CASE REPORT

The non-resolving lung cavity: a case of pulmonary cystic echinococcosis

Simon Mifsud,¹ Emma Louise Schembri,¹ Jonathan Gauci,² Adrian Mizzi,³ Charles Mallia Azzopardi,⁴ Josef Micallef²

¹Department of Medicine, Mater Dei Hospital, Msida, Malta

²Department of Medicine, Respiratory Division, Mater Dei Hospital, Msida, Malta

³Department of Medical Imaging, Mater Dei Hospital, Msida, Malta

⁴Department of Medicine, Infectious Disease Division, Mater Dei Hospital, Msida, Malta

Correspondence to

Dr Simon Mifsud, mifsudsimon@hotmail.com

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SUMMARY

The authors report a case of a 20-year-old woman who was diagnosed with pulmonary cystic echinococcosis. She was admitted to hospital with a 1-week history of unresolving cough, coloured sputum with occasional haemoptysis and fever despite oral antibiotics. Radiology revealed a cavitating right lower lobe lung abscess. After 4 weeks of treatment, follow-up radiology showed incomplete resolution. Bronchoscopy revealed a white, avascular cystic lesion in the right lower lobe and serology testing for *Echinococcus granulosus* was positive. Repeat imaging eventually confirmed the cystic lesion with the 'air bubble' sign. A thorough travel history, a high index of clinical suspicion and close follow-up are essential in making a diagnosis of pulmonary cystic echinococcosis.

BACKGROUND

Echinococcosis or hydatid disease is a parasitic infection caused by the metacestode stage of the tapeworm *Echinococcus*.¹ Pulmonary echinococcosis is an uncommon diagnosis, especially in non-endemic countries. Most cases remain asymptomatic for many years, leading to a delayed presentation and associated complications. The authors report a case of a non-resolving lung cavity which proved to be a case of pulmonary cystic echinococcosis.

CASE PRESENTATION

A 20-year-old female non-smoker presented to the emergency department in view of a 1-week history of cough productive of yellow sputum with

occasional haemoptysis, swinging fever up to 39°C and night sweats. Her symptoms did not improve on oral levofloxacin prescribed by her general practitioner. She was a Maltese national who regularly visited her extended family in Turkey.

Physical examination revealed fever of 39°C, tachycardia (116 bpm) and tachypnoea (24 breaths/min). She was normotensive and had an oxygen saturation of 97% on room air. Examination of the chest revealed signs of a right lower lobe consolidation.

Laboratory data revealed a leucocytosis of $13.99 \times 10^9/L$ with a neutrophil shift and a low eosinophil count ($0.06 \times 10^9/L$). C reactive protein was elevated at 236.8 (normal values: 0–10 mg/L). Serum creatinine, electrolytes and liver function tests were within normal limits. Peripheral blood cultures were negative. The chest X-ray (CXR) revealed a large cavitating lesion in the right middle and lower lung zones (figure 1).

The patient was started on intravenous piperacillin-tazobactam. CT scan of the thorax confirmed the presence of a cavitating lung abscess in the right lower lobe (figure 2). Sputum cultures, sputum for acid-fast bacilli, mycology investigations, galactomannan antigen and an autoimmune screen were negative. Following 7 days of intravenous antibiotics, she had improved markedly both clinically and biochemically. She was discharged on oral antibiotics for another 3 weeks with a plan to repeat a CXR in 4 weeks.

During her follow-up visit, the patient felt better but still had residual cough productive of yellow

Figure 1 Chest X-ray showing a large cavitating lung lesion projected over the right middle and lower zones.

Figure 2 CT thorax in lung window setting showing a thick-walled right lower lobe cavitating lesion containing an air fluid level.

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Figure 3 Repeat CT thorax in lung window setting showing a residual soft tissue mass in the right lower lobe.

sputum and occasional low grade fever. A repeat CXR showed a persistent opacity in the right lower zone. She was thus readmitted for further investigations and intravenous antibiotics. A CT scan of the thorax and abdomen revealed a 5 cm soft tissue mass with no residual cavitation in the right lower lung lobe (figure 3). The intra-abdominal organs were unremarkable. A flexible bronchoscopy revealed a white smooth avascular cystic lesion in the posterior basal segmental bronchus of the right lower lobe (figure 4). Cytology was negative for malignant cells.

Figure 4 Right posterior basal segmental bronchus lesion on flexible bronchoscopy.

Figure 5 CT thorax in mediastinal window setting, showing a thick-walled cystic lesion and the 'air bubble' sign.

The differential diagnosis for this persistent lesion included a lung abscess, tuberculosis, malignancy (primary or metastatic), aspergillosis and pulmonary hydatid cyst.

The case was discussed at the respiratory multidisciplinary team meeting, where the possibility of a pulmonary hydatid cyst was raised. Serological testing for *Echinococcus granulosus* proved positive at 1:256. On further questioning, the patient admitted to close contact with animals at her grandfather's farm in Turkey.

The patient was started on albendazole 400 mg twice daily; however, her cough persisted and a few weeks later, she suddenly woke up expectorating clear fluid mixed with haemoptysis. A repeat CT revealed a thick walled cystic lesion in the right lower lobe of the lung containing fluid and gas (figure 5). This radiological finding is known as the 'air bubble' sign and is a result of air entry inside the cyst due to erosion of a bronchiole. It is characteristic of a pulmonary hydatid cyst that has ruptured into the bronchial tree.

A cardiothoracic surgical opinion was sought and the patient was scheduled for a right thoracotomy and cystectomy 4 weeks after the initiation of albendazole therapy. During the surgery, there were multiple adhesions making complete surgical resection difficult. The cyst was opened and drained and marsupialised. Histology revealed the germinal membrane of the cyst (figure 6), with a mixed inflammatory infiltrate composed

Figure 6 Low power view of the cyst germinal membrane.

Figure 7 Low power view of suppurative inflammation in the adjacent lung tissue.

predominantly of neutrophils and eosinophils but without viable scolices. The adjacent lung parenchyma showed suppurative inflammation (figure 7).

OUTCOME AND FOLLOW-UP

Postoperatively, the patient continued treatment with albendazole and is planned to receive treatment for another 6 months. She is being followed up regularly on an outpatient basis and has remained well. Blood tests are within normal limits, and her CXR shows postoperative changes. A repeat bronchoscopy 4 months later showed clear bronchi, with no evidence of the cystic hydatid wall.

DISCUSSION

Echinococcosis or hydatid disease is a parasitic infection caused by the metacestode stage of the tapeworm *Echinococcus*.¹ The most common species responsible for human echinococcosis is *Echinococcus granulosus* followed by *Echinococcus multilocularis*.^{2,3} *E. granulosus* has a worldwide distribution and humans are frequently exposed to it especially in sheep-raising areas.^{3,4} *E. multilocularis* is confined to cold and wild ecosystems and the incidence of human infection is significantly less common than infection due to *E. granulosus*.⁵ Human echinococcosis is an incidental process as humans do not play an essential role in the echinococcal life-cycle.^{5,6}

Echinococcosis due to *E. granulosus* infection is an important public health concern in Turkey especially in rural areas.¹ In our case, the patient went to Turkey on regular occasions, where she was exposed to her family's farm animals including sheep and goats.

The life-cycle of *E. granulosus* involves a definitive carnivore host (dogs and other canines) and an intermediate herbivore host (sheep, cattle, pigs and goats).⁵ The mature adult tapeworm resides in the definitive host's small intestine. The gravid proglottids of the adult tapeworm shed embryonated eggs into the faeces of the definitive host. These eggs are surrounded by a tough protective shell and can survive for many weeks. To complete the life-cycle, these eggs are then ingested by herbivores (the intermediate host) when grazing on contaminated grounds. Once inside the intermediate host's intestine, the egg loses its shell and releases an oncosphere. The oncosphere penetrates the intestinal wall, following which it enters the portal venous system. These oncospheres then migrate to the liver

Figure 8 The life-cycle of *Echinococcus granulosus*.

where they may deposit and develop into cysts. These cysts enlarge and produce protoscolices that fill the cyst's interior. If the oncospheres are not filtered by the liver they may become trapped in the pulmonary circulation and cause cystic disease in the lungs. Uncommonly, the oncospheres may escape both liver and lung filters and disseminate into the systemic circulation. The definitive host becomes infected after consuming the cyst-containing organs of infected intermediate hosts. Once inside the definitive host, the protoscolices evaginate and attach to the intestinal mucosa where they then develop into the adult tapeworm.^{5,7,8} The life-cycle of *E. granulosus* is summarised in figure 8. Humans participate in this life-cycle by ingesting food that is contaminated with faecal material containing embryonated eggs from an infected carnivore.⁷ The authors present the case of a patient who probably contracted the disease by ingesting food which was contaminated with faecal material from an infected carnivore.

Most cases of human echinococcosis infection are initially asymptomatic as the parasite has developed mechanisms to avoid detection by the host's immune system.⁵ In fact, most cases of pulmonary echinococcosis are discovered incidentally on a chest radiograph.⁹ Symptoms usually become evident when complications occur, such as rupture of a cyst (which can lead to anaphylaxis),¹⁰ compression of vital structures, haemorrhage and secondary bacterial infection.¹¹ Uncommonly, intact pulmonary hydatid cysts result in non-specific symptoms including cough, chest pain and haemoptysis.¹² Lung cysts tend to expand at a slightly faster rate (1–5 cm/year) than liver cysts⁵ due to the greater elasticity of lung parenchyma.¹³

The patient's initial presentation with a productive cough and swinging fever was due to a bacterial infection causing consolidation and cavitation of the lung in close proximity to the cyst. This explains her initial response to antibiotic treatment with resolution of the abscess. One could then appreciate the presence of the cystic lesion with the 'air bubble' sign, confirming the presence of a complicated hydatid cyst.

The diagnosis of pulmonary cystic echinococcosis is based on the patient's history (with particular emphasis on travel history to endemic regions) and imaging findings.¹⁴ Serological tests provide supportive evidence for the diagnosis of pulmonary cystic echinococcosis.¹¹ While a chest radiograph is useful in detecting lung cysts in the majority of patients, a contrast enhanced CT scan of the thorax is often necessary as it allows for

better evaluation in complicated cysts and helps to rule out alternative diagnoses.⁹ In our case, the presence of the 'air bubble' sign was of significant importance in the diagnosis of hydatid disease. This sign has a high sensitivity and specificity for a complicated hydatid cyst.¹⁵

In countries where the disease is not endemic and exceptionally uncommon such as Malta, pulmonary echinococcosis can be a challenging diagnosis especially when the characteristic cystic lesions are not picked up on imaging. Pulmonary cystic echinococcosis requires a high index of clinical suspicion as there is no routine specific laboratory test. The typical eosinophilia usually occurs after leakage of some of the antigenic cystic fluid.⁵ Serological tests are unreliable as they produce many false-positive and false-negative results.⁵ In fact, several sources maintain that negative serological tests do not exclude the disease.^{5 11}

In addition to the travel history, a helpful pointer to the diagnosis in our case was the presence of a cystic lesion in the right lower lung lobe on repeat CT scan. This is a typical finding, since 60% of pulmonary cystic echinococcosis cases involve the right lung,^{5 7} and 60% involve the lower lobes.^{7 13} It is worth noting that absence of liver involvement does not make pulmonary cystic echinococcosis unlikely since only 5%–13% of patients present with simultaneous pulmonary and liver involvement.¹⁶

Treatment for cystic echinococcosis depends on various factors including age, number, size and location of cysts, cyst wall calcification and concomitant severe disease. The WHO guidelines on echinococcosis suggest the following treatment options for *E. granulosus* infection:

1. Surgery—indicated for cysts in the liver, lung, brain, kidneys, bones and other organs.¹⁰
2. Puncture, aspiration, injection and reaspiration—indicated for inoperable patients with cystic echinococcosis involving the liver, abdominal cavity, spleen, kidney and bones. This therapeutic modality should not be used in lung cysts.¹⁰
3. Medical treatment with benzimidazoles—indicated in inoperable patients with primary liver or lung echinococcosis and for those with multiple cysts in two or more organs.¹⁷ The benefit of benzimidazoles is largely dependent on the cyst wall thickness. The thinner the cyst wall, the more effective the medical treatment.¹⁰
4. Wait and observe approach—indicated for those with homogeneously calcified cyst walls.¹⁰

Surgery has the potential of removing the cysts and can hence lead to a complete cure.¹⁰ Presurgical use of benzimidazoles is recommended in liver cystic echinococcosis,¹⁷ but there is conflicting evidence with regards to preoperative benzimidazole use in pulmonary hydatid cysts. Albendazole therapy should be avoided preoperatively in lung cysts as this may reduce cyst wall tension, increasing the risk of rupture resulting in dissemination.^{17 18} It may be prescribed preoperatively in cases of ruptured pulmonary cysts.¹⁸ Postoperative use of benzimidazoles is beneficial as it can reduce the risk of postoperative recurrence.¹⁰ The WHO recommends regular monitoring of leucocyte counts when patients are being treated with benzimidazoles as these can lead to a non-reversible leucopenia.¹⁰

Learning points

- Pulmonary echinococcosis should be considered in the differential diagnosis of a cavitating lung lesion.
- A high index of clinical suspicion leads to early diagnosis and management of pulmonary cystic echinococcosis, thus preventing its potentially serious complications.
- Both medical and surgical measures have a role in the effective treatment of echinococcosis.

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