Intestinal Obstruction in Autosomal Dominant Polycystic Kidney Disease

Kaheita KAKINOKI, Yatsugi NODA, Masayoshi TAKAEDA, Masahiro KUBO, Keiichi MIZUHASHI,
Ichirou MIYAMOTO, Tetsuya NISHIDA and Yasuhiro TAKEMORI

Abstract

A 42-year-old woman with autosomal dominant polycystic kidney disease (ADPKD) was admitted to our hospital on April 29, 1999, with complaints of abdominal pain. A diagnosis of intestinal obstruction was reached on the basis of clinical findings and X-ray evidence. A computed tomography scan of the abdomen showed massively enlarged kidneys, especially the right kidney, which seemed to compress the small intestine. The patient underwent percutaneous aspiration of the largest cysts on the surface of the right kidney. The symptoms, in this rare case of intestinal obstruction by an enlarged kidney in ADPKD, were alleviated the day after the aspiration procedure. (Internal Medicine 41: 441–444, 2002)

Key words: ADPKD, percutaneous aspiration, small intestine, renal cyst, ileus

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is characterized by multiple bilateral cysts and kidney enlargement. However, the enlarged organs rarely directly interfere with the functioning of surrounding organ systems. We report here a case of ADPKD causing symptomatic intestinal obstruction.

For editorial comment, See p 415.

Case Report

A 42-year-old woman was admitted to our hospital on April 29, 1999, with complaints of abdominal pain and vomiting. Her father had died of renal failure. Four years previously the patient had been diagnosed with ADPKD. At that time, renal

insufficiency was evidenced by a blood urea nitrogen level of 40 to 50 mg/dl and a serum creatinine level of 3 to 4 mg/dl. However, she had not undergone any treatment because of an absence of symptoms until two days before admission, when she suddenly developed cramping abdominal pain and vomiting without any apparent cause. The pain had been continuing for two days, accompanied by nausea, constipation, accumulation of intestinal gas, and abdominal distention. As her condition did not improve, she was referred to our hospital.

An abdominal X-ray picture taken with the patient in the supine position revealed that the dilated small intestine contained an excessive amount of gas. A roentgenogram obtained with the patient in the upright position showed air-fluid levels in the small intestine (Fig. 1). Clinical findings plus the X-ray evidence led to a diagnosis of intestinal obstruction.

The patient's body temperature was 36.5°C, blood pressure 158/94 mmHg, and pulse 68/min. She was 152.0 cm tall and weighed 63.0 kg. Physical examination showed the patient to be obese and in pain. There was no sign of conjunctivitis, the conjunctivas were normal and of a good color, and the sclera showed no signs of jaundice. The lungs were clear in response to percussion and auscultation. Heart sounds were normal without any audible murmur. The abdomen was markedly distended and diffusely tender, while bowel sounds were loud and metallic. No organs or masses could be identified by palpation. In spite of the abdominal distention, the extremities were relatively lean. Neurologic examination results were negative.

The hemoglobin level was 10.1 g/dl, and the white cell count 6,500/mm³. Renal function studies showed blood urea nitrogen at 108 mg/dl, serum creatinine at 12.5 mg/dl, and creatinine clearance at 6.0 ml/min. The C-reactive protein level was normal. The urine was positive for protein (++) and occult blood (++); the sediment contained 6 to 10 red cells and 6 to 10 white cells per high-power field (Table 1). The patient was suffering from end-stage renal disease as a result of ADPKD.

A computed tomographic (CT) scan of the abdomen and pelvis showed that the bilateral kidneys were markedly enlarged due to the presence of enormous cysts occupying most of the abdomen and distorting other intraabdominal structures. The

From the Department of Internal Medicine, Toyama Rosai Hospital, Uozu

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Reprint requests should be addressed to Dr. Yatsugi Noda, the Department of Internal Medicine, Toyama Prefectural Central Hospital, 2-2-78 Nishinagae, Toyama 930-8550

Figure 1. (A) An abdominal X-ray picture taken with the patient in the supine position shows an excessive amount of gas in the small intestine, but little or no colonic gas is seen. (B) A roentgenogram obtained with the patient in the upright position reveals air-fluid levels in the small intestine.

Table 1. Laboratory Findings on Admission

<u>Hematology</u>		AST	17 IU/l	<u>Serological test</u>	
White blood cell	6,500/ μ l	ALT	13 IU/l	C-reactive protein	0.4 mg/dl
Neutrophil	72.8%	LDH	604 IU/l	<u>Tumor marker</u>	
Eosinophil	8.2%	ALP	195 IU/l		
Basophil	0.6%	γ -GTP	9 IU/l	CEA	4.0 ng/ml
Lymphocyte	13.0%	Total bilirubin	0.06 mg/dl	<u>Urinalysis</u>	
Monocyte	5.4%	Total amylase	101 U/l		
Red blood cell	334×10^4 / μ l	Blood urea nitrogen	108 mg/dl	Protein	100 mg/dl
Hemoglobin	10.1 g/dl	Creatinine	12.5 mg/dl	Sugar	0.10 g/dl
Hematocrit	30.9%	Uric acid	9.0 mg/dl	Urobilinogen	0.1 EU/dl
Platelet	30.8×10^4 / μ l	Na	144 mEq/l	Occult blood	(2+)
<u>Blood Chemistry</u>		K	3.7 mEq/l	Sediment	
		Cl	104 mEq/l	Red blood cell	6–10/HPF
Total protein	6.8 g/dl	Ca	3.3 mEq/l	White blood cell	6–10/HPF
Albumin	63.3%	P	9.3 mg/dl	Hyaline cast	(–)
α 1-globulin	4.1%	Fe	41 μ g/dl	Granular cast	(–)
α 2-globulin	11.6%	Creatinine clearance	6.0 ml/min	Squamous epithelium	(+)
β -globulin	8.6%				
γ -globulin	12.4%				

AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, LDH: Lactate dehydrogenase, ALP: Alkaline phosphatase, γ -GTP: gamma-glutamyl transpeptidase, CEA: carcinoembryonic antigen.

Figure 2. A computed tomographic scan of the abdomen and pelvis shows massively enlarged polycystic kidneys (A) and dilatation of the small intestine filled with fluid (B).

Figure 3. Barium enema shows medial deviation of the ascending and descending colons due to compression by the enlarged kidneys.

right kidney measured 25.0×20.5×12.0 cm and the left kidney 23.0×15.5×11.0 cm, so that enlargement of the right kidney was especially pronounced.

The small intestine was markedly dilated and filled with fluid, but without distention of the colon (Fig. 2). The exact location of the obstruction was not clear, but it seemed most likely that the large right kidney was compressing the small intestine.

On admission, conservative treatment was soon started with fasting and replacement of fluid. However, the symptoms progressively worsened and the abdomen became spastic during the conservative management. On the second day of hospitalization, percutaneous needle aspiration of the largest cysts on the surface of the right kidney was performed to alleviate the symptoms of obstruction. Under ultrasound guidance, an 18-gauge needle was introduced into the cysts through the right flank and a total of 1,130 ml of turbid yellow fluid was evacuated. The cells and culture of the fluid were normal. Abdominal pain and fullness were alleviated soon after cyst aspiration, and on the following day, the symptoms caused by intestinal obstruction had disappeared. An abdominal X-ray showed that the amount of gas in the small intestine had decreased, and a barium enema administered after the symptoms had vanished revealed an abnormal intestinal shape with medial deviation of the ascending and descending colons due to compression of the enlarged kidneys (Fig. 3). Finally, colonoscopic examination showed no obstruction of the colon. Despite relief of intestinal obstruction, however, the renal function did not improve and the serum creatinine level was still 11.4 mg/dl on the 15th hospital day. For this reason, an AV fistula was created for hemodialysis during the patient's hospital stay. The patient was discharged on the 38th hospital day and since then hemodialysis has been performed at our clinic. There has been

Table 2. Reported Cases of Intestinal Obstruction in Polycystic Kidney Disease

Case	Age	Sex	Author	Year	Location	Treatment
1.	53	F	Moadel E et al	1975	Duodenum	Not done
2.	Neonate	F	Garland N et al	1977	Duodenum	Nephrectomy
3.	64	F	Bednarz W et al	1992	Small intestine	Nephrectomy
4.	35	M	Fried LF et al	1998	Duodenum	Nephrectomy
5.	42	F	Present case	1999	Small intestine	Percutaneous needle aspiration

no recurrence 24 months after discharge.

Discussion

ADPKD is one of the most common hereditary disorders, characterized by multiple bilateral cysts and kidney enlargement. By age 50 the kidneys can be enormous, reaching up to 40 cm in length and 8 kg in weight (1). In spite of the large size of the kidneys, obstruction of surrounding organs has rarely been reported (2). Diverticulosis of the colon is generally recognized as a common extrarenal manifestation of ADPKD (3). However, little is known about intestinal obstruction as a result of ADPKD.

As far as we could determine, only five cases, including ours, of polycystic kidney disease complicated by intestinal obstruction have been reported, three of obstruction of the duodenum and two of the small intestine. All cases underwent nephrectomy (2, 4–7) except one who died of renal failure (6) (Table 2). In our case, conservative medical treatment for intestinal obstruction was started. Because the abdominal CT indicated that the right kidney was compressing the small intestine, the patient was treated with ultrasound guided percutaneous aspiration of cyst fluid. Cyst aspiration under ultrasound guidance is generally performed for ADPKD which is accompanied by pain and is unresponsive to medical management (8, 9). However, such cysts typically recur with pain within 3 to 6 months because of the relatively rapid reaccumulation of fluid (10). In the present case, however, there was no recurrence of the intestinal obstruction even though an abdominal CT scan performed 3 months after the cyst aspiration revealed reaccumulation of fluid in the cyst and enlargement of the kidney. Percutaneous needle aspiration of cyst fluid thus appears to be an effective treatment for the acute phase of intestinal obstruction in ADPKD.

If the present patient shows recurrence of intestinal obstruction, percutaneous instillation of iophendylate or alcohol following aspiration, cyst reduction surgery, or nephrectomy will need to be considered (10). Recently Ubara et al reported a patient with ADPKD who underwent successful transcatheter

arterial embolization (TAE) of the renal arteries, which led them to suggest that TAE was effective for shrinking enlarged kidneys without significant complications (11). TAE thus also seems to be an effective technique for preventing the relapse of intestinal obstruction in ADPKD.

Conclusion

We described a rare case of intestinal obstruction by an enlarged kidney with giant cysts associated with ADPKD. Aspiration of cyst fluid under ultrasound guidance seems to be valuable to try as an emergency procedure for acute intestinal obstruction in ADPKD.

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