Challenging Dilemma: Is it Rainstorm, Hailstorm, Snowstorm, or Snowball?

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Abstract

The increasing use of lung ultrasound has consistently shown its high sensitivity compared to stethoscopes and radiographs for acute inflammatory and cardiovascular pathologies. It saves time, is relatively cost-effective, bedside, and provides information on adjacent systems to the respiratory system while avoiding radiation exposure [1-6]. Lung ultrasound holds particular value not only in detecting abnormal lung conditions but often plays a decisive role in diagnosis or further diagnostic steps. Lesions near the thoracic wall can be identified using lung ultrasound, which provides good imaging for diagnosing lung diseases. Ultrasound examination of the liver may reveal concurrent liver involvement in up to 15% of individuals with lung echinococcus cyst [7,8]. In rare cases, the presence of fluid in the thoracic cavity is mistakenly identified through lung ultrasound as simple parapneumonic pleural effusion [9]. In this study, we aim to demonstrate the importance of lung ultrasound in resolving diagnostic dilemmas: plankton sign, hailstorm, snowstorm, or snowball [10-13] in a case of lung echinococcus cyst. The video image of the presentation of lung echinococcus cyst is nearly unique in our knowledge.

Video Link: https://www.gavinpublishers.com/assets/videos/RPReplay-Final1718883380.mp4

Keywords: Lung Ultrasound; Echinococcus Sand; Snowball Sign.

Introduction

In lung ultrasound examination, punctate, hyperechoic foci floating within the pleural effusion are referred to as the plankton sign. This sign may indicate underlying infection, hemothorax, or chylothorax [13]. However, hyperechoic foci swirling during ultrasound examinations in real-time may represent the “snowflake” sign, characteristic of echinococcus cyst sand [10-12].

Case Report

A 54-year-old patient presented to the pulmonologist with complaints of discomfort at the base of the right lung, a temperature of 37.1-37.5 degrees Celsius, a cough with scanty sputum, fatigue, and sweating. The symptoms began two months ago, starting with discomfort at the base of the right lung. Medical history revealed a non-invasive prostate intervention and treatment for a right lung pneumothorax one year ago. The patient was a non-smoker and had consumed 100 mg of alcohol daily for 20 years. Family history was unremarkable. Physical examination and blood tests showed normal results, except for reduced excursions of the right hemithorax compared to the left. A chest X-ray revealed increased basal pulmonary density in the form of opacity on the right lung, with a negative silhouette sign relative to the diaphragm. The
patient was admitted with a diagnosis of a right basal mass of undetermined nature.

Pending computed tomography examination, the patient underwent a lung ultrasound. The ultrasound quickly showed a hypoechoic density suggesting fluid between the liver’s diaphragm and the lung. However, tiny hyper-echo foci, clustered and gathered next to each other in a bright heterogeneous ball at the bottom of the hypoechoic image, surprised us as an image never seen before. This reminded us of the diagnostic challenges described in the literature: hailstorm versus snowstorm [10,11]. In this case, there was no floating or swirling of hyper-echo foci; the punctate foci didn’t move, and the image resembled a snowball, suggesting another rare image of pulmonary echinococcus. In the short axis view, volume reduction of the hypoechoic density and movements of the double-layered wall were found. Minimal pleural fluid was evident.

Lung CT with contrast revealed a cystic mass with lung subatelectasis caused by the cyst volume’s mass effect, suggesting a diagnosis of lung echinococcus. Minimal pleural effusion was evident. The serology test for echinococcus antigen, and the anatomopathological examination confirmed the diagnosis of cystic echinococcus. Treatment with albendazole began.

Discussion

Ultrasound examination of echinococcus cysts can clearly reveal the double wall in the periphery of the hypoechoic density, the hydatid sand, membranes, daughter cysts, and vesicles inside the hydatid cyst [14]. Hydatid cyst type I of the liver can be differentiated from simple liver cysts by ultrasound examination with 96% and 98% sensitivity and specificity, respectively. In comparison, CT can differentiate hydatid cyst type I with 80% and 62% sensitivity and specificity, respectively [15].

In our case, the ultrasound of the echinococcus cyst demonstrated a double-layered wall in the periphery of the hypoechoic density and hydatid sand that didn’t float, resembling the appearance of a “dirty snowball.” Ultrasound is useful for detecting hydatid cysts near the chest wall. A recent advancement in diagnosing pulmonary hydatid cysts is the recognition of the “wall sign”: a double-layered wall in single-cavity cysts and a double-layered septum in multilocular cysts. Research shows that the wall sign is nearly 100% specific for diagnosing pulmonary hydatid cysts [16].

“Hydatid sand” in echinococcal cysts is composed of scolecis, which are the larval forms of the Echinococcus parasite and can develop into new adult tapeworms. They appear as tiny, highly reflective (echogenic) particles on ultrasound and are crucial for diagnosing echinococcosis; membrane fragments, which are pieces of the germative layer and the inner cyst membrane (endocyst), and can appear as floating debris within the cyst; fluid elements inside the cyst, which contain substances produced by the parasite and fragments from the endocyst, contributing to the “hydatid sand” appearance; and the fibrous capsule, which surrounds the echinococcus cyst, separating it from surrounding tissues [10-12,14].

The “snowstorm” appearance in echinococcal cysts refers to the presence of hydatid sand swirling within the cyst. This hydatid sand consists of:

1. **Protoscolices:** Larval parasites with hooklets crucial to the Echinococcus tapeworms’ life cycle.
2. **Detached Hooks:** Tiny structures that anchor the parasite inside the cyst.
3. **Proligerous Vesicles:** Daughter cysts containing protoscolices.
4. **Endogenous or Exogenous Daughter Vesicles:** Vesicles that can form new hydatid cysts similar to the initial one.

These elements create the characteristic “falling snowflakes” appearance in hydatid cysts [10,11,12,14,15,17,18,19]. The “hailstorm” presentation, characterized by a “solid, heterogeneous echotexture...with multiple echogenic nodules of varying size” [11], is a sign of Echinococcus multilocularis [20]. Lung ultrasound data are crucial not only in suggesting and establishing the diagnosis of echinococcus cysts but also in determining the biological activity of hydatid cysts and guiding treatment approaches.

According to the WHO classification of cystic echinococcosis, there are five stages:

- **CE1 (Unilocular, Simple Cyst):** Simple cystic lesion with uniform fluid content. Ultrasound features: anechoic, well-defined cyst with no internal structures.
- **CE2 (Multivesicular, Multilocular Cyst):** Daughter cysts within the mother cyst, giving a honeycomb or wheel-like pattern. Ultrasound features: septated cysts with visible daughter cysts, creating a “rosette” or “honeycomb” appearance.
- **CE3 (Transitional Cyst)**
  - **CE3a (Early Transitional):** Cysts with detached endocyst, often described as having a “water lily” or “floating membrane” sign. Ultrasound features: mixed echogenicity with floating membranes within the cystic fluid.
  - **CE3b (Late Transitional):** Cysts with daughter cysts and partial detachment of the laminated membrane, described as having a “snowflake” appearance. Ultrasound features: multiple small echogenic structures (“snowflakes”) floating in the cystic fluid, indicating hydatid sand and detached membranes.
- **CE4 (Degenerative Cyst):** Degenerating cyst with heterogeneous contents. Ultrasound features: cysts with mixed
solid and liquid areas, showing partial calcification and reduced cyst wall thickness.

- **CE5 (Calcified Cyst):** Mostly or fully calcified cysts. Ultrasound features: solid-appearing structures with dense calcification [21].

Based on the viability of Echinococcus, cyst stages are classified into three clinical groups:

1. **Active Cysts:** Viable cysts in early stages, either unilocular (CE1) or multi-vesicular with daughter cysts (CE2).

2. **Transitional Cysts:** Cysts in an intermediate state (CE3), with the endocyst detaching (CE3a) or those primarily solid but containing daughter cysts (CE3b).

3. **Inactive Cysts:** Non-viable cysts showing signs of degeneration, solidification, and calcification (CE4 and CE5). These cysts are almost always non-viable [22].

A study employing magnetic resonance spectroscopy revealed distinct metabolic profiles between the two subgroups. The metabolic profile of CE3b cysts resembles that of viable stages like CE1 and CE2, whereas CE3a cysts may exhibit characteristics of either active or inactive stages. These findings align with studies that assess biological viability microscopically after cyst removal [23].

The primary factor influencing positive serology is whether complications such as rupture or infection/abscess have occurred, as these conditions release parasite antigens [24]. Tests using hydatid cyst fluid (HCF) are more sensitive (80-99%) but less specific (60-97%) in liver cysts, whereas tests using purified or recombinant proteins are more specific (80-100%) but less sensitive (38-93%) [25,26]. False negatives can occur with young CE1 cysts in the liver (30-58%), inactive CE4-CE5 cysts (50-87%), and in extra-hepatic cases, including up to 50% of patients with lung cysts and those with cysts in other areas. Patients with active or transitional cysts (CE2, CE3a, CE3b) have lower rates of sero negativity (5-20%), and those with multiple cysts are usually sero-positive [27-29]. The seropositivity rate is lower in pulmonary cysts than hepatic cysts. The sensitivity of serological testing varies from 50–60% in the pulmonary cyst and 85% to 98% in hepatic cysts. The sensitivity is higher in patients with multivorgan involvement, 90–100% [30]. Although cysts are categorized as active (CE1, CE2, CE3b), transitional (CE3a), or inactive (CE4, CE5), the integrity loss of the cyst structure—whether spontaneous or due to therapy is more closely related to positive serology than the biological viability or activity of the cyst itself [24].

**Conclusion**

Whenever faced with hypoechoic or anechoic ultra-sonographic images suggestive of liquid in the thoracic cavity, echinococcus should be included in the differential diagnosis. The “snowball,” double-layer wall, and reduction in the size of the hypoechoic image during the respiratory cycle are characteristic findings suggesting the diagnosis of lung echinococcosis. In the diagnosis of cystic echinococcosis, given the lower sensitivity of CT compared to LUS for detecting CE1 stage echinococcus, and the lower sensitivity of immunoserological tests, lung ultrasound (LUS) is the preferred examination for peripheral masses near the chest wall.

**References**


