Polycystic Liver Disease with Huge Infected Cyst Displacing the Pancreas, Inferior Vena Cava and Right Kidney

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ABSTRACT

Multiple liver cysts can be an isolated disease (isolated polycystic liver disease [PLD]) or they can be part of multi-organ involvement in other diseases, such as adult autosomal dominant polycystic disease (APD), which is the most frequently inherited polycystic disease affecting 1 in 1,000 of the population. About 33% of APD patients are expected to develop PLD. Cysts in the liver, as opposed to cysts in the kidney, grow dramatically in number and size. They are usually asymptomatic but may present with signs and symptoms of compression of adjacent organs as the cysts grow. We present a case of a 73 year-old female patient with PLD and a large (25 × 19 × 18 cm) infected cyst which caused the displacement of the inferior vena cava, right kidney, and pancreas. We also discuss the management of this patient along with a review of the literature.

Key words: Adult autosomal dominant polycystic kidney disease, liver cyst, polycystic liver disease

INTRODUCTION

Adult autosomal dominant polycystic disease (APD) is the most commonly inherited polycystic disease affecting 1 in 1,000 of the population. The majority of extra renal cysts in APKD are mainly located in the liver (33%), but the disease may also involve other organs.[1] Isolated polycystic liver disease (PLD) is rare, with an estimated incidence below 0.01%.[2] Both APKD and isolated PLD have an autosomal dominant inheritance pattern.[3] PLD usually has more severe liver phenotypes, especially in patients with a history of multiple pregnancies or prolonged exogenous estrogen exposure, which suggests a hormonal component.[4] PLD usually remains asymptomatic. Symptoms, if present, are usually nonspecific and are caused by increasing liver volume and adjacent visceral compression. Several acute complications may occur due to the sheer size of these cysts, which grow steadily and attain huge dimensions, due to displacement and compression of adjacent organs. A number of cases have been reported in which the cysts...
have attained enormous proportions (22 × 19 cm), resulting in acute complications such as pancreatitis, deep vein thrombosis, and acute renal failure. The present case describes a 73-year-old female patient with APD with PLD, with a hepatic cyst (25 × 19 × 18 cm), which caused the displacement of the right kidney, pancreas, and inferior vena cava (IVC).

This patient was known to have hypothyroidism, hypertension, and APD with chronic renal impairment. She presented with abdominal discomfort and supra pubic abdominal pain of one week’s duration. She also complained of nausea and vomiting of one day’s duration.

On examination her blood pressure was 117/76 mm/hg, pulse rate was 117/min, and a temperature of 36.8°C; there was no icterus. Cardiovascular, respiratory, and neurological system examinations were unremarkable. Her abdomen was distended, non-tender hepatomegaly. A computed tomography scan of the abdomen and pelvis without contrast revealed that both kidneys were enlarged with multiple variable sized cysts and hepatomegaly with multiple cysts, the largest of which was on the right side and measured approximately 25 × 19 × 18 cm, exerting a significant mass effect on the adjacent organs [Figures 1 and 2]. The laboratory results are shown in Table 1.

The cystic fluid was aspirated two days after starting antibiotics and the analysis of the fluid revealed white blood cells. However, a culture did not result in the growth of bacteria. As a result of the infected cysts, she was diagnosed with PLD. The patient was started on intravenous fluids. The following day, an ultrasound-guided pigtail was inserted to drain the fluid from the cyst; a total of and 3 liters of fluid was drained. After a period of one week, the patient improved clinically and was discharged.

**DISCUSSION**

APD is the most frequently inherited polycystic disease affecting 1 in 1,000 of the population. Approximately 33% of extrarenal cysts in APD are located in the liver, but various other organs may also be involved. Isolated PLD is rare, with an estimated incidence below 0.01%. Both APD and isolated PLD have an autosomal dominant inheritance pattern. Cysts in the liver grow to huge dimensions rapidly as opposed to renal cysts. PLD usually remains asymptomatic. Symptoms, if present, are usually nonspecific and are caused by increasing liver volume and adjacent visceral compression. Less than 5% of patients with PLD have acute medical complications. These complications include hemorrhage into the cyst, infection, rupture, portal hypertension, ascites, Budd–Chiari syndrome, and deep vein thrombosis.[5] The cysts in PLD may multiply in number and grow continuously to attain huge dimensions. Campos Franco et al. reported a case of PLD with a cyst measuring 22 × 19 cm, which displaced the right kidney.[6] A literature review has revealed very few cases of large cysts in PLD with acute complications due to pressure effects.

A previous report described a 70-year-old female who had a partial intestinal obstruction secondary to the multiple renal and hepatic cysts compressing the bowel.
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In another report, a 66-year-old man who presented with progressive lower limp edema and high jugular venous pressure was found to have compression of the right atrium and IVC due to a hepatic cyst which was revealed through an echo. Massive deep vein thrombosis in leg veins reaching the distal part of the IVC due to huge liver cyst has also been reported. A rare case of chronic obstructive pancreatitis due to a pancreatic cyst in a patient with APD has also been reported. Our patient, who was a 73-year-old female patient known to have APD with chronic renal failure and PLD, presented with abdominal pain and on investigation was found to have a giant (25 × 19 × 18 cm) infected liver cyst causing displacement of the adjacent structures. To the best of our knowledge, this is the largest liver cyst reported in PLD. Even though significant pressure effects were seen on right kidney, pancreas and IVC, no acute complications with these organs were seen. The management of compressive large liver cysts is by different modalities, depending on the cause and urgency of the situation. Aspiration of cyst fluid, ethanol sclerosis, and laparoscopic fenestration are possible treatment options. A liver transplant is another option if the cysts are multiple and compressing on adjacent organs. Surgical intervention may be needed in patients presenting with massive hepatomegaly or other complications such as cyst rupture, infection, hemorrhage, or obstructive jaundice.

**Table 1: Lab investigations**

| Variable                  | 2 months prior | At presentation | 2 weeks after | Reference range          |
|---------------------------|----------------|-----------------|---------------|--------------------------|
| White blood cell count    | 9.2            | 22.1            | 7.3           | 3.3-10.8×10⁹         |
| Hemoglobin                | 9.9            | 8.2             | 8.6           | 12-16 g/dL              |
| Platelet count            | 411            | 730             | 554           | 150-500×10⁹         |
| Blood urea nitrogen       | 7.9            | 15.6            | 7.9           | 1.7-8.3 mmol/L        |
| Serum creatinine          | 137            | 152             | 123           | 44-80 umol/L          |
| Serum sodium              | 134            | 128             | 133           | 135-147 mmol/L        |
| Serum potassium           | 4.4            | 5.5             | 5.4           | 3.5-5.1 mmol/L        |
| Serum chloride            | 97             | 97              | 97            | 98-106 mmol/L         |
| Total bilirubin           | 7              | 5.8             | 4             | 3-17.1 umol/L        |
| Alanine aminotransferase  | 18             | 17              | 12            | 5-35 IU/L            |
| Alkaline phosphate        | 356            | 573             | 441           | 35-187 IU/L         |

CONCLUSION

In conclusion, cysts in PLD may grow to enormous sizes and compress and compromise the function of adjacent organs and present with acute complications such as pancreatitis, renal failure, intestinal obstruction, and deep venous thrombosis.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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