Computed tomography imaging of complicated hepatic hydatid cysts

Poster No.: C-0466
Congress: ECR 2016
Type: Educational Exhibit
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Keywords: Parasites, Cysts, Diagnostic procedure, CT, Liver, Emergency, Abdomen
DOI: 10.1594/ecr2016/C-0466

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Learning objectives

CT is the imaging modality of choice for the diagnosis of hepatic hydatid cyst complications. Accurate interpretation of CT examinations requires:

• To understand the pathophysiologic mechanisms of hepatic hydatid cyst complications.
• To be oriented by clinical manifestations.
• And to recognize the different CT aspects of each complication

Our objectives are:

• To illustrate the different aspects of hepatic hydatid cyst complications.
• To evaluate the role of spiral CT in the detection of these complications.
Background

Hydatid disease is a parasitic infection mainly caused by the tapeworm Echinococcus granulosus (1, 2).

Humans become infected by ingesting eggs from E. granulosus either by eating contaminated food or through contact with dogs. The ingested embryos invade the intestinal mucosal wall, enter the portal circulation and develop into a cyst in the liver. Humans are intermediate hosts. In the liver, cysts grow to 1 cm during the first six months and approximately 2 - 3 cm annually thereafter, depending on the host tissue resistance (3).

Hydatid disease is a major endemic health problem in mediteranean region, and specifically in Tunisia (2).

The liver is the most commonly affected organ with an infestation rate of 60 - 75%. Specifically, the right hepatic lobe is affected in 80% of cases and the left lobe in 20% (4).

The hydatid cyst is composed of 3 layers (4):

- The outer layer is the pericyst, an avascular layer derived from modified host tissue and inflammatory cells.
- The middle layer is the laminated acellular membrane.
- The inner layer is the germinal layer which produces the laminated membrane and scolices.
- The 2 last membranes form the endocyst.

Natural history of the hepatic hydatid cyst can be divided into two phases (1):

The first phase is that of growth during which rupture can occur when the pressure of the hydatid liquid becomes more important than resistance of the hydatid wall (pericyst). Finally, the complications such as infection, acute allergic manifestations, jaundice are only the consequences of the rupture of the cyst wall.

The second phase is a phase of ageing and of progressive involution. It is the consequence of the overproduction of scolices and daughter cysts. During this phase, the hydatid cyst will be full of scolices and membranes which replace the hydatid liquid. Calcifications occur in the pericyst.

Hydatid disease is mostly asymptomatic and many hydatid cysts represent incidental clinical and radiological findings (4). Most symptomatic cysts are due to their complication.
Echinococcal cysts of the liver can cause complications in about 5 to 40% of cases. Some of them, in particular, can be fatal without early diagnosis and appropriate treatment (1).

Complications include two main categories (4):

- Rupture of the hydatid cysts more frequent, occurring in 20 - 50% of cases
- Secondary bacterial infection appearing in only 5 - 8% of cases.

Three types of rupture have been described (4, 5, 6):

- Contained.
- Communicating.
- And direct.

Contained rupture occurs when the endocyst ruptures and hydatid fluid escapes into the space between the pericyst and the endocyst resulting in the collapse of the endocyst. This collapse of the endocyst can be recognised on CT which shows detached undulating membranes inside the hydatid cyst without the reduction in the cyst size (« water lilly or serpent sign »)

Communicating rupture is the most common type of rupture, appearing in 44 - 64% of cases and it occurs when the content of hydatid cyst evacuate into the biliary radicles that have been incorporated by the pericyst. It is subdivided into two groups depending on the size of communication:

In the first subtype, which is more frequent, the communication concerns in small biliary radicles and it is established by small fissures or bile-cyst fistulas.

The second subtype is characterized by a wide perforation of the hydatid cyst into a main biliary radicle.

In both cases, hydatid fluid, hydatid sand and occasionally daughter cysts can be discharged into the biliary tree.

Direct rupture occurs when both the endocyst and the pericyst are torn so that the content of the cyst escapes into the peritoneal cavity, the thoracic cavity through the diaphragm, the mediastinum or occasionally a hollow organ such as the gastro-intestinal tract. It may occur spontaneously or result from trauma or surgery.

Secondary bacterial infection of the hepatic hydatid cyst is a rare complication due to avascularity of the pericyst and the lack of connection between the endocyst and the host.
vascular system. Essential prerequisite for the bacterial infection of the cyst is the rupture of both pericyst and endocyst, so bacteria can enter the cyst (4).
Findings and procedure details

In retrospective review (from January 2012 to August 2015), 27 patients with complications of hepatic hydatid cysts were enrolled.

There were 14 women and 13 men (sex-ratio: 0.9), ranging in age from 23 to 81 years (mean: 43.8 years).

Thirteen of them had a known medical history of hydatid disease, while 14 of them presented to our department as emergency cases.

The complaints at presentation were:

- Cholangitis in 14 patients.
- Sepsis in 4 patients.
- Upper abdominal pain in 9 patients.

All patients underwent CT examinations with high-resolution scanner before and after the administration of contrast agent, which revealed the following complications:

Rupture of the hepatic hydatid cyst in 21 patients (77.7%):

- into the biliary tree in 16 patients (59.2%).
- into the peritoneum cavity in 3 patients (11.1%).
- into both biliary tract and gastro-intestinal tract in 1 patient (3.7%).
- into the pleural cavity in 1 patient (3.7%).

Secondary infection of the cyst in 2 patients (7.4%)

Mass effect in 4 patients (14.8%):

- on bile tract in 1 patient (3.7%).
- on vessels in 2 patients (7.4%).
- on pancreas in 1 patient (3.7%).

*Fig. 1 on page 14 Fig. 2 on page 14 Fig. 3 on page 15 Fig. 4 on page 15*

Intrabiliary rupture of hepatic hydatid cysts is the most common complication with a reported incidence of 3 - 17% (59.2% of all complications in our study) (4, 7).
Communicating rupture of a cyst into the biliary system may occur through small fissures or bile-cyst fistulas (55% of cases) or through a wide perforation that allows access to a main biliary branch (5).

Rupture occurs mainly into the right hepatic ducts (55 - 60% of cases) and less frequently into the left ducts (25 - 30%) and the confluence or gall bladder (1, 4).

A rapid and accurate pre-operative diagnosis is essential. CT is an imaging method which provides such diagnosis based on characteristic findings (4, 5, 6):

- Discontinuity of the calcified or not hydatid cyst wall.
- Change in architecture of the cyst.
- Dilatation of the biliary tree with air or air-fluid level inside the cyst.
- Dilatation of the biliary tree with air or containing high density linear structures representing hydatid material.

However, simple dilatation of the biliary tree does not always indicate cyst rupture. It may result from direct compression of the biliary branches by the cyst.

The only direct sign of rupture into the biliary tree is the visualization of the cyst wall defect or of a communication between the cyst and a biliary radicle.

In cases of wide communication, CT demonstrates rupture in 77% of cases (5).

*Rupture of the hepatic hydatid cyst into the peritoneum cavity* Fig. 5 on page 16
The reported frequency of liver hydatid cyst rupture into the peritoneal cavity ranges from 1 to 16% (11.1% of all complications in our study) (8).

Rupture may result from trauma or may occur spontaneously from increased pressure of the cyst fluid (1).

The main risk factors predisposing to rupture include young age (because of traumatic events and higher prevalence of hydatid disease in children and adolescents than in adults), cyst diameter > 10 cm and superficial cyst location (8).

If the rupture is insidious, the release of brood capsules, scolices and even daughter cysts from a ruptured hydatid cyst into the peritoneal cavity leads to multiple cysts in the peritoneal cavity. This phenomenon is called « secondary echinococcosis » (1).

Rupture into the peritoneum cavity may present as acute abdominal pain. Antigenic fluid released into the peritoneal cavity and absorbed into the circulation may present with acute allergic manifestation (1).
Although the reported frequency of minor allergic reactions after rupture of the hydatid cysts ranges from 16.7 to 25%, the incidence of more severe reactions is 1 to 12.5% (8).

Abdominal pain, nausea, vomiting and urticaria are the most common symptoms (1, 9).

But in some cases, if the hydatid cyst contains bile due to associated rupture in biliary tree, the patient will present peritonitis or even hydatid choleperitonitis (1).

So, although rare, the rupture of hepatic hydatid cyst into the peritoneum cavity can be fatal and the diagnosis should be prompt because it requires emergency intervention.

CT is the imaging modality of choice which allows imaging of the entire abdominal cavity and pelvis and which provides the diagnosis based on specific findings (4, 5, 8):

Detached membranes, reduction of the cyst size, cyst wall discontinuity and change in the architecture of the hydatid cyst are the general imaging findings of rupture.

Specifically, CT shows the presence of daughter cysts inside the peritoneum cavity.

In case of peritonitis, apart from the imaging findings of rupture, fluid effusions in the peritoneum cavity, dilatation of mesenteric vessels and thickening of the peritoneum may also be detected on CT.

Finally, as a complication of direct rupture of hepatic hydatid cysts into the peritoneum cavity, CT shows the implantation of scolices in several organs leading to a condition called « metastatic hydatidosis ».

Rupture of the hepatic hydatid cyst into the gastro-intestinal tract Fig. 6 on page 16
The rupture of hepatic hydatid cyst into hollow viscus is an extremely rare complication with an estimated frequency of 0.5% and the most common site of perforation is into the stomach (5).

Only 0.29% of the operated hepatic hydatidosis were perforated into the gastro-intestinal tract (10).

The communication between hydatid cyst and digestive tract occurs when the cyst is located on the inferior surface of the liver. In these cases, the stomach and the duodenum may be affected too.

The clinical manifestations are various (5, 10):

Non specific signs as abdominal disconfort or pain, dyspepsia or fever

Hydatidorrhea or hydatidenteria (passage of hydatid membranes in the stools)

Hydatidemesis (presence of hydatid cyst as well as membranes in the vomit)

CT scan can be very useful for the diagnosis and can show (5, 10):
A cyst with an air-fluid level

An orally administered contrast material inside the cavity which is a close contact with the digestive tract

**Transdiaphragmatic rupture of the hepatic hydatid cysts** *Fig. 7 on page 17*

The transdiaphragmatic intrathoracic rupture of the hepatic hydatid cyst is a complication which occurs in 0.6 - 16% of patients with hepatic hydatid disease (4, 5).

Several factors, such as pressure gradient between thoracic and abdominal cavities, mechanical compression and ischemia of the diaphragm, sepsis in the hepatic cyst or chemical erosion by bile, participate in promoting intrathoracic evolution of hydatid cysts of the hepatic dome (1).

The bare area of the liver, specifically the posterior segments of the right hepatic lobe, is the most common route of transdiaphragmatic migration of hydatid material due to the lack of peritoneal covering in this area (4).

Surgical classification into 5 progressive stages has been proposed by R. Gomez et al. according to the degree of evolution of the diaphragmatic or thoracic (pleuropulmonary) involvement (11):

- **Grade 1** (adhered cyst): there are firm adherences between the surface of the cyst and the diaphragm but no diaphragmatic perforation
- **Grade 2** (hydatidic transit): cyst perforates the diaphragm, but there is little invasion of the thoracic cavity.
- **Grade 3** (pleurothoracicvesiculation- cyst in « sandglass »): cyst perforates the diaphragm and either grows inside the thoracic cavity or daughter vesicles become established.
- **Grade 4** (disease of the pulmonary parenchyma): cyst connects to the bronchial branches or there is simply compression and atelectasis of the pulmonary parenchyma.
- **Grade 5** (chronic bronchial fistula): this stage is the chronic aftermath stage, either postoperative or as a result of the spontaneous evolution of hydatid disease.

The clinical presentation is predominately pulmonary with abdominal symptoms being less frequent (1, 12):

Coughs, expectoration and dyspnea are present in 30% of cases.
A bronchobiliary fistula leads to hemoptysis and cyst expectoration

The diagnosis is performed with thoraco-abdominal CT scan which can show (4, 13):

- Pleural effusions with or without the characteristic image of a parasitic membrane floating on the pleural fluid or a pyopneumothorax particularly if the rupture into the pleura occurred after rupture into the bronches.
- Lung consolidation andatelectasis.
- The presence of air inside the hydatid cyst, strongly indicative of communication with the bronchial tree.
- The CT scan, in addition, may show a tear like connection between the abdominal and thoracic cavities, particularly on sagittal or coronal reconstruction.

**Secondary infection of the hepatic hydatid cyst** Fig. 8 on page 17

It occurs in 5 - 8% of cases (5).

Secondary bacterial infection of the hepatic hydatid cyst is due to the avascularity of the pericyst and the lack of connection between the endocyst and the host vascular system (4).

It occurs only after rupture of both the pericyst and endocyst (communicating and direct rupture), which allows bacteria to pass easily into the cyst (4, 5).

At clinical examination, infection usually manifests as a hepatic abscess with pain in the right hypochondrium, hepatomegaly and fever (1, 5).

CT is the preferred method for identifying cyst infection which may manifest as (4, 5, 14):

- Poorly defined masses, in contrast to the more clearly defined masses seen in uncomplicated cases.
- Gas or air-fluid levels within the cyst.
- Contrast-enhanced CT may reveal the typical high attenuation rim representing abscesses surrounding the lesion.
- Occasionally, patchy areas of contrast-enhanced liver parenchyma are seen in the vicinity of the lesion representing inflammatory changes.

**Mass effect on neighboring structures and organs**
This complication mainly occurs when the hepatic hydatid cysts reach large size due to an active growth phase (3).

Large hepatic hydatid cysts can compress neighboring tissue and cause mass effect.

Compression can concern vessels, biliary ducts or neighboring organs (3, 5, 15).

**Compression of the portal vein** with portal hypertension and thrombosis with secondary cavernomatosis and collateral circulation are rare and usually caused by cysts located in the caudate lobe and hepatic bifurcation.

Direct invasion by hydatid cyst contents is very unusual complication.

**Compression of the inferior veina cava or the hepatic veins** can be responsible of Budd-Chiari syndrome.

The CT scan can show ascite, hepatomegaly, hepatic heterogeneous enhancement, unopacified hepatic veins with or without tortuous intrahepatic collaterals.

**Large cysts in the hepatic parenchyma can cause biliary duct dilatation by either compression of a nearby duct by mass effect or by perforation into biliary ducts.**

The compression of nearby biliary ducts can cause microerosion within the bile duct wall and fistula formation.

**Compression can also concern other neighboring organs such as pancreas.**

Clinical presentation is variable depending on the location of compression.

Obstructive jaundice is caused by compression of the head of the pancreas and the common bile duct.

Compression of the body or tail of the pancreas is usually symptomless and CT scan can show a Wirsung duct dilatation.
**Fig 1:** 35 year-old woman presented with a right upper abdominal pain without fever nor jaundice. Enhanced CT in axial (a), coronal (b) and sagittal (c) plans shows an irregular cystic lesion in the right lobe of the liver (→) with dilatation of right intra hepatic ducts (↔) that indicates **intraduct rupture.**

**Fig. 1**

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**Fig 2:** 50 year-old woman presented with an upper abdominal pain. Enhanced CT shows discontinuity of the calcified cyst wall in the segment V of liver (→) with a dilatation of neighboring intrahepatic ducts (↔) that indicates **the rupture of the hepatic hydatid cyst into the biliary tree.**

**Fig. 2**
Fig. 3: 45 year-old woman presented with cholangitis. Enhanced CT shows three multivesicular hydatid cysts in segment IV of the liver, partially collapsed (→) with a communication with the biliary tree (→) which is is dilated (→) indicating the intrabiliary rupture of the hydatid cysts.

Fig. 4: 45 year-old man presented with cholangitis. Enhanced CT shows hydatid cyst in segment VI of the liver, partially collapsed, communicating with the biliary tree (→) and a dilatation of intra (→) and extra (→) hepatic biliary ducts indicating the intrabiliary rupture of the hydatid cyst.
**Fig. 5:** 35 year-old man presented with acute abdominal pain after road accident. Enhanced CT shows cyst wall discontinuity and changes in the architecture of the hydatid cyst (→) with a communication (→) and effusion (→) in hepatic subcapsular space that indicates the **intra peritoneal rupture of the hepatic hydatid cyst.**

**Fig. 6:** 60 year-old man presented with upper abdominal pain with fever. Enhanced CT shows the fistula (→) between the pylorus and the hepatic hydatid cyst containing air inside (→) that indicates the **rupture of the hydatid cyst into the stomach.**
Fig. 7: A 45-year-old man presented with thoracic pain and fever. Enhanced CT shows the transdiaphragmatic intrathoracic rupture of the hepatic hydatid cyst (→). The presence of air inside the hepatic cyst (→) is strongly indicative of communication with the bronchial tree.

Fig. 8: A 70-year-old man presented with a sepsis. Enhanced CT shows discontinuity of the calcified cyst wall (→) in the segment V of the liver containing air inside (→) that indicates secondary infection of the hydatid cyst.
Fig. 9: 62 year-old woman presented with an abdominal discomfort. Enhanced CT shows a voluminous hepatic hydatid cyst compressing the portal vein (→) which is laminated and its right branch (→) without signs of portal hypertension.

Fig. 10: 27 year-old man presented with un upper abdominal pain. Enhanced CT shows a well defined hydatid cyst in segment IV of the liver compressing the biliary convergence (→) with a discreet dilatation of biliary ducts (→).
Fig. 11: 40 year-old man presented with acute abdominal pain. Enhanced CT shows a voluminous hepatic hydatid cyst **compressing the body and the tail of pancreas** with a Wirsung dilatation upstream (→). The cyst compress also the **portal vein** (→) with portal hypertension and collateral circulation (→).

**Fig. 11**
Conclusion

Hydatid disease commonly affects the liver and it has many potential complications.

Some of them, in particular, can prove to be life-threatening and therefore should be early and accurately recognized.

CT is the imaging modality of choice for the diagnosis of different hepatic hydatid cysts complications and specific imaging findings, relating to each complication, allow definitive diagnosis and prompts operative intervention.
References


