CASE REPORT

HYDATID HEPATIC-BRONCHO-PLEURAL (HEPATO-PULMONARY) FISTULA CAUSED BY *ECHINOCOCCOSIS GRANULOSA*: A ZOONOTIC CASE REPORT

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ABSTRACT. A rare case is presented of a 58-year-old woman who developed a hepatic-bronchopleural fistula following a hydatid hepatic cyst complication. The hepatic-bronchopleural fistula was diagnosed when patient complained of severe repetitive attacks of productive cough of brown yellow to dark colour sputum biliptysis (bile in cough) with vomiting associated with night sweating and fever, for the past one month. She also complained of right side chest and abdominal pain for the past 3 years. This paper describes a unique case of hepatic-bronchopleural fistula caused by hydatid disease, emphasising the clinical feature and its surgical management. Hydatid disease is a zoonotic infection due to the damage caused by *Echinococcus granulosus*. *E. granulosus*, also called the hydatid worm, hyper tape-worm or dog tapeworm, is a cyclophyllid cestode that parasitises the small intestine of canids as an adult, but which has important intermediate hosts such as livestock and humans, where it causes cystic echinococcosis, also known as hydatid disease. It is hoped that this case report will add to the meagre case management reports of hepatic-bronchopleural fistula in literature.

Keywords: hydatid disease, hepatopulmonary fistula, hepatic-bronchopleural fistula, thoracotomy, *Echinococcosis granulosa*

INTRODUCTION

Hepatopulmonary fistulae, although rare, is considered a common complication of hydatid or amoebic liver disease. The fistula normally forms through trans diaphragmatic penetration, leading to rupture as a large cyst into the lower lobe of the lung. These fistulae may arise due to other secondary causes as well, such as congenital malformations, penetrating liver trauma, hepatobiliary surgery, biliary obstruction and most important, infective suppuration. Intrapulmonary rupture of a hepatic hydatid cyst is uncommon and the underlying cause is mostly the perforation from the right subphrenic space into the posterior basal segment of the right lower lobe (Virginio *et al.,* 2003). Other routes of fistula formation involve the presence of

an underlying infected biloma where the biliary stasis predisposes to an extensive suppurative process which leads to rupture and erosion through the diaphragm into the pleural space, bronchus or both (Palmer *et al.* 1996, Petrov *et al.* 2001).

CASE HISTORY

A 58-year-old Malaysian housewife complained of severe repetitive attacks of productive cough with brown yellow to dark colour sputum biliptysis (bile in cough) and vomiting associated with night sweating and fever, for the past one month. Past medical history showed the patient had complained of pain from the right side of the chest and abdominal pain for the past 3 years. There was no history of recent travel, significant comorbidities or positive family history. She was a non-smoker. On physical examination: she was conscious, oriented, febrile but not jaundiced. Blood pressure 130/80 mm Hg, pulse 95 beats per minute, respiration 26 breaths per minute, body temperature 39 °C. Chest auscultation revealed bronchial breathing sounds at lower zone of right lung. Abdominal examination, revealed mild tenderness in right hypochondrium. Laboratory Investigation results as follow: white blood cell count 14 × 10^3/µL, neutrophils 75%, erythrocyte sedimentation rate 101 mm/hr. Virus isolation was negative. Virus isolation was negative. Chest X-Ray: shows opacity shadow in lower zone of right lung, obliteration of right costo-phrenic angle (Figures 1 and 2).

A computerised tomography (CT) of chest and abdomen showed single hydatid cyst type 1 resting at the right lower zone of the right lung with subtle right-sided pleural effusion, adhering to segments VII and VIII of the right hepatic lobe (Figures 3 and 4). The provisional diagnosis was biliptysis due to broncho biliary fistula secondary to hydatid cyst. The diagnosis was discussed with the patient and her family. Patient consent for thoracotomy operation was set to be prepared for surgery.

Operation

Right posterolateral thoracotomy incision, right hepatic-broncho-pleural fistula with large infected cyst at right lower lobe filled with debris material (Figures 5 and 6). The fistula tract through diaphragm was excised, segmentectomy for the severely affected segment of the right lower lobe was followed by pericystectomy and evacuation, cleaning of the hepatic cyst in the liver through the diaphragm, a drain left in the sub-diaphragmatic space and the de-roofed liver cyst, closure of diaphragm and a right chest tube drain left too. The patient was put on intravenous antibiotics. Antihelminthic medical regimen was maintained post-operatively to prevent recurrence. Patient post-operative (PO) course was uneventful. Chest tube removed on the fourth PO day and the sub-diaphragmatic drain removed on the sixth PO day. Patient discharged home on the ninth PO day. X-ray of the chest one month post-operative, demonstrated good inflation of the right lung (Figure 11).

Histopathology

The histopathology examination of the resected lung segment and part of the

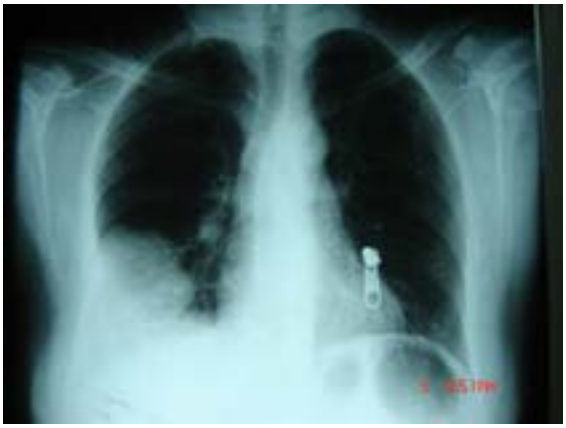

Figure 1. Plain chest radiograph posterior anterior view showing a right lower zone, right lung shadow, pre-operative (25 Jan 2007).

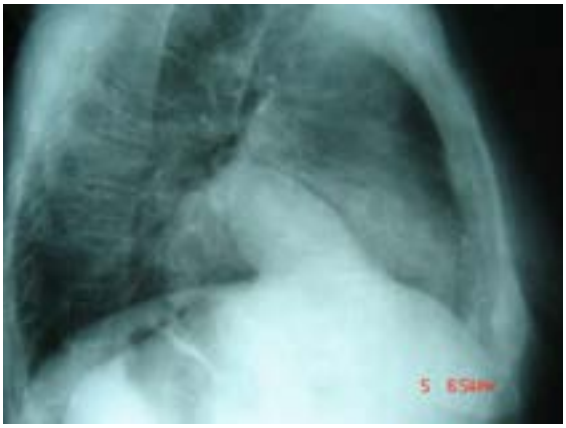

Figure 2. CXR, RL view, right lower zone, right lung shadow, pre-operative (25 Jan 2007).

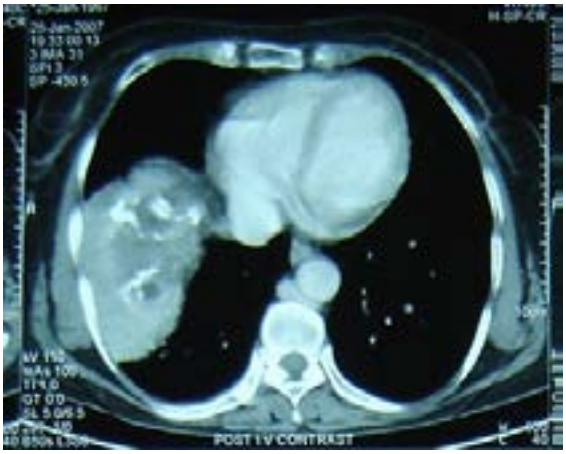

Figure 3. CT scan, 6×5 cm well defined cyst regular in outline in right lower zone of right lung, with subtle right side pleural effusion, pre-operative 25 Jan 2007.

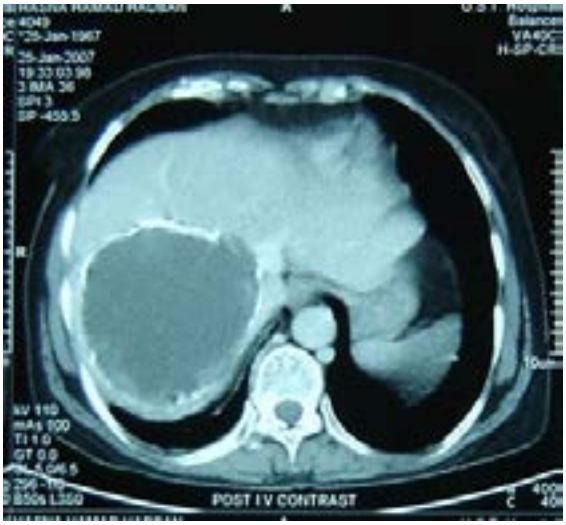

Figure 4. CT scan, well defined 8×6×5 cm hypodense cystic lesion with wall calcification at right lobe of liver, segment VII and VIII. With IV contrast, preoperative 25 Jan 2007.

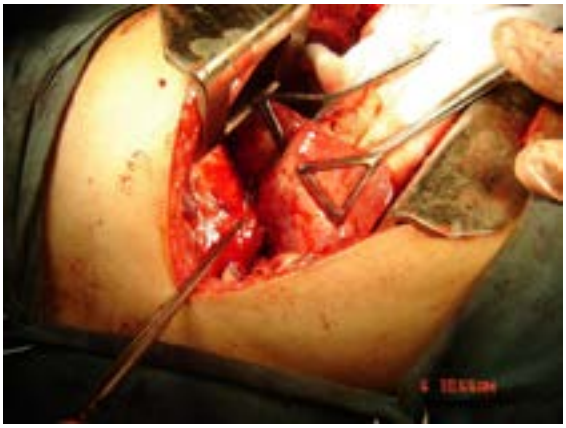

Figure 5. Operative view shows the connection between the right dome of liver and the right lower lobe, 4 Feb 2007.

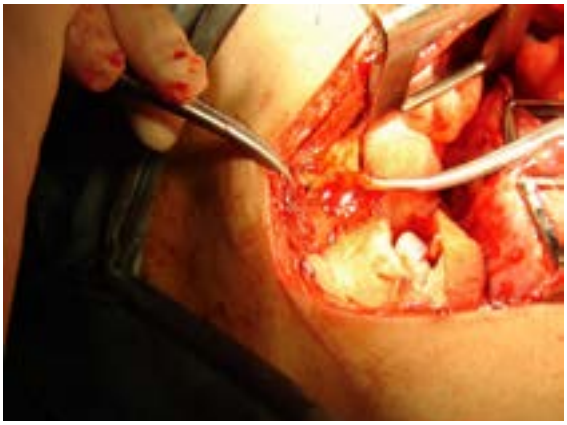

Figure 6. Operative view shows the opening of the fistula at the right dome of the liver after disconnecting it from the lung, 4 Feb 2007.

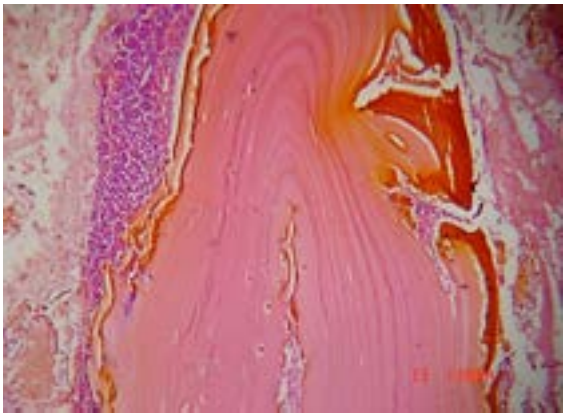

Figure 7. Photomicrograph of the excised degenerated hepatic hydatid cyst, the cyst is well demarcated from the liver parenchyma, note the pericyst layer of chronic active inflammation with fibrous capsule, a remarkable thick acidophilic chitinous layer of the hydatid cyst observed, ×20 H&E.

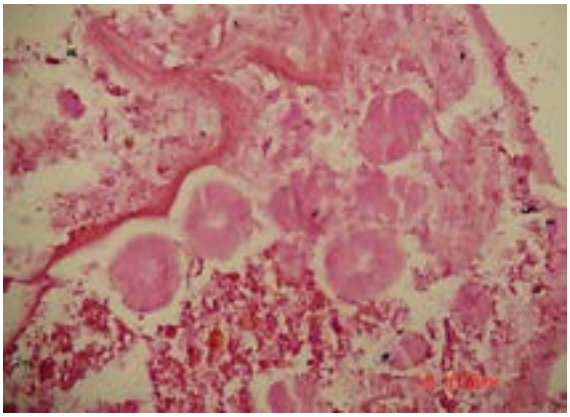

Figure 8. Photomicrograph shows degenerated hepatic hydatid cyst with non-viable germinal membrane and protoscolices, ×40, H&E.

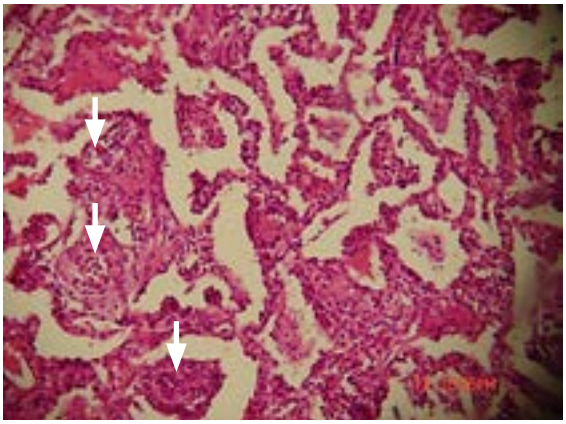

Figure 9. Photomicrograph of the excised lung segment shows predominantly an interstitial lymphocytic infiltrate associated with predominantly fibrotic pattern; note the widespread thickening of the alveolar septa by a cellular infiltrate. Early stage of non-necrotising granuloma like lesions was seen (arrows), ×20, H&E.

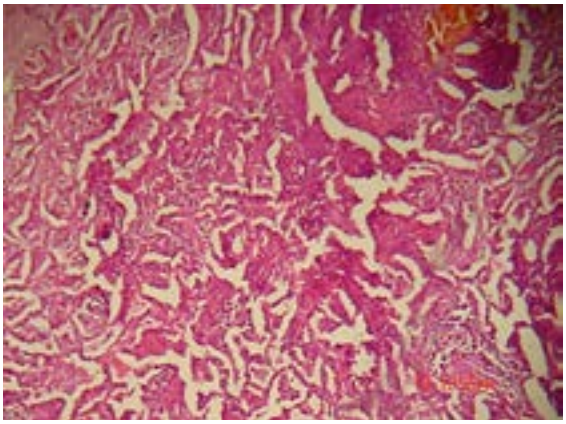

Figure 10. Photomicrograph of the excised lung segment demonstrating fibrotic non-specific interstitial pneumonia pattern. Note the uniform thickening of the alveolar walls and chronic inflammation. ×10, H&E.

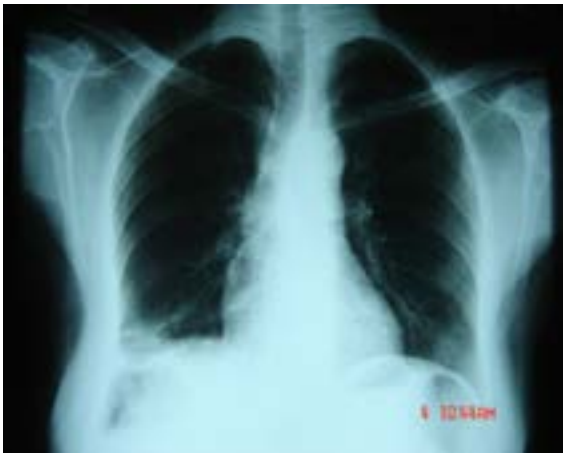

Figure 11. CXR post-operative, 5 Mar 2007, one month post-operative.

hepatic cyst wall showed non-specific chronic active inflammation with broncho-biliary fistula due to hepatic hydatid cyst infestation (Figure 7). Hepatic hydatid cyst showed degeneration, the cyst is well demarcated from the liver parenchyma and pericyst layer of chronic inflammatory tissue with fibrous capsule and thick chitinous layer of the cyst (Figure 8). The lung segment showed predominantly an interstitial lymphoplasmacytic infiltrate associated with predominantly fibrotic pattern, a widespread thickening of the alveolar septa by cellular infiltrate (Figure 9). The septal widening is due to a mild to moderate infiltrate of lymphocytes with scattered plasma cells, with minimal associated fibrosis. Poorly formed non-necrotising granulomas are evident (Figure 10). The lung segment demonstrated fibrotic non-specific interstitial pneumonic pattern. The alveolar walls showed uniform thickening and chronic inflammation.

DISCUSSION

As the hydatid cyst was located on top of the right lobe of liver and has been infected most probably by haematogenous root or after eroding a biliary radicle. It erodes through the diaphragm, leading to adhesion of the bases of right lower lobe of lung, then initiating an inflammatory reaction and pneumonitis and erode a segmental bronchus, ending with hepatic-bronchopleural fistula formation.

It is well known that most of the hydatid cases are asymptomatic and only reveal across the medical attention when the cyst is incidentally found or become infected. As in this case, the hepatic-bronchopleural fistula revealed after the constitutional symptoms of fever, tachycardia, shortness of breath, abdominal and chest pain was evident.

Pulmonary hydatid are incidentally found on chest X-rays of patients who have sustained trauma, some are diagnosed in cases of pathologic fracture of a long bone, other cases have been diagnosed in breast, thyroid gland, parotid gland, muscles, heart, spleen and even discovered as a space occupying lesion in the brain (Dahniya *et al.,* 2001, Utkan *et al.,* 2000).

It has special importance when it affects heart, lung and spleen, as the hydatid cyst will not calcify with time as in liver. For that reason, especially in lung and spleen, even a simple trauma may result in rupture of the cyst and either anaphylactic reaction may occur or dissemination of the scolices. This may lead to the formation of multiple cysts and provoke adhesions. It may end with intestinal obstruction, a condition diagnosed as malignant hydatidosis in areas where it is endemic. However, this study had found no record of hydatid disease in Malaysia.

An important measure to advocate proper control for this disease is urgently needed. This positive intention may be achieved in collaboration with the World Health Organisation through a worldwide health promotion campaign to eradicate the *E. granulosus* from the primary (definitive) hosts and to control infected intermediate human and animal hosts (Karamustafaoglu *et al*., 2012; Borrie and Shaw, 1981; Gulamhussein *et al.,* 2015).

CONCLUSION

Surgical treatments are by conventional surgery or by minimal invasive technique such as video-assisted thoracic surgery (VATS) technique and laparoscopic surgery. All procedures proved its efficacy in treating the different types of hydatid disease and its complications, but still prevention is better than cure. Eradication of the causative agent *E. granulosus* should be the target and priority aim. *E. granulosus* is ingested and attaches to the mucosa of the intestines in the definitive host such as dogs and there the parasite will grow into the adult stages. Adult *E. granulosus* release eggs within the intestine which will be transported out of the body via faeces. When contaminated waste is excreted into the environment, intermediate host has the potential to contract the parasite by grazing in contaminated pasture, perpetuating the cycle. In the case of human infection, living in close proximity to dogs infected with *E. granulosus* can cause hydatid cysts such as this case study. Control of stray dog population and maintaining good personal hygiene can prevent such infections.

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