

Partial intestinal obstruction: a rare complication of autosomal dominant polycystic kidney disease. Case report and review of the literature.

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ABSTRACT

Autosomal dominant polycystic kidney disease is a common disease with an estimated prevalence of 1 in 400-1000 people. It is a multisystem genetic disorder characterised by multiple bilateral kidney cysts, often accompanied by cysts in the liver and other organs. It is the cause of 5-7% of end-stage renal disease on chronic haemodialysis and can lead to various renal and extrarenal complications. We report an unusual complication of this condition that has rarely been described in the available literature – a case of partial intestinal obstruction in a 70-year-old male on chronic haemodialysis due to polycystic kidney disease, with multiple typical associated manifestations. The patient was hospitalised because of cyst haemorrhage and infection. He complained of constipation accompanied by abdominal distension and pain. Ultrasonography and computed tomography showed multiple cysts of large size and partial intestinal obstruction due to cyst compression of the bowel. This was relieved by unilateral right nephrectomy.

Key-Words:

Autosomal dominant polycystic kidney disease (ADPKD); haemodialysis; nephrectomy; partial intestinal obstruction.

INTRODUCTION

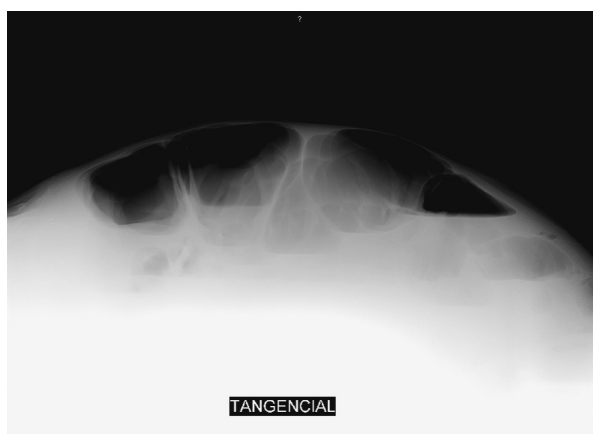
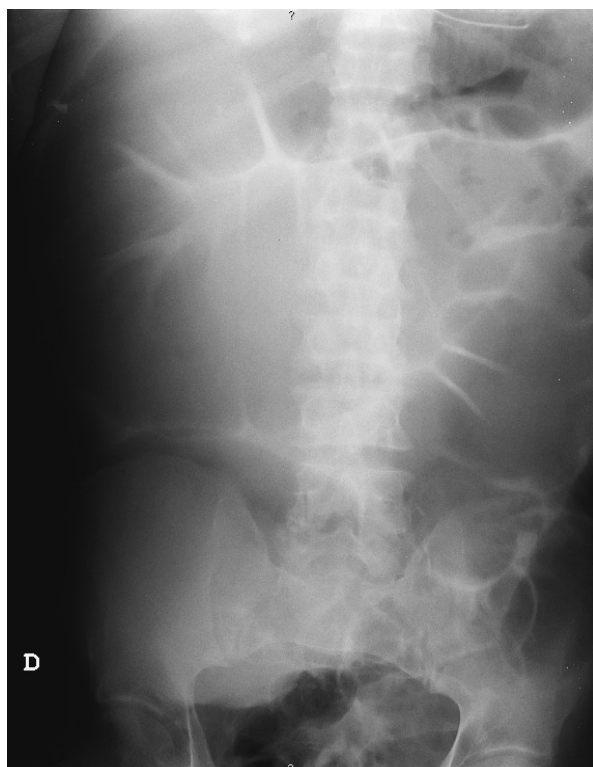
Autosomal dominant polycystic kidney disease (ADPKD) is a common disease (1:400-1000 people). It is usually asymptomatic, but it may become clinically evident with renal or extrarenal manifestations¹. It can present with arterial hypertension, flank pain, haematuria and progressive chronic renal disease. It can be complicated by infected or bleeding cysts. Its major extrarenal complications are intracranial aneurysms in 5-20%, hepatic and pancreatic cysts, valvular heart disease in 25% (mitral valve prolapse and aortic incompetence), colonic diverticula, and abdominal wall and inguinal hernia¹⁻³. Kidney enlargement resulting from the expansion of cysts in patients with ADPKD is continuous and quantifiable. It is associated with decline of renal function, and compression of local structures can lead to gastrointestinal symptoms, and intestinal compression, necrosis or obstruction⁴⁻⁷.

We report a rare clinical case of constipation and abdominal discomfort secondary to partial intestinal obstruction in a patient with ADPKD. Other cases reported in the literature are reviewed in the discussion.

CASE REPORT

A 70-year-old male presented on 1st September 2008 to the emergency unit of the hospital with a four-day history of fever ($>39^{\circ}\text{C}$) and abdominal distension. He also complained of progressive chronic constipation.

ADPKD had been diagnosed 16 years previously and had progressed to end-stage renal disease, treated by haemodialysis, in December 1999. He had known extrarenal manifestations of the disease: hepatic cysts, mitral and aortic valve disease, heart failure and hypertension.



Top: erect. Bottom: supine. Both showing marked gaseous abdominal distension.

Figure 1

Abdominal X-Rays in 2 positions (3/9/2008)

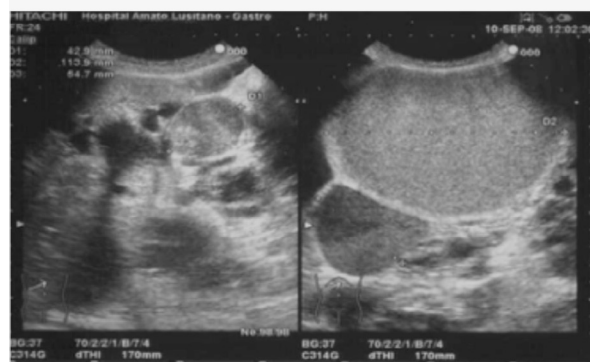
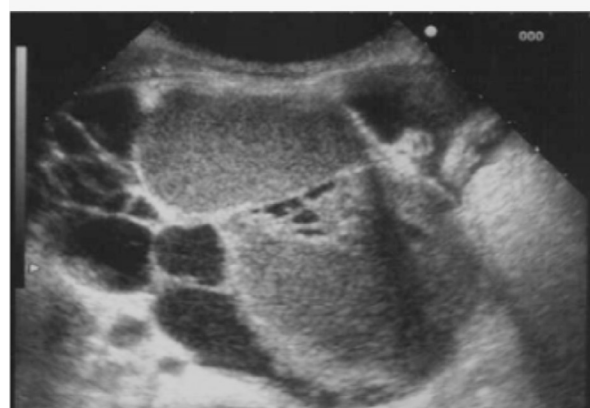


Figure 2

Abdominal ultrasonography (6/9/2008). Uncountable cysts in liver (largest in right side of the liver measuring 78 mm) and kidneys (largest 11 cm). There are 5 apparently bleeding cysts and 2 apparently infected cysts in the left kidney (these have heterogeneous content).

On admission he had a fever of 39.6°C and a distended and tympanic abdomen with mild left-sided tenderness. The remaining physical examination did not reveal any notable findings. Blood analysis showed anaemia (Hb 8.9 g/dL) and a very high C-reactive protein (>90 mg/dL). There was no leukocytosis. Urinalysis showed haematuria. There were no changes on chest X-ray. Screening for hepatitis B and C, cytomegalovirus and Epstein Barr virus was negative. No parasites were found.

As he was a patient on chronic haemodialysis with a past history of infected cysts, he was admitted to the nephrology department. Urine and blood cultures were performed and empiric broad-spectrum antibiotics were started based on the high probability of a new infected cyst. However, a few days later, the clinical picture persisted with pain, constipation, constant fever and abdominal distension. At that

time an abdominal X-ray was performed and showed multiple bowel air-fluid levels (Fig. 1).

Abdominal ultrasonography showed polycystic disease of the kidneys and liver with huge haemorrhagic and infected cysts (Fig. 2). To better characterise the cysts and the clinical picture he underwent an abdominal computed tomography scan which showed that the large size of the renal cysts had led to compression of the intra-abdominal organs, particularly the terminal part of the large bowel, leading to the rare complication of chronic partial intestinal obstruction (Fig. 3).

The infection started to improve with appropriate antibiotics. Constipation was partially relieved with abdominal massages and enemas, with a small quantity of stools of decreased consistency eliminated. However, the option of nephrectomy

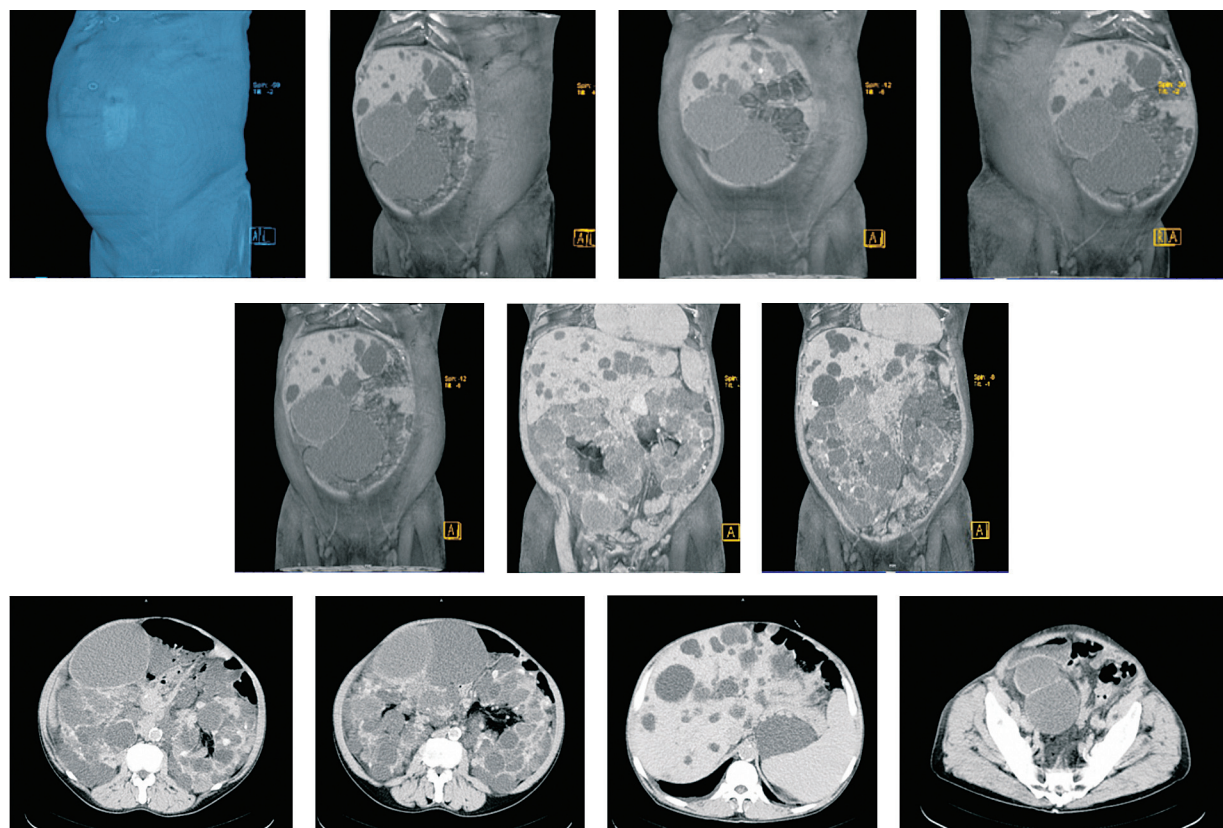


Figure 3

Abdominal computerised tomography (10/9/2008): uncountable liver and kidney cysts that fill most of abdominal cavity. Intra-abdominal organs and adjacent structures are compressed by the large cysts.

was considered to avoid the recurrence of infection and haemorrhage and to decompress the intra-abdominal organs, allowing free bowel movements and resolution of the partial intestinal obstruction.

On 13th October 2008 the patient underwent successful unilateral right nephrectomy and has since had total remission of gastrointestinal symptoms for over two years of follow-up.

■ DISCUSSION

ADPKD is characterised by progressive renal disease due to continuous growth of the cysts, and may have renal and extrarenal complications. This patient had large hepatic and renal cysts, end-stage renal disease needing chronic haemodialysis, heart failure with valve disease, bleeding and infected cysts and an unusual clinical picture of chronic partial intestinal obstruction.

Normally the bowel is flexible and can modify its shape and position easily inside the abdomen but, in this patient, huge cysts filled most of the abdominal cavity and led to compression of the surrounding organs, as documented in the images. Similar to this clinical picture, there are reported cases of very large polycystic kidneys⁴ weighing more than 20 Kg.

In addition to the recurring infections and haemorrhage, the patient had had a reduced quality of life for an extended period of time due to chronic constipation caused by partial extrinsic mechanical bowel occlusion that impeded the normal intestinal progression of stools and gas. Extensive investigation excluded a paralytic ileus, and the patient's final and uncommon diagnosis was that of chronic partial intestinal obstruction.

A review of the available references in electronic libraries reveals that intestinal obstruction as a complication of ADPKD, whether on chronic dialysis or not, has been rarely described.

The first nephrectomy of a polycystic kidney causing intestinal obstruction was mentioned as being in 1905 by Dr. W.A. Giles⁸. After that, in

1914 there was the first written report of an operation to remove an abdominal mass that was the cause of intermittent attacks of abdominal pain, vomiting and chronic constipation. Only during the surgery was the real cause of the symptoms found to be a huge left renal cyst⁹. In 1958 there was a brief description of a nephrectomy that relieved an acute large bowel obstruction⁸. In 1962 there was a case report of intestinal obstruction caused by ADPKD¹⁰ and in 1975 a patient with acute pancreatitis associated with duodenal obstruction caused by a right renal large cyst, and successfully relieved by its resection, was described¹¹. In 1976 there was a description of partial jejunal obstruction with jaundice secondary to a simple renal cyst, relieved with aspiration of cyst fluid¹². One year later there was another description of duodenal obstruction in a newborn secondary to kidney cysts¹³.

There are two more case reports of acute small bowel partial occlusion: one of duodenum¹⁴ in 1998, and another of the small intestine¹⁵ that resolved with percutaneous aspiration of the larger kidney cysts in 1999. In 2008 there was a report of a patient with an acute abdomen as the result of strangulation and necrosis of the intestine due to mesenterium ischaemia caused by enlarged kidney cysts; this was treated by proximal jejuno-transverse colostomy¹⁶. More recently, in 2009, there was a clinical report of a 97-year old woman with external colic compression by a right cortical renal cyst of about 15 cm in diameter, and associated symptoms of asthenia, loss of appetite, nausea and intermittent diarrhoea, that was also relieved with cyst drainage¹⁷. In 2010, there was another clinical report of a giant cyst that caused an acute abdomen due to acute small bowel occlusion. It resolved with excision of the mass⁷. There are also some other articles in the literature that report similar cases of bowel partial obstruction caused by large kidney cysts¹⁸.

In total, we found just 13 available clinical reports of intestinal compression by renal cysts (Table I). Five of the cases had chronic abdominal complaints but only two of the reports described the treatment by nephrectomy of chronic partial intestinal obstruction due to large kidney cysts^{8,9}.

Percutaneous aspiration of cysts may help manage severe pain due to haemorrhage or compression but

Table I

Case reports of intestinal compression due to kidney cysts in ADPKD patients

Author	Year	Diagnosis	Treatment
Giles WA ⁽⁸⁾	1905	Chronic intestinal obstruction	Nephrectomy
Beyers EC ⁽⁹⁾	1914	Chronic intestinal obstruction	Nephrectomy
Bedford PW ⁽⁸⁾	1958	Acute intestinal obstruction	Nephrectomy
Willox SW ⁽¹⁰⁾	1962	Subacute intestinal obstruction	n.a.
Bubrick MP ⁽¹¹⁾	1975	Acute bowel obstruction/acute pancreatitis	Partial nephrectomy
Sanella NA ⁽¹²⁾	1976	Acute partial jejunal obstruction (jaundice)	Cyst drainage
Porterfield GN ⁽¹³⁾	1977	Acute duodenal obstruction	n.a.
Zeliak NV ⁽¹⁸⁾	1993	Chronic intestinal obstruction	n.a.
Fried LF ⁽¹⁴⁾	1998	Acute small bowel partial obstruction	n.a.
Kakinoki K ⁽¹⁵⁾	2002	Acute small bowel partial obstruction	Cyst drainage
Yoshikawa T ⁽¹⁶⁾	2008	Acute intestinal necrosis	Colostomy
Vos B ⁽¹⁷⁾	2009	Chronic partial intestinal obstruction	Cyst drainage
Arruda PFF ⁽⁷⁾	2010	Acute small bowel obstruction	Nephrectomy
Gonçalves J	2012	Chronic partial intestinal obstruction	Nephrectomy

n.a. (information not available)

has no effect on long-term outcome. Surgical decompression of large cysts may provide effective symptomatic relief. Both open and laparoscopic surgical approaches have been described⁶.

Unilateral or bilateral nephrectomy in ADPKD is considered to be a therapeutic option best avoided if possible. However it is mandatory when a patient with ADPKD is on dialysis and has chronic abdomen pain that requires narcotic medication, recurrent urinary tract infection, insufficient abdominal space to hold a proposed kidney transplant, renal cell carcinoma or chronic haematuria requiring ongoing transfusions^{7,19-21}. In addition to these formal indications, nephrectomy will also relieve cosmetic deformation of the abdomen and may increase the quality of life²².

This review and rare case report of a partial intestinal obstruction associated with ADPKD highlights that this complication should be considered in the differential diagnosis of a patient with ADPKD (whether on dialysis or not) and persistent constipation. The second point of interest is to underline that nephrectomy led to total relief of symptoms.

Conflict of interest statement. None declared.

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