Sub-Acute Intestinal Obstruction – A Rare Complication in a patient with Autosomal Dominant Polycystic Kidney Disease: A Case Report

Nazish Naseer, Sonia Yaqub.

Section of Nephrology,
Department of Medicine,
Aga Khan University,
Stadium Road,
Karachi, Pakistan

Abstract:

An 82 year old gentleman, with autosomal dominant polycystic kidney disease (ADPKD) presented with abdominal pain and distension associated with vomiting and constipation. On examination he was an obese, ill looking male with a distended abdomen and bilateral palpable flank masses. Gut sounds were sluggish. Small and large bowel loops were found to be dilated on abdominal x-ray. Computed tomography scan of the abdomen showed grossly enlarged kidneys occupying almost whole of the abdomen pushing small bowel loops anteriorly. Based on clinical and radiological findings a diagnosis of sub-acute intestinal obstruction was made. Patient was managed conservatively (i.e. with NG tube and rectal decompression).

This case highlights intestinal obstruction as a rare complication of ADPKD.

Keywords: *Autosomal dominant polycystic kidney disease (ADPKD), Intestinal obstruction*

Corresponding author:

Dr. Sonia Yaqub
Assistant professor
Director Nephrology Residency Program
Department of Medicine
Aga Khan University
Karachi
Email: Sonia.yaqub@aku.edu

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Introduction:

Autosomal dominant polycystic kidney disease (ADPKD) is a common disorder, occurring in approximately 1 in every 400 to 1000 live births.¹ It is a multisystem disorder characterized by formation and enlargement of cysts in kidneys and other organs.² Clinical manifestations can range from mild abdominal discomfort to more serious complications like cyst hemorrhage, recurrent cyst infections. Clinically significant extra-renal complications like acute pancreatitis, cholangitis, peptic ulcer bleeding, bowel necrosis and rarely intestinal obstruction has also been reported in literature.³ We here report a case of a patient with adult polycystic kidney disease who presented with sub-acute intestinal obstruction.

Case presentation:

An 82 year old gentleman a known case of ADPKD, diagnosed at the age of 40 years, presented to the emergency department with three days history of generalized colicky abdominal pain. It was associated with non-bilious, non-projectile vomiting, decreased appetite and relative constipation. He denied any history of fever, weight loss, hematemesis, melena, hematuria or lower urinary tract symptoms. On examination he appeared to be in distress due to pain. His BP was 133/85 mmHg, pulse 100 beats per minute and a temperature of 36.8 C. He appeared dehydrated. His abdomen was distended and non tender. Gut sounds were sluggish. Both kidneys were palpable. His respiratory, cardiovascular and neurological examination was normal.

On presentation his Hemoglobin was 11.2g/dl, white cell count of 13.8 x 10⁹/L. Blood urea nitrogen was 68 mg/dl, serum creatinine of 2.8mg/dl (baseline Cr: 2 mg/dl), creatinine clearance of 22 ml/min and with a serum potassium of 4.5 mmol/L. His C-reactive protein was 9.9 mg/dl.

Liver function tests, coagulation profile, amylase and lipase were normal.

Abdominal X-ray showed significantly dilated small and large bowel loops without any evidence of pneumoperitoneum (figure 1).

ADPKD & Intestinal Obstruction

Computed tomographic (CT) scan of abdomen with intravenous contrast was done which showed grossly enlarged kidneys (right kidney measuring 36x27x22 cm in size and left kidney measuring 39x28x20cm in size) with almost complete replacement of renal parenchyma by the cysts (Figure 2). Polycystic kidneys were occupying almost whole of the abdomen pushing the bowel loops anteriorly.

A diagnosis of sub-acute intestinal obstruction secondary to massively enlarged polycystic kidneys was made. Patient was managed conservatively with intravenous fluids, analgesics (Paracetamol and Tramadol), NG tube and rectal decompression. NG was kept on low volume suction. He initially responded to the conservative approach and passed stool but after removal of rectal and NG tube he again developed the symptoms. Both were then re-introduced and total parenteral nutrition was started.

A multidisciplinary meeting including a general surgeon, Urologist and the primary nephrologist was arranged and after a detailed discussion and literature review, surgical options like nephrectomy / Cyst reduction surgery and their risks were discussed but the patient and his family. They opted for conservative management due to very high risk associated with anesthesia and surgical procedure.

Figure 1. Abdominal X-Ray

Showing significantly dilated small bowel and large bowel as compared to previous x-ray. No definite evidence of pneumoperitoneum. Arrows

Figure2 (A-C). Axial and coronal non-enhanced CT showing bilateral enormously enlarged kidneys (long arrows) replaced entirely by hypodense cysts occupying most of the abdominal cavity leaving little room for bowel loops which are compressed and anteriorly displaced. Few dilated fluid and air distended bowel loops are seen in anterior abdomen (short arrows)

ADPKD & Intestinal Obstruction

Discussion

ADPKD is one of the most common hereditary cystic disorders characterized by multiple cysts and kidney enlargement. It is the 4th most common cause of End stage renal disease.¹ The number of cysts increase with age and so does the kidney size which in turn contributes to the sign, symptoms and complications of the disease.² Although renal cysts is a common manifestation, ADPKD can also present with many other systemic manifestations and extra-renal features. These include cysts in organs other than kidney (liver: 94%, seminal vesicle: 40%, pancreas: 9%, arachnoid membrane: 8%, and spinal meningeal, 2%) and connective tissue abnormalities (mitral valve prolapse: 25%, intracranial aneurysms: 8%, and abdominal hernia: 10%).³ Abdominal pain can be a diagnostic challenge in patients with ADPKD. Although cysts related complications like cyst hemorrhage, rupture and infection can give rise to abdominal pain but non renal sources of pain unrelated to cystic complications like diverticulitis, cholangitis, peptic ulcer, bowel obstruction / necrosis etc. should also be considered.

Intestinal obstruction is a very rare complication of ADPKD. Only a few cases are reported in literature so far. Normally the bowel is flexible and their shape and position can be modified easily inside the abdomen but huge cysts can lead to compression of the bowel loops and adjacent structures.⁴ In such cases, patients can present with acute abdomen or with sign and symptoms of complete or partial intestinal obstruction.^{5,6} The goal of treatment is removal of pressure from compressed bowel loops by the enlarged renal cysts. Patients have previously been managed with cyst aspiration, partial excision, and unilateral nephrectomy.^{4,5,7}

Conclusion:

There can be a number of causes of abdominal pain in ADPKD patients. Intestinal obstruction is a rare complication of this disease and should always be kept amongst the differentials of abdominal pain while dealing with a patient of polycystic kidney disease.

Conflict of Interest Statement: none declared.

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