Bilateral emphysematous pyelonephritis in autosomal dominant polycystic kidney disease

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ABSTRACT

Emphysematous pyelonephritis (EPN) is an uncommon infection of renal parenchyma with gas-forming organisms. EPN is usually seen in patients with diabetes mellitus (DM). The association of EPN with conditions other than DM is extremely rare. We report a case of autosomal dominant polycystic kidney disease with bilateral EPN who had to undergo bilateral nephrectomy.

Key words: Autosomal dominant polycystic kidney disease, emphysematous pyelonephritis, nephrectomy

Introduction

Emphysematous pyelonephritis (EPN) is a rare, severe gas-forming infection of renal parenchyma and its surrounding areas with life-threatening potential if left untreated. The EPN has no diagnostic clinical features and gas could be demonstrated only in minority on plain abdominal radiographs. The computed tomography (CT) scan is an important investigation; it can not only confirm the diagnosis but also show the extent of disease and help in planning the treatment and assessment of the response.

Case Report

A 43-year-old man, who is known to have autosomal dominant polycystic kidney disease (ADPKD), systemic hypertension, and chronic kidney disease (on conservative management) for the last five years, was admitted in our Institute with symptoms of bilateral flank pain, fever, and macrohematuria of two days duration. He had a progressive worsening of renal functions over five years.

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On examination, he was conscious, oriented, pale, toxic, and febrile (Temperature: 103 degree F). Pulse rate was 120/minute and regular, Blood pressure was 130/70 mmHg, and respiratory rate was 26/minute. He had bilateral enlarged kidneys, which were bimanually palpable with irregular surface and associated with tenderness on both sides.

Investigations: Urine showed bacteriuria and pyuria, and urine culture grew *Escherichia coli* (100,000 cfu/ ml). He had anemia (Hb: 8.2 g/dl), neutrophilic leukocytosis (15 000 cells/cu.mm with 85% neutrophils), normal blood sugars, and severe renal failure (S. creatinine: 10.4 mg/dl; B. urea: 277 mg/dl). Ultrasonogram of abdomen showed bilateral enlarged kidneys with numerous cysts, with some cysts having internal echos within. Multi detector computed tomography (MDCT) of abdomen (plain + contrast) confirmed bilaterally enlarged kidneys with numerous cysts and also showed multiple cystic lesions scattered in both lobes of liver. Both kidneys had multiple hemorrhagic cysts with a few showing multiple air pockets bilaterally [Figure 1a and 1b]. Other intra-abdominal organs were normal on ultrasonogram as well as MDCT. The diagnosis of ADPKD with bilateral emphysematous pyelonephritis (EPN) class 4[1] with severe renal failure was made.

Hemodialysis was initiated through a left radiocephalic areriovenous fistula (AVF) which had been created earlier. He was started on sensitive antibiotics as per urine culture report (Piparacillin-Tazobactam). As he continued to have high-grade fever after 7 days of Piparacillin-Tazobactam, this antibiotic was stopped and was started on injection Meropenem in appropriate doses.

The MDCT of abdomen repeated after 2 weeks showed persistence of with air pockets in both kidneys. Patient continued to have fever and loin pain. Gastrosurgery and Urology units evaluated him and bilateral nephrectomy was advised as he did not respond to an appropriate antibiotic course. He underwent bilateral nephrectomy. Forty-eight hours after bilateral nephrectomy, he became afebrile and macrohematuria subsided. At present, he is being continued on thrice weekly hemodialysis and is planned for renal transplantation at a later date.

Macroscopic examination of kidneys showed multiple cystic spaces with necrotic areas [Figure 2a and 2b]. On light microscopy, the cysts were lined by flat to cuboidal epithelium. Intercystic spaces had periglomerular fibrosis and glomerulosclerosis. Extensive necrosis with predominantly neutrophilic infiltrates was seen in the interstitium. Tubules showed dilatation and thyroidization at places. Renal pelvis showed areas of necrosis and inflammation. There was no evidence of granulomas, dysplasia, or malignancy. The above features were consistent with diagnosis of ADPKD with pyelonephritis.

Discussion

EPN has been defined as a necrotizing infection of the renal parenchyma and its surrounding areas that results in the presence of gas in the renal parenchyma, collecting system, or perinephric tissue. EPN occurs almost exclusively in patients with diabetes mellitus (DM), but occasionally, it is seen in those without DM and in patients with obstruction to pelvicalyceal system.[1-5] Kelly and MacCallum reported the first case of gas-forming renal infection (pneumaturia) in 1898.[6] EPN has been classified according to the location of gas accumulation on computed tomography (CT) imaging; Class 1: Gas in the collecting system only (emphysematous pyelitis); Class 2: Gas in the renal parenchyma without extension to the

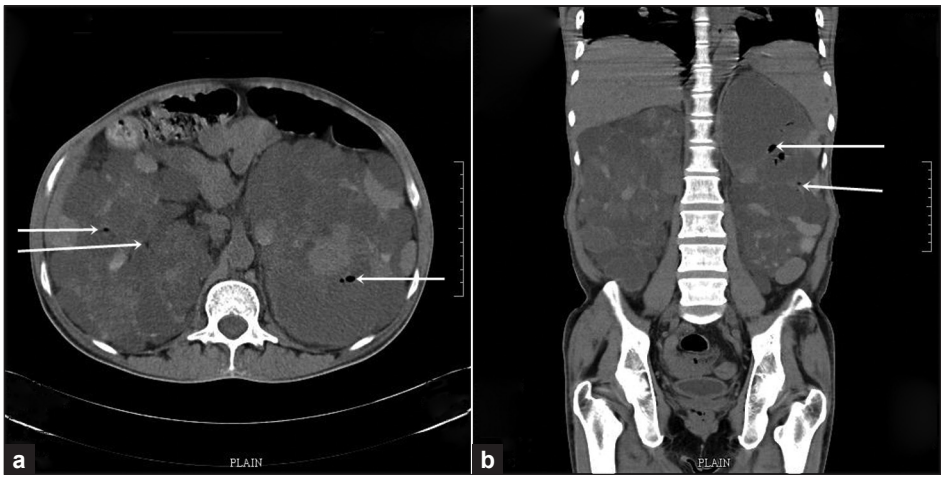

Figure 1: MDCT of abdomen showing bilaterally enlarged kidneys with numerous cysts. Both kidneys had multiple hemorrhagic cysts with a few showing multiple air pockets (arrows) bilaterally [Figure 1a - transverse and 1b - coronal view]

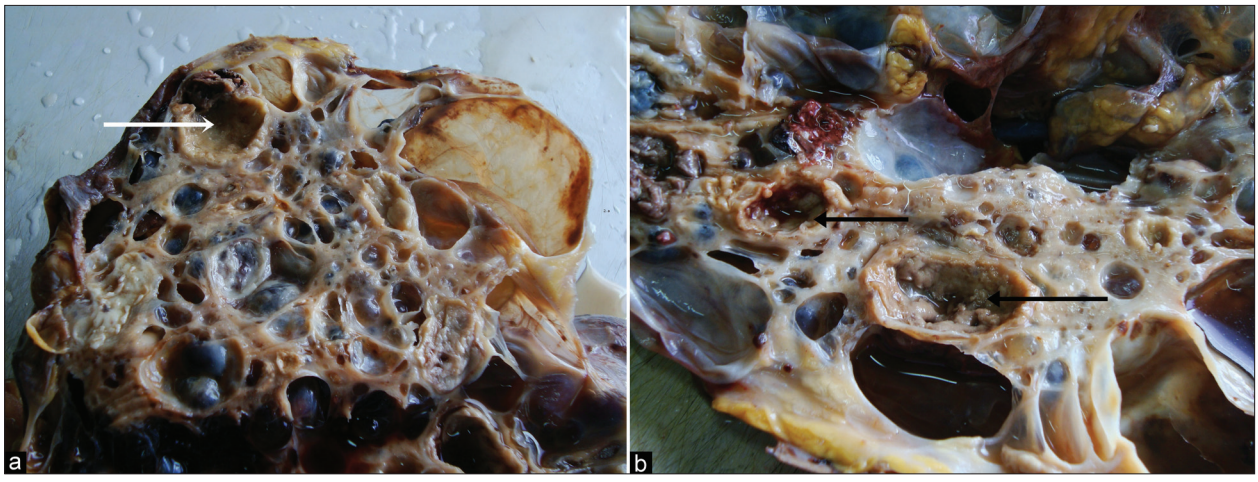

Figure 2: Gross specimen of kidneys [Figure 2a - left kidney and 2b - right kidney] showing enlarged kidneys with numerous cysts (multiple cysts showing hemorrhage). Both kidneys also show infected cysts with necrotic tissue in the cyst wall (arrows)

extrarenal space; Class 3A: Extension of gas or abscess to the perinephric space; Class 3B: Extension of gas or abscess to the pararenal space; and Class 4: Bilateral EPN or solitary kidney with EPN.[1]

The mechanisms postulated for the gas formation include fermentation of glucose through the glycolytic pathway (most Enterobacteriaceae, e.g., *E. coli, K. pneumoniae*, and *Proteus*) and butyric fermentation (*Clostridium*); these processes give rise to H_2 as an end product.[1,6] Huang and Tseng have postulated four factors that may be involved in the pathogenesis of EPN, including gas-forming bacteria, high tissue glucose level, impaired tissue perfusion, and a defective immune response in DM.[1]

The most common clinical manifestations of EPN are not different from the usual classic triad (i.e., fever, flank pain, and pyuria) of symptoms of upper urinary tract infection (UTI). However, thrombocytopenia (46%), acute renal failure (35%), disturbance of consciousness (19%), and shock (29%) can be the initial presentation, especially in severe cases or in patients who have not had an early diagnosis and management of EPN.[1-5,7] The diagnosis of EPN is made by demonstrating gas in renal or perinephric tissue by plain x-ray (seen only on 33%), renal ultrasonography, or by MDCT of abdomen. The MDCT of abdomen is helpful to confirm the diagnosis and to assess the extent of disease as well.[1,7,8]

Ahlering *et al*., Pontin *et al*., and Shokeir *et al*. had concluded that vigorous resuscitation and appropriate medical treatment should be attempted, but nephrectomy should be considered, if indicated, for successful management of EPN.[9-11] In a case series (one of the largest) published by Huang and Tseng, the mortality rate in patients who received antibiotic treatment alone was 40%.[1] All patients of classes 1 and 2 EPN were treated using a percutaneous catheter drainage (PCD) or ureteral catheter combined with antibiotic treatment survived.[1] The success rate of management by PCD combined with antibiotic treatment was 66%.[1] Patients with more severe EPN (classes 3 and 4), with fewer than two risk factors (thrombocytopenia, acute renal function impairment, disturbance of consciousness, or shock) were successfully treated using PCD combined with antibiotic treatment. The patients with two or more risk factors had a significantly higher treatment failure rate (92% *vs* 15%, $P<0.001$).[1] Of those who had an unsuccessful treatment using a PCD along with antibiotics and underwent subsequent nephrectomy, 90% survived. Our patient is one the very few cases (six) of ADPKD with EPN to be reported so far.[2-5,7,8] Of the six cases available online (Pubmed and Google), three patients responded to conservative management,[2-4] one required open drainage of infected cysts,[5] and two[7,12] needed unilateral nephrectomy. To the best of our knowledge, this is the first case of ADPKD with bilateral EPN to be reported which required bilateral nephrectomy, as he did not respond to sensitive antibiotics given for an appropriate duration.

Conclusions

EPN is a rare, gas-forming infection of renal parenchyma and its surrounding areas. EPN is most commonly seen in patients with DM and common organisms implicated are *E. coli* or *K. pneumoniae*. EPN is an extremely rare complication in nondiabetics. Prompt diagnosis and appropriate medical treatment with or without PCD should be attempted in all cases, but nephrectomy should be done for successful management of EPN if response to conservative measures is inadequate. Bilateral nephrectomy was a life-saving treatment for our patient who had chronic kidney disease due to ADPKD.

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