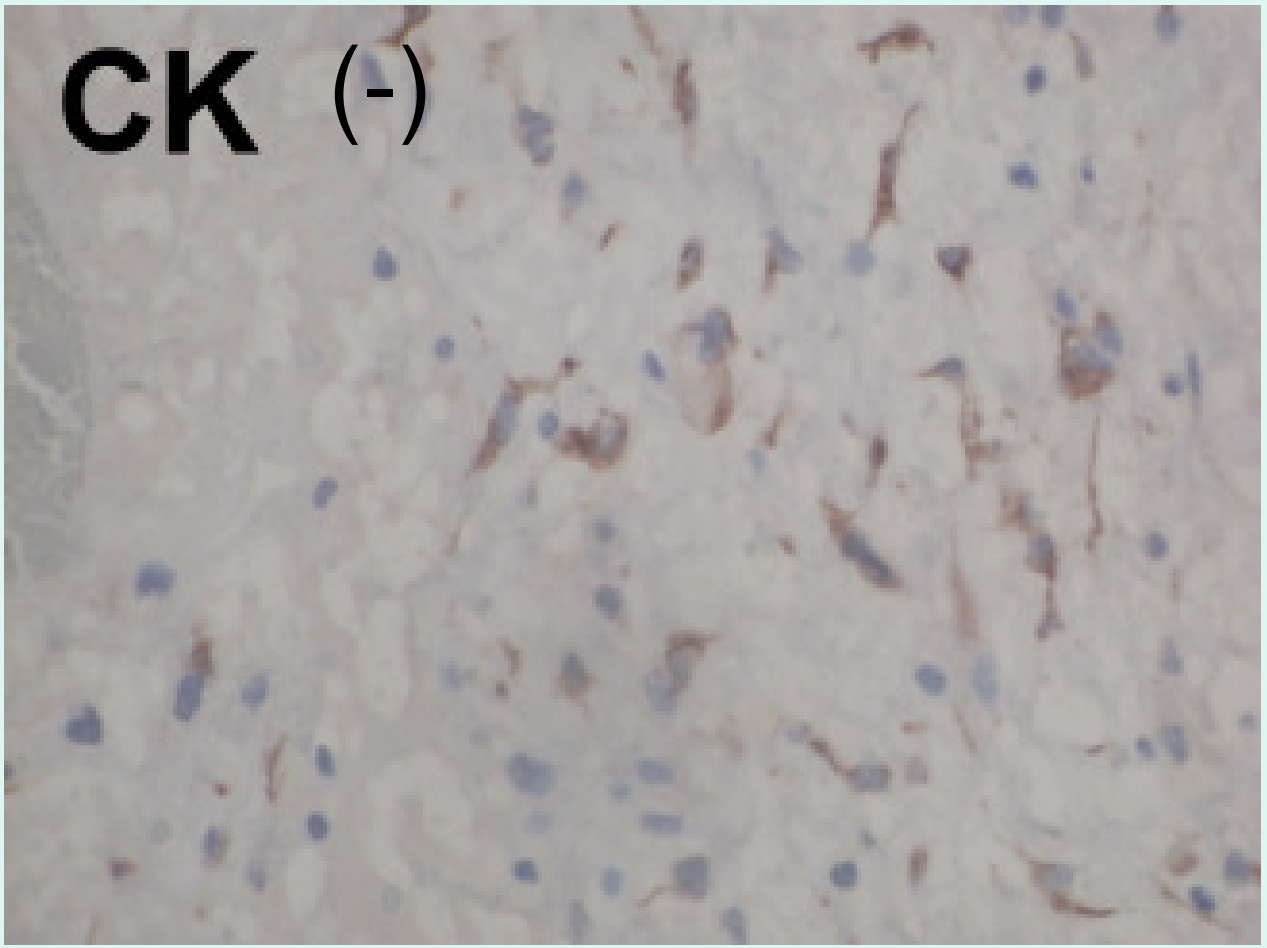

CLINICAL ASSESSMENT
Ultrasonography
CYST/S
DAUGHTER CYSTS
NO DAUGHTER CYSTS
Hydatid serology
Positive
Negative
HYDATID CYST/S
CONGENITAL CYST/S
RESECT or Albendazole
OBSERVE
ABSCESS/ES
Amebic serology Stools for amebae
Negative
Positive
PYOGENIC ABSCESS/ES
AMEBIC ABSCESS/ES
ANTIBIOTICS & DRAINAGE
METRONIDAZOLE ± drainage
TUMOR/S
? BENIGN
? MALIGNANT
? HEMANGIOMA
OTHER LESIONS
Radionuclide blood-pool scan, MRI or CT with contrast bolus
BIOPSY or FNA
Positive
Negative
CT, MRI ARTERIOGRAM
HEMANGIOMA
BENIGN TUMOR or TUMOR-LIKE LESION
OBSERVE
RESECT or OBSERVE
α-fetoprotein
Non-diagnostic
Diagnostic
CT or MRI Arteriogram
HEPATOCELLULAR CARCINOMA
CT or MRI Arteriogram
Resectable
Not resectable
Resectable
Not resectable
RESECT
CHEMOTHERAPY
RESECT
Chemotherapy
Ethanol injection
Tumor reduction or resection
Transplant

Surgical finding (95-4-26)

- well-defined tumor about 12x10x10 cm located at S4 ,S5,S6 segment, hard in consistency with yellowish color , without capsule
- Dissection between liver and vessel (IVC and portal vein) segmentectomy partial S4 and total S5,S6 with cholecystectomy
- minimal ascites


TMUH06-3267


Vimentin (+)


CK (-)

Pathology finding

- Liver, segment 4, 5, 6, segmentectomy, undifferentiated sarcoma ,Gallbladder, cholecystectomy, no tumor involvement

Undifferentiated embryonal sarcoma

- First described as a clinicopathologic entity in 1978 , Before that mesenchymoma, primary sarcoma of the liver, fibromyxosarcoma and malignant mesenchymoma
- The fourth most common malignant tumors of the liver in the children
- Typically diagnosed after 6 years of age with a decline in incidence after 10 years of age and 50% of the patients were between the above ages

Clinical presentation

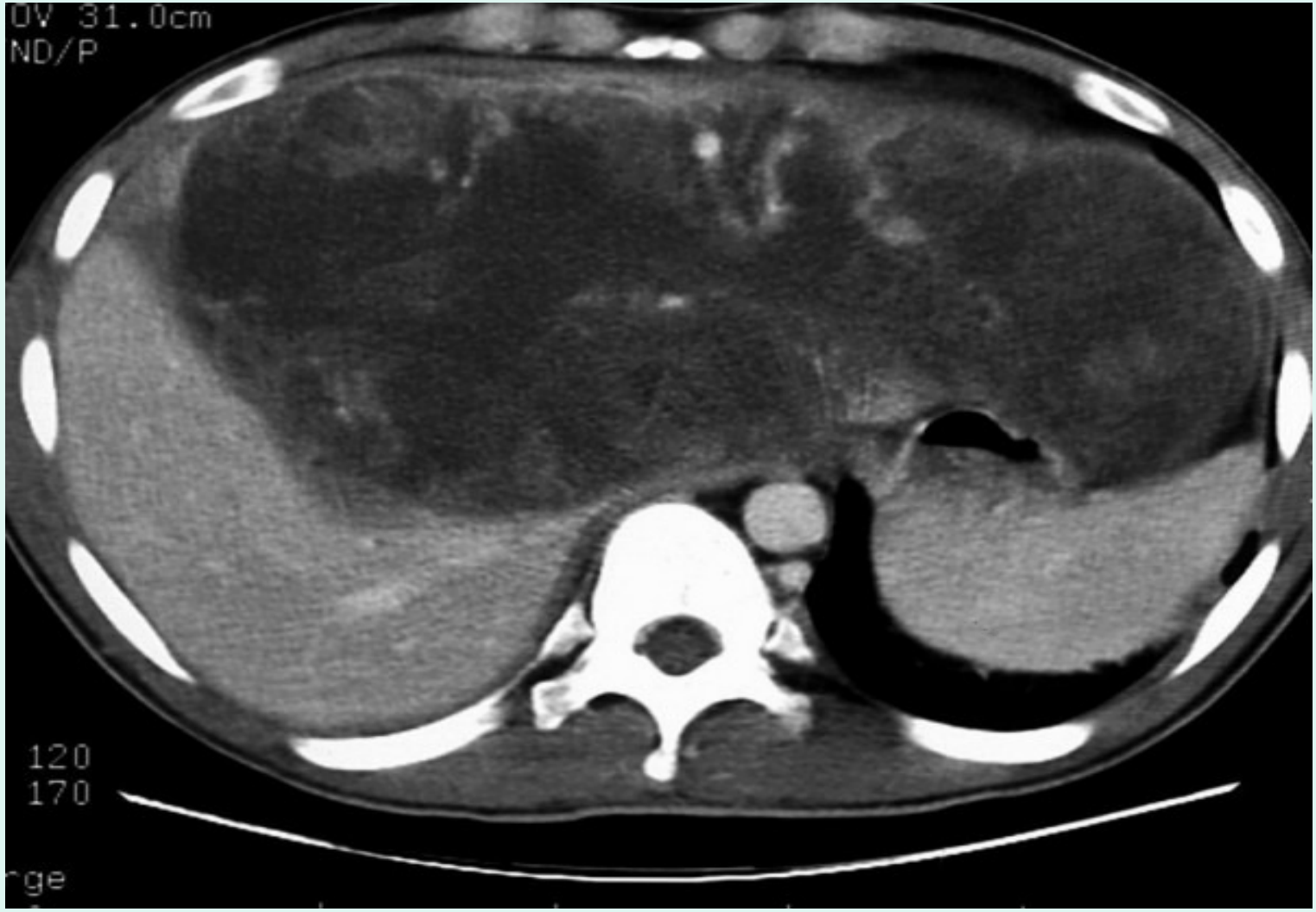
- Abdominal mass with or without upper abdominal pain or swelling

- Fever is probably related to the hemorrhage and necrosis found in the majority of these tumors , of possible significance is the absence of jaundice.

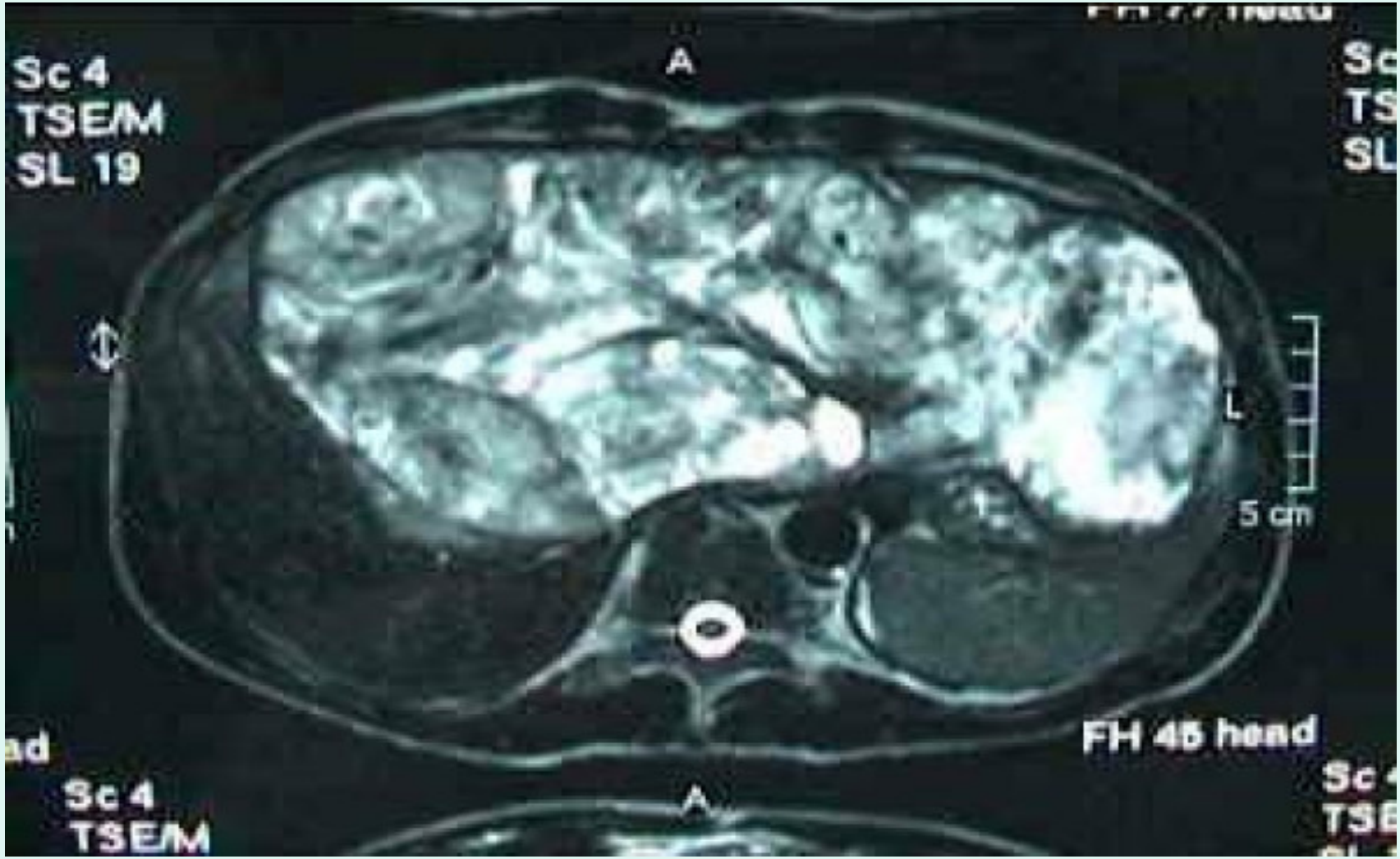
- UESL has no relation to hepatitis or liver cirrhosis, no disturbance of hepatic function. Laboratory studies are non-specific, and the α -fetoprotein is not increased

Imagine diagnosis

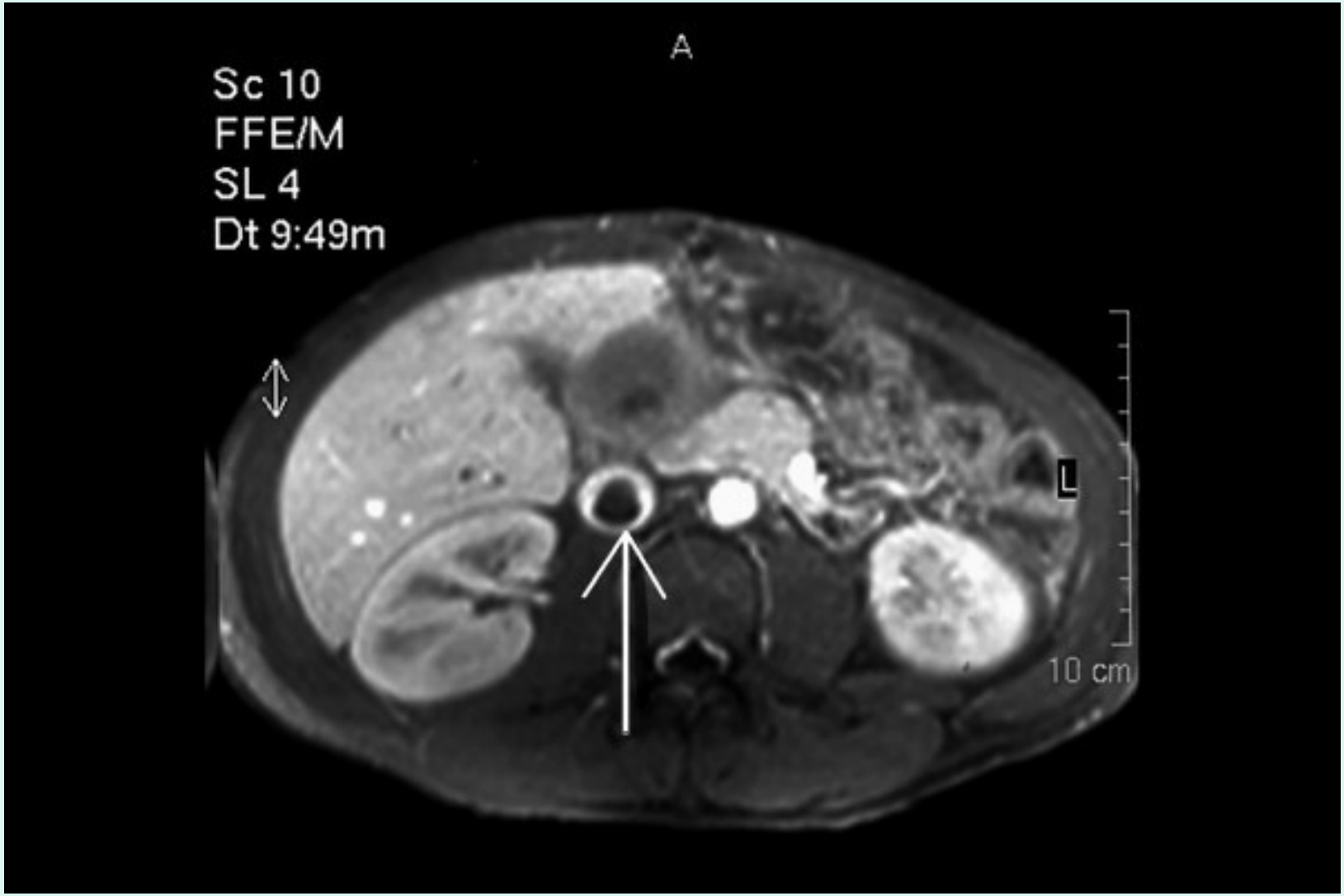
- Radiographs of the abdomen are usually normal
- The lesion can be detected by ultrasound, CT and MRI. MRI localizes the lesion more accurately than the other methods
- It also can detect vascular invasion, biliary obstruction and hilar adenopathies



CT of the abdomen demonstrating hepatomegaly and a large hypodense lesion of the liver with multicystic appearance with septations and solid portions.



MRI in T2-weighted and Short Time Inversion Recovery (STIR) sequences revealing hyperdense areas with intermixed hypodense septa



MRI revealing a thrombus in the inferior vena cava (the arrow shows the thrombus)

Pathologic diagnosis

- Tumor size often exceeds 10 cm and can be as large as 30 cm. single, well-demarcated, soft, globular mass that frequently has cystic, gelatinous, hemorrhagic and necrotic foci.

- Microscopic examination reveals a pseudocapsule surrounding a neoplasm, composed predominately of spindle, oval, or stellate cells with ill-defined cell borders.

- Abundant myxoid stroma that contains many thin-walled veins.

- Vimentin and the "histiocytic" determinants (alpha-1-antitrypsin and alpha-1-antichymotrypsin) have been the only consistent immunohistochemical markers expressed by this tumor.

Treatment

- Evaluable treatments included surgery, hepatic arterial ligation, hepatic transplantation, and combinations of surgery and/or chemotherapy and radiation therapy.

- Radical resection of the tumor is the optimal treatment of choice

- pre- and/or post-operative systemic chemotherapy (with cisplatin, andriamycin, cyclophosphamide) and/or radiotherapy, when necessary, can remarkably improve patient's survival

Prognosis

- The prognosis for UESL has been poor to until recently and majority of the patients died of tumor recurrence or metastasis within two years after the initial operation

- The major impediments in achieving long-term, disease-free intervals, is local recurrence in the upper abdomen and distant metastases

- Metastases to lung, pleura and peritoneum are common; invasion of the vena cava with extension into the right atrium rarely occurs

- The tumor does not produce any characteristic serum markers to permit monitoring of subclinical recurrences, the second-look laparotomy after the completion of chemotherapy should be considered