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CASE REPORT

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Complete removal of a ruptured pulmonary hydatid cyst during conscious sedation bronchoscopy: A case report and literature review

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Abstract

The complete removal of a pulmonary hydatid cyst by bronchoscopy occurs rarely in clinical practice. We describe a 22-year-old male originally from Lebanon, with suspected hydatid cyst rupture on computed tomography chest after experiencing sudden onset fevers and cough whilst taking empiric anthelmintic therapy. Bronchoscopy revealed white gelatinous material in the posterior segment of the left lower lobe. The complete membranes of a hydatid cyst were removed with grasping forceps. Histologic examination confirmed the diagnosis of echinococcosis.

KEYWORDS
bronchoscopy, echinococcosis, pulmonary hydatid cyst, removal

INTRODUCTION

Hydatid cyst disease (cystic echinococcosis) is a parasitic infection caused by the dog tapeworm *Echinococcus granulosus*. The diagnosis of hydatid cyst disease is usually based on a combination of epidemiological risk factors, imaging and serology.[1] We present a case where the diagnosis of isolated pulmonary hydatid cyst disease was confirmed by bronchoscopy during which the intact membranes of a ruptured hydatid cyst was completely removed. Clinical presentation, diagnosis and management of pulmonary hydatid disease are discussed.

CASE REPORT

A 22-year-old man who recently migrated from Lebanon was incidentally found to have two well defined rounded lesions on chest x-ray during a routine medical examination for his visa application in Australia (Figure 1A). Subsequent computed tomography (CT) chest showed a 32 × 25 mm, well circumscribed, smooth walled, fluid density lesion in the left lower lobe (Figure 1B) and a similar 15 × 10 mm lesion in the right upper lobe (Figure 1C). CT abdomen & pelvis did not show any liver lesions. He was asymptomatic and had no significant past medical history. He had lived in a rural environment in Lebanon, with exposure to livestock including chickens, cows, goats, sheep and dogs.

Physical examination was normal. Baseline blood tests were unremarkable, with a normal eosinophil count of 0.1 $\times 10^9$/L (normal value: 0.0–0.5). Hydatid serology was negative at two separate laboratories using both indirect hemagglutination (IHA) and chemiluminescent immunoassay (CLIA). Pulmonary function tests were normal. He was reviewed in a tertiary hospital infectious diseases clinic and a diagnosis of probable hydatid disease was made based on epidemiological risk factors

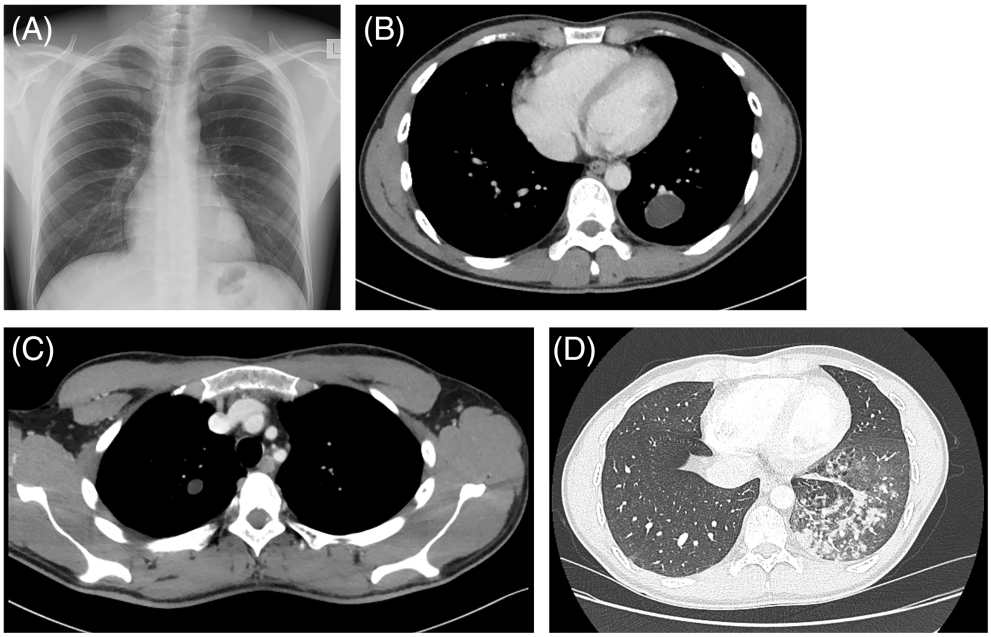

FIGURE 1 (A) Chest x-ray showing two well defined rounded lesions in the right upper zone and retrocardiac region. (B) Computed tomography (CT) chest showing a well circumscribed, smooth walled, fluid density lesion in the left lower lobe. (C) CT chest showing a rounded lesion in the right upper lobe (D) CT chest with new scattered nodular infiltrates suggestive of cyst rupture.

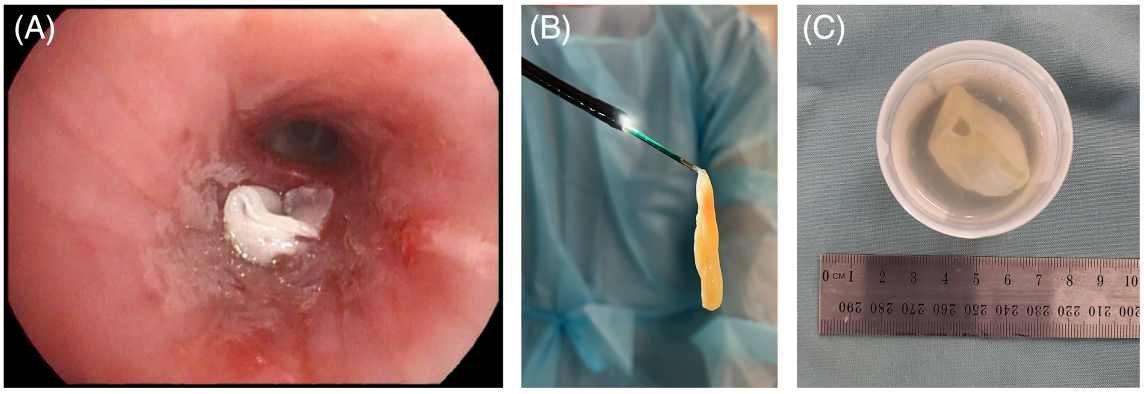

FIGURE 2 (A) Bronchoscopic image showing white gelatinous material in the posterobasal segment of the left lower lobe (LB10a). (B) Hydatid cyst membrane held with grasping forceps via bronchoscope. (C) Hydatid cyst membrane inside specimen jar.

and typical pulmonary CT features despite the negative serology. Shortly after review he commenced empiric albendazole 400 mg twice daily. One month into treatment, he experienced sudden onset fevers, rigours, myalgias and a productive cough of 2 days duration. Progress CT chest showed the left lower lobe cystic lesion had decreased in size, with scattered surrounding nodular infiltrates and ground glass opacities suggestive of cyst rupture (Figure 1D).

Flexible bronchoscopy was performed via the left nares with a white gelatinous material visualized in the posterior subsegment of the posterobasal segment of the left lower lobe (LB10a) (Figure 2A). The right upper lobe cyst was not identified. The lesion was grasped with flexible forceps and a complete membranous sack was removed en-bloc along with the bronchoscope as one unit through the nose (Figure 2B,C). Microscopy of the bronchial washings demonstrated hooklets consistent with *E. granulosus*. Histologic examination of the specimen demonstrated laminated membranes of a hydatid cyst. Progress CT chest showed complete resolution of both lung lesions at 3 months of treatment. Subsequent hydatid serology was positive (CLIA) and he developed a transient eosinophilia with a peak of 0.8 $\times 10^9$/L post-cyst rupture. He continued albendazole 400 mg twice daily and will be monitored with 6 monthly CT scans to monitor for new cyst occurrence. If no new cysts develop, albendazole will cease after 1 year of treatment. Following treatment cessation, he will be followed up with routine CT scans for 5 years to monitor for recurrence.

DISCUSSION

Human echinococcosis is a zoonotic parasitic infection caused by the larval stage of the cestodes (tapeworms) of the genus *Echinococcus*. Cystic echinococcosis (CE) also known as hydatid cyst disease, is caused by *E. granulosus* of which dogs are a definitive host. Alveolar echinococcosis disease is caused by *E. multilocularis*. CE is an underestimated global public health problem, particularly for rural communities.[2] Hydatid cysts are composed of three layers. The outer layer (pericyst) is host derived, followed by two parasite derived layers, with a thick outer laminated layer (exocyst) and an inner germinal layer (endocyst) from which protoscoleces and daughter cysts can form.[1] CE is transmitted to humans through the ingestion of hydatid eggs in dog faeces from contaminated food, water or close contact with dogs. The embryos hatch in the small intestine and penetrate through the intestinal wall to enter the portal circulation to the liver where they can deposit. Otherwise, they pass through the hepatic sinusoids, through to the inferior vena cava and settle in the lungs or other organs. Embryos can also enter the lungs through the lymphatic system or directly into the tracheobronchial tree by inhalation of air contaminated with *Echinococcus* eggs.[1,3,4] CE is endemic worldwide, but in Australia only *E. granulosus* is present at a low prevalence.[4,5]

CE is commonly asymptomatic and discovered incidentally on imaging. CE typically presents as a solitary cyst, though multiple cysts or multiorgan involvement can occur. Multiple cysts can result from rupture of a pre-existing cyst or from haematogenous spread. The liver is the most common site of involvement in 70% of cases of hydatid disease, followed by the lungs in 10%–30%.[1,4] Patients with pulmonary hydatid cyst (PHC) should be screened for the presence of hepatic cysts due to the high incidence of coexistence.[3,6] PHCs are usually unilateral (80% of cases) and predominantly involve the lower lobes (60% of cases).[7] Symptomatic PHCs are often a consequence of cyst rupture or due to compressive effects of cysts larger than 5 cm on adjacent structures.[6] Cyst rupture can be spontaneous or associated with trauma, presenting with sudden onset cough, chest pain or fever. If hydatid material is expelled into the airway and expectorated, it can result in a salty or peppery taste. Cyst rupture can also result in hypersensitivity reactions including fever, urticarial rash, wheezing and anaphylaxis. Other complications of PHCs include secondary bacterial or fungal infection, pneumothorax, haemoptysis, the development of new cysts due to haematogenous parasite dissemination and recurrent pulmonary embolism in rare cases.[1]

The diagnosis of PHC is usually based on a combination of epidemiological risk factors, imaging and serology. Uncomplicated PHCs appear on chest x-ray as homogenous, sharply defined, round or oval opacities. On CT they appear as cystic lesions with smooth walls of variable thickness with homogenous internal contents with fluid density.[1,6] Cyst rupture can produce several radiographic features that include the 'air-crescent' or 'meniscus sign', which results from the erosion of the cyst into adjacent bronchioles with the entrance of air between pericyst and exocyst, producing a radiolucent crescent in the upper part of the cyst. When there is partial emptying of the cyst fluid, a collapsed endocyst can sometimes be seen floating on top of the remaining cyst fluid producing the 'water-lily sign' or 'Camelotte sign'.[8] In contrast to extrapulmonary cysts, PHCs do not undergo calcification and daughter cyst formation is rare.[1,6] Serologic tests have sensitivity of 85%–95% for hepatic cysts but only 50%–60% for pulmonary cysts, so it is confirmatory rather than 'rule out' in pulmonary cysts.[9,10] Percutaneous aspiration of pulmonary cysts is not currently recommended due to the risk of cyst rupture, dissemination of cyst contents and anaphylaxis but there are several reports of the percutaneous approach being successful in the treatment of pulmonary hydatid cysts.[10–13]

Pulmonary hydatid cysts can be treated medically and/or surgically depending on factors such as size, location and symptoms. Currently, there are no clinical trials comparing the different treatment modalities and hence treatment is individualized. Medical therapy may be appropriate for small cysts (<5 cm) and those with contraindications to surgery, using the benzimidazoles such as albendazole or mebendazole. Albendazole is the preferred agent due to better bioavailability. The recommended dose of oral Albendazole is 400 mg twice daily with a fatty meal in adults or 10-15 mg/kg per day (max 800 mg daily) in two divided doses in children.[14,15] The optimum duration of albendazole is unclear but should be given for a minimum of 3–6 months.[16] Praziquantel, an isoquinolone, has been used in combination with albendazole preoperatively or following spillage of cyst contents during surgery.[17] There is no consensus on praziquantel dose or frequency, hence routine use of combination therapy is not undertaken.[3] The initial degenerative changes seen in pulmonary cysts with medical treatment is cyst rupture, which usually occurs between the second and fourth weeks of treatment. Most pulmonary cysts resolve by 5–14 months after treatment initiation. Fibrotic changes can be seen as long term sequelae on CT in the prior cyst locations.[18] Surgery is the principal treatment approach for patients who can undergo surgery, as it can be curative. Care must be taken to minimize intraoperative spillage of cyst contents to prevent parasite dissemination and recurrence. This can be achieved by injecting scolicidal solutions into the cysts or packing the operative field with sponges soaked in scolicidal agents such as 20% hypertonic saline, povidone-iodine or 95% ethanol.[19] Pre-operative treatment with benzimidazoles is not recommended for larger pulmonary cysts as it is thought to soften the cyst wall and lead to cyst rupture.[10]

The complete removal of a pulmonary hydatid cyst via bronchoscopy is a rare event, with nine prior cases reported.[20–26] In seven of the nine cases, the diagnosis of PHC was unexpected. Bronchoscopy is not routinely performed for the diagnosis of PHC and is used to evaluate atypical radiology.[27] In the seven cases where a PHC was unexpectedly found, the dominant CT findings were atelectasis and focal consolidation. In our case and the two cases

reported by Hejazi et al., the diagnosis of PHC was suspected as the CT findings were consistent with cyst rupture and bronchoscopy was performed to confirm the diagnosis. The most specific finding on bronchoscopy is the presence of a whitish-yellow gelatinous endobronchial membrane.[3,28] Bronchoscopy is otherwise helpful by identifying protoscoleces or hooklets from bronchial washings or demonstrating on biopsy the characteristic hydatid membrane appearance on histologic examination. Methods for PHC removal include the use of bronchoscopic suction or grasping forceps, as was in this case, to remove the specimen with the bronchoscope as a single unit.[25] A saline injection (M. E. Hejazi Method) technique has been described for PHCs adherent to the bronchial wall that cannot be removed with bronchoscopic suction alone.[26] We suggest that bronchoscopy should be strongly considered to evaluate suspected ruptured PHCs and that cyst removal be attempted if possible. As the PHC has already ruptured, the risk of anaphylaxis is low. If an unruptured PHC is encountered, it has been suggested that the cyst contents should be aspirated using a cytology needle before extraction to minimize spillage of hydatid fluid.[25] Early removal of the PHC membrane may minimize the inflammatory stimulus and development of bronchial scar tissue which has been described as long term sequalae.[25]

In conclusion, pulmonary hydatid cysts are often discovered incidentally, and the diagnosis is based on a combination of patient epidemiological risk factors, radiology and serology. Surgery remains the treatment of choice for large cysts but medical management can be considered for small cysts. Flexible bronchoscopy should be performed in suspected ruptured pulmonary hydatid cysts for diagnostic clarification and may offer therapeutic options through cyst removal.

AUTHOR CONTRIBUTIONS

Kevin Ziyi Wen and Ricky Tanujaya Lim drafted the initial manuscript. All authors have reviewed and approved the final version of the manuscript.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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