Bilateral Emphysematous Pyelonephritis and Emphysematous Cystitis with Autosomal-Dominant Polycystic Kidney Disease: Is Conservative Management Justified?

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Introduction

Emphysematous pyelonephritis (EPN) is a rare, necrotizing form of acute renal infection that results in the presence of gas in the renal parenchyma, the perinephric tissue, or collecting system [1]. Almost 60–70% of cases are associated with uncontrolled diabetes mellitus, with or without obstructive uropathy and superimposed infection with gas forming organisms [2]. Facultative anaerobes that are able to ferment glucose to lactate and CO₂ such as Escherichia coli, Proteus, Pseudomonas, Klebsiella, Aerobacter are possible causative organisms, with E. coli being the commonest [2–4]. We report a case of bilateral EPN and emphysematous cystitis occurring with autosomal-dominant polycystic kidney disease (ADPKD).

Case Report

A 64-year-old male, a known case of ADPKD and renal failure, presented with worsening of renal function and fluid overload. He had a history of insulin-dependent diabetes mellitus, hypertension, ischemic heart disease, and multiple cerebrovascular accidents, and had previously undergone coronary artery bypass grafting.

Physical examination revealed an ill-appearing man with normal consciousness, vital signs, and blood pressure. Auscultation
of the chest revealed bilateral crepitations. Examination of the abdomen revealed bilateral palpable kidneys, which were not tender.

Laboratory evaluation showed normal white cell and platelet count, hemoglobin 78 g/l (normal 130–170), serum creatinine 522 μmol/l (normal 62–115), blood glucose 31 mmol/l (normal 3.9–6.1) and marked pyuria. Ultrasonography showed bilateral polycystic kidneys and no perinephric collection. Urine was sampled for bacterial culture and management was instituted with control of blood glucose, maintenance of fluid balance with dialysis, and empirical antibiotics (cefotaxime). Urine culture showed no growth and following dialysis transient hematuria was observed. Seven days after admission he developed fever, turbid urine, neutrophilic leukocytosis, and elevated blood glucose. He still had no loin pain or tenderness and was hemodynamically stable. Computerized tomography (CT scan) showed bilateral polycystic kidney with multiple pockets of air in both renal parenchyma and collecting systems (fig. 1); there was no air in the perinephric region and the urinary bladder wall and lumen showed air streaks. In view of his hemodynamic stability, absence of loin tenderness, and multiple medical risk factors, it was decided to continue conservative management with antibiotics. Repeated urine culture grew *E. coli* and *Staphylococcus aureus*, and antibiotics were changed to tazocin and vancomycin according to the sensitivity test. He showed good clinical response 4 days later, when his urine became clear and serum creatinine was stable at 430 μmol/l without dialysis. Fever subsided after 1 week and repeated CT scan showed marked reduction in the air in the right kidney and complete resolution of the air in the left kidney and bladder. Antibiotics were continued and CT scan was repeated after 4 weeks, which showed complete resolution of the air in both the kidneys, and in the bladder (fig. 2). He was discharged 42 days after admission on oral antibiotics and was doing well at 3 months’ follow-up.

**Discussion**

EPN occurring with ADPKD is very rare with only 4 cases being previously reported [5–8]. Although concurrent EPN and emphysematous cystitis has been reported before [9], to our knowledge, this is the first case to document bilateral EPN and emphysematous cystitis occurring with ADPKD. The etiology of EPN in ADPKD is not clear. It has been suggested that the preexisting damaged
renal tissue and impaired vascular supply might be a precipitating factor, as certain bacteria can use the necrotic tissue as a substrate for gas formation [6]. The hyperglycemia in septic diabetic patients and impeded host response favors the bacterial overgrowth [3]. Ureteral instrumentation leading to seeding of bacteria into cysts has also been reported to be the cause in 1 case [5].

Mortality reportedly occurs in 20% of unilateral EPN and 50% of bilateral EPN, so appropriate management is essential [3]. Conservative medical treatment was preferred in early reports of EPN [1]. Later series showed a better survival with early aggressive surgical treatment (nephrectomy) in comparison to medical treatment (75 vs. 29%) [5] and decreased antibiotic perfusion was suggested to be the cause of failure of medical treatment [2]. Prompt diagnosis and surgical intervention was therefore suggested to be the most effective way of managing EPN [3, 5]. However, in patients with bilateral disease and in patients with associated medical risk factors the morbidity associated with this approach was higher. An alternative medical approach with broad-spectrum antibiotics, maintenance of fluid balance, control of blood glucose and intensive circulatory support was suggested [2, 4]. Factors stated to be essential for success of medical management include early diagnosis, no obstructive uropathy, and adequate perfusion visualized on renal flow scan. Persistence of localized gas within the renal parenchyma and/or the pyelocalyceal system is not per se a discouraging sign [1]. Huang and Tseng [10] suggested antibiotic treatment combined with percutaneous catheter drainage, wherever necessary, in patients with localized EPN and in patients with extensive EPN with a more benign manifestation (less than two of the following four risk factors including thrombocytopenia, acute renal function impairment, disturbances of consciousness, and shock).

Treatment in the 4 reported cases of EPN with ADPKD included bilateral nephrectomy in 1, unilateral nephrectomy in 1, conservative management in 1, and not defined in 1 [5–8]. Our patient was managed conservatively despite extensive disease and outcome was good, probably because of early diagnosis, presence of only one risk factor, and appropriate antibiotic therapy. The type and duration of antibiotic therapy cannot be generalized due to limited experience. It appears that in the early phase of EPN, and when the patient is hemodynamically stable, conservative management can be successful. However, the patient should be closely monitored, both clinically and radiologically, and percutaneous catheter drainage or surgical intervention carried out whenever necessary.

**Conclusion**

This case demonstrates that conservative management along with close monitoring can be a successful modality in the treatment of EPN occurring with ADPKD despite extensive disease, and is an acceptable alternative to percutaneous catheter drainage or surgical intervention.

**References**