Hepatobronchial fistula caused by hydatid disease

The Dunedin Experience 1952–79

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ABSTRACT Despite intensive hydatid control measures in New Zealand, hepatopulmonary fistula resulting from infection by the *echinococcus granulosus* still occurs. Although the patients may quickly become debilitated from coughing bile and pus and associated septic complications, appropriate surgical therapy is usually effective. The exact diagnosis may be obscure, but it is helped by a high index of suspicion. A retrospective study of eight patients seen over a 27-year period is presented, and principles of management are outlined. Where biliary hypertension is not present adequate evacuation of the intrahepatic cysts, obliteration of the cyst space, freeing of the adherent lung, and closure of the pulmonary fistula(e) usually give satisfactory long-term results. Pulmonary lobectomy or segmental resection is seldom required.

Hepatopulmonary fistula is an uncommon condition, the most common causes being hydatid or amoebic disease of the liver with transdiaphragmatic penetration and rupture into the lower lobe of the lung. However, fistulae secondary to penetrating trauma, congenital lesions, and pyogenic suppurative with associated biliary obstruction have also been described.¹ Intrapulmonary rupture of a hepatic hydatid cyst is uncommon and usually the perforation is from the right subphrenic space posteriorly into the posterior basal segment of the right lower lobe.¹ In this report eight cases of hydatid hepatopulmonary (HHP) fistula seen over a period of 27 years are presented. The cases emphasise the merits of having a high index of suspicion in making the diagnosis, and of using aggressive surgery for the subdiaphragmatic disease coupled with a more conservative approach for the associated pulmonary lesion.

Patients and methods

A retrospective analysis was made of the cases of bronchobiliary fistula secondary to hydatid disease managed in the Southern Regional Thoracic Surgical Unit, Dunedin in the 27-year period 1952–79. There were eight patients—two

adult females, one female child, and five adult males. The child was aged 3 years, and the adults on admission were aged between 33 and 71, the mean age being 57 years.

Case reports

CASE 1

A 3-year-old child was seen in 1952 with right upper quadrant abdominal pain and an abdominal mass. She had had several episodes of right lower lobe pneumonia, and was thought to have bronchiectasis. Her Casoni test was positive, and on clinical grounds she was thought to have a fistula between the liver and the right lung.

At operation a hydatid cyst in the left lobe of the liver was removed, but while a communication between the right lobe of the liver and the diaphragm was being explored, the child sustained a cardiac arrest and died. Her death was thought to be the result of anaphylaxis and there was no necropsy.

CASE 2

A 69-year-old woman seen in 1962 underwent general anaesthesia for surgical correction of tic douloureux. Bile was then noted in the anaesthetic equipment, and this was aspirated and a tracheostomy performed. Later she became jaundiced and plain radiographs revealed a calcified hepatic hydatid cyst. A bronchogram showed

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a fistula between the lung and a cavity in the right lobe of the liver. Operation was declined and her symptoms settled over several months. However, she was troubled with intermittent bouts of coughing up bile and hydatid debris for four more years. She was eventually admitted in 1966 with a fractured femoral neck. Her biliary fistula re-opened and soon after surgical fixation of the fracture she developed bronchopneumonia and died.

CASE 3

In 1962 a 38-year-old man attended with a history of coughing up bile and pus. He was febrile and had lost one stone in weight over two months. Chest films revealed a high right hemidiaphragm and right lower lobar bronchiectasis. He had a positive Casoni test, a positive hydatid complement fixation test, and a 7% blood eosinophilia.

A clinical diagnosis of HHP fistula was made and he underwent thoracotomy. The adherent lung was dissected off the diaphragm and the pulmonary fistulae were oversewn. The fistulous tract through the diaphragm was identified and the hydatid cyst lying in the right lobe of the liver was evacuated of its contents. The upper half of the pericyst, which was in part calcified, was excised, the cavity was swabbed with 2% formalin solution and the space obliterated with a series of mattress sutures placed between the diaphragm and the lower half of the pericyst. The pleural cavity was temporarily drained with a water seal drain. He made an uneventful recovery and nine years later underwent cholecystectomy without incident.

CASE 4

A 71-year-old woman seen in 1963 had a 20-year history of pulmonary hydatid disease for which she had had multiple operations. In 1963 she began coughing up bile and yellow pus. She became febrile and over three months lost a stone in weight. A sinus developed in an old thoracotomy wound and through this a sinogram revealed a bronchocutaneous and a hepatopulmonary fistula. At thoracotomy an infected hydatid cyst of the liver was curetted out and the pericyst was subtotally removed, complete removal being impossible because of haemorrhage. The liver cavity was obliterated with interrupted linen mattress sutures between the diaphragm and the remaining pericyst tissue as in case 3. The pulmonary fistula was closed and the posterior basal segment of the right lower lobe was oversewn. In addition two other uninfected hydatid cysts

lying in front of the first liver cyst were evacuated and swabbed with 10% formalin. The resulting cavities were obliterated with mattress sutures, and finally the pleurocutaneous sinus tract was completely excised. One year later she was well, and she has since died of unrelated causes.

CASE 5

In 1971 a 73-year-old man underwent cholecystectomy and choledochotomy. Two small hydatid cysts were removed from the common bile duct and an operative cholangiogram revealed a hydatid cyst of the right lobe of the liver communicating with the biliary tree. Four months later he began to cough up green sputum and lost a stone in weight. He was not jaundiced but his hydatid complement fixation test was positive and chest films revealed a collapsed right lower lobe. HHP fistula was suspected and thoracotomy was undertaken. The diaphragm was incised, the hydatid cyst was curetted out, and again the upper half of the pericyst was removed. The resulting liver cavity was swabbed with 2% formalin and obliterated by infolding the diaphragm as in cases 3 and 4. The fistulae into the posterior basal segment of the right lower lobe were oversewn, and when the lower lobe inflated readily pulmonary resection was considered unnecessary. He was well three years later and has since died of unrelated causes.

CASE 6

In 1971 this 51-year-old man was reviewed. In 1970, at a peripheral hospital, a hydatid cyst had been excised from the right lobe of the liver. After this he had developed a subphrenic abscess. This was drained, after which he had become profoundly unwell, coughing green sputum and losing four stone in weight over three months. He was also troubled with severe right shoulder-tip pain. After his transfer to this unit a liver scan showed a mass in the right lobe of the liver and bronchography revealed a fistula from a residual liver cavity into the right chest. At thoracotomy dense adhesions between the right lung and diaphragm were divided. The fistula led to a large infected hydatid cyst occupying the upper surface of the right lobe of the liver. The cavity was evacuated, the upper half of the pericyst was removed and the cavity obliterated as in the previous cases. Because of dense adhesions the right middle and lower lobes were decorticated before oversewing the fistulous tract. A sinus persisted for three months but three years later he was leading a normal life and had regained his lost weight.

CASE 7

This 65-year-old man was referred in 1978. In 1940 a hydatid cyst had been removed from the right lobe of the liver and in 1976 he had undergone cholecystectomy and removal of a stone from the common bile duct. At the same time a second hydatid cyst was removed from the right chest wall. In 1978 he was admitted to hospital for investigation of haemoptysis and a mass in the right lower lobe. The differential diagnoses were pulmonary infarct, carcinoma of the lung, bronchiectasis, Wegener's granuloma, and hydatid cyst. At thoracotomy in March 1978 the mass was found to be surrounded by dense adhesions fixing it to the diaphragm. Right middle and lower lobectomy was performed and the specimen was later shown histologically to contain hydatid elements.

Two months later a hydatid cyst was discharged from his thoracotomy wound. He was observed for a year, but in May 1979, after another cyst had been discharged from the wound, a sinogram was performed. This showed a fistula between a hydatid cyst in the liver and the right pleural cavity. The space above the diaphragm and the cyst in the liver were separately approached via limited rib resections. The two spaces were evacuated, swabbed with 2% formalin solution, and packed with Vaseline gauze. Healing by granulation was complete within two months.

CASE 8

In 1979 a 33-year-old man presented with right-sided chest pain, fever, lethargy, and a seven pound weight loss over two months. He was coughing grey sputum. Liver function tests showed an elevated alkaline phosphatase, and although his hydatid serology tests were negative, he had a 10% eosinophilia. A liver scan revealed a mass in the right lobe, and his chest films showed a high right diaphragm. A clinical diagnosis of HHP fistula was made and at thoracotomy the infected cyst in the apex of the right lobe of the liver was curetted. The cavity was packed with Vaseline gauze and marsupialised. No fistula was then detected, but a later sinogram revealed a fine communication between the residual hepatic cavity and the anterior basal segment of the right lower lobe. This area was drained, as for an empyema, and eventually healed by granulation tissue forming along the drainage tube track.

Discussion

Hydatid disease may occur in all age groups, and involve any body cavity, organ, or site. The com-

plication of hepatopulmonary fistula is rare. Kourias² found this complication in only 2% of 1198 cases of hydatid disease. Dew³ observed that suppuration of the cyst commonly preceded rupture, but in the case report of Matar *et al*⁴ there was no obvious cyst infection and the fistula was thought to have been caused by biliary hypertension from stones blocking the common bile duct.

In three of our eight cases the hydatid cysts had become abscesses. In one other case, not explored surgically, there may have been cyst infection, but in the remaining four cases there was no evidence of gross suppuration. In none of the cases was there biliary tract obstruction at the time of fistulation. Apart from cyst infection and biliary hypertension other factors which may predispose to rupture and fistula formation are the fact that intrathoracic pressure is lower than intra-abdominal pressure,¹ and the tendency of the pericyst to erode adjacent structures.⁵

Rupture usually occurs on the right, a fistula developing between a cyst in the right lobe of the liver and the basal segments of the right lobe of the lung. Yacobian,⁶ however, in a series of six patients with HHP fistula, had one patient who fistulated into the left chest. In the present series all fistulae were from the right lobe of the liver into the right lung, but in two cases the anterior basal segment of the right lower lobe was involved rather than the more usual posterior basal segment.¹ In both cases where the anterior basal segment was involved the diagnosis proved to be difficult and the fistulae were missed on initial exploration. After postoperative sinograms revealed fistulous tracts, re-exploration was required.

The HHP fistula patient may present coughing up bile or thick pus, but depending on the size of the communication hydatid debris or cysts may also be expelled. There is often pyrexia and occasionally marked weight loss. In the present series the fever was seldom above 38°C, but in two cases the weight loss was gross, two men losing two and four stone in weight respectively. With such substantial weight loss HHP fistula may mimic a malignancy.

The appearance of multiple opacities in the form of a "smoke stack"⁷ on chest films is highly suggestive of an HHP fistula, but the demonstration of a tract on a sinogram or a bronchogram is pathognomonic.⁶ Other less specific radiological findings include a high right hemidiaphragm or evidence of right basal bronchiectasis.⁵ In the present series three patients had radiological evi-

dence of bronchiectasis and two had a high right hemidiaphragm. Contrast radiography was invaluable. In patients 4, 7, and 8 sinograms revealed the fistula and in patients 2 and 6 bronchography delineated the tract. In two of the more recent patients scanning showed the hepatic lesion, but these findings were not as useful as contrast radiography in diagnosing the fistulas.

Because the fistula is usually inaccessible from the abdomen, surgical correction required a thoracic approach, which allowed the coexisting lung disease to be managed simultaneously.⁵ McConchie,⁷ Boyd,¹ and Matar *et al*⁴ stressed that the control of the subdiaphragmatic hydatid disease is fundamental for eradicating the fistula permanently. There are, however, several other critical aspects of management.

Biliary hypertension must be diagnosed and managed before surgical correction of the fistula.⁵⁻⁸ If there is obstruction to bile flow the fistula is most unlikely to remain closed after operation. In the present series persistent fistulation secondary to biliary obstruction was not a problem. Two patients had a common bile duct exploration before their HHP fistula treatment but none of the others had been jaundiced or were known to have gallstones.

The hepatic cavity should be obliterated and, if this is not possible, adequately drained. A separate, lower incision has been used by some authors to drain the cavity externally.⁵ In our series two patients required this, and on both occasions a limited rib resection was used. The four remaining cases treated surgically had the hydatid cyst evacuated and the upper half of the pericyst excised, allowing the cavity to be obliterated with non-absorbable mattress sutures placed between residual pericyst and diaphragm. Previous painting of the cavity with scolical solution is also important to prevent recurrent disease.

Reventos *et al*⁵ indicated that pulmonary resection was seldom necessary in patients with HHP fistula, a view also held by Matar *et al*.⁴ In the present series, management of the subdiaphragmatic disease was by dissection of the lung from the diaphragm, and suturing the fistula. In only one case—where the exact nature of the lower lobe mass could not be determined at thoracotomy—was pulmonary resection required. However, bile is a powerful irritant to the bronchial mucosa and

with added infection this irritation may cause severe pulmonary damage necessitating resection of part or all of a lobe.⁶⁻⁹⁻¹⁰ In four of the six cases of HHP fistula reported by Yacoubian⁶ a pulmonary resection was deemed necessary.

If there is a substantial cutaneous leak of bile in the postoperative period, or if after operation the patient coughs up bile, biliary hypertension must be excluded.⁵

Operative mortality varies from five to almost 50%.⁵ In the present series seven of the eight patients were treated surgically and there was only one surgical death. The remaining six patients were discharged from hospital well and have been followed for a period of three months to 10 years. The patient who refused operation gives some clue to the natural history of HHP fistula. Although her initial symptoms abated over some months, she later had many relapses coughing up bile, pus, and hydatid debris. Eventually this led to right lower lobe bronchiectasis and when an operation for a fractured neck of femur became necessary this proved fatal.

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