Imaging in pulmonary hydatid cysts

Mandeep K Garg, Madhurima Sharma, Ajay Gulati, Ujjwal Gorsi, Ashutosh N Aggarwal, Ritesh Agarwal, Niranjan Khandelwal

Mandeep K Garg, Madhurima Sharma, Ajay Gulati, Ujjwal Gorsi, Niranjan Khandelwal, Department of Radiodiagnosis and Imaging, Post Graduate Institute of Medical Education and Research, Chandigarh 160012, India

Ashutosh N Aggarwal, Ritesh Agarwal, Department of Pulmonary Medicine, Post Graduate Institute of Medical Education and Research, Chandigarh 160012, India

Author contributions: All authors equally contributed to this paper with conception and design of the study, literature review and analysis, drafting and critical revision and editing, and final approval of the final version.

Conflict-of-interest statement: None.

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Correspondence to: Dr. Mandeep K Garg, Additional Professor, Department of Radiodiagnosis and Imaging, Post Graduate Institute of Medical Education and Research, Sector 12, Chandigarh 160012, India. gargmandeep@hotmail.com
Telephone: +91-172-2756380
Fax: +91-172-2744401

Received: September 25, 2015
Peer-review started: October 3, 2015
First decision: October 27, 2015
Revised: March 5, 2016
Accepted: March 22, 2016
Article in press: March 23, 2016
Published online: June 28, 2016

Abstract

Hydatid disease is a zoonosis that can involve almost any organ in the human body. After the liver, the lungs are the most common site for hydatid disease in adults. Imaging plays a pivotal role in the diagnosis of the disease, as clinical features are often nonspecific. Classical radiological signs of pulmonary hydatid cysts have been described in the literature, aiding in the diagnosis of the disease. However, complicated hydatid cysts can prove to be a diagnostic challenge at times due to their atypical imaging features. Radiography is the initial imaging modality. Computed tomography can provide a specific diagnosis in complicated cases. Ultrasound is particularly useful in peripheral lung lesions. The role of magnetic resonance imaging largely remains unexplored.

Key words: Pulmonary; Hydatid; Cyst; Radiography; Computed tomography

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Core tip: The lungs are the second-most common site for hydatid disease in adults. The classical radiological signs of pulmonary hydatid cysts have been described in the literature. In this article, we have comprehensively reviewed the various radiological signs and presentations of pulmonary hydatid cysts. We have also described atypical imaging manifestations of the disease. Radiologist should be familiar with the spectrum of the imaging findings in pulmonary hydatid cysts.

Garg MK, Sharma M, Gulati A, Gorsi U, Aggarwal AN, Agarwal R, Khandelwal N. Imaging in pulmonary hydatid cysts. World J Radiol 2016; 8(6): 581-587 Available from: URL: http://www.wjgnet.com/1949-8470/full/v8/i6/581.htm DOI: http://dx.doi.org/10.4329/wjr.v8.i6.581

INTRODUCTION

Hydatid disease is caused by Echinococcus tapeworm in
the larval stage. In humans, Echinococcus granulosus is responsible for the most common type of hydatid disease. Although hydatid disease has been known to exist in humans for almost 2000 years, Rudolph in 1808 first used the term “hydatid” to describe echinococcosis. “Hydatid” is a Greek word (literal meaning: Watery vesicle), which aptly describes the characteristics of the disease. Hydatid disease is endemic in sheep-rearing countries, particularly in the Mediterranean region, Africa, South America, the Middle East, Australia, and New Zealand. The liver is the most common organ to be involved in adults (75%), followed by the lungs (15%) in the pediatric population, while in the pediatric population, the lungs are the most common site of involvement. Uncomplicated hydatid cysts of the lungs are usually asymptomatic, while complicated cysts present with nonspecific clinical features like coughing, chest pain, and hemoptysis. Imaging thus plays a pivotal role in the diagnosis of the disease. Although typical imaging findings have been well described in the literature, radiologists should also be aware of atypical imaging findings that can occur secondary to complications.

ETIOPATHOGENESIS

A definitive host for E. Granulosus is dog (or other carnivores), which harbor adult worms in their small bowel. Eggs released by the worms are excreted in faeces. Excreted ova are ingested by an intermediate host (most commonly sheep). When in the intestine of an intermediate host, ova develop into hexacanth embryos and reach the liver through portal circulation. Embryos in the liver develop into cysts. Humans are accidental hosts and become part of this lifecycle through contact with a definitive host or the ingestion of water or vegetables contaminated with echinococcal ova.

The hydatid cyst wall is composed of three layers: The outermost layer, known as the pericyst, is formed by the protective response of the host tissue; the acellular middle laminated layer (occasionally called the ectocyst) allows for the passage of nutrients; the innermost germinal layer (known as the endocyst), produces scolices toward the inner side and laminated membrane on the outer side. Hydatid cysts contain many daughter vesicles resembling bunches of grapes.

The lungs are the most common site in the pediatric population and the second-most common site in adults. Lung hydatid cysts have certain peculiar characteristics compared to cysts in other locations. The lungs facilitate the cyst’s growth due to negative pressure and their compressible nature. As a result, hydatid cysts grow in the lungs three times faster than in the liver. In addition, calcification (occurring in only 0.7% of cases) and daughter cyst formation in lung hydatids are very rare.

CLINICAL FEATURES AND COMPLICATIONS

Most pulmonary hydatid cysts are acquired in childhood and remain asymptomatic for a long period of time. Uncomplicated hydatid cysts are usually diagnosed incidentally on chest X-rays. Chest pain, dyspnea, dry coughing, and hemoptysis can occur due to the mass effect caused by larger cysts. Acute-onset chest pain, coughing, hemoptysis and anaphylactic reactions may suggest cyst rupture. The expectation of membranes and hydatid sand is diagnostic of a ruptured hydatid and has been described in 31%-38% of cases.

A hydatid cyst rupture is the most common complication occurring in up to 49% of cases. Ruptures may be contained (by detachment of the pericyst from the endocyst), communicating (with the bronchus) and direct (rupture of all membranes with spillage of contents). All hydatid cysts carry the risk of rupture and are classified as complicated hydatid cysts. The degeneration of cyst membranes is responsible for rupture, which in turn is determined by several factors, such as the age, chemical reactions, and defense mechanisms of the host. Moreover, the risk of rupture increases with the size and number of cysts. Antihelminthic therapy and percutaneous aspiration are also known to cause cyst rupture and fatal complications. Cyst rupture can occur into bronchus (manifesting as coughing with sputum containing hydatid sand and membrane fragments) or the pleural cavity (manifesting as pneumothorax, effusion, and emphysema). Occasionally pleural seeding from live scolex during a rupture can cause secondary pleural disease. Rarely, a cyst can rupture into the vena cava and present as a recurrent pulmonary embolism.

Infection is the most common complication of cyst rupture, clinically presenting with features of lung abscess. On rare occasions, a pulmonary hydatid cyst can also involve the thoracic wall.

IMAGING FEATURES

The lungs are the second-most common site for hydatid cysts in adults. The lower lobes are the most common location in the lungs (in 60% of cases) with the right basal lobe being more common. In 30% of cases, there is more than one cyst, and they can be bilateral in 20% of cases. X-ray and computed tomography (CT) are the usual imaging modalities used. Ultrasound can be beneficial in peripheral lesions and to assess pleura.

Radiography

An uncomplicated hydatid cyst appears as a well-defined homogenous radio-opacity on a chest X-ray. Differential diagnoses on a chest X-ray include fluid-filled cysts, benign tumors, carcinoma, metastases, and inflammatory masses. The appearance of cysts has been compared to cannon balls in anteroposterior projection and to rugby balls in lateral projection. Cysts can assume polycyclic configuration due to pressure from adjacent structures. Notching can also occur in cysts, giving them a bilobed appearance. The loss of a spherical shape on an X-ray with the appearance of small depression (resulting in a reniform shape) may
Implied bronchial rupture and has been called the "slot sign" [12]. Simple hydatid cysts are sharply demarcated from adjacent lung parenchyma. However, atelectatic and reactive changes in the adjacent lung can cause the loss of sharp margins of cysts on an X-ray, thereby mimicking pneumonia or carcinoma [35,36]. Multiple large masses in the lungs are pathognomonic for hydatid cysts [36] (Figure 1B).

As described earlier, calcification is very rare. Hydatid cysts can erode a bronchus with the introduction of air between the pericyst and the endocyst appearing as a radiolucent rim around the cyst on an X-ray known as the "crescent sign" [36] (Figure 2). However, this sign is not specific for hydatid cyst and can be seen in mycetoma, bronchogenic carcinoma, blood clot and pulmonary artery aneurysm.

Hydatid cysts can erode a bronchus with the introduction of air between the pericyst and the endocyst appearing as a radiolucent rim around the cyst on an X-ray known as the "crescent sign" [36] (Figure 2). However, this sign is not specific for hydatid cyst and can be seen in mycetoma, bronchogenic carcinoma, blood clot and pulmonary artery aneurysm [37]. With the introduction of more air, the endocyst shrinks and ruptures with the introduction of air into the endocyst. At this stage, an air fluid level is seen in the endocyst with a radiolucent rim between the pericyst and the endocyst, known as the "cumbo sign" or the "double arch sign" [33,35,36,38]. With further collapse of the endocyst, the membranes floating in the remaining fluid are known as the "water lily sign" or the "camoulette sign" [35,36,38]. With the rupture of the endocyst, daughter cysts may appear as round radio-opacities at the bottom of cysts, giving them a "rising sun" appearance [18]. With coughing out of membranes, the pericyst can become empty (the "dry cyst sign"), appearing as air-filled cysts on X-rays [18]. With superadded infection, an air fluid level can appear in hydatid cyst, mimicking a lung abscess (Figure 3). All these signs have been summarized in Table 1.

The classical radiographic signs described above are not always present in complicated hydatid cysts. Thus, complicated cysts can be difficult to diagnose on routine radiography, and CT is usually required to establish the accurate diagnosis.

**CT**

CT features of uncomplicated hydatid cysts: Uncomplicated hydatid cysts appear as well-circumscribed fluid attenuation lesions with homogenous content and smooth, hyperdense walls. Unlike hydatid cysts of the liver, calcification and daughter cyst formation are rare in lung hydatids [12-15]. Intact hydatid cysts can be difficult to differentiate from other pulmonary cysts [23]. Associated hydatid cysts with calcification and daughter cysts can also be seen in liver, thereby providing clue to the diagnosis (Figure 4).
CT features due to contained ruptures of the endocyst: Bronchial erosion caused by the cyst can cause the appearance of crescents of air between the pericyst and the endocyst (the crescent sign). Occasionally, air may dissect through the posterior aspect of membranes without anterior extension. This causes the appearance of air crescents along the posterior aspect of lesion, known as the "inverse crescent sign" [18,39]. Blebs of air can be seen between the pericyst and the endocyst, which implies impending rupture and are called "signet ring sign"[39]. Small intracystic air foci can be seen at the periphery of cyst, between the pericyst and the endocyst ("air bubble sign"), which can cause the shrinkage of the endocyst and rupture[16,40] (Figure 5).

CT features due to complete ruptures of the endocyst: Due to the continuous accumulation of air between the pericyst and the endocyst, the endocyst ruptures with the collapse of membranes. An air fluid level within the endocyst and the crescent of air between the endocyst and the pericyst results in an onion peel appearance or the "cumbo sign"[18]. After the expectoration of cyst fluid, collapsed membranes can be seen within the cyst, known as the "whirl" or the "serpent sign"[18,21] (Figures 6 and 7B). With complete collapse, the crumpled endocyst appears as a wavy membrane floating on fluid, known as the "water lily sign"[13,18,21] (Figure 8). The detached and crumpled endocyst may settle in the most dependant part of the cavity, resulting in a "mass within the cavity"[16] or the "incarcerated membrane sign"[12]. After the complete expectoration of the cyst fluid and membranes, the cyst appears only air filled, known as the "dry cyst sign"[12,18].

CT features due to pleural rupture: The most

Table 1  Classical signs of pulmonary hydatid cyst on chest X-ray

| Uncomplicated hydatid cyst                                      | Complicated hydatid cyst                  |
|-----------------------------------------------------------------|------------------------------------------|
| Well circumscribed round radio-opacity                          | Crescent sign                            |
| (resembling canon ball on AP and rugby ball on lateral projection) |                                          |
| Polycyclic and bilobed appearance                               | Cumbo or double arch sign                 |
| Slot sign (impending rupture)                                   | Water lily or camelotte sign              |
|                                                                | Rising sun sign                           |
|                                                                | Dry cyst sign                             |

AP: Anteroposterior.
common imaging finding after a pleural rupture of a hydatid cyst is hydrothorax or hydropneumothorax. Occasionally, membranes can be seen floating in pleural effusion [12]. It should be noted that pleural thickening and effusion can also occur secondary to a reactive inflammatory response [29] (Figure 7).

**CT features due to infection:** Increases in the size and number of pulmonary hydatid cysts increase the risk of infection [26]. A superadded infection is the most common complication of a ruptured hydatid cyst [44]. Increases in the cyst wall thickness with enhancement (the “ring enhancement sign”) (Figure 5) and the density of content are seen in cases of superadded infection of hydatid cysts [26,41]. In addition, the appearance of air bubbles (the “air bubble sign”) (Figures 5 and 7A) and an air fluid level within the cyst also indicate infection [41].

**Other complications:** Rarely, hydatid cysts can invade the thoracic wall. On CT, a cystic lesion can be seen in the thoracic wall showing communication with a lung cyst (Figure 9) [28]. Sometimes cysts can rupture into the inferior vena cava and lead to recurrent pulmonary embolism. Occasionally, cysts can directly extend into the pulmonary artery (Figure 10). Various CT signs in pulmonary hydatid cysts have been summarized in Table 2.

**Ultrasonography in hydatid cysts**
Ultrasound can prove useful in cases of peripheral hydatid cysts located along the chest wall. Recently, the “wall sign” has been described in pulmonary cysts [41].

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**Table 2** Computed tomography signs described in pulmonary hydatid cyst

| Signs of contained rupture                  | Crescent sign          |
|--------------------------------------------|------------------------|
| Inverse crescent sign                      |                        |
| Signet ring sign                           |                        |
| Air bubble sign                            |                        |
| Signs of cyst rupture                      | Cambo sign             |
| Serpent sign                               |                        |
| Swirl sign                                 |                        |
| Water lily sign                            |                        |
| Mass within a cavity sign                  |                        |
| Incarcerated membranes sign                |                        |
| Dry cyst sign                              |                        |
| Signs of cyst infection                    | Air bubble sign [3]    |
| Air bubble sign                            |                        |
| Ring enhancement sign                      |                        |
| Air fluid level                            |                        |

1Indicated impending rupture; 2 can be seen in contained rupture as well as superadded infection.
Though a variety of signs have been described on imaging, complicated hydatid cysts can present with atypical imaging findings. CT is the imaging modality of choice, especially in complicated hydatid cysts, and can provide an accurate diagnosis by demonstrating the internal characteristics and morphology of the lesion. Thus, radiologists should be well aware of the typical and atypical imaging features of the disease.

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