ACG CASE REPORTS JOURNAL

CASE REPORT | LIVER

Hepatopleural Fistula with Empyema Thoracis: A Rare Complication of Autosomal Dominant Polycystic Kidney Disease

Jeerawat Maytapa, MD[1], Kessarin Thanapirom, MD, MSc[1,2], Sombat Treeprasertsuk, MD, PhD[1,2], Piyawat Komolmit, MD, PhD[1,2], Bundit Chaopathomkul, MD[3], and Pinit Kullavanijaya, MD[1]

[1]*Division of Gastroenterology, Department of Medicine, Faculty of Medicine, Chulalongkorn University and King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Bangkok, Thailand*
[2]*Center of Excellence in Liver Diseases, King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Bangkok, Thailand*
[3]*Department of Radiology, Faculty of Medicine, Chulalongkorn University and King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Bangkok, Thailand*

ABSTRACT

We report a 70-year-old man with autosomal dominant polycystic kidney disease (ADPKD) who presented with right-sided extended-spectrum beta-lactamases *Escherichia coli* empyema thoracis. Chest and abdominal computed tomography showed hepatopleural fistula. The patient refused a surgical operation and was treated with tube thoracotomy, percutaneous drainage of dominant liver cyst, and intravenous antibiotics. His symptoms improved after 2 months of nonsurgical treatment.

INTRODUCTION

Polycystic liver disease (PCLD) is a rare inheritable disorder, which may occur in conjunction with or as an independent entity from autosomal dominant polycystic kidney disease (ADPKD). PCLD is the most common extrarenal manifestation of ADPKD. Typically, most patients with PCLD and ADPKD are asymptomatic. However, as patient lifespans are extended by medical interventions such as widespread use of dialysis and renal transplantation, symptoms have become more common. Symptoms in patients with PCLD may develop from the mass affecting the adjacent structures, including blood vessels and the bile duct, or from complications related to the cysts such as infection, hemorrhage, or rupture.[1-8]

CASE REPORT

A 70-year-old man presented with right-sided pleuritic chest pain, progressive dyspnea, and low-grade fever over a 1-month period. He denied a history of weight loss and trauma. He did not smoke or drink alcohol. He was diagnosed 30 years ago with ADPKD and PCLD. He also had chronic renal failure. One month prior to this admission, he had an infected liver cyst treated with 7-day intravenous ceftriaxone and percutaneous aspiration. Pus culture revealed no organism. His father and sister were also diagnosed with ADPKD. On examination, the patient had a low-grade fever (37.5°C) and tachypnea (34 breaths/min). There was decreased breath sound with dullness on percussion of the right hemithorax. Abdominal examination showed tender hepatomegaly and bilateral enlarged kidneys. Emergency endotracheal intubation was performed due to ventilatory failure.

Laboratory data showed white blood cell (WBC) 23,870 $\times$ 10^6 cells/L, predominantly neutrophil (87.3%) and lymphocyte (8.4%), and a platelet count of 210 $\times$ 10^9 platelets/L. Liver function test showed total protein 7 mg/dL,

ACG Case Rep J 2018;5:e2. doi:10.14309/crj.2018.2. Published online: January 3, 2018.

Correspondence: Kessarin Thanapirom, Division of Gastroenterology, Department of Medicine, Chulalongkorn University, Rama IV Road, Patumwan, Bangkok, Thailand, 10330 (tkessarin@hotmail.com).

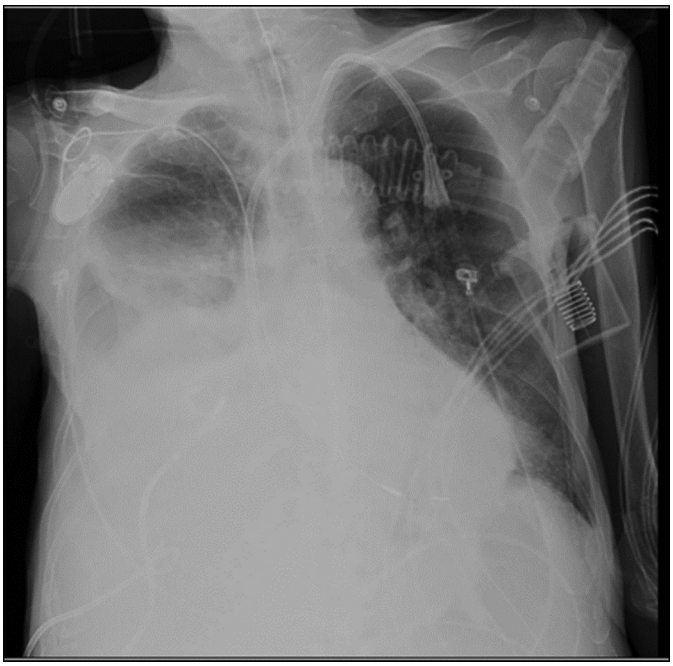

Figure 1. Chest x-ray showing loculated right pleural effusion.

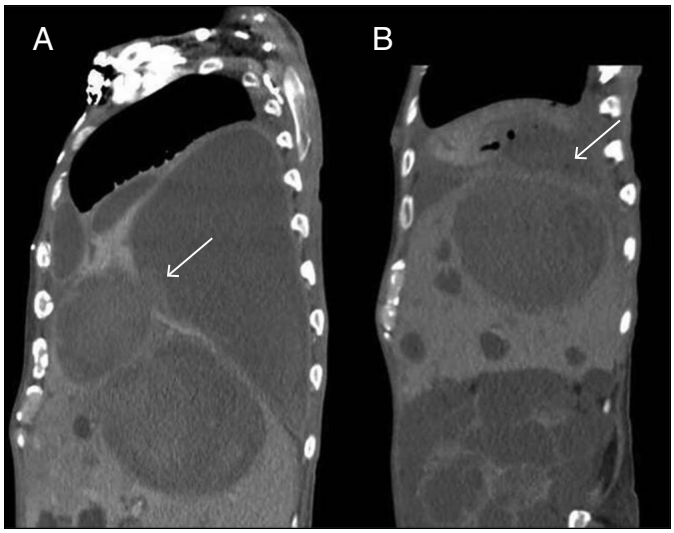

Figure 2. (A) Contrast-enhanced computed tomography (CECT) of chest and abdomen showing multiple liver cysts with a fistula between the liver cysts and the right pleural cavity (arrow). (B) Follow-up CECT after treatment showing decreased size of the liver cysts and diminished hepatopleural fistula (arrow).

albumin 2.8 mg/dL, total bilirubin 1.1 mg/dL, direct bilirubin 0.55 mg/dL, aspartate aminotransferase 27 IU/L, alanine aminotransferase 18 IU/L, and alkaline phosphatase 240 IU/L. Chest x-ray revealed large right pleural effusion (Figure 1). Computed tomography of the chest and abdomen showed innumerable cystic lesions of the liver and both kidneys. The largest liver cyst, measuring 25 × 13 cm, was connected to the right pleural cavity via a fistula resulting in massive right pleural effusion (Figure 2). There was no evidence of pneumonia by imaging. Pleural fluids from thoracocentesis showed frank pus with exudative profile (WBC 349,114 × 10^9 cells/L, neutrophil 84%, protein 4.7 mg/dL, and lactate dehydrogenase 37,814 U/L). Pleural fluid culture identified extended-spectrum beta-lactamases *E. coli*. The patient refused surgery and was treated with intercostal drainage of the right pleural effusion, percutaneous drainage of the large liver cyst, and intravenous meropenem for 2 months. Follow-up chest CT after treatment showed only a small amount of the right pleural effusion and improvement of the hepatopleural fistula. Repeated pleural and cystic fluid culture showed no microorganisms. Both drainage catheters were removed without subsequent complication. He recovered well and was discharged after 60 days of hospitalization.

DISCUSSION

Although the majority of patients with adult PCLD show no symptoms, significant symptoms may occur in some. These symptoms include abdominal pain, abdominal distension, and a sensation of fullness from the enlargement of the liver. Common complications of PCLD are infection, hemorrhage (intracystic or hemoperitoneum), rupture, torsion of liver cyst, and fistula formation. There have been several reports of rare complications due to compression of the adjunct structures by the dominant liver cyst, including portal hypertension, Budd-Chiari syndrome, hepatic venous outflow, inferior vena cava obstruction, and obstructive jaundice.[7] Pleural effusion associated with ADPKD or PCLD has also been previously reported.[1,7,9] Possible mechanisms of pleural effusion formation are thought to be due to cystopleural fistula or disruption of the local capillary permeability from pleural inflammation.[7]

A case of transudative pleural effusion with cystopleural fistula in an ADPKD patient has been previously described.[2] To our knowledge, ours is the first reported case of an adult with PCLD and ADPKD complicated by empyema due to the rupture of an infected liver cyst and hepatopleural fistula, which responded to conservative treatment without the need for surgery. The most common cause of empyema is pulmonary infection.[1] Nevertheless, this case presented no evidence of pneumonia on imaging. The mechanism of empyema in this case might be due to the rupture of a complicated liver cyst and hepatopleural fistula formation. Previous percutaneous drainage may be the predisposing factor for hepatopleural fistula formation in this patient. The most common causes of hepatopleural fistula and pleural empyema are amoebic liver abscess or hydatid cyst.[10] Biliary tract obstruction secondary to tumors and acute cholecystitis are unusual causes of hepatopleural fistula.[11,12]

There are several therapeutic approaches for managing PCLD-associated complications, including aspiration, sclerotherapy, fenestration, segmental hepatic resection, or liver

transplantation.[13,14] Surgery remains the gold standard treatment for hepatopulmonary fistula.[11] Other treatment options for the management of hepatopleural fistula include thoracotomy, adequate subcostal drainage of the hepatic bed, secure closure of the diaphragmatic perforation, and decortication for the lung.[15] The success rates of surgical and nonsurgical treatments for hepatopulmonary fistula were 78.6% and 93.3%, respectively.[10] Our case has established that conservative treatment is a treatment option for patients who decline surgery or are unsuitable for surgery. Our patient refused surgery because of the concerns about surgical complications. After 60 days of combined tube thoracotomy, percutaneous cyst drainage, and antibiotic treatment, the patient recovered well and was discharged without complication.

DISCLOSURES

Author contributions: J. Maytapa and K. Thanapirom wrote the manuscript. K. Thanapirom, S. Treeprasertsuk, P. Komolmit, B. Chaopathomkul, and P. Kullavanijaya edited the manuscript. K. Thanapirom is the article guarantor.

Acknowledgments: The authors thank Dr. Peera Jaru-Ampornpan from the National Center for Genetic Engineering and Biotechnology (BIOTEC) for English language editing.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received July 15, 2017; Accepted November 8, 2017

REFERENCES

1. Heffner JE, Klein J. Parapneumonic effusions and empyema. *Semin Respir Crit Care Med*. 2001;22(6):591–606.
2. Peterson M, Aboudara M. Hepatic cystopleural fistula. *Am J Respir Crit Care Med*. 2014;189(2):e4–e5.
3. Woolnough K, Palejwala A, Bramall S. Polycystic liver disease presenting with an exudative pleural effusion: A case report. *J Med Case Rep*. 2012;6(1):107.
4. Van Keimpema L, De Koning DB, Van Hoek B, et al. Patients with isolated polycystic liver disease referred to liver centres: Clinical characterization of 137 cases. *Liver Int*. 2011;31(1):92–8.
5. Tahvanainen E, Tahvanainen P, Kääriäinen H, Höckerstedt K. Polycystic liver and kidney diseases. *Ann Med*. 2005;37(8):546–55.
6. Arnold HL, Harrison SA. New advances in evaluation and management of patients with polycystic liver disease. *Am J Gastroenterol*. 2005;100 (11):2569–82.
7. Chauveau D, Fakhouri F, Grünfeld J-P. Liver involvement in autosomal-dominant polycystic kidney disease therapeutic dilemma. *J Am Soc Nephrol*. 2000;11(9):1767–75.
8. Torres VE, Harris PC, Pirson Y. Autosomal dominant polycystic kidney disease. *Lancet*. 2007;369(9569):1287–301.
9. Ihara K, Naito S, Yamaguchi W, et al. Hepatic cyst infection in an autosomal dominant polycystic kidney disease patient diagnosed by right pleural effusion. *Intern Med*. 2014;53(12):1355–9.
10. Liao G-Q, Wang H, Zhu G-Y, Zhu K-B, Lv F-X, Tai S. Management of acquired bronchobiliary fistula: a systematic literature review of 68 cases published in 30 years. *World J Gastroenterol*. 2011;17(33):3842–9.
11. Kontoravdis N, Panagiotopoulos N, Lawrence D. The challenging management of hepatopulmonary fistulas. *J Thorac Dis*. 2014;6(9):1336–9.
12. Allison M, Milkins S, Burroughs A, Rogers H, Thomas H. Bronchobiliary fistula due to acute cholecystitis in a suprahepatic gall bladder. *Postgrad Med J*. 1987;63(738):291–4.
13. Drenth JP, Chrispijn M, Nagorney DM, Kamath PS, Torres VE. Medical and surgical treatment options for polycystic liver disease. *Hepatology*. 2010;52(6):2223–30.
14. Temmerman F, Missiaen L, Bammens B, et al. Systematic review: The pathophysiology and management of polycystic liver disease. *Aliment Pharmacol Ther*. 2011;34(7):702–13.
15. Ferguson TB, Burford TH. Pleurobiliary and bronchobiliary fistulas: Surgical management. *Arch Surg*. 1967;95(3):380–6.