Case Report

# Pulmonary echinococcosis presenting as a pulmonary mass with fever and haemoptysis; a case report

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**Key words:**
- pulmonary echinococcosis
- Echinococcus granulosus
- Hemoptysis

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**SUMMARY. Hydatidosis, which is caused by larval stage of *Echinococcus granulosus*, is the most frequently encountered type of anthropozoonosis, and it has been known for centuries. Due to the traditional close association between sheep, dogs and humans, *Echinococcus granulosus* is endemic in many parts of the world and particularly the Mediterranean countries. The case is reported of a 68 year-old man, a farmer and non smoker, who presented with a 32 day history of haemoptysis and fever. On chest X-ray a mass was observed at the hilum of the right lung. He underwent full clinical, laboratory, and endoscopic evaluation, which was unrevealing. During right thoracotomy a right lower lobectomy was performed that included the mass. Frozen section of the mass was negative for malignancy and the final histopathological examination established the diagnosis of a ruptured pulmonary echinococcus cyst with abscess formation. The postoperative course was uneventful and the patient was discharged from hospital on the ninth postoperative day being treated with albendazole *per os*. Pulmonary echinococcosis should be included in the differential diagnosis of a lung mass in patients presenting with haemoptysis, especially if there are domestic animals in their environment. *Pneumon 2010, 23(2):180-183.***

## INTRODUCTION

Echinococcosis or hydatid disease is an anthropozoonosis that has been known for centuries[1]. It is caused by larvae, which are the metacestode stage of the tapeworm Echinococcus. The vast majority of infestations in humans are caused by *E. granulosus*, which is the commonest type in the Mediterranean countries, including Greece[2]. Humans are exposed less frequently to *E. multilocularis*, which causes alveolar echinococcosis.

Although it can involve almost every organ of the body, the lung is the second site in frequency, following hepatic infestation, in adults and is the

predominant site for hydatidosis in children. The lung facilitates the growth of the cyst due to its compressible nature and the presence of negative pressure[3,4]. Clinically, a small hydatid cyst in the lung often causes no problems and may remain asymptomatic, but a large cyst may cause pressure symptoms such as chest pain, cough, dyspnoea and haemoptysis, as well as allergic reactions including anaphylaxis[5,6]. Rupture of a pulmonary hydatid cyst into a neighbouring or more remote cavity, either spontaneous or iatrogenic, is termed a "complicated cyst" and is associated with higher postoperative morbidity and mortality[7].

Classically, the chest X-ray shows an intact cyst as a sharply demarcated round-to-oval homogenous mass of variable size[6,8]. Computed tomography (CT) offers high sensitivity and specificity for the definitive diagnosis of the disease[9].

The disease is usually encountered in the younger age groups, in most cases before the age of 40 years. Regarding gender, there appears to be a predominance in males, and based on its aetiology, there is a higher incidence in rural populations[4].

## CASE REPORT

A 68 year-old man, a farmer, was transferred to the chest hospital for further evaluation of fever, haemoptysis and a lung mass revealed on the chest X-ray.

The patient had presented to the local general practitioner 32 days earlier complaining of a few episodes of minor haemoptysis (mostly blood-tinged sputum). The physician prescribed an oral antibiotic, but when the haemoptysis persisted chest X-ray and computed tomography (CT) revealed a mass in close contact with the hilum of the right lung. The mass had a maximum diameter of 5 cm, irregular margins and a mixed content of solid and cystic elements. No enlargement of lymph nodes was observed on the CT (Figures 1-3). The patient was admitted to the provincial hospital four days later with fever and continued haemoptysis, from where he was transferred after five days to the chest hospital for further evaluation and management.

The patient was not a smoker and did not report any weight loss. He had a history of hypertension and coronary artery disease. His maximum temperature did not exceed 38.2°C. On physical examination crackles were heard over the base of the right lung. The heart sounds were dual without audible murmurs, the blood pressure was 140/90 mmHg and pulse rate 80/min. There were no abdominal or neurological findings and no oedema or palpable lymph nodes. Ear, nose and throat examination was negative for bleeding.

Laboratory tests showed: Hct 41.8%, Hgb 13.9 gr/dl, PLT 430,000, WBC 15,300 with 75.3% neutrophils and 14.9% lymphocytes, blood glucose 102 mg/dl, blood urea 40 mg/dl, creatinine 1.1 mg/dl, SGOT 11 U/L, SGPT 45 U/L, LDH 114 U/L, GGT 23 U/L, Tbil 0,4 mg/dl, Na 134, K 3.9 and INR 1.11. The urine examination was within normal limits. The sputum microscopy was negative for *Mycobacterium tuberculosis*. Blood gases revealed pH 7.43, $pO_2$ 84 mmHg, $pCO_2$ 36 mmHg, with $SO_2$ 97% on air, and the spirometry values were: $FEV_1$ 2.82 (104%), FVC 3.20 (92%).

CT scan of the brain and upper abdomen, together with isotope scanning of the bones, showed no abnormal findings. The bronchoscopy revealed gross bleeding and a thickening of the mucosa at the entrance of the medial basal bronchus of the right bronchial tree. The rest of the upper and lower respiratory tract was normal. Brushing of the suspicious area was accomplished and bronchial secretions sent for cytological examination were negative for malignancy, but inflammatory cells were found.

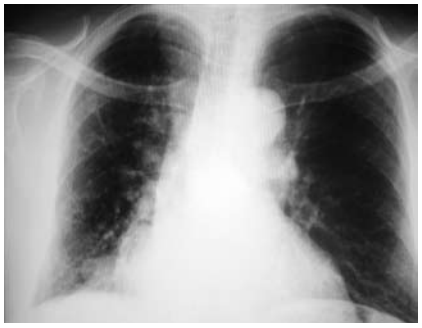

**FIGURE 1.**

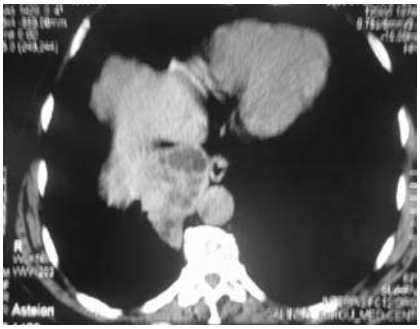

**FIGURE 2.**

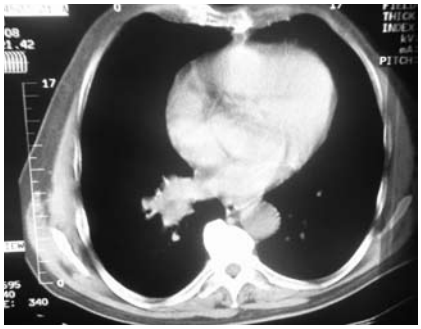

**FIGURE 3.**

The patient underwent exploratory thoracotomy ten days after admission. Following right posteriolateral thoracotomy and lysis of adhesions, a right lower lobectomy, including the mass, was performed. Frozen section was negative for malignancy and following lymph node dissection one chest tube was inserted and the chest was closed. In the final histopathological report the diagnosis of a ruptured echinoccocus cyst with small surrounding abscesses was established. All lymph nodes were negative for malignancy, showing only chronic reactive inflammatory changes.

The postoperative course was uneventful and the patient was discharged on the ninth postoperative day. With the establishment of the diagnosis treatment with oral albendazole was given for three months. At follow up 10 months after surgery the patient was doing well.

## DISCUSSION

Of patients with cystic echinococcus, 85–90% show single organ involvement and >70% harbour a solitary cyst. The liver is the most common site of cyst formation, followed by the lung, which accounts for 10–30% of cases, and various other sites (usually spleen, kidney, orbit, heart, brain and bone) in 10% of cases[10, 11]. In children, the lung may be the commonest site of cyst formation[12]. Of all patients with lung hydatid cysts, about 20–40% also have liver cysts.. Pulmonary hydatid disease affects the right lung in almost 60% of cases[13-17]. The patient presented here had a solitary lesion in the right lung.

Most intact lung cysts are discovered incidentally on chest X-ray. Occasionally, an unruptured cyst results in cough, haemoptysis and/or chest pain.[16-19] Mediastinal cysts may erode into adjacent structures causing bone pain, haemorrhage or airflow limitation. Symptomatic hydatid disease of the lung, however, most often follows rupture of the cyst. The cyst may rupture spontaneously or as a result of trauma or secondary infection. Cyst rupture may be associated with the sudden onset of cough and fever. If the contents of the cyst are expelled into the airway, expectoration of a clear salty or peppery tasting fluid containing fragments of hydatid membrane and scolices may occur[20].

Generalized symptoms of hydatid disease can result from the release of antigenic material and secondary immunological reactions that develop following cyst rupture. Fever and acute hypersensitivity reactions ranging from urticaria and wheezing to life-threatening anaphylaxis are the principal manifestations. Although allergic episodes may develop quite commonly after cyst rupture, fatal anaphylaxis is uncommon[21]. In the reported case, the patient's symptoms were fairly mild. There was no allergic reaction nor any severe pain and only slight bleeding. Hydatid disease would not be the first disease to consider with this mild and fairly common clinical presentation

Calcification, which usually requires 5–10 years for development, occurs quite commonly with hepatic cysts but rarely with pulmonary cysts, and bone hydatid cysts do not undergo calcification. Total calcification of the cyst wall suggests that the cyst may be nonviable[11].

In the majority of cases, a combination of imaging and serological methods usually yields the diagnosis of cystic echinococcosis. The most useful diagnostic method in pulmonary hydatid disease is the plain chest X-ray[14]. The typical chest radiographic appearance of uncomplicated pulmonary hydatid disease is that of one or more homogeneous round or oval masses with smooth borders, surrounded by normal lung tissue. Pulmonary hydatid cysts may range between 1 and 20 cm in diameter[22]. Large cysts can shift the mediastinum, induce a pleural reaction or cause atelectasis of adjacent parenchyma. Calcification of pulmonary cysts is rare, but the chest X-ray may reveal calcification of hepatic cysts.

CT scan with contrast may demonstrate a thin enhancing rim if the cyst is intact. CT scanning can elucidate the cystic nature of the lung mass and provide accurate localization for the planning of surgical treatment of complicated cysts[23]. In the case presented, the imaging studies are of special interest. Both the plain chest X-ray and the CT showed a pulmonary mass with irregular margins, no calcification, and a mixed content of solid and cystic elements. These imaging characteristics do not tally with hydatid disease, but conversely they are consistent with neoplastic disease.

Immunodiagnostic testing was not performed because of the low positivity of enzyme-linked immunosorbent assay or indirect haemagglutination test (only 50% of patients with pulmonary hydatidosis and >90% of patients with hepatic cysts test positive[24]). Testing for specific antibodies and antigens, such as specific immunoglobulin G1 or G4, antigen 5 (arc 5 test), and antigenB[30] are unfortunately time consuming, expensive and are not performed in this hospital.

The patient expressed a wish to be operated on, in order for a definitive diagnosis to be established and possible final treatment to be given, although for a long time clinical and laboratory/imaging evaluation had failed to confirm the diagnosis. Surgery, for patients who are

fit enough, is considered the treatment of choice since the parasite can be completely removed and the patient cured. Surgery is indicated in most cases of pulmonary cystic echinococcosis as operative mortality is low (1–2%), morbidity rates are acceptable and the recurrence rate is low (1–3%)[4,14,17,26].

Medical therapy with benzimidazoles is valuable for disseminated disease, including secondary lung or pleural hydatidosis, for poor surgical risk patients and when there is intraoperative spillage of hydatid fluid. Adjunctive chemotherapy before and after surgery appears to reduce the risk of recurrence by inactivating protoscolices, while preoperative treatment reduces the tension of the cysts for easier removal[27]. Treatment generally should begin more than 4 days prior to surgery and be continued for 1–3 months[28].

Albendazole is preferred because it has better bioavailability. It is given at a dosage of 10– 15 mg/kg body weight per day in two divided doses, and the usual dose is 800 mg daily. A newer benzimidazole compound, oxfendazole, has been studied and preliminary results suggest it may be a more effective compound[29].

In conclusion, hydatid disease presents with a variety of symptoms and sometimes with special imaging features. Because it is endemic in Greece, this disease should always be included in the differential diagnosis of haemoptysis and fever, especially in patients who live in agricultural areas.

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