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Retroperitoneal cyst hemorrhage in polycystic kidney disease

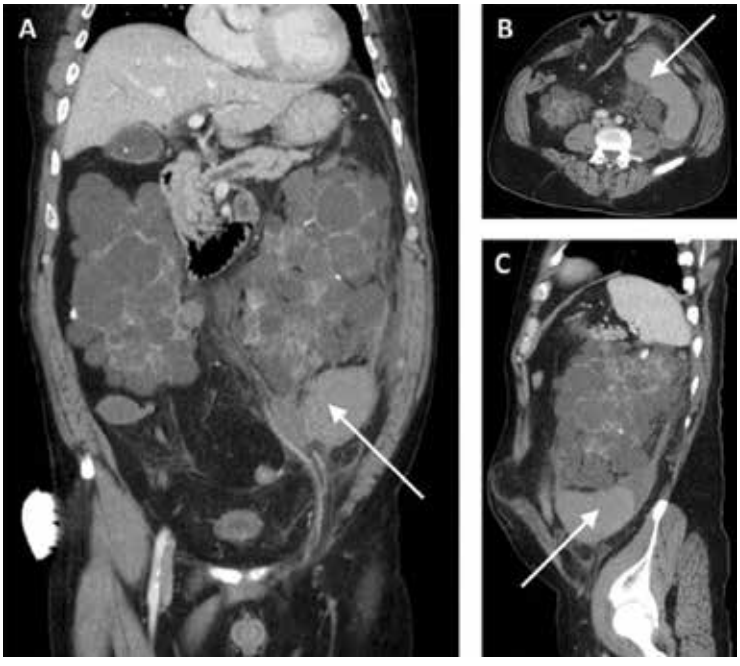

FIGURE 1. Computed tomography reveals acute hemorrhagic rupture of a cyst in the left kidney into the retroperitoneal space (arrows).

A 59-year-old man with autosomal dominant polycystic kidney disease (ADPKD), end-stage renal disease on hemodialysis, hypertension, and diverticulosis presented with acute pain in the left lower abdomen. The pain began 4 days previously, was dull and nonradiating, was relieved partially with hydrocodone-acetaminophen, and had no clear exacerbating factors. Two days before presentation, he developed a fever with chills. He reported no recent dysuria, diarrhea, hematuria, hematochezia, or melena. He had not been taking anticoagulants or nonsteroidal anti-inflammatory drugs, and he had no history of heavy lifting or trauma.

His temperature was 38.5°C (101.3°F), blood pressure 141/60 mm Hg (normal for this patient). On examination, his left lower quadrant was tender with voluntary guarding. Also present was a reducible ventral hernia, which was not new.

His hemoglobin level was 10.6 g/dL (reference range 13.0–17.0), which had dropped from a previous value of 13.7 g/dL.

Computed tomography of the abdomen and pelvis revealed a ruptured retroperitoneal hemorrhagic cyst (**FIGURE 1**) in the inferior aspect of the left kidney extending into the fascia of Gerota.

Since his vital signs were stable, he was managed supportively during his hospitalization with intravenous fluids, serial hemoglobin checks, and analgesia. He was eventually discharged home in good condition.

CYST HEMORRHAGE IN POLYCYSTIC KIDNEY DISEASE

ADPKD is a relatively common, inherited systemic disease that leads to cyst formation, primarily in the kidneys but also in the liver (94%), seminal vesicles (40%), pancreas (9%), arachnoid membrane (8%), and spinal meningeal area (2%).[1]

In addition to cyst formation in multiple organs, ADPKD can have extrarenal manifestations such as connective-tissue abnormalities (including mitral valve prolapse) (25%), abdominal hernia (10%), and intracranial aneurysm (8%).[1] Management of extrarenal complications of ADPKD is discussed in detail elsewhere.[2]

The estimated prevalence of ADPKD is 1 of every 400 to 1,000 live births. However, given that ADPKD is often clinically silent, it is diagnosed during the lifetime of fewer than

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half of people who have it.[3]

Most ADPKD cases are caused by mutations in either the *PKD1* or *PKD2* gene.[4,5] Although the mechanism of cyst formation in ADPKD is still unclear, it is known that *PKD1* and *PKD2* encode proteins called polycystin-1 and polycystin-2, respectively. Polycystin-1 is a membrane protein found in renal tubular epithelia, hepatic bile ductules, and pancreatic ducts. Polycystin-2 is involved in cell calcium signaling and has been identified in the renal distal tubules, collecting duct, and thick ascending limb. Mutations in *PKD1* and *PKD2* are thought to contribute to cyst formation, with *PKD1* mutations associated with earlier onset and more severe development of renal and extrarenal cysts.

Cyst hemorrhage

Hemorrhage of renal cysts is a well-known complication, occurring in up to 70% of patients with ADPKD.[6] Renal cyst hemorrhage often presents clinically as flank pain with point tenderness or hematuria, or both. Flank pain results from hemorrhage into a cyst with consequent distention of the renal capsule, whereas hematuria results from rupture of a cyst into the collecting system.

Spontaneous nonfatal retroperitoneal cyst hemorrhage, as in our patient, is rare. Indeed, in one series reviewing the abdominal computed tomographic findings of 66 patients with ADPKD, only 2 patients (3%) had perinephric hematomas in the absence of recent trauma.[6]

Management of cyst hemorrhage is primarily conservative. Pain associated with cyst hemorrhage is managed conservatively with bed rest, intravenous hydration, and analgesics (but not nonsteroidal anti-inflammatory drugs).

Hematuria is also managed conservatively with bedrest and intravenous hydration, and most episodes of hematuria are self-limiting and last 2 to 7 days. However, if excessive bleeding occurs, the patient may be at risk of urinary tract obstruction from clot formation. If obstruction occurs and persists beyond 2 weeks, then ureteral stenting may be necessary. In rare cases of prolonged, severe bleeding with extensive subcapsular or retroperitoneal hematomas, patients require hospitalization, transfusion, or percutaneous transcatheter embolization of the renal artery. If such efforts are not successful, surgery, including nephrectomy, may be required to control the hemorrhage.[2]

Other causes of abdominal pain

In addition to renal cyst hemorrhage, the differential diagnosis of abdominal pain in a patient with ADPKD includes cyst enlargement causing stretching of the renal capsule or traction on the renal pedicle, cyst infection, nephrolithiasis, pyelonephritis, and rarely, tumors including renal cell carcinoma.

Unlike cyst rupture and hemorrhage, which are associated with point tenderness, cyst infection often manifests as diffuse, usually unilateral flank pain with associated fever, nausea, malaise, and leukocytosis. Our patient had none of these except for fever, which can also occur in cyst hemorrhage.

Nephrolithiasis occurs in up to 35% of patients with ADPKD,[7] but no kidney stones were seen on computed tomography in our patient.

Pyelonephritis was unlikely in our patient, given that he had no significant white blood cells in his urinalysis and no leukocytosis.

Abdominal and pelvic imaging did not reveal any tumors in our patient. ■

Fewer than half of people with ADPKD are diagnosed with it in their lifetime

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