Polycystic liver disease with right pleural effusion

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Abstract. Polycystic liver disease (PCLD) is a condition in which multiple cysts form in the hepatic parenchyma. The polycystic liver disease is also an autosomal dominant disorder (ADPLD) caused by a mutation in a gene that encodes a protein hepatocystin. PCLD has a prevalence count of 1,200,000 people in the people of America. PCLD occurs ± 24% of patients in the third decade of age to 80% by the sixth decade. Women tend to get larger cysts and more and correlated with the number of pregnancies. The following case report of a woman, 51-years-old who was treated at Haji Adam Malik hospital Medan with a diagnosis of polycystic liver disease with right pleural effusion. Some literature has reported complications of the polycystic liver disease but rarely reported with pleural effusion presentation. The patient had already undergone a puncture of pleural fluid and after three weeks of treatment condition of the patient improved and permitted to be outgoing patient.

1. Introduction
Poly Cystic Liver Disease (PCLD) is a rare condition in which multiple cysts form in the hepatic parenchyma. The polycystic liver disease is also an autosomal dominant disorder (ADPLD) caused by a mutation in a gene that encodes a protein hepatocystin. Given two genes that are mutated in PCLD, PRKCSH gene, and gene SEC63. Mutations of these genes can be less than a third of cases.[1,2,3,4] PCLD has a prevalence count of 1,200,000 in the people of American. It is approximately 24% of patients in the third decade of life to 80% in the sixth decade of life, but the kidney disease is the dominant clinical course. Cysts also appear in the pancreas, spleen and, less often, other organs.[1,2] Women presented the larger and more numerous cysts and are commonly known as high age, typically in the fourth or fifth decade of life, number and frequency of pregnancies and severity of renal disease.[1,2,5]

Diagnosis of PCLD is by using ultrasound, CT scan, or MRI.[1,2] Polycystic liver disease was with 95% of asymptomatic patients. The remaining 5% come with symptoms which are local effects of mass suppression polycystic liver or stretching or compression of the structure of other organs.[1,5] There have been several reports in the literature describing the complications of the polycystic liver disease. However, the presence of right pleural effusion has rarely been described.[6]

2. Case report
The patient is a 51-year-old woman, Javanese, hospitalized in Adam Malik hospital with chief complaint shortness of breath within one month before admission. Fever persists for one week earlier with coughing and whitish sputum without haemoptoe. Right upper abdominal mass was since three years ago without any symptomatic complaints such as abdominal pain, but in the last one month, the mass felt even more extensive. Pale and weakness were felt within one week before admission. There
are no spontaneous bleeding, history of hypertension and diabetes mellitus, history of tuberculous medicine, and the family history of malignancy or with the same symptoms.

Patient vital signs were componsentis, blood pressure 120/80mmHg, pulse 94x/minute, respiratory rate 28x/minute, temperature 37.4°C. On physical examination, inferior palpebral conjunctiva was pale, diminished breath sound on the right middle-lower lobe lung, infiltration on the right upper lobe lung, liver were palpable on 10cm below arcus costae, 10cm below processesxyphoides, blunted, irregular, without abdominal tenderness.

Laboratory results showed Hb 9.8gr%, Leukocyte 20,660/mm3, Platelet 235,000/mm3. Erythrocyte morphology showed a normochromic normocytic anemia, Ferritin e 483.5ng/ml, SI 15mg/dL, TIBC 129µg/dL, reticulocyte count 2.46%, LED 100mm/hour, Ureum 26.10mg/dL, Creatinin 0.56mg/dL, Total bilirubin 0.55mg/dL, direct bilirubin 0.39mg/dL, alkaline phosphatase 241U/L, AST 14U/L, ALT 12U/L, γ-GT 131U/L, HbsAg (-), anti HCV (-), Blood sugar 127.5mg/dL. Electrolytes Na 137mEq/L, K 4.1mEq/L, Cl 104mEq/L, Albumine 2.5g/dL, Globuline 3.4g/dL. Blood gas analysis showed pH 7.429, pCO₂ 34.9, pO₂158.4, HCO₃ 22.6, Total CO₂ 23.7, BE -1.4, SaO₂ 99.4 %.

Thorax photo showed a picture of pneumonia which we differential diagnosed with the specific process with right pleural effusion. AFBDS/3X were negative. Abdominal ultrasound showed multiple cysts on the liver. Abdominal CT Scanning with contrast showed multiple cysts on liver, spleen and bilateral kidney with right pleural effusion. Sputum culture showed no bacterial growth. The urinary culture showed Klebsiella sp. >100,000CFU/ml urine which was sensitive with amikacin, meropenem, cefmetazole, piperacillin-tazobactam. Cytology pleural effusion showed C3 smear with atypical cells, not confirmative as malignancy. Pleural effusion analysis was exudative. The culture of pleural effusion showed no bacterial growth. Pleural effusion was negative for acid-fast bacilli (AFB).

Figure 1. The photo of thorax before and after pleural effusion puncture.
Patient was diagnosed with polycystic liver disease + right pleural effusion + pneumonia + urinary tract infection + anemia due to chronic disease. Treatment including diet 1700kcal, Oxygen 1L/min, IVFD NaCl 0.9% 10gtt/min, Meropenem IV 1gr/12hour, Ambroxol syrup 5xCl, Paracetamol 3x500mg, pleural effusion puncture was 1000cc. After three weeks of treatment, the patient’s condition improved and permitted to be discharged without any needs of follow-ups.

3. Discussion
Poly Cystic Liver Disease (PCLD) is a condition in which multiple cysts form in the hepatic parenchyma. It is on clinical attention in adulthood. PCLD usually presents autosomal dominant polycystic kidney disease (ADPKD) but can appear as isolated polycystic liver disease.[1,2,4] The
polycystic liver disease is an autosomal dominant disorder (ADPLD) may be caused by a mutation in a gene that encodes a protein hepatocystin. Two genes are known to be related to PCLD. PRKCSH is the first gene, which encodes for the β-subunit of glucosidase II, an N-linked glycan processing enzyme in the endoplasmatic reticulum. It is on 19p13.2-p13.1. SEC63 gene is the second gene, which encodes a component of the protein translocation machinery in the endoplasmatic reticulum. It is on 6q21-q23. These facts suggest a role for co-translational protein-processing pathways in maintaining epithelial luminal structure and implicate (non-clarial) endoplasmatic reticulum proteins in PCLD. Mutations of genes can be in less than one-third of the cases.

PCLD has a prevalence count of 1:200,000 people of American. It is approximately 24% of patients for the third decade of life to 80% in the sixth decade of life, but the kidney disease is usually becoming the clinical course. The Adult disease is inherited as an autosomal dominant disorder, and infantile disease as an autosomal recessive disorder.

Results in embryonic hepatic maldevelopment with failed involution of interlobular bile ducts, and cysts of other organs such as the kidney, pancreas, lung, and spleen around 50% of patients. Women have larger and more numerous cysts, and correlation with the number of pregnancies has been found.[1,2,5]

In this case, the patient was frequently asymptomatic, although large or multiple cysts may result by continuing pain due to stretching, mass effect, or compression of other structures. Nausea from stomach compression and jaundice from bile duct obstruction may develop.

There are reports in the literature describing the complications of polycystic liver disease due to compression of mass effect of the massive cyst, but thoracic complication such as pleural effusions have been reported as a complication of the polycystic liver disease. Vanerpemct al. reported a case of symptomatic large right-sided pleural effusion complicating polycystic liver disease and requiring intervention. It was also associated with ascites and attributed to an abdomino-pleural communication. Kerry Woolnough et al. reported a case of recurrent right pleural effusion related to polycystic liver requiring surgical intervention.[2,6,7]

In this case, chief complaints were shortness of breath and abdominal enlargement due to right pleural effusion et causa polycystic liver disease.

Developing of exudative pleural effusions when there is a change in the permeability of local capillaries or the pleural surface. Common causes are a malignancy and parapneumonic effusions. Lower common triggers include pulmonary infarction, rheumatoid arthritis, autoimmune disorders, asbestosis, and pancreatitis.[6,8]

On cytological pleural effusion is not a confirmative malignancy. Sputum and others pleural effusion analyses exclude tuberculosis infection.

The pathogenesis ways of a right-sided exudative pleural effusion in association with the polycystic liver disease gets unclear, with no mechanism in the literature. Kerry Woolnough et al. suggested that pleural effusion was not recurrent after the main hepatic cyst had been removed. Therefore, they said that the recurrent right-sided pleural effusion occurred as a direct consequence of the mass effect of the main hepatic cyst displacing and deforming the right hemidiaphragm. In this turn, led to a disruption of the local capillary permeability and pleural inflammation, resulting in a persistent exudative effusion.[6,8]

In this case, size of cysts and compression effect to adjacent structure organ (diaphragm) complication of polycystic liver disease caused a right pleural effusion. There were no communication defects between peritoneal and pleural cavity on abdominal CT Scan with contrast, thereby excluded a fistula and diaphragm defect or liver rupture, of which excluded as the mechanism of pleural effusion associated with the polycystic liver disease.

Palpation may suggest an enlargement size or massive liver. The texture may be nodular and firm as the large cysts (adult polycystic disease) are enlarged, smooth, and firm (infantile polycystic disease).[2]

In this case, on abdominal palpation were found massive hepatomegaly, swollen abdomen, but without pain.
The polycystic liver disease has many natural histories, with the majority of patients seen to remain asymptomatic with normal liver function tests, although serum alkaline phosphatase and GGTP levels may elevate. A raised right hemidiaphragm may be evident on a plain x-ray of the chest in patients with severe PCLD. The diagnosis of PCLD is sure by ultrasound, CT, or MRI. The cysts range in diameter from a few millimeters to 10 cm or more. Cysts are in line with biliary-type epithelium and filled with a fluid similar to the bile-salt independent fraction of bile. Cysts also may be appeared in the kidney, pancreas, lung, and spleen in up to 50% of patients.[1,2,6]

In this case, liver biochemical test results were normal, with increased alkaline phosphatase serum and GGTP levels. Renal function test was still normal. Abdominal ultrasound and CT Scan with contrast showed multiple cysts in the liver, kidney, and spleen with varying diameter size from few millimeters to 11 cm.

It does not have a significant idea the effective medical therapy. Medication options based on sign and symptoms, the cyst anatomy, and the cyst distribution. When the cystic disease is asymptomatic, non-therapy is needed. If there are only cysts near the surface of the liver, symptoms may be handled by aspiration, cyst sclerosis by ethanol injection, surgical the cysts laparoscopically or by open laparotomy, or surgical cystojejunostomy. Lower the amount collections of cysts can be resected, while the severe diffuse symptomatic disease has been treated with liver transplantation. Less than 5% of patients have acute medical complications. These consist of cyst hemorrhage, rupture, infection, uterine prolapse due to displacement, obstructive jaundice, portal hypertension, transudative and exudative ascites and Budd-Chiari syndrome. Treatment should be in case of persistent symptoms and complications.[2,7,9] In a retrospective journal series review of 53 patients with a polycystic liver disease and Complications need intervention in this category of patients; cyst bleeding was 12.5%, cyst rupture was 12.5% and cyst infection was 30%. Portal hypertension was 2.5% of the patients, and 5% received a liver transplant. Biliary and inferior vena cava obstruction, chronic abdominal pain, and symptomatic abdominal distension are also criteria to offer surgical intervention.[6]

In this case, pleural effusion puncture for her symptoms. The prognosis is excellent in those asymptomatic or when operation or aspiration can easily control cysts. The prognosis depends in part on the severity of cystic disease in other organs like the renal, where renal failure may be a dominant risk.[1,2,5]

In this case, renal function test was within normal limits, and liver function test was increasing of alkaline phosphatase (241U/L) and γ-GT (131U/L). Right pleural effusion had been puncture to decrease her symptoms. The patient was discharged without any needs for follow-ups.

4. Conclusion
We have reported a case of the polycystic liver disease with right pleural effusion based on clinical features, laboratory, and radiology examination. Patients already do puncture of pleural fluid and patient went home.

References
[1] Feldman M, et al. 2006 Fordtran’s gastrointestinal and liver disease: pathophysiology / diagnosis / management, 8th edition vol 2 (Philadelphia: Saunders-Elsevier) chapter 94
[2] Friedman S L, et al. 2003 Polycystic liver disease essentials of diagnosis, current diagnosis & treatment in gastroenterology 2nd edition (McGraw-Hill) pp 809-11
[3] Karimbeg A A, et al. 2006 Multiple cysts in the liver autosomal dominant polycystic liver disease Netherland J. Med. 64(6) 199-201
[4] Morgan D E, et al. 2006 Polycystic liver disease: multimodality imaging for complications and transplant evaluation Radio. Graph. 26(6) 1655-68
[5] Peter R M N Liver and biliary tract disorder GI liver secret 3rd edition Chapter III pp 312-3
[6] Woolnough K, et al. 2012 Polycystic liver disease presenting with an exudative pleural effusion: a case report J. Med. Case Rep. 6(107)
[7] Li T J, et al. 2008 Treatment of polycystic liver disease with resection-fenestration and a new classification I World J. Gastroenterol. 14(32) 5066-72
[8] Halim F H 2012 Efusi pleura dan penyakit-penyakit dengan efusi pleura Kompendium tatalaksana penyakit respirasi kritis paru PERPARI Jilid I pp 279-88
[9] Caetano-Junior E M, et al. 2006 Laparoscopic management of hepatic cysts Surg. Laparosc. Endosc. Percutan. Tech. 16(2) 68-72
[10] Yang G S, et al. 2004 Combined hepatic resection with fenestration for highly symptomatic polycystic liver disease: A report on seven patients World J. Gastroenterol. 10(17) 2598-601