Case Report

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Hydatid hepatopleural fistula causing biliothorax: minimally invasive approach with ERCP stent placement and chest tube drainage: a case report

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Introduction and importance: Biliothorax and hydatid bilio-bronchial fistula (HBBF) are rare complications of hydatid cysts of the liver with high perioperative mortality.

Case presentation and clinical discussion: The authors here report the case of a patient with a right massive pleural effusion with evidence of HBBF in imaging studies, who underwent surgical resection of a hydatid cyst of the liver 8 years ago. The patient was managed with intercostal chest tube drainage for biliothorax and endoscopic sphincterotomy with biliary stent placement for the re-establishment of internal biliary drainage, which ultimately allowed the fistulous tract to heal without any major surgical intervention.

Conclusion: This article focuses on the successful management of the HBBF at our institution. Our findings emphasize the significance of early diagnosis and the criticality of implementing aggressive and early interventions to minimize complications and fatalities. By examining this specific case and reviewing existing literature, the authors have determined that endoscopic treatment appears to be a viable, secure, and efficacious option for addressing postoperative HBBF.

Keywords: biliothorax, case report, ERCP (endoscopic retrograde cholangiopancreatography), hydatid bilio-bronchial fistula (HBBF), hydatid cyst

Introduction

An abnormal connection between the pleura of the lungs and the hepatic parenchyma is called a hepatopleural fistula. Heptaopleural fistula is an uncommon complication of a hydatid cyst of the liver. Between the years 1914 and 1961, there were 1198 cases of liver hydatid cysts, as per the statistics from one of the hospitals in Greece. Of these, 0.77% ruptured into the pleural cavity and created a hepatopleural fistula[1]. Hydatid cyst intrapleural rupture is a significant consequence that can cause intense pain, cyanosis, dyspnea, and collapse. The development of empyema with bile-stained fetid pus and hydatid material is possible[2]. The diagnosis entails radiographic tests, though the fistula can also be found with percutaneous transhepatic cholangiography (PTC) and endoscopic retrograde cholangiopancreatography (ERCP)[3]. Surgery remains the main treatment in congenital or acquired broncho-biliary fistulas. However, interventional radiology and gastroenterological techniques (endoscopic sphincterotomy, endoscopic drainage, and endoscopic stenting) are alternatives to surgical intervention for acquired fistula[4].

Here, we present a case of a 23-year-old male diagnosed with a right hepatopleural fistula with a hydatid cyst of the liver and a proximal common bile duct fistula. Endoscopic retrograde cholangiography and stent placement were done for the common bile duct fistula to re-establish the biliary flow tract and, in turn, decrease biliary hypertension.

This case report has been reported in line with the Surgical Case Report (SCARE) Criteria[5].

HIGHLIGHTS

- Hepatopleural fistula refers to the formation of an abnormal connection between the liver and the pleural cavity, allowing the passage of fluid, bile, or air between the two areas. It most commonly occurs as a complication of liver abscesses, trauma, or surgical interventions.
- Patients may present with pleuritic chest pain, dyspnea, cough, or recurrent pleural effusion.
- Diagnosis involves a combination of clinical suspicion, imaging studies (such as CT scans or ultrasound), and analysis of pleural fluid. The presence of bile in the pleural fluid confirms the diagnosis.
- With the advancement in minimally invasive procedures endoscopic retrograde cholangiopancreatography (ERCP) alleviating biliary obstruction along with pleural drainage is considered to be a very successful approach in managing cases.

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Case presentation

A 23-year-old male was referred to our hospital, one of the tertiary-level hospitals, for the need of an ERCP for a stent exchange that was placed 3 months ago for biliary drainage.

The patient gives a history of fever, dry cough, and generalized myalgia for 3 days. Fever was acute on onset and intermittent type, associated with chills and rigors. The maximum temperature was not documented. He also gives a history of a dry cough that was later productive, yellowish, and nonblood stained.

The patient has had a history of intravenous drug abuse for 5 years. There is no history of similar illness in other family members, and there is no significant genetic or psychosocial history in other family members. The patient gives no history of chronic illnesses like diabetes mellitus, hypertension, or tuberculosis. There is no history of allergic history to drugs and any foreign bodies like dust, or wool.

On examination, vitals were stable, with an oxygen saturation of 93% at room temperature, a pulse of 80 beats per minute, systolic and diastolic pressures of 120 mmHg and 80 mmHg, a respiratory rate of 16 breaths per minute, a temperature of 37°C. On chest examination, there was asymmetrical chest expansion with respiration. On percussion, there was decreased tactile fremitus and dullness noted over the right lung. There was decreased air entry in the right lung with decreased vocal resonance.

Investigations

During his further work-up, his chest roentgenogram was suggestive of a right-sided massive pleural effusion. Ultrasound-guided pleural fluid tapping was planned (Fig. 1A).

Ultrasonography (USG) showed a collapse of the right lung with moderate septate pleural collection, and diagnostic pleural tapping was performed (Fig. 2).

On diagnostic tapping, straw-colored pleural fluid was obtained and sent for analysis. Biochemical analysis of pleural fluid showed a whole blood count of 9600/mm^3 with 96% neutrophils, glucose of 34.0 mg/dl, albumin of 2.46 g/dl, ADA of 20.85 U/l (reference range: <30) total protein of 5.22 g/ml (total protein level in serum of 6.8 g/dl) with a ratio of pleural protein and serum protein being greater than 0.5, lactate dehydrogenase (LDH) of 4896.5 U/l (LDH level in serum of 259.03 U/l) with ratio of pleural LDH and serum LDH exceeding 0.6. aerobic and anaerobic cultures of pleural fluid had no growth, and LDH levels were out of proportion, that is 4896.5 U/l. Cytopathology reports of the pleural fluid showed the presence of degenerated mesothelial cells and inflammatory cells consisting of neutrophils and lymphocytes. No atypical cells were noted and the smear was negative for malignant cells (Table 1). Gene Xpert ruled out tuberculosis.

Contrast-enhanced computed tomography (CECT) of the chest showed a gross hypodense pleural collection in the right pleural cavity causing a near-total collapse of the right lungs with a mild contralateral mediastinal shift (Fig. 3).

Intercostal tube drainage was done using a 28F chest tube for pleural effusion. A bilious collection of pleural fluid was observed. On further inquiry, the patient's party gave the history of a hydatid cyst of the liver and the removal of the cyst (surgical) 8 years back. The Echinococcus IgG antibody test was positive (2.40 COI) (reference 0–0.8).

The CECT chest, abdomen, and pelvis were planned. Impressions on CECT showed the gross hypodense pleural collection in the right pleural cavity, causing a near-total collapse of the right lung with a mild contralateral mediastinal shift and a minimal right subphrenic collection (Fig. 3).

The patient also underwent magnetic resonance cholangiopancreatography (MRCP).

Findings on MRCP showed heterogeneous collections with multiple ill-defined thin membranes in the right pleural cavity and the collapse of the visualized right lung. Areas of diffusion restriction were noted within the collection. The minimal, heterogeneously hyperintense collection was also noted in the right subphrenic space. A few ill-defined areas were noted within the collection, which were likely detached membranes or residual hydatids. Those lesions were closely abutting segments VII and VIII of the right lobes of the liver.

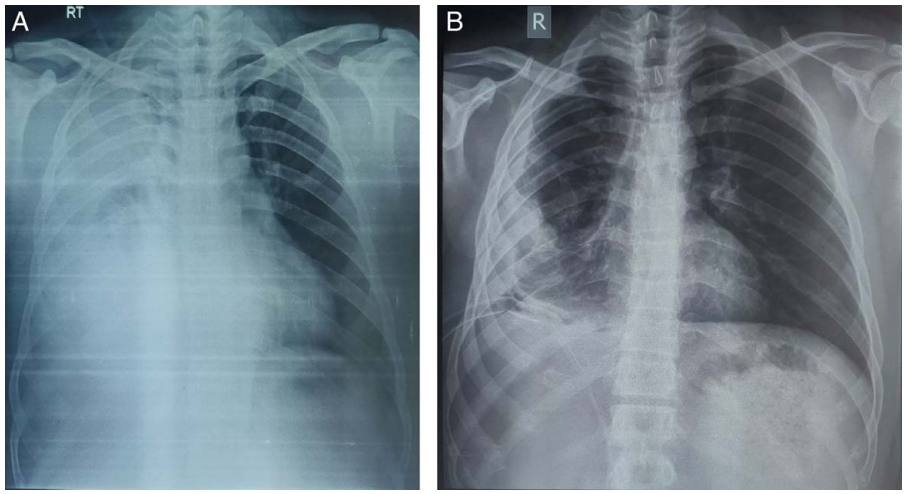

Figure 1. A, Chest radiography PA view: showing increased radio-opacity over right lung field with obliterated right costo-phrenic angle-likely right-sided massive pleural effusion. B, Chest radiography PA view following intercostal tube drainage for right-sided massive pleural effusion showing resolving right-sided pleural effusion.

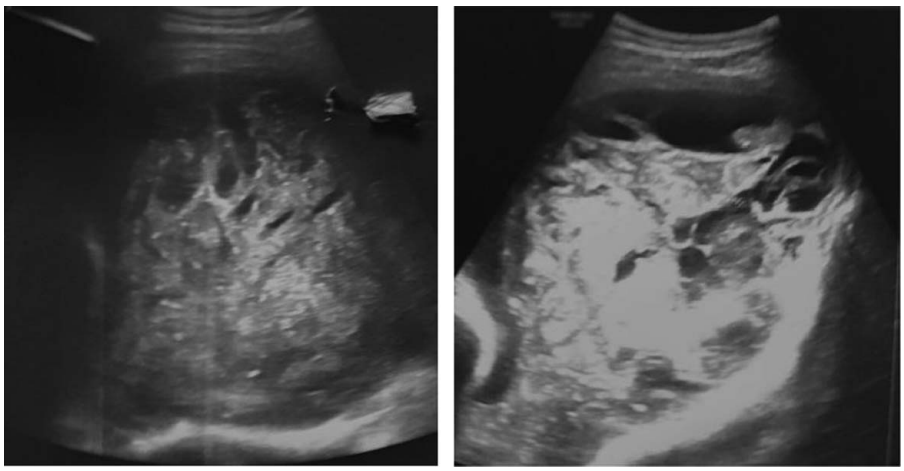

Figure 2. USG abdomen and pelvis showing collapse of the right lung with the moderate septate pleural collection.

A defect measuring nearly 5 mm was noted in the posterior aspect of the right hemidiaphragm communicating between the pleural cavity and subphrenic collection, suggesting trans diaphragmatic rupture of the hydatid (Fig. 4).

An ERCP was then performed to rule out a pleurobiliary fistula.

The patient underwent an ERCP that revealed a proximal CBD fistula following leakage of contrast from the proximal CBD (Fig. 5). Endoscopic sphincterotomy and biliary stent placement were performed to improve biliary drainage. An 8F × 8 cm double pigtail plastic stent was deployed over the common hepatic duct (CHD) with a good flow of bile (Fig. 5). He was then advised to follow-up after 3 months for a stent exchange. He was discharged with oral medications including tablet cefuroxime axetil, albendazole, paracetamol, and pantoprazole.

Stent exchange could not be performed during the subsequent visit due to decreased oxygen saturation, so he was referred to our center for ERCP and stent exchange.

The patient came with an intercostal chest drain (ICD) in situ for re-ERCP. ICD placement was done at the nearby hospital a few days back for the pleural fluid drainage after he clinically deteriorated with features of respiratory distress following a chest radiography. Baseline investigations were sent at our hospital that showed raised C-reactive protein (CRP) with normal RFT and LFT. Cardiothoracic and Vascular Surgery (CTVS) consultation was done for leakage of fluid from the intercostal drain site (ICD). On local examination, the suture site was infected with purulent discharge from a nonfunctioning chest tube. A right-sided chest drain was exchanged with a 28F chest tube at another site with the repair of the previous site. A repeat chest radiography was done to look for the proper placement of the chest tube. Immediate drainage of around 600 ml of bile was evacuated, and the drained fluid had a bile color. About 1050 ml of the drain was collected within the first 24 h of chest tube placement. The patient's dyspnea, tachycardia, and chest pain were relieved after the chest tube insertion. A chest radiography showed right-sided lung expansion. As the patient's condition was stabilized and there were no signs of empyema that required surgical intervention, we decided to treat the patient conservatively with close vital signs monitoring, chest tube output surveillance, chest radiography, the addition of antibiotics, and control of the source.

The CECT abdomen and pelvis were repeated. ERCP was planned. During ERCP, a cholangiogram revealed normal extrahepatic and intrahepatic bile ducts with contrast flow proximally from the right duct, suggesting communication. A 7 Fr 10 cm straight biliary plastic stent was placed in the right hepatic duct. A good flow of bile was noted (Fig. 6). That was very efficient in stopping the bile leak into the right chest cavity, as it was obvious that after the procedure there was no more bile drainage into the right pleural space. The patient's respiratory symptoms were improving, and the chest tube was removed after 7 days with a total drain of about 2 l. We were successful in managing the biliothorax with the steps that we followed.

The patient was then discharged with oral medications in a clinically stable condition. He was advised to make a subsequent visit after 3 months for a stent exchange.

Provisional diagnosis

Right hepatopleural fistula with hydatid cyst of the liver with proximal CBD fistula.

Table 1

Reports of diagnostic pleural fluid tapping.

S. N	Tests	Results	Units	Reference interval
1.	Color	Straw color		
2.	WBC	9600/mm^3	Cells/cumm	
3.	Sugar	34.0	Mg/dl	
4.	Protein	5.22	g/ml	
5.	Albumin	2.46	g/dl	
6.	LDH	4896.5	U/l	
7.	ADA	20.85	U/l	< 30: Negative > 40–60: Positive
8.	Cytopathological	Negative for malignant cells		

ADA, adenosine deaminase; LDH, lactate dehydrogenase.

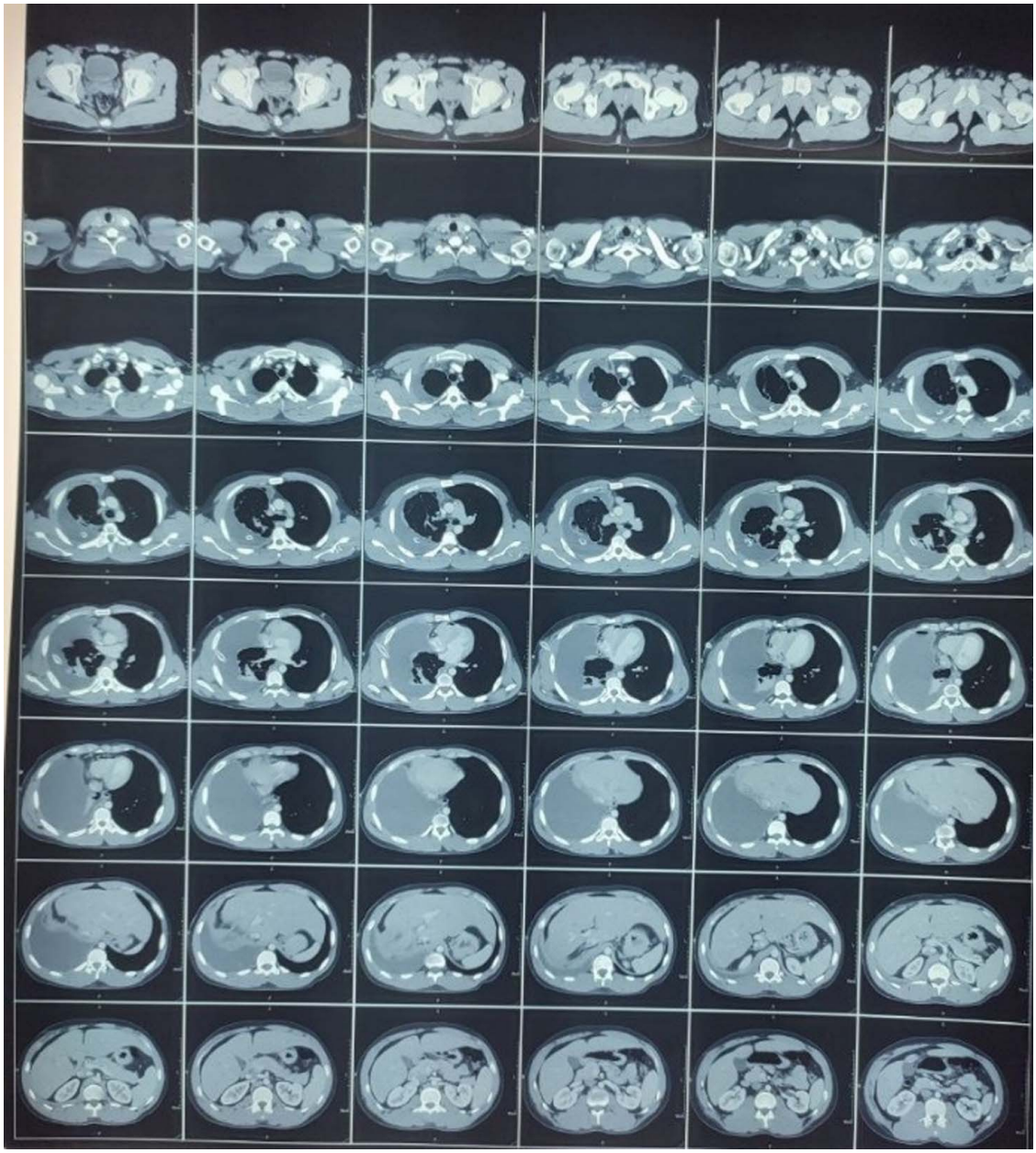

Figure 3. CECT chest, abdomen, and pelvis showing gross hypodense pleural collection in the right pleural cavity causing a near-total collapse of right lungs with a mild contralateral shift.

Outcome and follow-up

Regular follow-up examinations confirmed the absence of complications or recurrences.

Discussion

Hydatid cyst disease (echinococcosis) is a parasitic infection caused by the cestode worms *Echinococcus granulosus* and *E. multinocularis*. It is endemic in developing countries and also in the pastoral lands of the Mediterranean region and South America, where livestock is raised in association with dogs[6].

Although hydatid disease most frequently affects the liver, it can affect practically any organ in the body, except the hair, teeth, and fingernails. In adults, the lungs are the second most frequent location of hematogenous spread and probably the most common location in children[3,7]. Although the lung parenchyma is the most frequent location in the thorax, it can also grow in any extrapulmonary area, including the chest wall, pleural cavity, fissures, mediastinum, heart, and vascular systems[8].

Hepatopulmonary fistulae, although rare entities, are a common complication of hydatid or amebic liver disease with trans diaphragmatic penetration and rupture into the lower lobe of the lung as a large cyst. The presence of PBF (pleural biliary fistula)

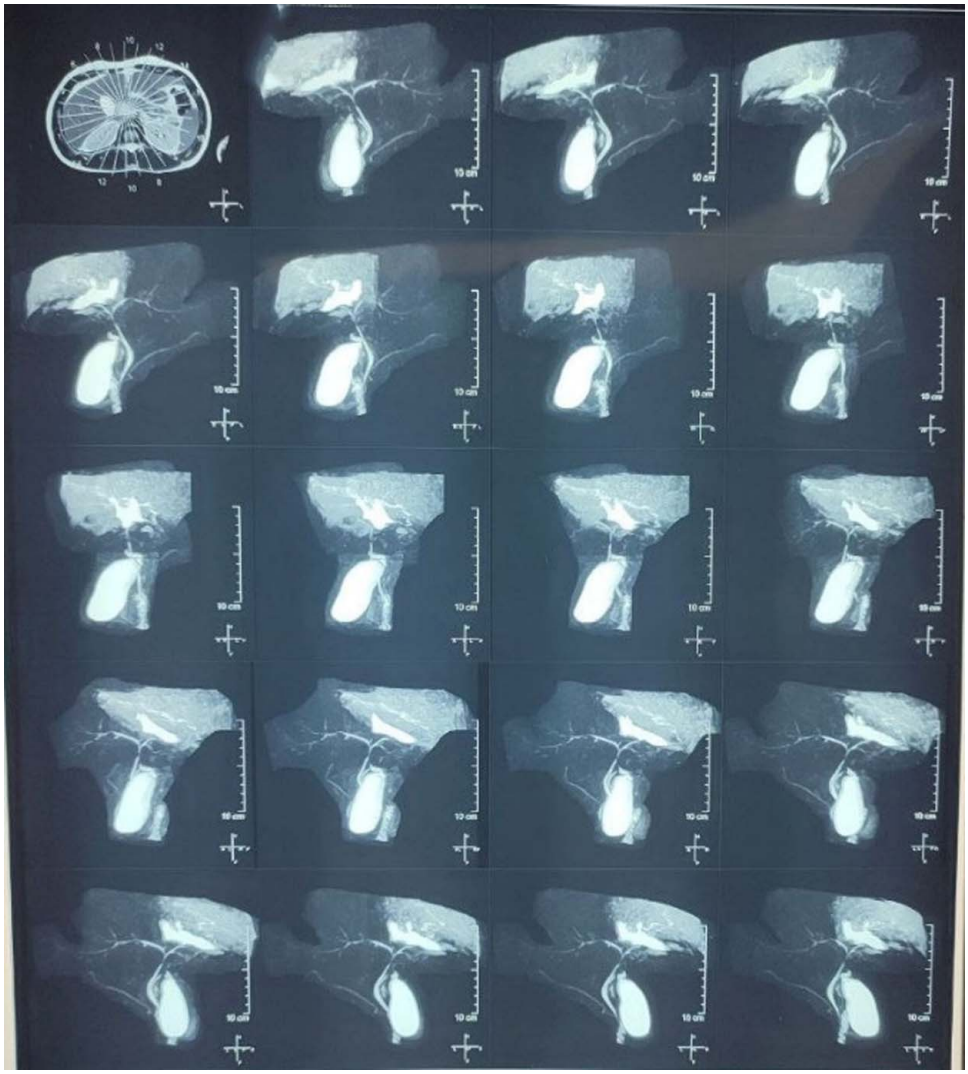

Figure 4. MRCP showing heterogeneous collections with multiple ill-defined thin membranes in the right pleural cavity and the collapse of the visualized right lung. A few ill-defined areas were noted within the collection, closely abutting segments VII and VIII of the right lobes of the liver, which was likely detached membranes or residual hydatids. A defect measuring nearly 5 mm was noted in the posterior aspect of the right hemidiaphragm communicating between the pleural cavity and subphrenic collection, suggesting trans diaphragmatic rupture of the hydatid.

secondary to choledocholithiasis, trauma, congenital malformations, or iatrogenic operations is identified as the primary etiology of biliothorax. Hepatic parasitic infestation, radiofrequency ablation of liver tumors, and biliary stent migration are further uncommon reasons that have been discussed in the literature[9].

In one report, 2% of the total 1198 cases of hydatid disease had this complication[7].

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[7,10]. Theory suggests that biliary hypertension following any obstruction in the biliary tract, along with small perforations in the diaphragm, leads to fenestration of contents from the peritoneal cavity into the pleural space. The defect will mature over time if the fluid (bile) is not drained[11].

Thus, biliary hypertension must be diagnosed and managed before surgical correction of the fistula[7].

Before a rupture, there are no particular symptoms. Hemoptysis, dyspnea, pleuritic chest discomfort, epigastric discomfort, coughing due to diaphragmatic irritation, and moderate expectoration are all possible symptoms. More persistent pain can be found in the liver or shoulder. Attacks of jaundice are reported in about one-third of patients. In 56% of the cases, there was an abrupt rupture followed by a sharp tearing sensation. Depending on the size and location of the fistulous tract, patients typically begin releasing cysts in later stages[10]. Bilioptysis, or the presence of bile in pleural effusions, is a pathognomonic sign of a hepatopulmonary or biliopulmonary fistula[10].

If untreated, the patient's respiratory state can quickly deteriorate due to fluid-induced lung volume loss as well as an inflammatory reaction of the bile that has a direct corrosive effect on the pleural layers, which can lead to ARDS[9,11].

Although imaging may be helpful for diagnosis, fistulae are often difficult to identify[3].

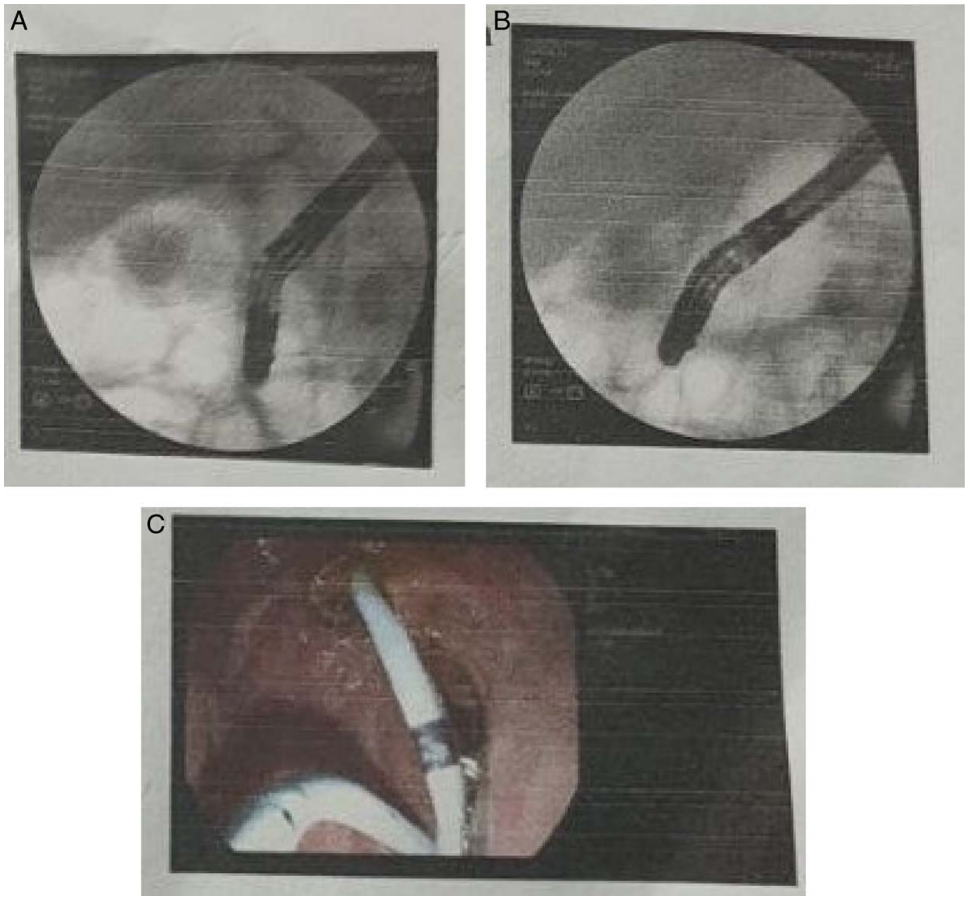

Figure 5. A-C, ERCP findings: Cholangiogram demonstrating contrast leakage originating from the proximal common bile duct (CBD) with no observable filling defects. Additionally, an 8F × 8 cm double pigtail plastic stent is visible, successfully deployed over the common hepatic duct (CHD), ensuring a smooth and unimpeded flow of bile.

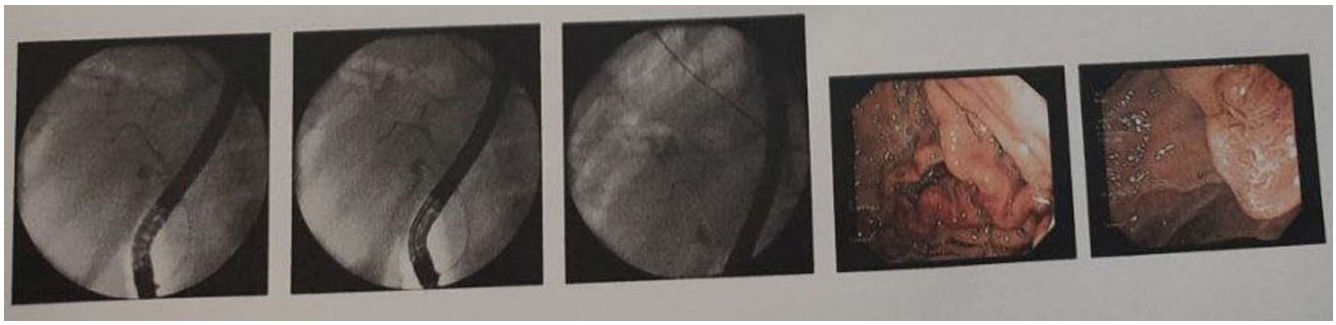

Figure 6. ERCP findings: Cholangiogram revealed: normal extrahepatic and intrahepatic bile ducts with contrast flow proximally from the right duct, suggesting likely communication. Previously placed stent was removed and A 7 Fr 10 cm straight biliary plastic stent was placed in RHD to ensure appropriate biliary drainage.

The diagnostic tests of choice include PTC, ERCP, magnetic resonance cholangiography (MRC), and computed tomography (CT)[4].

A chest radiography revealing pleural effusion after a liver or biliary procedure should raise the question of the possibility of a bilothorax.

The presence of green-colored pleural fluid with elevated levels of bilirubin is a clue to the diagnosis of biliothorax. The diagnosis is based on the demonstration of bile in pleural fluid with a ratio between the pleural bilirubin and the serum greater than one, with a range of 1.4–11.6. A total pleural fluid bilirubin level of 6.8–57.78 mg/dl was reported in five of the fifty-two patients[12,9].

ERCP and PTC are useful for the detection of fistulae. Despite their potential limitations, CT and MRI are nonetheless helpful diagnostic techniques for identifying fistulous tracts. In our case, we suspected HBF based on pleural fluid analysis and CT findings[3]

There is currently no standard treatment for HBF. Maintaining low pressure at the opening of the fistula is vital, which can be accomplished by drainage. This can be obtained by using a chest tube, ERCP, PTC, or surgical excision of the fistula tract. Clinicians prefer less invasive and nonsurgical treatment as the recovery rate is higher than that with traditional surgery, except in refractory cases where traditional surgery is necessary[3].

The initial treatment approach for biliothorax involves a combination of actions, including draining the fluid from the pleural space, identifying the site of the bile leakage, and relieving the biliary obstruction to promote closure of the bile-pleural fistula through endoscopic techniques like ERCP or percutaneous cholangiography[11].

In a series by Lakranbi M *et al.* ERCP with sphincterotomy was performed as a first-line treatment, and this allowed the bilio-bronchial fistula to heal in 60% of cases. In one case study, surgical intervention was found to be performed in 40% of cases with the persistence of FBB[13].

In another study conducted by Senturk *et al.*, various therapeutic modalities (endoscopic sphincterotomy, endoscopic stent, nasobiliary drainage, and medical therapy) were applied to treat HBBF in multiple cases. However, according to their findings, endoscopic stenting was identified as the most effective approach for treating HBBF[4].

ERCP with stenting represents an emergent therapeutic method in the setting of a biliary or bronchial fistula[13]. By implementing internal biliary drainage through the placement of a stent, the healing of the fistulous tract can occur without the need for extensive surgical intervention[4]. This approach is particularly beneficial in cases where there is significant biliary communication that is challenging to close. Internal drainage offers the dual advantage of effectively draining the biliary fistula and addressing the residual cavity, making it a safe and valuable procedure[14].

Maintaining low pressure at the openings of the bile-pleural fistula is crucial, and this is typically achieved through the placement of a thoracostomy tube or by performing biliary decompression using ERCP before surgery. Several studies have suggested that the fistula can spontaneously close (up to 60% in cases of post-traumatic fistulas) following endoscopic or subcutaneous drainage of the biliary system.

The most difficult aspect of surgical management in these patients is the reconstruction of the damaged boundary between the chest and diaphragm caused by the fistula. In cases where there are significant defects, the usual approach involves utilizing nearby tissue such as intercostal muscle or pericardial fat to seal and restore continuity[10]. Lung lesions resulting from chronic intrapulmonary rupture are irreversible, and in such cases, lobectomy may be required as a necessary treatment option[2]. Hepatopulmonary fistulas, despite being nonmalignant, pose a significant risk of mortality, with rates as high as 10.3%. The main cause of this mortality risk is primarily associated with surgical complications[10].

In our case, endoscopic sphincterotomy with endoscopic stent placement was done for HBBF due to hydatid disease.

Conclusion

Bilothorax is a rare exudative pleural effusion. The first steps of management should include fluid drainage and antibiotic treatment to prevent further respiratory complications such as empyema and respiratory distress.

With the advancement in minimally invasive procedures, percutaneous drainage of the biliary tree and ERCP alleviating biliary obstruction along with pleural drainage is considered to be a very successful approach in managing a lot of bilothorax cases.

In this case, ERCP stent placement provided effective biliary decompression and closure of the fistulous tract, complemented by chest tube drainage. This case highlights the importance of minimally invasive techniques in the management of biliothorax and the need for long-term follow-up to ensure favorable outcomes.

Ethical approval

Ethics approval is not required for case reports at our institution. Name of ethics committee: IRC-KUSMS.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal on request.

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Author contribution

All authors contributed in writing the paper, reviewing and editing, revising it critically for important intellectual content.

Conflicts of interest disclosures

The authors declare that they have no competing interest.

Research registration unique identifying number (UIN)

1. Name of the registry: Research registry.
2. Unique identifying number or registration ID: researchregistry 9173.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): https://www.researchregistry.com/browse-the-registry

Guarantor

Sumina Mainali.

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Data sharing is not applicable to this article.

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