Imaging Spectrum of Simple and Complicated Hydatid Disease

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Learning objectives

- To assess the imaging features of simple and complicated hydatid disease in different anatomic locations.
- To illustrate an imaging guide of the hydatid cyst in correlation with the development course of the parasite.

Background

Definition. Echinococcosis is a parasitic disease caused by larval forms of the genus *Echinococcus* found in the small intestine of carnivores. Two closely related species of cestode are pathogenic for humans: the most frequently *E. granulosus*, the causative agent of cystic hydatid disease (or Cystic echinococcosis, CE), and the less common, though more invasive, *E. multilocularis*, which causes Alveolar echinococcosis (AE). [1]

Incidence. Hydatidosis is a worldwide zoonosis, occurring predominantly in countries of South and Central America, African part of the Mediterranean area, the Middle East and some sub-Saharan countries, Russia and China, and a well-known phenomenon in Europe, with a high prevalence of *E. granulosus* infection extending from the Iberian Peninsula to the Balkans. Reports from Bulgaria, Corsica, southern France, Italy, Greece, Portugal, Spain and Romania have highlighted the hyperendemic nature of the disease. [2]

Etiopathogeny. *E. granulosus* requires two hosts for completion of their life cycle (Fig. 1 on page 4). Free eggs are passed in the feces of the definitive host (the dog, hyena or dingo for the *E. granulosus*), being ingested by an intermediate host, in which the metacestode stage and protoscoleces develop. The cycle is completed if the metacestode and protoscoleces are eaten by a suitable carnivore. Although the natural intermediate carriers are sheep, deer or cattle, humans may accidentally become intermediate hosts through contact with a definitive host (usually a domesticated dog) or ingestion of contaminated water or vegetables. The larval tapeworms reach portal circulation through the intestinal wall to form the hydatid cyst, establishing the liver as the most common organ affected (75%), while the remaining can reach almost any organ through secondary hematogenous dissemination, as a complication after spontaneous, trauma-induced cyst rupture or during invasive treatment procedures. [3]

The histology of a typical hydatid cyst demonstrates three layers: (1) an outer fibrous capsule (pericyst) composed of modified host cells that form a rigid protective zone, (2) a laminated acellular membrane (ectocyst) that allows passage of nutrients and is easily
ruptured, which predisposes to infection, and (3) an inner germinal layer (endocyst) where scolices (larval stage) and laminated membrane are produced. The thickness of these layers depends on the tissue in which the cyst is located. The layers tend to be thick in the liver, less developed in muscle, absent in bone, and sometimes visible in the brain. Cyst fluid is clear, has a neutral pH, may contain scolices and hooklets, and is antigenic, being able to cause eosinophilia or anaphylaxis. Daughter vesicles are small spheres that contain the protoscolices. When vesicles rupture within the cyst, scolices pass into the cyst fluid and form a white sediment known as hydatid sand. [4]

**Diagnosis.** Diagnosing the hydatid infection is complex and requires multiple paraclinical, laboratory investigations and imaging techniques (X-ray, ultrasound, CT and MRI), which usually confirm the clinical suspicion.

**Imaging procedures.** The imaging appearance of the hydatid cyst is various and it depends on its evolutive stage.

Sonography is the most sensitive technique for the detection of membranes, septa, and hydatid sand within the cyst.

The WHO standardized classification of *Echinococcus granulosus* based on ultrasonographic images comprises 5 stages within the normal development course of the parasite [6]:

- an initial undifferentiated cystic lesion (CL) stage - unilocular, simple cyst, without visible wall;

- cystic *echinococcosis* (CE) stage 1- (active) unilocular, fluid-filled, cyst-like structure with internal hydatid sand and a visible wall;

- cystic *echinococcosis* (CE) stage 2 - (active) multilocular fertile cyst with daughter cysts and/or matrix develop;

- cystic *echinococcosis* (CE) stage 3 - (transitional) degenerative cyst, with heterogeneous content and detached laminated membrane from the cyst wall;

- cystic *echinococcosis* (CE) stage 4 - (transitional) solid content heterogeneous cyst;

- cystic *echinococcosis* (CE) stage 5 - (inactive) thick calcified wall cyst.

Computed tomography or magnetic resonance are particularly useful in extra abdominal disease, in obese patients, in case of complicated cysts, demonstrating cyst wall defects and the passage of contents through a defect, as well as in planning of surgery or interventional therapy. Computed tomography best demonstrates cyst wall calcification, cyst infection and peritoneal seeding.
MR imaging is superior in demonstrating irregularities of the rim, that probably represent incipient detachment of the membranes and better defines the anatomic relationship of the lesion with the adjacent structures and helps in surgical planning. [5,14]

Prognosis and management. In CE, prognosis is generally good, with complete cure with total surgical excision without spillage. Surgery remains the mainstay in the treatment of hepatic hydatid disease, especially for multivesicular and complicated cyst. Occasionally, formal hepatic resection may be required.

Indications for hepatic surgery include large liver cysts with multiple daughter cysts; single liver cysts, situated superficially which may rupture spontaneously or after trauma; superinfected cysts; cysts communicating with biliary tree and/or exerting pressure on adjacent vital organs; brain, heart and kidney cysts; spinal and bone cysts. [6,15]

In contraindicated cases for surgical removal of the cyst, treatment of hydatid cyst has several alternatives such as PAIR (Puncture, Aspiration, Injection of an heminthicide and Reaspiration), chemotherapy or ‘wait and observe’ approach.

Indications for PAIR are: non-echoic lesion # 5 cm in diameter; cysts with daughter cysts, and/or with detachment of membranes; multiple cysts if accessible to puncture; infected cysts and also pregnant women; children > 3 years old; patients who fail to respond to chemotherapy alone; patients in whom surgery is contraindicated or who refuse surgery. [6]

Images for this section:
Fig. 1: Life cycle of *E. granulosus*. (A) Adult parasite. (B) Domestic dog as principal definitive host; wild canids (dingo, hyena etc.) can be involved in the cycle. (C) Proglottid with eggs. (D) Egg with oncosphere. (E) Infection of humans. (F) Sheep as principal intermediate hosts; other ungulates are of lower significance. (G) Sheep liver with cysts.
Imaging findings OR Procedure details

Imaging analysis.

The authors highlight through a retrospective study conducted in our institution between 2004-2012, the hydatid disease with its multiple imaging appearances and different locations, reviewing a total number of 73 patients diagnosed with hydatid disease (54 with hepatic involvement, followed by 6 pulmonary, 3 splenic, 2 renal, 3 intra and retroperitoneal, 2 mediastinal/cardiac, 2 soft tissue and 1 cerebral cases of hydatid cysts), as well as demonstrating radiological appearances of hydatid cyst complications, such as rupture (5 cases), infection (3 cases) and compression (2 cases), biliary communication (4 cases), transdiaphragmatic migration (2 cases) illustrated with representative cases.

The radiological diagnosis was confirmed by reviewing clinical and surgical records.

Ultrasoundography was an excellent tool for the primary diagnosis, in abdominal, pelvic and soft tissue locations of hydatic cysts, for detecting cystic membranes, septa, hydatid sand (51 patients being diagnosed with this method) and also for the therapeutic decision.

Although simple hydatid cysts (Fig. 2 on page 10, Fig. 3 on page 10) do not demonstrate internal structures, multiple echogenic foci due to hydatid sand (Fig. 4 on page 11) may be seen within the lesion on US, by repositioning the patient, the echogenic foci fall to the most dependent portion of the cavity ("snowstorm sign") [4]. Multivesicular hydatid cysts manifest as well-defined fluid collections with multiple septa representing the walls of the daughter cysts (honeycomb pattern) (Fig. 11 on page 18). When daughter cysts (typical peripheral located within the mother cyst) are separated by the hydatid matrix (a mixed echogenicity material), they demonstrate a "wheel spoke" pattern. Detachment of the endocyst from the pericyst is probably related to degeneration, decreasing intracystic pressure, host response, trauma, or response to therapy. Complete detachment of the membranes inside the cyst is seen as "water lily sign" on US (Fig. 5 on page 12).

Computed tomography may display the same findings as US. Cyst fluid usually demonstrates water attenuation (0-30 HU) and daughter vesicles usually contain fluid with a lower attenuation than that of the fluid in the mother cyst. Cyst calcification usually occurs in the cyst wall and is usually curvilinear or ringlike. During the natural course of an HC, complete calcification occurs. Death of the parasite is not necessarily indicated by calcification of the pericyst, but it is implied by complete calcification (Fig. 6 on page 13). Intravenous administration of contrast material is necessary when complications (such as infection and communication with the biliary tree) are suspected.
**MRI** shows the characteristic low-signal-intensity rim of the hydatid cyst on T2-weighted images (Fig. 13 on page 20) that represents the outer layer of the hydatid cyst (pericyst), which is rich in collagen and is generated by the host. When present, daughter cysts are seen as cystic structures attached to the germinal layer that are hypointense relative to the intracystic fluid on T1-weighted images and hyperintense on T2-weighted images.

Extrahepatic hydatid lesions have nearly identical imaging features, including the presence of cyst wall calcification, daughter cysts, and membrane detachment (Fig. 7 on page 14, Fig. 8 on page 15, Fig. 9 on page 16, Fig. 10 on page 17).

**Sites of involvement.**

**LIVER.** Hydatid disease primarily affects the liver and typically demonstrates characteristic imaging findings. The hydatid cyst can have nonspecific appearance (Fig. 2 on page 10) and may be difficult to distinguish from a simple epithelial cyst on the basis of imaging findings alone. [7]

**LUNG.** The lungs are the second most frequent secondary involvement site of hematogenous spread in adults and probably the most common site in children (Fig. 3 on page 10), incidentally diagnosed at chest radiography, after being asymptomatic for a long period of time. Pulmonary hydatid cysts may grow large, up to 20 cm in diameter, because of lungs' compressibility, and may show specific CT imaging findings (Fig. 9 on page 16). [8]

Atypical locations may involve almost any anatomic site due to hematogenous dissemination of hydatid disease. [9]

**PLEURA.** Pleural involvement is generally secondary to lung involvement or may be due to an hydatid cyst that arises in the liver and ruptures into the chest (Fig. 14 on page 21).

**SPLEEN.** The spleen is the third most common site after the liver and lungs (Fig. 8 on page 15), with an incidence that has been reported to be from 0.9% to 8%, and it develops most frequently secondary to systemic dissemination or intraperitoneal spread from ruptured liver hydatid cysts.

**RENAL.** Renal involvement occurs in 3% of cases, are frequently solitary and located in the cortex (Fig. 8 on page 15) and usually remains asymptomatic for many years. [10]

**INTRA AND RETROPERITONEAL.** Intraperitoneal hydatid disease is secondary to spontaneous or iatrogenic rupture of hepatic, splenic, or mesenteric cysts (Fig. 7 on page 14). Isolated retroperitoneal localisation is also rare and usually secondary to the involvement of liver (Fig. 7 on page 14). [13]
**BRAIN.** Brain involvement is rare (Fig. 10 on page 17), with reported incidence of 1-2% of patients with hydatid disease and most frequently in paediatric population. The most common symptoms are headache, vomiting, and focal neurologic deficits. Computed tomography and magnetic resonance imaging are the first-line diagnostic tools. Intracranial hydatid cysts of the brain are usually solitary, supratentorial and located within the distribution of the middle cerebral artery. [11]

**Other sites.** Hydatid disease may involve almost any anatomic site due to hematogenous dissemination. Unusual locations include diaphragmatic involvement (Fig. 8 on page 15), the heart (Fig. 9 on page 16), soft tissue (Fig. 10 on page 17) and mediastinum (Fig. 10 on page 17, Fig. 14 on page 21).

**Complications.**

**Rupture and biliary communication.** Local complications such as rupture, which may cause biliary obstruction, infection, dissemination, and anaphylaxis are more important than the mass effect of the enlarging lesion. Intrahepatic rupture (spontaneous or related to trauma) may be contained, with endocyst detachment and rupture (visualised as floating membranes) and intact pericyst; communicating, when it implies the passage of the cyst contents into the biliary tree; or direct rupture, when both the pericyst and endocyst rupture occurs, with free spillage of hydatid material. Biliary communication of hydatid cysts is very common (Fig. 13 on page 20) and can be explained by the fact that during cyst growth, small biliary radicles are incorporated into the pericyst; imaging studies demonstrate a cyst wall defect or a communication between the cyst and a biliary radicle, hydatid material filling the biliary radicles or common bile duct, an air-fluid level within the cyst. Dilatation of the biliary tree may result from direct compression of the biliary branches by the cyst, and does not always indicate cyst rupture. [12] Hydatid cyst may also rupture into adjacent spaces, establishing fistulae (see Fig. 10 on page 17 of a mediastinal hydatid cyst fistulised into descendent thoracic aorta) or loco-regional dissemination (see Fig. 14 on page 21 of a hepatic hydatid cyst ruptured into the pleural and mediastinal cavity).

**Compression** of the portal vein is rare, but not uncommon (Fig. 12 on page 19).

**Bacterial superinfection** of hydatid cyst may occur and is always secondary to rupture (Fig. 12 on page 19).

**Pulmonary consolidation.** Complications of lung hydatid cysts include rupture of the cyst into the parenchyma (producing pulmonary consolidation surrounding the cyst- see Fig. 12 on page 19) and pleural cavity, or recurrent acute pulmonary embolism, a rare complication that can be seen when a cyst directly involves the vena cava. [8]

**Peritoneal Seeding** is almost always secondary to previous surgery for hepatic disease, although spontaneous, asymptomatic microruptures of hepatic cysts into the peritoneal
cavity are not uncommon (Fig. 7 on page 14, Fig. 14 on page 21). Cysts may be multiple with different localisation anywhere in the peritoneal cavity. [13]

Images for this section:
**Fig. 1:** Life cycle of *E. granulosus*. (A) Adult parasite. (B) Domestic dog as principal definitive host; wild canids (dingo, hyena etc.) can be involved in the cycle. (C) Proglottid with eggs. (D) Egg with oncosphere. (E) Infection of humans. (F) Sheep as principal intermediate hosts; other ungulates are of lower significance. (G) Sheep liver with cysts.

**Fig. 2:** Multiple noncomplicated pulmonary and hepatic hydatid cysts in adult patient. (a,b) Left pulmonary well-defined rounded mass with hydric intensity on chest Xray and low attenuation on NECT; (c,d) Hepatic low attenuated lesions with slightly calcified wall, surgery confirmed as hydatid cysts, Note the right hemidiaphragm elevation (arrow).
**Fig. 3:** Noncomplicated pulmonary hydatid cyst in pediatric age. (a) Chest Xray shows a well-circumscribed, masslike lesion in the right hemithorax in a 14-year-old boy; (b) Axial (c) coronal NECT shows a hypoattenuating, cystic-like mass that occupies almost the entire right lung and displays anteriorly the right lower lobe bronchus (arrow).
**Fig. 4:** Different stages of hepatic hydatid cysts. (a,b) Axial CECT scan in a 35-year-old man that shows (a) right hepatic lobe hypoattenuating mass that contains daughter vesicle (V) and (b) a left hepatic lobe well defined simple cystic lesion with right anterior portal vein compression (white arrow); (c,d) Stage 2 WHO classification cystic lesion visualised as an anechoic mass with no Doppler signal, daughter vesicle and echogenic material in the most dependent portion of the cavity (hydatid sand- S).
Fig. 5: Hydatid cyst with detached membrane. (a) NECT and (b) US examination of a right lobe hepatic hydatid cyst with a dense circular area of increased attenuation within the cyst representing detached membranes ("water lily sign"); (c,d,e) Axial and coronal T2WI of hepatic and perisplenic multiple hydatid cysts in a 23-year-old woman.
Fig. 6: Calcified hydatid cysts (a,b) NECT showing (a) a low attenuation interhepatogastric lesion with thick wall calcifications (black arrow) and (b) a completely calcified (inactive) hydatid cyst (red arrow) in a 62-year-old patient. Ringlike pattern calcification representing the pericyst (black arrows), visualised on routine chest Xray in same patient.
**Fig. 7:** Different involvement sites in hydatid disease. (a) Axial CECT demonstrating disseminated intraperitoneal hydatid cysts with internal daughter vesicles (v) associated to liver disease in a 50-year-old woman that had undergone previous surgery for hepatic HD; (b,c) Axial and sagittal MR images revealing a hyper T2WI signal mass (arrow) with internal septae displacing rectum, confirmed as a rectovesical hydatid cyst in a 25-year-old woman complaining of constipation; (d) Right iliac fossa multivesicular hydatid cyst in a young man.
Fig. 8: Different involvement sites in hydatid disease. (a) Axial T1WI and (b) SS-FSE short TE showing right renal hydatid cyst with multiple peripheral daughter vesicle cysts (v) in a 47-year-old patient; (c) Unilocular splenic hydatid cyst with floating membranes (black arrowhead) depicted on NECT; (d) Axial CECT showing a right anterior diaphragmatic cystic lesion (black arrows) with slightly hyperintense internal septae.
**Fig. 9:** Multiplanar images obtained in a 25-year-old woman show (a) Axial thoracic NECT and (b,c) cardiac MRI (Cor T2 FSE fat sat and +C Cor T1 FSPGR) of a pericardial hydatid cyst (black star); (d,e) NECT scan in the same patient shows multiple pulmonary hydatid cysts (white stars) with "water lily sign" created by collapsed and crumpled endocysts (black arrow) floating freely in the most dependent part of the cyst.
Fig. 10: (a) Cerebral hydatid disease. Axial CECT scan revealing a cystic multiloculated mass (C) with small wall calcifications without perilesional edema in the right temporo-parietal lobe. Note also the mass effect; (b) Posterior mediastinal hydatid cyst with aortico-cystic fistula; (c,d) Soft tissue laterocervical right sided cystic lesion in a patient with history of hepatic and peritoneal hydatid disease.
**Fig. 11:** Complicated stage 2 hydatid cyst. (a,b,c) NECT and (d) US examination of the liver in a 26-year-old woman showing a giant hydatid cyst with multiple lower intensity daughter vesicles (v), with ill-defined, interrupted contour and subcapsular extravasation of the content (black arrow); (e) CECT in a 36-year-old man with hepatic hydatid disease that reveals a hypointense cystic lesion with internal detached membrane and loss of sharp delineation of the posterior margin with a distinct linear intraparenchymal extravasation of cystic content.
Fig. 12: Complicated hydatid disease. (a) Plain Xray and (b) Chest CT scan performed few weeks later after sudden onset of thoracic pain, fever and leukocytosis in a patient known with hydatid disease, revealing left pulmonary hydatid cyst ruptured into a segmental bronchus with gas content due to bronchial evacuation; note the adjacent alveolar consolidation - aspiration pneumopathy (red arrow); (c) Contrast enhanced axial CT in a patient with no previous surgical intervention showing two rounded intrahepatic lesions with gas content suggestive for infected hydatid cysts (red small arrows) and large multivesicular hydatid cyst with low-attenuation daughter cysts with mass effect on portal vein bifurcation (blue arrow); (d) Axial CT scan in a patient with a history of hepatic hydatid disease showing a cystic right hepatic lobe mass with interrupted anterior contour (black arrow) and intraparenchymal leakage of the content.
**Fig. 13:** Billiary communication. Case of a 40-year-old patient with acute onset of jaundice and pain in the right upper quadrant. (a) Axial T2-weighted MR image reveals a right hepatic lobe hydatid cyst (C) with detached hypointense linear membrane, ill-defined posterior wall (white arrow), (b,c) Secondary biliary dilatation (small arrows) and inhomogeneous signal of common bile duct (d) that indicates leakage of cystic fluid into the biliary ducts; (e) Endoscopic retrograde cholangiopancreatography demonstrates the communication between the cyst and a biliary radicle.
Fig. 14: Transphrenic rupture. Right lobe hepatic hydatid cyst with (b,c) transdiaphragmatic rupture (short arrows) and seeding into the right pleural space (d), mediastinal space (e) and peritoneum (f) in a 40-year-old patient that had undergone surgery for liver hydatid cyst (image a - before surgical intervention); Axial CECT and coronal reformatted images that show right pleural effusion with multiple daughter vesicles (v) visualised as rounded hypointense lesions disseminated into the entire right pleural cavity.
Conclusion

- Hydatid disease is still endemic in certain parts of the world. It primarily affects the liver, but secondary Echinococcosis can develop in almost any anatomic location due to hematogenous dissemination; the lungs are the second most frequent site and brain, heart and soft tissue are atypical involvement sites.
- Depending on the stage of cyst growth, hydatid disease can have the appearance of a simple cyst or can have typical imaging findings (presence of daughter cysts, detached membrane or wall calcifications).
- Complications can be local: rupture, infection or compression, abdominal/thorax extension or complications due to systemic dissemination.

References


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