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Complicated Hydatid Cyst and “Air Bubble” Sign: A Stepping-Stone to Correct Diagnosis

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 32
Final Diagnosis: Complicated hydatid cyst
Symptoms: Cough with expectoration and fever for the last 4 months
Medication: Tab. Albendazole
Clinical Procedure: —
Specialty: Pulmonology

Objective: Unusual clinical course





Background: Hydatid cyst, or Echinococcosis, is an important helminthic zoonotic disease in humans that commonly affects the liver and lungs. Uncomplicated hydatid cysts, seen as round opaque lesions on chest radiography, are easily diagnosed, whereas complicated cysts (infected and or perforated) may change the radiographic appearance of the hydatid cyst, causing an incorrect diagnosis and delayed treatment. Although in radiology many signs have been described, the “air bubble” sign, seen in the mediastinal window of CECT as a single or multiple small rounded radiolucent areas with sharp margins within the periphery of a solid mass lesion, is being recognized as a sign with high sensitivity and specificity in the diagnosis of complicated hydatid cysts.

Case Report: A 32-year-old female on anti-tubercular treatment for the past 3 months without any improvement was admitted to our hospital. CECT of the chest revealed a mass-like lesion with the “air bubble” sign. After 15 days the patient had a vigorous bout of coughing, leading to expectoration of pieces of whitish yellowish gelatinous membrane for the next 3 days. The ELISA result for Echinococcus was highly positive. On the basis of the “air bubble” sign, positive serology, and expectorated pieces of the membrane, the patient was diagnosed as having a complicated hydatid cyst.

Conclusions: Due to the varied presentations of complicated hydatid cyst, the knowledge and awareness of various signs in radiology associated with the hydatid cyst, in particular the “air bubble” sign, is imperative in making a prompt and accurate diagnosis of a complicated hydatid cyst.

MeSH Keywords: Echinococcosis, Pulmonary • Helminthiasis • Lung Diseases

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/892621>

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Background

The Echinococcus (E) tapeworm, a common helminth infecting carnivores such as dogs and wolves, is known to cause disease in humans, who act as an intermediate host when people accidentally become infected by the eggs of the worm. Four species are recognized: *E. granulosus*, *E. multilocularis*, *E. vogeli*, and *E. oligitharus*. Of these, the vast majority of the infestations in humans are caused by *E. granulosus* [1]. After the liver, the lungs are the second most commonly affected organ [2]. The growth of hydatid cysts is slow and may remain undiagnosed for long periods. Uncomplicated cysts, seen as round opaque lesions on chest radiography, are easily diagnosed. However, infected and or perforated cysts, known as “complicated” cysts, may change the radiographic appearance of the hydatid cyst, causing an incorrect diagnosis and delayed treatment [3–5]. Imaging modalities and serology establish the diagnosis in most cases. However, diagnosis of a complicated hydatid cyst is very difficult and usually delayed. Over time, certain radiological presentations/signs have been identified that are contributive in confirming the diagnosis of complicated hydatid cyst. An important finding on CECT is the “air bubble” sign. It is reported to be highly sensitive and specific in confirming the diagnosis of a ruptured and infected hydatid cyst [6].

Case Report

A 32-year-old woman presented to our department with complaints of fever and cough with expectoration for the past 4 months. The fever was low-grade and intermittent, and the cough was accompanied with expectoration, was moderate in quantity and mucopurulent in character. Three months earlier, she had been put on anti-tubercular treatment on the basis of her lesions on X-ray (Figure 1), despite her sputum for AFB (Acid fast bacilli) being negative, in the form of category 1 DOTs (directly observed therapy short course) under RNTCP (Revised National Tuberculosis Control Programme) without any improvement. General physical examination showed a moderately built and nourished anemic female. She was febrile, alert, with a pulse rate of 92/min, respiratory rate of 22/min, and blood pressure of 110/76 mm Hg. Examination of all other organs were essentially normal. Examination of the respiratory system revealed impaired percussion note, decreased vesicular breath sounds, and vocal resonance over the right clavicular and supra scapular region. Her latest x-ray chest (Figure 2) showed no change in the opacity in the right upper zone as compared to her earlier x-ray. Other than a hemoglobin of 9.5% gm, her hematological and biochemical tests were non-revealing. Her sputum for AFB (acid-fast bacilli) and gram stain were negative. Her sputum, sent for culture for pyogenic organisms and BACTEC for AFB, showed no growth.

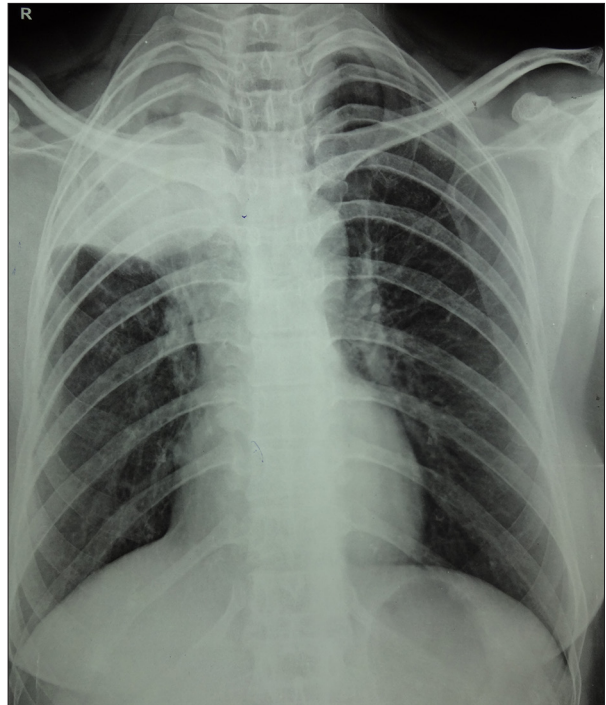


Figure 1. Chest X-ray PA view showing a homogeneous opacity in the right upper zone.



Figure 2. Chest X-ray PA view showing a homogeneous opacity in the right upper zone.

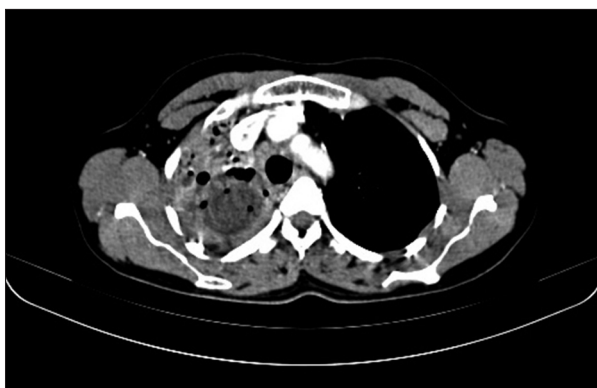


Figure 3. Chest CECT mediastinal window showing the hydatid cyst with "air bubble" sign. The surrounding parenchyma also shows secondary infection.



Figure 5. Coughed-up pieces of the whitish-yellowish membrane of the hydatid cyst.

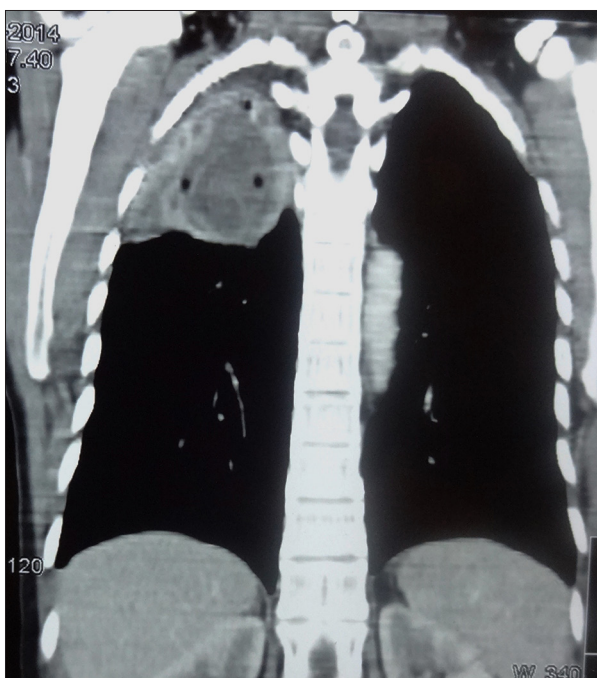


Figure 4. Chest CECT (coronal view) showing the hydatid cyst with "air bubble" sign.

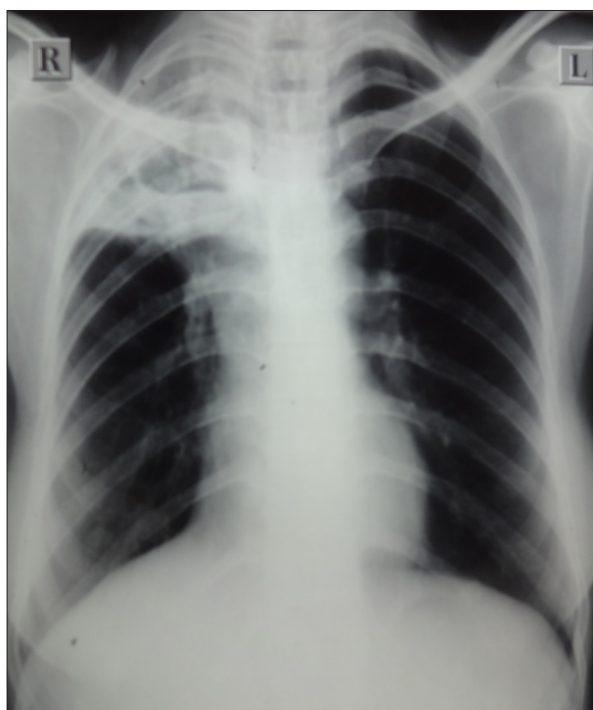


Figure 6. Chest X-ray shows a ruptured hydatid cyst masked by surrounding pneumonitis and an air-fluid level mimicking lung abscess.

Her contrast-enhanced computed tomography (CECT) chest showed a mass-like lesion (attenuation value 24 HU) with air bubbles and secondary infection in the surrounding parenchyma (Figures 3 and 4). A CT-guided trans-thoracic FNAC (fine-needle aspiration cytology) was attempted and 10 ml of pus was aspirated. Various tests, gram stain, AFB stain, and fungal stains were negative. The culture for pyogenic organisms and BACTEC for AFB showed no growth. The patient was put on broad-spectrum antibiotics and treated for non-resolving pneumonia. After 14 days, although the patient showed symptomatic improvement, she did not show radiological improvement; thus, we planned to do bronchoscopy. On the night prior to the bronchoscopy, she had an episode of vigorous coughing

with expectoration of whitish yellowish membranous material (Figure 5), suggesting a ruptured hydatid cyst in the lung. The patient continued to expectorate pieces of similar membranous material for the next 3 days. Histopathology of this membranous tissue confirmed it to be the outer layers of a hydatid cyst. Her latest x-ray showed a ruptured cyst in the form of a lung abscess obscured by the surrounding pneumonitis (Figure 6). ELISA for Echinococcus, which was advised after the patient expectorated membranous material, turned out to be highly positive – 45.47 U/ml (N <8 U/ml). Searching through the literature, we came across the "air bubble" sign and its importance in the diagnosis of complicated hydatid cyst, and

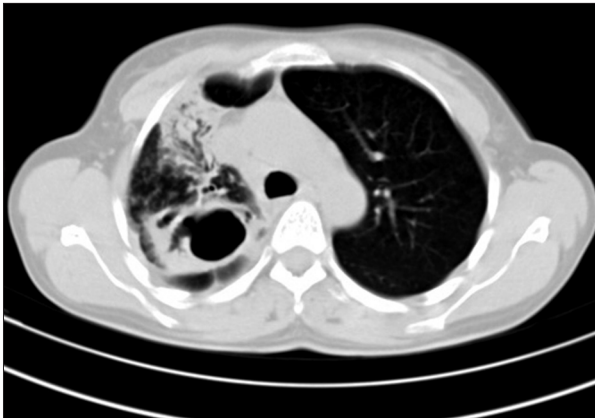


Figure 7. Chest CECT showing an “empty cyst” sign.

indeed we were able to appreciate this sign on the patient's CECT (Figure 3). Ultrasonography of the abdomen revealed no abnormality of any organ. Further, there was no history of any pet dog. On the basis of clinical, radiological, and serological findings, she was diagnosed as having a complicated hydatid cyst. The patient refused surgery and received medical treatment consisting of albendazole 400 mg twice daily for 3 weeks, to be repeated after a gap of 15 days for at least 3–4 cycles. The patient then showed marked symptomatic and radiological improvement. After 1 month, a repeat CECT chest was done, which showed an empty cyst/cavity, also known as the “empty cyst” sign (Figure 7). She is on regular follow-up and continuing medical treatment.

Discussion

Pulmonary hydatid disease is a serious problem in many countries, including several Mediterranean countries, New Zealand, Australia, North America, South America, Central America, and Asia [7]. Infection with the larva of the tapeworm of the genus *Echinococcus* is responsible for hydatid disease, also known as echinococcosis or hydatidosis. Although, dogs and some wild carnivores are the primary hosts, it spreads in humans, who are the intermediate host, by incidental direct contact with infected dogs or their feces. The disease is characterized by cyst formation in various organs of the body, with the liver being the most common site of infection, followed by the lungs. The hydatid cyst colonizes the right lung more than the left lung and in most instances the cyst is solitary [8,9]. The simultaneous involvement of the liver and lung is quite uncommon but when it occurs, the right lung is affected in 97% of the cases [10]. In our case the hydatid cyst was located in the upper lobe of the right lung.

The clinical presentation and the complications associated with pulmonary hydatid cyst depend on whether the cyst is simple or complicated. Most patients with simple pulmonary hydatid

cyst are asymptomatic, but depending on the size and its location in the lung, it can produce some nonspecific symptoms such as cough, chest pain, and hemoptysis [11]. A complicated hydatid cyst is a cyst that has ruptured into the bronchus or pleural cavity, with or without infection [12]. The patients with complicated cyst, other than cough, dyspnea and chest pain present with symptoms such as expectoration of cystic contents (salty sputum and grape skin-like membrane), persistent hemoptysis, productive sputum, and fever [12].

Routine hematological and biochemical tests are unhelpful in the diagnosis of hydatid disease. Casoni test, Weinberg complement fixation test, and total eosinophils counts cannot be relied upon [1]. Serology and imaging modalities in combination secure the diagnosis in most cases. With a sensitivity of 85.3%, immunoglobulin G enzyme-linked immune-sorbent assay (ELISA) test is the most sensitive serological test in detecting pulmonary hydatid disease [13].

Among the various imaging modalities, x-ray of the chest is the initial investigation of choice. Unruptured pulmonary hydatid cyst usually shows as a homogenous round or oval-shaped mass with smooth borders surrounded by normal lung tissue on chest radiograph; many times it can be an incidental finding, with the x-ray ordered for some other ailment [14]. As a consequence of cyst rupture, the complicated hydatid cyst may simulate a lung abscess, malignant tumor, tuberculosis, and other infected cystic lesions of the lung [5,6], which complicate the clinical picture further, as also seen in our case. Among the different radiological signs of ruptured hydatid cyst, “crescent” or “meniscus” sign, “double arch” or “Cumbo's” sign, “water lily” sign or sign of “camelotte”, “daughter cyst” or “rising sun” sign, “serpent” sign, “Monod's” sign, and “air bubble” sign are important signs [15].

Contrast-enhanced computed tomography (CECT), is an important imaging modality, especially in evaluation of the complicated hydatid cyst. Of the various signs associated with hydatid cyst, the “air bubble” sign is reported to be very sensitive and specific (85.7% sensitivity and 96.6% specificity) in establishing the diagnosis of complicated hydatid cyst [16]. It is best seen in the mediastinal window as single or multiple small, rounded radiolucent areas with sharp margins within the periphery of a solid mass lesion. Dissection of air between the pericyst and parasitic membrane due to erosion of a bronchiole by an expanding cyst is thought to be responsible for this sign [17]. Compared to classical CT signs, the “air bubble” sign is more accurate in the correct diagnosis of ruptured, infected, solid hydatid cysts [16]. The presence of this sign in our case, along with a positive serology, helped us in making the correct diagnosis. Moreover, attenuation values in infected cysts are considerably higher (above 20 HU) than in unruptured cysts. This solid density of the infected hydatid

cyst makes it very difficult to differentiate from an abscess or neoplasm [18]. Solid appearance on CT scan prevents the correct differentiation between hydatid disease and malignant tumor. Pursuit of correct diagnosis leads to further invasive diagnostic investigations such as bronchoscopy, transthoracic needle aspiration, and abdominal and cranial CECT [6]. The mass-like appearance with a high attenuation value (24HU) on CECT and our lack of awareness/knowledge regarding the “air bubble” sign led us to undertake certain avoidable investigations and delayed the diagnosis. In a few cases, follow-up CECT scan obtained 2–3 months later shows an empty cyst after complete expectoration of the contents, known as the “empty cyst” sign [19]. This sign was also appreciated in our case in the follow-up CECT obtained 1 month later.

Bronchoscopy is not the preferred investigation in patients with pulmonary hydatid cyst who present with a typical clinical picture and radiological appearance. However, it may be performed when a tumor is suspected or when the radiological picture is atypical [20], as in our case. Specific and non-specific bronchoscopic findings for pulmonary hydatid cysts have been described in adults, a whitish-yellow gelatinous membrane being the solitary specific finding [21]. We had also planned to do bronchoscopy, but decided against it once the patient expectorated whitish – yellowish membranous material, a common finding in the ruptured hydatid disease.

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Regarding management, surgery is the treatment of choice for patients who are fit and willing to undergo surgery [22]. The surgical options for lung cysts include lobectomy, wedge resection, pericystectomy, intact endocystectomy, and capitonnage [20]. Medical therapy with benzimidazoles is beneficial in disseminated disease, including secondary lung or pleural hydatidosis and in poor surgical risk patients [23]. Albendazole, the preferred benzimidazole because of its better bioavailability, is given at a dosage of 10–15 mg/kg body weight in 2 divided doses and the usual dose is 800 mg daily. Therapy is most often indicated for a minimum of 3–6 months [24].

Conclusions

By reporting this case, we would like to emphasize that the knowledge and awareness of various signs in radiology associated with the hydatid cyst, in particular the “air bubble” sign, is imperative in making a prompt and accurate diagnosis of a complicated hydatid cyst.

Conflict of interest

The authors declare that there is no conflict of interest, financial or otherwise, related to the publication of this study or its findings.