Pulmonary cystic echinococcosis in a farm worker - do not biopsy the benign nodule

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ABSTRACT

Background: Pulmonary hydatid cystic disease or pulmonary cystic echinococcosis is caused by the tapeworm Echinococcus granulosus. The cysts are usually incidental findings of well-defined round lesions on chest radiographs or on a computer tomography which can rupture and cause symptoms.

Case Presentation: This case report presents a complicated clinical course of pulmonary hydatid cystic disease in a 50-year-old farm worker, in whom the diagnosis was not initially thought of.

Conclusion: The discussion highlights the importance of considering this disease in the differential diagnosis of any pulmonary cystic lesion, as significant complications arose because of the investigations performed.

Keywords: Nodule, cystic echinococcosis, hydatid lung disease, case report.

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Background

For the Echinococcus granulosus (EG) parasite, definitive hosts (those supporting adult or sexually reproductive parasites) are household carnivores, such as dogs. Dogs excrete infectious eggs which then pass to intermediate hosts (organisms supporting immature or non-reproductive parasites). Those are sheep and cattle. Humans are accidental dead-end intermediate hosts, from which infectious agents are not transmitted and are infected via the fecal-oral route or direct contact with definite hosts [1-3].

The parasites travel by hematogenous and lymphatic routes to visceral organs. A fluid-filled cyst then develops at the site of organ entry. The cyst has an outer layer (pericyst), formed by the protective response of the host. The middle layer (ectocyst) is acellular to allow passage of nutrients. The inner layer (endocyst) produces the scolices (the anterior ends of tapeworms, bearing suckers and hooks for attachment) [1-3].

The liver is the most affected in adults (75%), followed by the lungs (15%). The cysts may remain asymptomatic for years until compressive symptoms or rupture occur [3]. Hydatid cyst disease is endemic in sheep and cattle-rearing areas, such as Australia, New Zealand, Southern Europe, Turkey, the Middle East, India, and South and Central America. It is increasingly recognized in Western Europe in farming communities [1].

Case Presentation

A 50-year-old female patient presented with a 4-week history of left-sided chest pain. There were no red flag symptoms. She had never smoked, had no occupational dust exposure, and no past medical history. She was a nurse and lived on a farm with dogs, sheep, and cattle. She had a normal examination and observations. Full blood counts and inflammatory markers were normal. Her chest radiograph (CXR) showed a homogenous well-defined rounded opacity in the left lower lobe (Figure 1). Her blood tests were all normal. A contrast-enhanced computer tomography (CT) was performed. Figure 2 shows a cystic lesion in the posterolateral aspect of the left lower lobe measuring 45 × 30 mm. Cysts were noted in the right lobe of the liver. The local cancer multidisciplinary team meeting felt that the lesion was a benign pseudo-tumor, with an encysted effusion or a post-inflammatory lesion close to the pleura. The patient wanted to have the lesion biopsied for a definitive diagnosis rather than just having repeated scans to monitor any potential growths. Hence, a CT-guided fine needle aspiration (FNA) was performed under local anesthetic. Clear fluid was aspirated and sent for microbiological and histopathological analysis. After the biopsy, the patient developed progressive breathlessness. A CXR showed an iatrogenic left

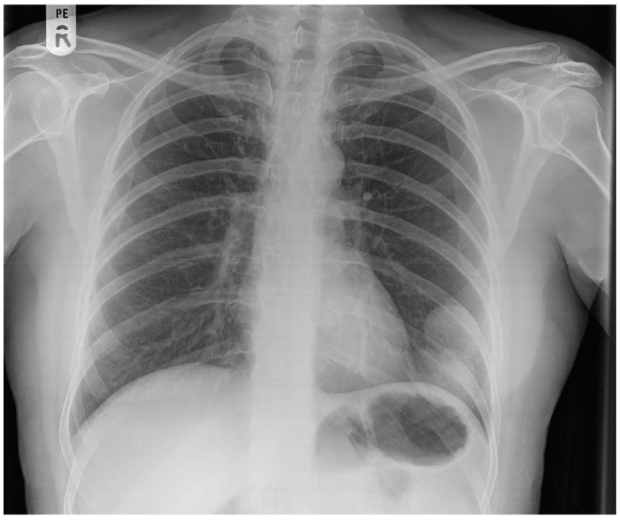

Figure 1. *CXR showing a homogenous well-defined rounded opacity in the left lower lobe.*

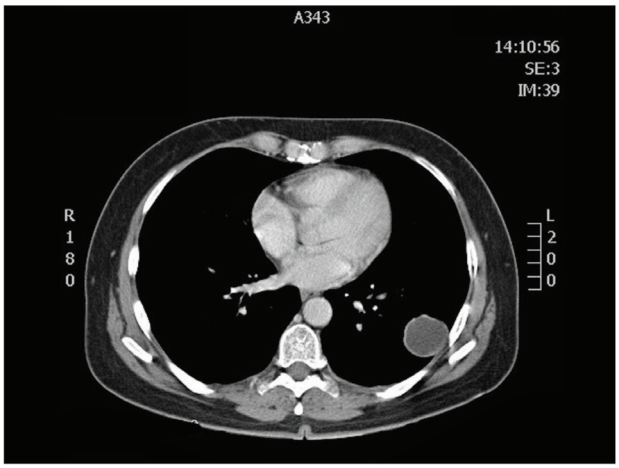

Figure 2. *CT scan showing the cystic lesion in the posterolateral aspect of the left lower lobe.*

hydro-pneumothorax which was aspirated with good clinical and radiological resolution. Cytology showed numerous rounded scolices from the brood capsule (one of the secondary scolex-containing cysts that are proliferated from the lining of a hydatid) and elongated hook-like structures which were in keeping with parasitic EG infection. There were no malignant cells seen. Cultures for fungus, legionella, mycobacteria, and extended anaerobic organisms were all negative.

Albendazole 400 mg twice daily and praziquantel 1.25 g in two doses, 6 hours apart, once weekly were started. Over the following 2 weeks, the patient developed lethargy, anorexia, a dry cough, and further dyspnea. On review, she had a temperature of 38°Cand a heart rate of 110 beats per minute, with normal oxygen saturations and blood pressure. A bedside thoracic ultrasound showed a moderate left-sided pleural effusion with multiple septations. A CT scan showed a large left-sided pleural effusion with a loculated hydro-pneumothorax and passive lung collapse around enlarged cystic lesions within the lung itself (Figure 3A-D). Aspiration of the pleural fluid was unsuccessful. Her white cell count was 23.2 × 10^9/l (4-11) and C-reactive protein was 152 mg/l (<5). Neutrophil count was 16.1 × 10^9/l (2-7.5) and eosinophil count was normal at 0.06 × 10^9/l (0-0.4).

Cyst contents had spilled into the pleural space, resulting in empyema. There was also the possibility of a superadded bacterial infection, so broad-spectrum intravenous antibiotics were started. Anti-helminthic medications were continued. A liver magnetic resonance (MR) imaging scan showed liver cysts not to be hydatid and benign in nature.

A multidisciplinary meeting about her case involved respiratory and infectious disease specialists and cardiothoracic surgeons. Given the extent of the pleural disease, failure to aspirate any fluid and the potential lack of response due to poor pleural penetration of anti-parasitic medications, she underwent left lung decortication and closure of cyst cavity via a left posterolateral thoracotomy. Intra-operatively, 1 L of pale yellow, clear fluid with fibrinous debris contained in two cysts was removed from the pleural space. Decortication of the thickened lower pleura managed to completely free the lung and achieved good lung expansion. The pleural space was washed with 20% hypertonic saline, and the cysts were filled with iodine for 15 minutes and closed by capitonnage.

Postoperative course was uneventful. CXRs showed a fully expanded lung and a resolving pleural effusion. Praziquantel 20 mg /kg twice daily was continued for 14 days post-operatively and albendazole 10 mg/kg twice daily for 1 year. Her wound healed well and repeat CXRs showed minimal left lung volume loss.

She was followed-up by the infectious diseases team at 3, 6 and 12 months. Her hydatid enzyme-linked immunosorbent assay (ELISA) test was only showed a weak positive reaction at 12 months and her albendazole was then stopped. Her indirect hemaglutination (IHA) hydatid assay at 12 months also showed an equivocal result of titer levels of less than 1:320. She only ever had an ELISA and IHA test at 12 months. She was then discharged from follow-up.

Discussion

Hydatid cysts are normally asymptomatic, and their clinical presentation depends on their size, location, and relationship to any organs. Lung cysts present non-specifically with cough, chest pain, dyspnea, and malaise. They can rupture into the bronchial tree (with expectoration of clear, salty, or peppery-tasting fluid-containing hydatid membranes) or into the pleural cavity. Dissemination of cyst material can cause anaphylaxis. On CT or MR imaging, uncomplicated pulmonary cysts appear as well-rounded lesions with homogenous content whose density is close to water. Calcification and daughter cyst formation are rare in lung hydatids. El Fortia et al. [4] reported that thoracic ultrasound can demonstrate the “wall sign”:

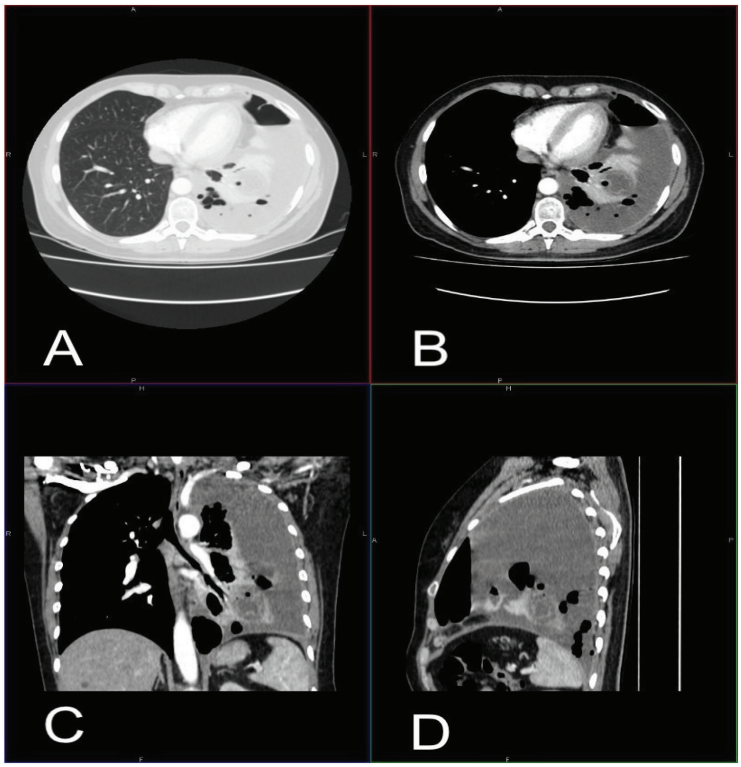

Figure 3. *(A-D) Coronal and sagittal CT scan with lung windows showing a large left-sided loculated hydro-pneumothorax and a passive lung collapse around the enlarged cystic lesions within the lung itself.*

a double-layered wall in univesicular cysts or a double-layered septum in cases of multivesicular cysts has a specificity approaching 100% for pulmonary hydatid cysts. Peripheral blood eosinophilia occurs in less than 15% of the cases and is more frequent with cyst leakage. Our patient never demonstrated eosinophilia. An ELISA or IHA test can be used as an initial screen but are positive in only approximately 50% of the pulmonary disease and almost always negative in isolated simple pulmonary cysts. Serological testing has a lower specificity and sensitivity in the presence of other parasitic infections, cancer, and autoimmune disorders [3,5-7,8,9]. The Casoni test was once a major hypersensitivity-based skin test used to detect hydatid disease but is only 63.8% sensitive and 47% specific, and has thus been superseded by serological testing [10].

Observation of cysts is acceptable but in case of diagnostic uncertainty, needle biopsy or cyst resection may be performed [3]. However, while fine needle aspiration (FNA) is a sensitive diagnostic tool for nodules, it is not recommended for pulmonary hydatid cysts due to the risk of content rupture and subsequent infections, anaphylaxis reactions, and dissemination of the disease [5, 8,11,].

Hydatid lung disease was not part of the initial differential diagnosis and the patient wanted to have a biopsy of the lesion. Hence, the FNA caused the rupture of the cyst contents into the pleura and all the other complications ensued. If there had been a clinical suspicion of a hydatid cyst, serological testing and clinic-radiological correlation would have perhaps been enough.

A single benzimidazole (BMZ) is efficacious in small uncomplicated cysts. Monotherapy was not considered suitable here due to potential risk of cyst rupture. Combining albendazole with praziquantel results in higher scolicidal and anti-cyst activity and has better outcomes [12].

For uncomplicated cysts, puncture, aspiration of cyst contents, injection of protoscolicidal agent for at least 15 minutes and reaspiration (PAIR) in combination with chemotherapy is preferable. PAIR is performed under ultrasound or CT and is repeated until the cyst contents run clear [3,7,11,12]. This was not appropriate here given the extent of her pleural disease. Surgery aims for completely cyst excision via a thoracotomy or video-assisted thoracotomy, with maximum preservation of lung tissue. Spillage of cyst content should be minimized intraoperatively. Capitonnage is recommended for obliteration of the residual cavity and pericyst by placing multiple mattress sutures in the cavity wall [11].

All patients postoperatively should receive a BMZ as recurrence rates approach 11% without. The optimal duration is not known but is believed to be a minimum of 3-6 months [3,5,8].

For an empyema, the British Thoracic Guidelines suggest that aspiration of fluid should be attempted (this failed here due to the significant number of loculations) and a chest drain is inserted if the pH of the fluid is less than 7.2 or frank pus is aspirated [13]. Antibiotics should be tailored to any positive microbiological growth. However, a direct approach to surgery was the clinician-preferred and patient-centered option here and yielded excellent results.

Conclusion

EG is a parasite that is passed to humans from dogs and cause hydatid cysts. Pulmonary hydatid cysts are usually incidental findings on CXRs or CT scans. A high index of suspicion is required, with the medical history a very important aspect of the investigative pathway. Pulmonary hydatid cysts can rupture into the pleural space or the bronchial tree. Serological testing for pulmonary hydatid cysts is neither specific nor sensitive. Management of pulmonary hydatid cysts can involve observation, aspiration of the cyst, surgery, and anti-parasitic medications for at least 6 months.

What is new?

It is well known that pulmonary hydatid cysts should not be biopsied. We made an error and wish to educate others about this.

List of Abbreviations

BMZ	benzimidazole
CT	computed tomogram
CXR	chest radiograph
ELISA	enzyme-linked immunosorbent assay
FNA	fine needle aspiration
IHA	indirect hemaglutination
PAIR	puncture, aspiration of cyst contents, injection of protoscolicidal agent for at least 15 minutes and reaspiration

Funding

None.

Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this case report.

Consent for publication

Written informed consent was taken from the patient.

Ethical approval

Ethical approval is not required at our institution for publishing an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Female, 50 year old
2	**Final diagnosis**	Disseminated hydatid disease
3	**Symptoms**	Chest pain initially
4	**Medications**	None initially. Albendazole for a year
5	**Clinical procedure**	Biopsy of lung nodule and VATS procedure for empyema
6	**Specialty**	Respiratory and Infectious diseases