

A Thai Man with a Abdominal Palpable Mass for 5 Months

Chanunta Hongthanakorn, M.D.
Siwaporn Chainuvati, M.D.

ABSTRACT

GIST is an uncommon disease but it commonly metastases to liver. We reported a 42 year old Thai man who presented with enlargement abdominal palpable mass that had been observed for the previous 5 months. Diagnostic imaging revealed multiple varying size of liver cystic lesions and post contrast ring enhancement. There were thickened wall and contained fluid content. There were bowel mass under pancreas and Lt. renal vein. Both lesions had the same characteristic as liver lesions. Endoscopic examination revealed large polypoid ulcerative lesion about 3.5 cm in diameter at jejunum (100 cm from dental line). Histopathologic tissue presented multiple spindle cell and immunohistochemical stains for CD117 and S100 were positive. He was diagnosed as GIST with liver metastases. He received 400 mg per day of Imatidib treatment. After 5 months of treatment, his clinical conditions were improved and the liver lesions' size was decreased.

Key words : GIST, malignant cystic lesion, cystic liver metastases

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A 42 year-old married Thai man, an employee living in Chaipayum

Chief complaint: he had a abdominal palpable mass for 5 months

Present illness

He had been healthy prior to this illness.

5 months prior to admission (PTA), he had abdominal discomfort. He noted periodic abdominal pain and noted a large mass at right upper quadrant. He had no fever and no yellow eyes. He had no icterus ,no pale stool. He went to see a local doctor and under-

went liver aspiration. He was prescribed some medications without any clinical improvement.

2 months PTA, he experienced more right upper abdominal pain. He noted increase in size of abdominal mass. He had weight loss 5 kg in 2 months. He had no bowel habit change. He had no pitting edema at both legs.

2 weeks PTA, he had severe right upper abdominal pain then he came to Siriraj hospital.

Past History

He had been healthy

Personal History

He used to drink half a bottle of liqueur twice a week for 10 years

He quit smoking for 5 years

He denied any intravenous drug use.

Family History

His aunt was dead from liver cancer.

Physical examination

Vital sign: T 36.5 °c, BP 110/70 mmHg, P 80/min, RR 20/min

GA: good consciousness, hyposthenic build, mildly pale, no jaundice, no cyanosis, no edema, no sign of chronic liver disease

HEENT: hyperpigmented spot at angle of lips

CVS and RS: WNL

Abdomen: distended upper abdomen, bulging mass at RUQ 15 cm below Rt. costal margin, lobulated surface, cystic consistency, not tender, span 22 cm, no bruit, no shifting dullness, no fluid thrill, bowel sound normal

PR: no mass, no rectal shelf, no melena

NS: WNL

LN: not palpable

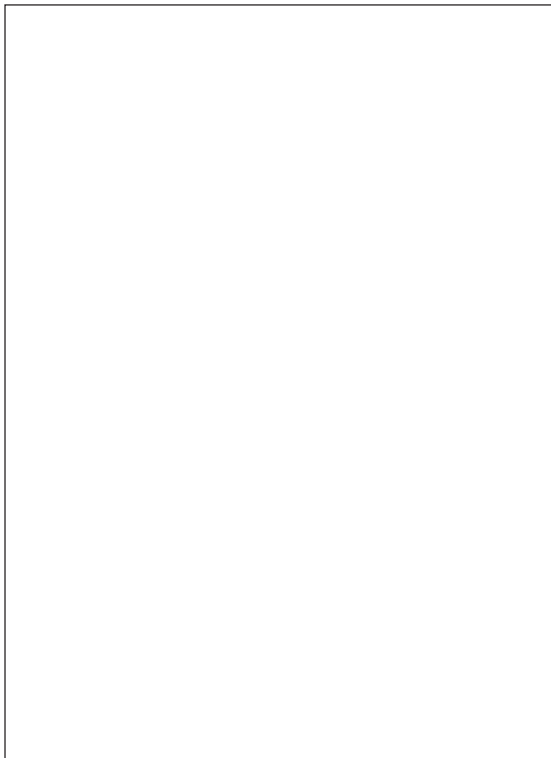


Figure 1 Liver ultrasonography showed multiple hypodensity areas at both lobe of liver (likely liver cyst)

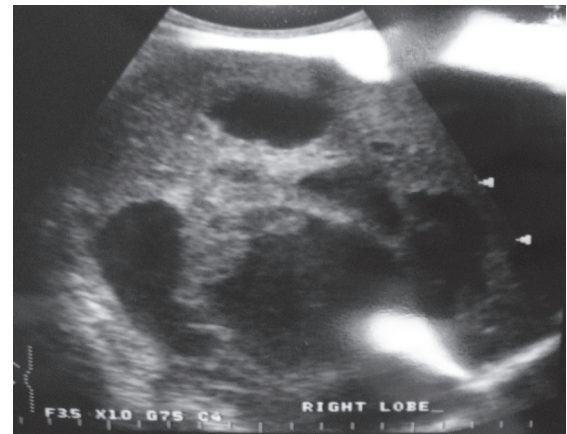


Figure 2

Investigations

CBC: Hb 6.3 gm/dl, Hct 20%, MCV 64 fL, WBC 5,700/uL (N 63%, L 28%, Mo 4%, E 4%, B 1%) Plt 439,000/uL

BUN: 16 mg/dl, Cr 0.7 mg/dl

LFT: TB 0.6 mg/dl, DB 0.3 mg/dl, AST 45 U/L, ALT 61 U/L, ALP 346 U/L (normal 39-117), GGT 558 U/L, Alb 3.4 g/dl, Glob 6.4 g/dl

U/S upper abdomen (Figure 1): showed marked hepatomegaly. There were multiple varying sizes of liver cystic lesions, some had septations and contained fluid content. There was no splenomegaly, no ascites and both kidneys were normal.

CT scan abdomen (Figure 3, 4): there were multiple varying sizes of liver cystic lesions with post con-

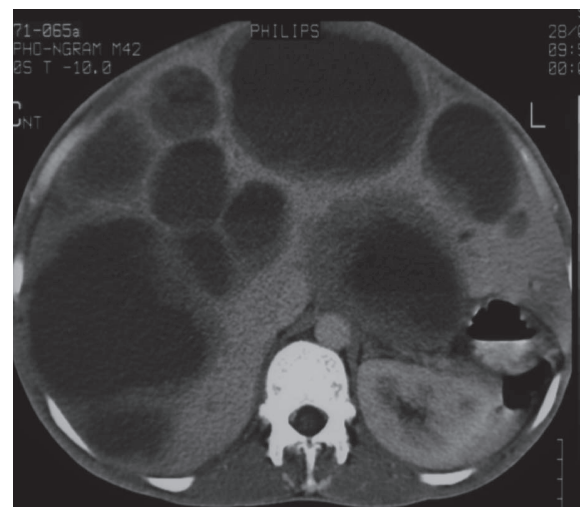


Figure 3 Contrast CT scan showed multiple varying sizes of liver cysts at both lobe, with thickened wall and fluid contained.

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trast ring enhancement. There were thickened wall and contained fluid content. There were bowel mass under pancreas and Lt. renal vein. It had the same characteristic as the lesion in the liver. There was no splenomegaly and ascites seen.

GI follow through showed the stomach was pliable without ulcer or thickened fold. Pylorospasm with dilatation of stomach was noted. Duodenal bulb showed deformity. Large polypoid mass was detected at proximal jejunum without evidence of bowel obstruction. Antegrade filling of contrast medium into large bowel within normal transit time was demonstrated.

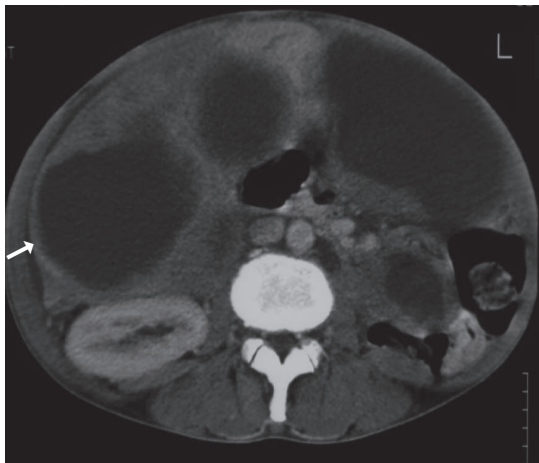


Figure 4 Contrast CT scan showed small bowel cystic mass lesion size 3.5 cm in diameter, thickened wall and some peripheral rim enhancement (arrow)

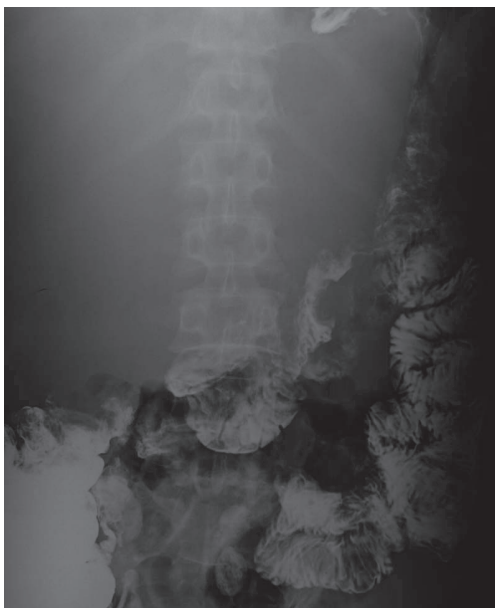


Figure 5 GI follow through showed irregular and narrowing lumen at proximal jejunum

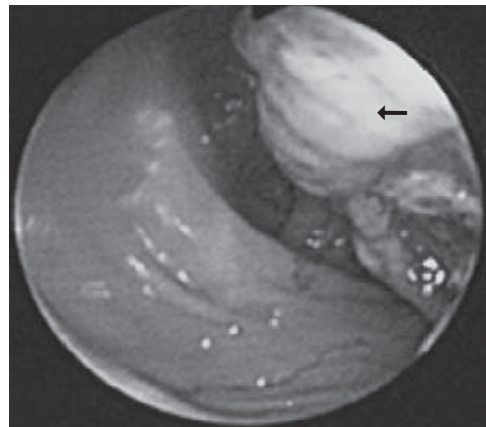


Figure 6 Endoscopic finding showed large polypoid ulcerative mass at proximal jejunum

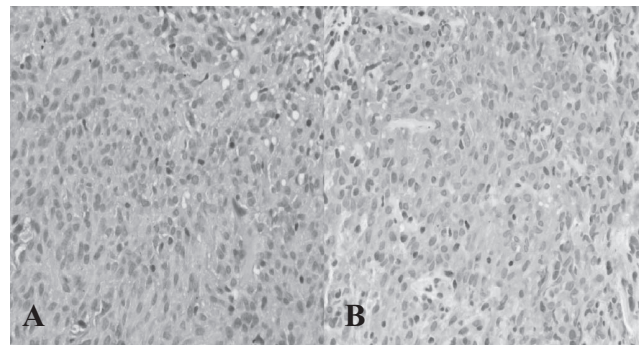


Figure 7 A: H&E stain showed multiple spindle cell
B: CD117 stain showed positive

Enteroscopy was done and revealed large polypoid ulcerative mass at proximal jejunum. (100 cm from dental line) Biopsy was done at the lesion. Pathohistologic finding showed multiple small spindle cells and immunohistochemistry stained for CD 117 and S100 were positive

DISCUSSION

A 42-year old man had presented with abdominal distention and physical examination revealed right upper abdominal cystic consistency mass. To further characterize an origin and nature of this mass, ultrasound was performed and showed multiple liver cystic lesions with contained-fluid.

Differential diagnosis of multiple liver cystic lesions (Table 1) in a middle aged man with 5 kg weight loss within 2 months are the malignant neoplasm, inflammation and to a lesser extent a benign neoplasm such as polycystic liver disease and hemangioma.

Table 1 CT and MRI imaging of cystic liver lesions.⁽⁷⁾

Type of lesion	Critical clinical features	Key CT and MR imaging features
Development		
Hepatic cyst	Asymptomatic, coexistence of renal cystic disease in ADPLD*	Homogeneous, round, regular, no wall no enhancement
Bile duct hamartoma	Asymptomatic	Homogeneous, rim enhancement, all lesions smaller than 1.5 cm in diameter
Caroli disease	Asymptomatic or symptoms of cholangitis	Cyst communication with the biliary tree, central dot sign, septa
Neoplastic		
Undifferentiated sarcoma	Occurs in older children and young adults, symptomatic	Large solitary lesion, enhancing solid components, calcifications
Biliary cystadenoma or cystadenocarcinoma	Occurs in middle-aged women, usually symptomatic	Multilocular, mural nodules, fibrous capsule, calcifications, variable signal intensities
Hepatocellular carcinoma	Liver cirrhosis, history of embolization	Hypervascular solid part, capsule, signs of cirrhosis
Cavernous hemangioma	Asymptomatic	Peripheral nodular enhancement, large lesion
Cystic metastases	Known primary tumor (colorectal, sarcoma, carcinoid, etc)	Rim enhancement, multiple
Metastases from ovarian cancer	Known ovarian carcinoma	Capsular implants
Inflammatory		
Abscess	Fever, sepsis	Presence of air, double target sign, enhancing wall
Hydatid cyst	Positive serologic results or clinical history	Calcifications, daughter cysts, pericyst
Miscellaneous		
Subcapsular pseudocyst	Pancreatitis	Occurs in left liver lobe, signs of pancreatitis, thin capsule
Intrahepatic hematoma	Liver trauma, known hepatocellular adenoma	Fluid attenuation at CT, methemoglobin at MR imaging, signs of trauma
Intrahepatic biloma	Liver trauma, biliary surgery	No capsule, no septa, no calcifications

*Autosomal dominant polycystic liver disease

Most liver cysts are found incidentally on imaging studies and tend to have a benign course. Cystic neoplasms are estimated to comprise approximately 5% of liver cysts.⁽¹⁾ Malignant cystic neoplasms of liver are primary liver lesions eg. cystadenocarcinoma, cystic type cholangiocarcinoma, and metastatic lesions. Inflammatory cause of liver is echinococcosis. Imaging in this case is most likely due to hepatic metastases because CT scan showed multiple cystic lesions with fluid inside and suspected cystic mass at jejunum.

Hepatic metastases are very common but a variety of nonspecific appearance have been reported.⁽²⁾ Most common metastases are solid but some have cystic appearance.⁽²⁾ Common cystic metastases include colon cancer, ovarian cancer, melanoma and carcinoid.⁽³⁾

History may be helpful. Two pathologic mechanisms can explain the cystic appearance. Firstly, hypervascular metastatic tumors with rapid growth may lead to necrosis and cystic degeneration.

This mechanism is found in metastases from neuroendocrine tumors, sarcoma, melanoma and subtypes of lung and breast cancer.^(2,4) Secondly, cystic metastases may be found in mucinous adenocarcinoma, such as colorectal or ovarian carcinoma.⁽³⁾

Ovarian metastases commonly spread by peritoneal seeding. CT of metastases is revealing primary hypo to isodensity lesions. Margins can be ill or well-defined, depending on their type and size. Calcifications are common with gastrointestinal, ovarian, breast and renal metastases.^(4,5)

CT imaging in this case demonstrated multiple hepatic cystic lesions and a small bowel cystic mass. Both lesions have contained fluid content. The primary lesion was thought to be from a small bowel with metastases to liver. We then proceeded enteroscopy and multiple biopsies were obtained from polypoid ulcerative lesion, it showed spindle cell infiltration which expressed CD117 suggestive of GIST. Contrast-CT scan of GIST which may be rim like or uniform, showed hyperdensity enhancing masses, although very large GIST may be less homogeneous due to necrotic, hemorrhagic, or degenerating components.^(8,9) GIST commonly metastasizes to liver and peritoneum, less frequently to lung, pleura and bone.⁽¹⁰⁾ This case is not typical for liver metastatic presentation because the lesions have contained-fluid content. In generally, this presentation can be observed in patient who received chemotherapy or contained intervention. The fluid are from necrosis or hemorrhage.⁽⁶⁾

Cystic type cholangiocarcinoma has a late onset about sixth decade. Symptoms are non-specific as abdominal pain, weight loss and jaundice. Ultrasound finding is a hyperechoic mass. Noncontrast CT finding is a large hypodense mass with irregular border. There is a characteristic of early rim enhancement with slow concentric filling. Delayed CT contrast enhancement of tumor is used to differentiate cholangiocarcinoma from HCC.^(4, 11, 12) The CT findings of liver lesions of this case was not typical for cholangiocarcinoma since they contained fluid and there was a soft tissue mass at small bowel which is suspicious for a primary lesion.

Cystic multiple septated hepatocellular carcinoma is a far less common form. HCC is the most common primary malignant liver neoplasm in Southeast Asia, Japan, and Italy.⁽⁴⁾ HCC is usually presented in sixth to seventh decade of life. Previous history may be cirrhosis, hepatitis B infection, hepatitis C infection, heavy alcohol drinking. Alpha-fetoprotein level may be increased. CT scan may present well-defined intrinsic tumor, such as hypervascularity of the solid part as, a capsule, with vascular or biliary invasion.^(7,13) Liver cirrhotic sign is demonstrated.⁽¹³⁾ This case did not have any features of HCC on imaging.

Biliary cystadenocarcinomas are rare less than 0.4% of liver neoplasm.^(14,15) It is a slow growing tumor, occurring in elderly, and has more equal sex distribution.⁽⁴⁾ Biliary cystadenocarcinoma is a malignant transformation of a cystadenoma and difficult to dif-

ferentiate from it. Biliary cystadenocarcinoma has slightly better prognosis compared to cholangiocarcinoma. Histopathology is helpful.⁽¹⁶⁾ Ultrasound finding typically appears as a hypoechoic lesion with thickened irregular wall and occasionally showed internal echoes representing debris and wall nodularity. CT scan finding is low attenuated mass, which may be uni- or multiloculated mass up to 30 cm in size with wall enhanced with contrast administration.⁽¹³⁾

Intrahepatic hydatid cysts are infected by ingestion of eggs of tapeworm, Echinococcal species, either by eating contaminated food, sheep or from contact with dogs.^(7,17) Endemic areas are in the Mediterranean basin, South and Central America, Middle east and other sheep-raising countries.^(17, 18) Right lobe is predominant 60-80% and more than 80% of cases have one cyst.⁽¹⁹⁾ Symptoms are variable and non specific such as nausea, vomiting, right upper quadrant pain, biliary obstruction. Biochemical analysis, usually stains eosinophilia, and a serologic test is positive about 25% of patients.⁽²⁰⁾ The most common ultrasound finding is an anechoic smooth, round cyst, which is difficult to distinguish from a benign cyst. When daughter cysts are present, characteristic internal septation results. Floating hydatid sand may be seen. CT scan usually appears as a well defined hypoattenuating cyst with a distinguishable wall.⁽²⁰⁾ This case had history of eating sheep 5 years ago but Echinococcal disease has very long incubation period. Other findings did not support this diagnosis.

CONCLUSION

GIST with multiple cystic liver metastases lesion is common, however, fluid-filled cystic lesions is rare presentation without any history of chemotherapy or intervention.

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