A 60-year-old man presented with complaints of gradually increasing abdominal distension for 8 days with decreased urine output for 5 days. Seven days earlier, a local doctor had tapped one liter of clear straw-colored ascitic fluid. There was no history of hematuria or pyuria. He also complained of constipation for the last 3 days. He was passing flatus, and there was no history of vomiting, melena, jaundice, fever, cough, or bleeding from any site. He also gave a history of significant anorexia. He was being treated by a local doctor for dyspepsia for 15 years. He was not a known diabetic, hypertension, alcoholic or a smoker.

One year back, he was admitted with features of acute abdomen. Based on clinical and investigative findings, he was diagnosed to have a sealed duodenal perforation and was managed conservatively. At ultrasonography, he was detected to have multiple hepatic and renal cysts. He was subsequently diagnosed to be having autosomal dominant polycystic kidney disease (ADPKD). His serum creatinine level was 4.0 mg/dL, which subsequently came down to 2.7 mg/dL. He was discharged with advice to follow up at the Nephrology Clinic.

On examination, he was conscious but disoriented. His pulse rate was 82/min, regular, and blood pressure 120/70 mmHg. He did not have anemia, jaundice, cyanosis or clubbing. He was detected to have pedal edema. He had essentially normal cardiovascular examination except for an end-systolic murmur. Except for scattered wheezing, his respiratory tract examination was normal.

On abdominal examination, he was had bilaterally ballotable renal lump. He had clinically detectable free fluid. There was no guarding or tenderness. Bowel sounds were sluggish. Central nervous system examination was normal.

**Investigations**

Hemoglobin 12.5 g/dL, TLC 9,800/dL (80 polymorph, 16 lymphocyte, basophil 2, monocyte 2). Peripheral blood smear showed normochromic normocytic RBC with adequate platelet. Serum electrolytes: Na⁺ 140 mEq/L, K⁺ 7.1 mEq/L. Urea 310 mg/dL, bilirubin 0.7 mg/dL, random blood sugar 118 mg/dL, amylase 100 U/L.

**Ultrasonography:** Liver enlarged with numerous small and large cysts; intrahepatic biliary radicals, common bile duct and portal vein were normal. Pancreas, spleen and gall bladder were also normal. Kidneys bilaterally enlarged with multiple cysts of varying sizes. There was distortion of the normal renal parenchymal echotexture and loss of cortico-medullary distinction. Presence of ascites was confirmed, and there was internal echogenicity with septations. Paracentesis revealed frank pus.

**Clinico-pathologic conference**

**Polycystic liver and kidney disease with ascites and sepsis**

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**Course in hospital**

Emergency medical management of hyperkalemia was instituted. The patient was started on broad-spectrum antibiotics. Nephrology consultation was sought and patient was planned for urgent dialysis. However the patient developed hypotension refractory to inotropes and his sensorium worsened; he sustained cardiac arrest from which he could not be revived.

**Treating unit’s diagnosis**

ADPKD, chronic renal failure (CRF) with hyperkalemia and encephalopathy. Pyoperitoneum probably secondary to duodenal ulcer perforation. Cause of death: hyperkalemia

**Discussion**

**Clinical protocol**

The diagnosis of ADPKD is based on the Ravine criteria according to age:¹ <30 years: 2 cysts in 1 or 2 kidney; 30-59 years: 2 cysts in each kidney; >60 years: 4 cysts in each kidney. Presence of hepatic cysts adds to the diagnosis.

Once renal dysfunction sets in, a majority of cases progress to end-stage renal disease (ESRD); 50% would develop ESRD by 60 years of age. Factors predicting rapid progression to ESRD are i) male sex, ii) age, iii) large kidney size, iv) hypertension, v) albuminuria and vi) PKD-1 phenotype. Common causes for development of acute renal failure in such a setting are obstruction of the ureter either by a clot, an impacted stone or compression by a cyst in the renal hilum. Ultrasonography had not revealed any unusual dilatation of the ureter in this case.

The incidence of liver cyst in ADPKD increases proportionately, and by 50 years co-existing cysts in liver and kidney are observed in 80% of patients. Liver dysfunction is rare in polycystic liver disease.

The ascites could be multi-factorial. It could be the effect of volume overload in CRF. Although hepatic venous outflow obstruction needs to be considered, ultrasonography had revealed normal portal vein and inferior vena cava. The patient possibly developed bacterial infection following the peritoneal tapping done 7 days back. The absence of fever, abdominal pain or tenderness and leukocytosis could be ex-
plained on an immune-compromised state.

Colonic diverticular disease has been reported in ~75% of ADPKD patients with CRF. Infection of these diverticuli could lead to perforation peritonitis, and the incidence increases with age. Spontaneous rupture of superficially located cysts following peritoneal dialysis is a well-known phenomenon. There could have also been cyst hemorrhage and rupture, which would be associated with hemorrhagic ascitic fluid, hematuria and loin pain.

**Final clinical diagnosis**
- ADPKD with liver cysts
- acute-on-chronic renal failure, volume overload, hyperkalemia, encephalopathy
- secondary peritonitis following duodenal ulcer perforation
- sepsis

**Cause of death**
Hyperkalemia leading to cardiac arrhythmia

**Pathology protocol**

At autopsy, there was gross abdominal distension. The peritoneal cavity contained 1500 mL of purulent fluid. Post-mortem culture of the fluid grew staphylococci and acinetobacter. Serosa along the full length of the intestine was dull with fibrous tags. Section from the intestine revealed fibrinous serositis, indicating an acute inflammatory process. Both small and large intestine had essentially normal histology. Duodenum was normal on gross and microscopic examination. Stomach showed histological evidence of mild superficial gastritis, no ulcer or erosion. Esophagus was grossly discoloured and histology showed diffuse erosive esophagitis.

Liver was enlarged and weighed 1700 g. Its capsule was covered by fibrinous exudates, more so on the inferior aspect. There were many cysts, almost completely replacing the whole right lobe and partly the left lobe, and extending to the hilar region (Fig 1). Some of the cysts on the inferior aspect were superficially located and a few appeared to be collapsed. The cysts size ranged from 5-40 mm. They had smooth and shiny wall; some of the superficially located cysts had transluscent wall. Most cysts contained clear or brownish fluid. Though the bile duct, portal vein and hepatic artery could be traced out, they appeared distorted and collapsed with gross distortion of the anatomy due to compression by the cysts located at the hilum.

On histology, the cysts showed lining by flattened to tall columnar biliary-like epithelial cells. Some of the cysts were filled with eosinophilic granular material. Most cysts were placed back-to-back to each other with no visible liver parenchyma in between, but wherever there was liver parenchyma it appeared compressed, resulting in hepatocyte atrophy. Portal tracts entrapped in between the cysts showed complete disorganization of architecture and many portal tracts contained clusters of dilated and immature looking bile ducts (von Meyernberg complexes); some were filled with bile concretion (Fig 2). Many of the portal tracts showed porto-portal bridging fibrosis and bile duct proliferation. The hilar structures had normal histology. Fibrinous exudate was seen over the inferior surface of the liver capsule, with gram-positive bacterial colonies. The inferior vena cava was patent.

Kidneys weighed 690 g together. They were grossly enlarged and cystic in appearance (Fig 3). The outer surface was studded with multiple cysts;
some contained translucent and clear fluid, whereas some had discoloured and hemorrhagic fluid. Cut sections of the kidneys showed near-total replacement of the renal parenchyma by the cysts, with no distinctive landmark for cortical or medullary region. The cysts varied from a few mm to 30 mm in size. They had smooth and shining cyst wall. Little renal parenchyma could be identified between the cysts. The pelvic calyceal system was not well defined though ureters could be seen arising from the renal hilum. The right ureter was dilated at the middle portion and filled with blood clot.

Histology revealed the cysts with lining of various types of epithelial cells, i.e., single layer of flattened cells to cuboidal to columnar cells with micropapillary projections into the cystic cavity. Some of the cysts contain eosinophilic hyaline material. The residual renal parenchyma between the cysts showed preserved glomeruli, tubules and blood vessels. Some of the tubules were atrophic with patchy interstitial fibrosis. Muscular blood vessels showed reduplication of internal elastic lamina, mild medial hypertrophy and intimal plaque formation.

Heart was enlarged and weighed 300 g. Aortic valve was unicuspid unicommissural type (Fig 4). The valve cusps were retracted, exposing the coronary ostia. The unicommissure was placed to the left of the left coronary valve cusp. Opposite to this, there was a highly placed raphe forming a dome-like appearance about 3 mm wide. The two coronary artery ostia were placed on either side of the commissure. The valve cusp appeared irregularly thickened, nodular and calcified. Both the coronary ostia were normally placed within the coronary sinuses, with normal caliber. The left ventricles showed concentric hypertrophy with focal thinning along the anterior wall. The left ventricular cavity was dilated, with subendocardial discoloration. Left ventricular anterior wall and anterior papillary muscle showed thinning, with organized mural thrombi underlying the anterior wall of the left and right ventricles. The left outflow tract showed endocardial sclerosis. All the three coronary arteries showed atheromatous plaque, and the left anterior descending artery was occluded by an organized thrombus. Histology confirmed the thinned out and discolored anterior wall of the left ventricle to be an area of old infarct, replaced by fibrosis.

Spleen was of normal size, weighing 80 g. Sections showed inflammatory cell exudate over the capsule with gram-positive organisms. Lungs bilaterally showed features of diffuse alveolar damage and patchy bronchopneumonia. Fibrin thrombi were seen within the small muscular pulmonary artery. Rest of the organs were essentially normal. No cyst was found in any other organ. The major blood vessels showed grade II atherosclerosis; inferior vena cava and other major veins were patent.

Clinical association between ADPKD and mitral valve prolapse has been frequently reported in literature.\textsuperscript{2,3} Association with aortic valve anomaly is rare.
Enterothorax mimicking pleural effusion

A 44-year-old woman presented with acute dyspnea since seven days. She had a history of Barrett carcinoma (pT1, pM0, R0) and subsequent resection of the distal esophagus 15 months earlier. Thyroidectomy had been done for Graves' disease four years earlier. Clinical examination at admission was unremarkable except for reduced breath sounds at the left basal thorax. Blood count and biochemistry indicated hypochromic anemia and latent hypothyroidism. Chest X-ray showed a shadow at the left base suggesting pleural effusion. Upper GI endoscopy under fluoroscopic guidance revealed an enterothorax, besides marked reflux esophagitis (Fig). The patient underwent immediate surgery and the small bowel was repositioned. This case demonstrates, as is well known, that enterothorax, most likely due to the prior esophageal surgery in this patient, can mimic pleural effusion on chest X-ray. As an alternative to computed tomography, endoscopy combined with radioscopy might be helpful in establishing the diagnosis.

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Fig: Endoscopic and radioscopic examination of upper gastrointestinal tract. Panel A and B show position of endoscope near gastric outlet. Arrow marks diaphragm. Panels C and D: Endoscope is positioned in jejunum and thoracic projection of endoscope is documented by fluoroscopy.

References


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The case was discussed in the Student CPC forum of the institute. Ramesh Chilal Kashinath, Junior Resident, Internal Medicine, presented and discussed the clinical protocol; Saugata Banerjee, Junior Resident, Pathology, presented and discussed the pathology protocol. The case was compiled and edited by Kim Vaiphei and Surinder Rana

Final autopsy diagnosis
- Autosomal dominant polycystic kidney disease with liver cysts
- Bicuspid aortic valve with three-vessel disease and old myocardial infarction of the anterior wall
- Diffuse alveolar damage with bilateral bronchopneumonia
- Diffuse esophageal ulceration
- Ascites with secondary peritonitis

Clinico-pathologic conference

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