Hydatid disease: not only in liver and lung. A pictorial review

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Learning objectives

A pictorial review of Hydatid cysts not located in lung or liver.

Purpose:

- To illustrate the spectrum of imaging findings of hydatid disease (HD) in unusual anatomic locations.

- Recognize the appropriate imaging methods for each location, its advantages and limitations.

- To discuss specific radiologic signs, differential diagnosis and pitfalls in every site.
Hydatid disease is a parasitic disease caused by Echinococcus granulosus. In the past, it was restricted to endemic areas but nowadays has a worldwide distribution due to immigration from endemic areas. So, continuing medical education about CT images from HD is necessary not only for radiologists working in endemic areas also for young radiologists or for those that work in non-endemic areas. When cysts are in lung or liver, radiological diagnosis is usually easy but when cysts are located in other organs, diagnosis could be difficult specially in non-endemic areas because HD is not the first diagnosis managed by radiologist. We present our experience in non-lung non-liver hydatid cysts.
Findings and procedure details

The central nervous system (fig 1-4)

Cerebral HC is extremely rare, accounting for only 2% of all intracranial masses, even in countries where this disease is endemic.

HCs are mostly located in the territories of the middle cerebral artery but can be seen anywhere within the brain. Most cysts are supratentorial. The parietal lobe is most frequently involved. Cerebral HC is more common in children than in adults.

Imaging findings:

Cerebral HC generally appears unilocular and is isointense or isoattenuating relative to cerebrospinal fluid. Fine peripheral enhancement can be seen in the fibrous capsule.

The lack of surrounding edema and the marked mass effect make it easy to distinguish cerebral HC from abscess and cystic tumor.

The presence of a hypointense rim, especially on T2-weighted MR images, is characteristic of HC of the brain.

Cerebral HC is generally solitary but may be multiple when it ruptures spontaneously or due to trauma or surgery.

Multivesicular cysts are rare in the brain.

Calcification occurs in less than 1% of cases.

The mass effect of HC is very prominent in the brain, and symptoms are generally due to compression of vital cerebral structures.

HCs located in proximity to the cortex can protrude into the meninges and calvaria and cause erosion.

Complications: Rupture and infection

When HCs are infected, the lesions show enhancement after contrast material injection, and differential diagnosis is sometimes difficult.

In the cerebral cisterns, the most likely pathway of dissemination is hematogenous spread to the meninges and rupture into the subarachnoid space.

Differential diagnosis:
Arachnoid cysts, porencephalic cysts, and epidermoid tumors: Arachnoid cysts and porencephalic cysts are neither spheric nor entirely surrounded by brain tissue.

HCs can be differentiated from brain abscess and cystic astrocytoma by the absence of significant rim enhancement, perifocal edema, and mural nodules.

**In the latero-cervical region (fig 5)**

HCs of the neck are rare, with only a few cases reported in the literature.

HCs in thyroid tissue can become large because of the compressibility of the thyroid gland and adjacent tissues. However, knowledge about the imaging features of HCs in thyroid tissue is inadequate due to their relative rarity.

Radiologic signs are usually nonspecific. The diagnosis is greatly facilitated with ultrasonography, CT, and MRI.

The ultrasonography is highly efficient in detecting germinal vesicles in cystic lesions, which is important for a preoperative diagnosis of hydatidosis. Although cystic echinococcosis typically consists of a single unilocular cyst, 20-30% of cases present with multiple cysts in the same or multiple organs.

CT-scan and MRI are complementary studies. They provide a precise assessment of the extension into the soft tissues and the calcifications of the peripheral rim of the cyst. The signal from the cysts is inhomogeneous of low intensity on T1-weighted and high intensity on T2-weighted images. HD may mimic benign or malignant tumours, cysts, abscess, and other lesions.

**The mediastinum (fig 6)**

The larvae of HCs are rarely present in the mediastinum, although approximately 108 cases have been reported in the literature.

Like HCs located elsewhere in the body, mediastinal HCs affect both sexes equally and can be seen in patients of all ages. The symptoms of mediastinal HC depend on the size and location of the cyst and the involvement of adjacent structures.

It may be primary or can spread through the mediastinum during the rupture of lung HC.

Mediastinal involvement may be solitary or multiple, depending on the type of occurrence. **The imaging appearance:** can vary from type I to type III. Type II and type III HCs in the anterior mediastinum should be differentiated from thymoma and teratoma that have mostly necrotic components.
CT and MR imaging are the preferred examination methods for the evaluation of mediastinal HC.

**Heart:**

Isolated cardiac hydatid cysts are rare, accounting for 0.02%-2% of all cases. The larvae reach the myocardium via the coronary circulation. The most frequent area of involvement is the left ventricle (60%), presumably reflecting the dominance of the left coronary circulation. This is followed by the right ventricle (10%), pericardium (7%), pulmonary artery (6%), left atrial appendage (6%), and the interventricular septum (4%). Another route is by rupture of pulmonary cysts into the pulmonary vasculature.

**Interventricular septum: (fig 7)**

Cysts of the interventricular septum may present with outflow tract obstruction and conduction abnormalities. Serologic tests are of limited value in diagnosis. Enzyme-linked immunosorbent assay has sensitivity and specificity of 94% and 99%, respectively, and along with the indirect hemagglutinin test, it has proved to be the best test for follow-up. Transthoracic echocardiography is the investigation of choice. Computed tomography best shows wall calcification. Magnetic resonance imaging is the technique of choice for follow up. Specific signs of hydatid cyst include calcification of the cyst wall, the presence of daughter cysts, and membrane detachment.

Early surgical removal is the treatment of choice.

**Hydatid pulmonary embolism: (fig 8)**

Clinical manifestations of the hydatid pulmonary embolism are not specific although hemoptysis is the most frequent sign. In the absence of a medical history of a visceral hydatid cyst, clinicians should use all the means necessary to reach such a diagnosis, ie, pulmonary embolism is due to hydatid cyst. The diagnostic investigation of patients with suspected hydatid pulmonary embolism should involve a two-dimensional echocardiography, a spiral CT scan and a MRI.

Echocardiography is the investigative procedure of choice for studying cardio pericardial hydatidosis but it rarely enables direct visualization of the pulmonary embolus. With transoesophageal echocardiography, it is possible to visualize massive emboli in the central pulmonary arteries.

Spiral CT scan and MRI have been used successfully in the diagnosis of hydatid cysts of the lungs and the heart. However, MRI has an advantage over spiral CT-scan in examination of the heart and the great vessels, because with images in multiple phases it gives a more complete anatomical picture. Nowadays, MR-angiography in hydatid pulmonary embolism whether performed before or after surgery, yields good results.
**Bone (fig 9)**

Bone involvement is also rare (0.5%-2% of cases). The most commonly involved bone structures are the spine (35% of cases), pelvis (21%), femur (16%), tibia (10%), ribs (6%), skull (4%), scapula (4%), humerus (2%), and fibula (2%).

Pericyst formation does not occur in bone, and the cyst has a much thinner wall. Because of this fact and the rigid nature of bone, the cyst cannot assume its typical shape (sphere). The cyst enlarges in an irregular branching fashion along the path of least resistance.

Over time, the parasite replaces the osseous tissue and destroys the cortex. It then spreads from bone to surrounding tissue such as muscle and the spinal cord.

Extraosseous HCs may calcify, whereas intraosseous HCs rarely show calcification. Parasitized bone has heterogeneous medium to low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted images.

CT can demonstrate the calcification in extraosseous HCs.

In the spine, HD simulates tuberculosis spondylitis or chronic osteomyelitis. Lack of osteoporosis and sclerosis in involved bone, absence of damage to intervertebral disk spaces and vertebral bodies, paraspinal extension, and (in the thoracic spine) involvement of contiguous rib are the most common features of spinal HD.

Bone HD in other sites manifests as different-sized areas of pure osteolysis that may become confluent and cause thinning of the cortex. In such cases, HD becomes symptomatic when HCs cause a pathologic fracture or spread to surrounding soft tissues.

**Primary muscular hydatid cyst (fig 10-11)**

Soft-tissue HD occurs in 2.3% of patients living in endemic areas. The growth of the cyst within a muscle is difficult because of the contractility of muscles and the presence of lactic acid.

The affinity for muscles of the neck and trunk and at the root of limbs could be explained by the increased vascularity and decreased activity of these muscle groups.

Primary muscular HD is rare, with only isolated cases being described in the literature.

HD in the soft tissues may have various appearances. Unilocular type I HCs, unilocular cysts with daughter vesicles or detached membranes (type II), and calcified cysts (type III) can be seen. Multiple HCs can be seen due to spontaneous rupture or to trauma or surgery.
Edema or acute inflammation caused by compression of or allergic reaction in soft tissue adjacent to the cyst is uncommon but may be seen.

A low-signal-intensity rim is evident on T2-weighted MR images; this is not a common finding in HCs located elsewhere in the body.

**Intra dural extra medullar (fig 12)**

Spinal HD accounts for less than 1% of all HD. The thoracic spine is most frequently involved (50% of cases), followed by the lumbar (20%), sacral (20%), and cervical (10%) spine.

Spinal HD is classified into five groups: intramedullary, intradural extramedullary, extradural intraspinal, vertebral, and paravertebral.

The first three groups of HD are rare.

Calcification is also rare in spinal HD.

Radiological studies are usually inconclusive but may be helpful in the diagnosis of hydatid disease. Spherical cysts with peripheral calcification may be seen on a plain radiograph and are indicative of hydatid cyst.

CT scan and MRI are the current diagnostic modalities of choice. Irregular bony erosions along with multilocular, non-enhancing flattened sausage-shaped lesions with very thin, non-septated walls are the hallmark of vertebral hydatid disease.

Extraosseous lesions are usually unilocular with thicker cystic walls and acquire a spherical shape due to the lack of resistance from the hard bone tissue during their growth; calcifications may develop as the disease progresses.

Magnetic resonance imaging shows cyst which generally have two dome-shaped ends, which usually have no debris in the lumen, and usually look like flattened sausages, with thin, regular walls without septations. Intradural cysts may be single or multiple; extradural cysts are always multiple and involve the bone.

On MRI, the lesions appear isointense to the cerebrospinal fluid (CSF) (hypointense on T1-weighted images and hyperintense on T2-weighted images)

**Peritoneum and Retroperitoneum (fig 13-14)**

Although a few unusual cases of primary peritoneal involvement have been described, peritoneal HCs are almost always secondary to hepatic involvement. The overall prevalence of peritoneal involvement in cases of abdominal HD is approximately
13%. Most of these cases are related to previous surgery for hepatic HC, although spontaneous, asymptomatic microruptures of hepatic cysts into the peritoneal cavity are not uncommon (12% of cases).

CT and MR imaging are the modalities of choice in the evaluation of all abdominal structures. HCs are generally multiple and can arise anywhere in the peritoneal cavity.

The differential diagnosis can be made easily owing to the characteristic appearance of daughter cysts, but unilocular type I HCs may be difficult to differentiate HCs from mesenteric cysts or intestinal duplication cysts.

Any form of HC can be seen in the peritoneal cavity. Moreover, HCs can be located anywhere in the peritoneal cavity (eg, in a herniated umbilical sac).

Isolated retroperitoneal HCs are also rare and are secondary to the involvement of other organs-especially the liver-or to surgery.

The imaging appearances were similar to those of HCs located elsewhere in the body. Any type of HC can be seen in the retroperitoneum.

**Ovarian HD** (fig 15) is rare, with only a few reported cases in the literature. Involvement of the ovaries is generally secondary to peritoneal spread of daughter cysts due to rupture of a liver HC. Isolated primary ovarian involvement has been reported. Ovarian HCs may remain asymptomatic for a long time and be discovered incidentally or may cause irritation or compression symptoms. It is very difficult to differentiate HCs from other ovarian lesions that may appear to be mostly cystic (cystadenoma, cystadenocarcinoma) on the basis of imaging findings alone.

HCs may be unilocular and can mimic an ovarian cystadenoma, especially in women of reproductive age. Daughter cysts can simulate septal structures that may be seen in cystadenocarcinoma.
Fig. 1: Axial CT scan of the brain: completely calcified hydatid cyst in the left temporo-occipital lobe non modified after injection of contrast medium.

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**Fig. 2:** sagittal T1 and axial T2 weighted MR image of the brain: mass of heterogeneous signal on all sequences, without enhancement after intravenous injection of contrast

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**Fig. 3:** Axial CT scan of the brain: unilocular hydatid cyst in the right frontoparietal lobe (a) and in the left frontoparietal lobe with membrane detachment (b) exerting a significant mass effect on the ventricular system and midline.
**Fig. 4**: Axial CT scan of the brain after injection of contrast medium: Hydatid cyst with floating membrane in the left occipital lobe with important mass effect. It shows a peripheral enhancement and perilesional edema.

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Fig. 5: Ultrasound images: multiple cystic and hypoechoic nodules of varying size within the left thyroid lobes. One of these nodules reached approximately 5 cm in diameter and appeared to be heavily calcified. The patient underwent surgery and the diagnosis of HD was made.

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**Fig. 6:** Fig 6: Axial CT scan of the thorax before and after injection of contrast medium: mediastinal hydatid cyst with daughter cysts without sign of mediastinal compression.

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**Fig. 7:** Fig 7: Axial CT scan of the thorax without injection of contrast medium and axial ultrasound image: Hydatid cyst with peripheral calcification in the inter ventricular septum without hemodynamic modifications.

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**Fig. 8:** Fig 8: Axial CT scan of the thorax after injection of contrast medium: Hydatid pulmonary embolism

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**Fig. 9:** Chest radiograph: osteolytic multi lacunar lesion blasting the anterior arch of the 6th right coast. Axial CT scan of the Chest with injection of contrast medium: osteolytic hypodense process with cortical breaking of the 6th right coast.

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**Fig. 10:** Ultrasoundography of the left thigh: well-defined heterogeneous soft tissue mass in the adductor magnus muscle containing numerous cystic formations.

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**Fig. 11:** a- MRI. Coronal T1-weighted image: multiple daughter cysts appear as hypointense signals (arrows), relative to surrounding intracystic fluid (arrowhead), b- MRI. Coronal T2-weighted image: cyst formation in the adductor compartment, the daughter cysts have a hyperintense signal (white arrow), c- MRI. Left thigh axial T1-weighted image with fat saturation: diffuse pericystic enhancement (arrows)

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**Fig. 12:** Axial T2 weighted MR images: multiple HC of the right peri vertebral soft-tissue associated with intra ductal multilocular HC. Sagittal T1 and T2 weighted MR
Multiple multilocular intra-ductal lesions hypointense on T1 and hyperintense on T2 with peripheral enhancement after gadolinium injection. There is a compression of the terminal marrow and roots of the cauda equina.

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**Fig. 13:** Fig 13 and 14: Axial CT scan of the abdomen after injection of contrast medium: unilocular peritoneal and retroperitoneal hydatid cyst.

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Fig. 14: Fig 13 and 14: Axial CT scan of the abdomen after injection of contrast medium: unilocular peritoneal and retroperitoneal hydatid cyst.

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Fig. 15: Axial CT scan of the pelvis after injection of contrast medium: unilocular left sided ovarian hydatid cyst.

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Conclusion

The classic imaging findings of the hydatid disease are well known, but the atypical manifestations can make the diagnosis difficult. The hydatid disease should be included in the differential diagnosis of a cystic lesion, found anywhere in the body, especially when they occur in endemic regions. Familiarity with unusual locations and its imaging findings are essential to make a prompt and accurate diagnosis. The imaging method used depends on the affected organ and the evolution of the cyst.
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