Case Report **OPEN ACCESS**

Intraperitoneal rupture of renal cyst in autosomal dominant polycystic kidney disease

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Abstract

Context: To report a rare case of spontaneous rupture of an infected renal cyst into the peritoneal cavity. **Case Report:** We report a case of 66-year old man with autosomal dominant polycystic kidney disease and kidney dialysis who had suffered from intestinal obstruction and peritoneal syndrome for 2 days associated with purulent urine. An exploratory laparotomy found multiple hepatic cysts and bilaterally enlarged polycystic kidneys. We noted left renal cyst infected and ruptured into the peritoneal cavity causing general peritonitis. The patient died from septic shock and hemodynamic failure. **Conclusion:** The peritoneal rupture of infected renal cyst is an exceptional and serious complication of polycystic kidney disease. Medical and surgical treatments are urgent to prevent lethal complications.

Keywords: Kidney cyst, autosomal dominant polycystic kidney disease, ruptured infected cyst, peritonitis

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Introduction

Multiple renal cysts occur commonly in autosomal dominant polycystic kidney disease and are generally considered to have little clinical significance [1, 2]. Rupture of an infected renal cyst is uncommon causing retroperitoneal sepsis, and peritonitis due to infected cyst rupture is even more unusual. To our knowledge, after revue of the world literature, this is the first case reported. Early diagnosis and surgical treatment are crucial to prevent lethal complications.

Case Report

A 66-year-old man with known polycystic kidney and liver disease and kidney dialysis of ten years' duration was admitted to the emergency department complaining of acute abdominal pain. The patient had a seven days history of left flank pain, fever and vomiting. He suffered for the two recent days from a severe diffuse pain and obstipation. Physical examination revealed a temperature of 38, 5°C, enlarged and painful left kidney, diffuse tenderness and purulent urine. On laboratory investigation, white blood count was 10300/mm^3 and serum creatinine concentration was 624 µmol/ml. Urine culture isolated *Escherichia coli.* X-ray abdominal exam showed small bowel dilation. Enhanced CT scan revealed multiple hepatic cysts and bilaterally enlarged polycystic kidneys. It showed a communication between heterogenous cyst in left kidney and pericolic space with soft tissue thickening in the left mesocolon, there was also an increased amount of peritoneal fluid (Figure 1).

The diagnosis of peritonitis was suspected and the patient underwent a laparotomy exploration. We noted a generalized peritonitis and a 150 ml purulent collection from a ruptured cyst of a giant polycystic left kidney (Figure 2).

This collection was covered by the mesentery and the left mesocolon. We proceeded to a peritoneal lavage and drainage after debridement. Bacteriol analysis of the collection isolated *Escherichia coli.* The patient did not

improve with despite intensive medical support and parenteral antibiotic. He died on the 3rd post-operative day from a septic shock and hemodynamic instability.

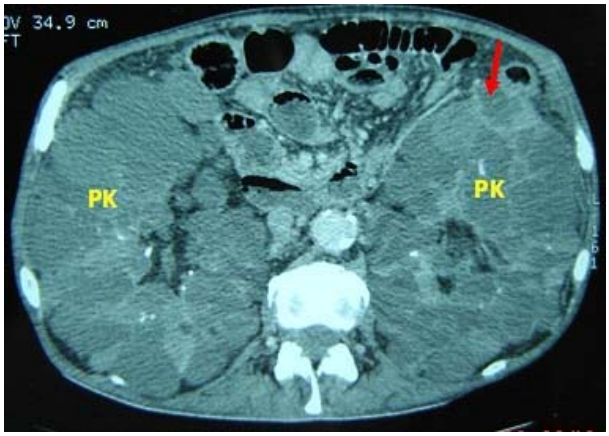

Fig. 1 Enhanced CT scan showed bilaterally enlarged polycystic kidneys (PK), heterogeneous cyst in left kidney (red arrow) and thickening pericolic space.

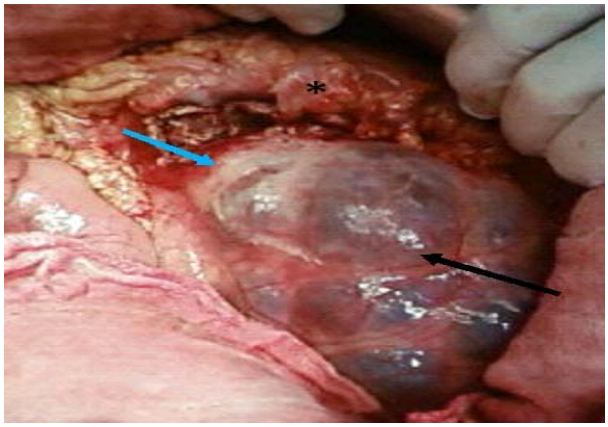

Fig. 2 Note the giant polycystic left kidney (black arrow) and a purulent collection from a ruptured cyst into the peritoneal cavity (blue arrow) after removing the adherent mesentery and left mesocolon (asterisk).

Discussion

Polycystic kidney disease (PKD) is an inherited disorder that involves bilateral renal cysts without dysplasia. Polycystic kidney disease is broadly divided into two types: autosomal recessive polycystic kidney disease (ARPKD), previously known as infantile polycystic kidney disease, and autosomal dominant polycystic kidney disease (ADPKD), previously known as adult polycystic kidney disease [1, 2]. ARPKD is characterized by cystic dilatation of renal collecting ducts associated with hepatic abnormalities of varying degrees, including biliary dysgenesis and periportal fibrosis. All patients with ARPKD have congenital hepatic fibrosis (CHF), which may have more severe clinical manifestation than the renal disease. The portal hypertension secondary to the CHF can be clinically debilitating, with splenomegaly, varices, and gastrointestinal hemorrhage. Autosomal dominant polycystic kidney disease differs from autosomal recessive polycystic kidney disease in that cysts associated with autosomal dominant polycystic kidney disease develop anywhere along the nephron. Upon clinical presentation, kidneys are usually enlarged, with numerous, large, round nodules on the external surface of the kidney, causing the loss of its original reniform shape. ADPKD occurs at an incidence of approximately 1/500 to 1/1000 live births [3, 4]. It affects all races. Age-adjusted sex ratios greater than unity (1·2–1·3) suggest a more progressive disease in men than in women [2]. The genes responsible for ADPKD were localized to the short arm of chromosome 16 (*PKD1*) in 85% of cases and the long arm of chromosome 4 (*PKD2*) in most of the remaining cases. The proteins encoded by *PKD1* and *PKD2* are polycystin 1 and polycystin 2, respectively. These proteins are expressed in the developing kidney, and their functions overlap considerably. The dysfunction of these proteins is thought to be pathogenetically responsible for the manifestations of ADPKD, primarily by renal ciliary dysfunction.

Patients with ADPKD are usually diagnosed and become symptomatic in adulthood [1, 2]. The diagnosis is based on genetic testing by linkage or sequence analysis to identify *PKD1* and *PKD2* mutations [2]. Fifty percent of patients with ADPKD will develop end-stage renal disease requiring replacement therapy by dialysis or kidney transplantation when they reach their 50s [2, 4, 5]. It is a multisystem disorder characterized by progressive cystic dilatation of both kidneys with variable extrarenal manifestations. These manifestations include cysts in other organs: liver (up to 90%), seminal vesicles (40% of men), pancreas (5%) and arachnoid membrane (8%) [1, 2]. Other abnormalities are also reported such as intracranial aneurysms and dolichoectasias, aortic root dilatation and aneurysms, mitral valve prolapse, and abdominal wall hernias [1, 3, 6].

There are several complications which can develop with ADPKD including: abdominal pain, renal masses, kidney stones, hematuria, hypertension, urinary tract infections, manifestations of stroke secondary to cerebral hemorrhage of ruptured aneurysms, hepatic cysts, Cardiovascular system manifestations, inguinal, abdominal, and umbilical hernias, as well as colonic diverticulae [2, 4, 5, 7]. Infection of a cyst within a polycystic kidney is a serious complication of ADPKD, potentially leading to abscess, sepsis and death. Female preference and enteric organism predominance suggest that these renal infections are acquired retrograde from the urinary bladder [3]. The rupture of the infected cyst is reported always into the pyelocaliceal system with extravasation in the retroperitoneal space [1, 2, 8, 9]. The intraperitoneal rupture has never been described world-widely. Clinical presentation is that of peritonitis: fever, vomiting, abdominal pain or tenderness. In this context, the pain may be localized initially in the flank or upper abdominal quadrant, if the infectious process is not contained, the pain becomes diffuse. The severity of this sepsis state (dehydration, acidosis) is complicated by the renal insufficiency occurring with ADPKD. Abdominal ultrasound and enhanced CT scans can show multiple renal cysts with signs of infection, peritoneal abscesses and

increased amounts of peritoneal fluid. MRI and PET scan has been shown to be helpful in detecting infected cysts [6, 12]. Surgery remains the important therapeutic modality after preoperative medical support including broad-spectrum parenteral antibiotic coverage, correction of potential electrolyte and coagulation abnormalities, intensive care with hemodynamic, pulmonary, and renal replacement support. The follow up of this pathology is important to prevent such complication. Thus, cysts that are painful, infected, bleeding or causing an obstruction may need to be drained. Some authors suggest the surgical removal of one or both kidneys if there is not a residual renal function [5, 10, 11].

Conclusion

The peritoneal rupture of infected renal cyst in polycystic kidney disease is unforeseen complication. The prevention either by medical or surgical options represent the treatment of choice.

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