

3-7-2023

## Emphysematous pyelonephritis necessitating bilateral radical nephrectomy: A case report and literature review.

Christine E Wamsley

Arianna Morton

Christopher G Roth

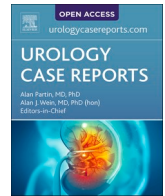
Joseph K Izes

Li Li

Follow this and additional works at: <https://jdc.jefferson.edu/tjucompmedctrfp>Part of the [Nephrology Commons](#), and the [Urology Commons](#)

### [Let us know how access to this document benefits you](#)

This Article is brought to you for free and open access by the Jefferson Digital Commons. The Jefferson Digital Commons is a service of Thomas Jefferson University's [Center for Teaching and Learning \(CTL\)](#). The Commons is a showcase for Jefferson books and journals, peer-reviewed scholarly publications, unique historical collections from the University archives, and teaching tools. The Jefferson Digital Commons allows researchers and interested readers anywhere in the world to learn about and keep up to date with Jefferson scholarship. This article has been accepted for inclusion in Computational Medicine Center Faculty Papers by an authorized administrator of the Jefferson Digital Commons. For more information, please contact: [JeffersonDigitalCommons@jefferson.edu](mailto:JeffersonDigitalCommons@jefferson.edu).



Inflammation and infection

# Emphysematous pyelonephritis necessitating bilateral radical nephrectomy: A case report and literature review

Christine E. Wamsley<sup>a</sup>, Arianna Morton<sup>a</sup>, Christopher G. Roth<sup>b</sup>, Joseph K. Izes<sup>c</sup>, Li Li<sup>a,\*</sup><sup>a</sup> Department of Pathology and Genomic Medicine, Thomas Jefferson University Hospital, Philadelphia, PA, United States<sup>b</sup> Department of Radiology, Thomas Jefferson University Hospital, Philadelphia, PA, United States<sup>c</sup> Department of Urology, Thomas Jefferson University Hospital, Philadelphia, PA, United States

## ARTICLE INFO

## Keywords:

Emphysematous pyelonephritis  
Diabetes mellitus  
*Escherichia coli*  
Radical nephrectomy

## ABSTRACT

Emphysematous pyelonephritis (EPN) is a severe, acute necrotizing infection of the kidney with gas accumulation in the renal parenchyma, collecting ducts, and/or perirenal tissue. We report a case of EPN in a 66-year-old male with uncontrolled diabetes mellitus (DM), urinary tract infection and septic shock necessitating emergent bilateral radical nephrectomy. Morphologically, the kidney parenchyma revealed severe acute bacterial pyelonephritis with extensive abscesses, gas cysts, necrosis and foci of infarctions. The uninvolved background cortex parenchyma exhibited diabetic nephropathy (DN), Class IIB. Post-operatively, the patient remained dialysis-dependent with multiple re-admissions for recurrent multi-drug resistant *E. coli* pyocystitis, anemia and urinary retention.

## 1. Introduction

Emphysematous pyelonephritis (EPN) is a rare, potentially life-threatening condition caused by necrotizing inflammation of the renal parenchyma, collecting duct system and perirenal tissues.<sup>1</sup> While most commonly associated with uncontrolled diabetes mellitus (DM), cases have also been reported in patients with urinary tract abnormalities and immunocompromised states.<sup>2</sup> The gram-negative, facultative anaerobic microorganism *Escherichia coli* (*E. coli*) remains the most-often implicated pathogen.<sup>1</sup> The most common presenting symptoms of fever, flank pain and pyuria are non-specific and may mimic those of acute pyelonephritis.<sup>2</sup> Therefore, high clinical suspicion and appropriate imaging must be utilized to accurately diagnose this condition, which is associated with high morbidity and mortality if left untreated.<sup>1,3</sup> Here, we report a case of EPN in a male patient with underlying uncontrolled DM and urinary tract obstruction.

## 2. Case presentation

A 66-year-old man with DM and benign prostatic hyperplasia presented with six weeks of fatigue and three days of suprapubic tenderness. Urinalysis showed protein 1+, glucose 3+ and blood 3+, but no nitrites or leukocyte esterase. Ultrasound demonstrated moderate

hydronephrosis of the right kidney and urinary retention. Foley catheter yielded 2.3 L of urine, with improved symptoms. The patient returned one week later with lethargy, hyperglycemia (blood glucose 840 mg/dL) and continued dysuria. Urine and blood cultures were positive for multidrug-resistant *E. coli*. His hemoglobin A1c was 13.6%. Computed tomography (CT) showed extensive gas within the bilateral renal parenchyma and the left renal sinus (Fig. 1A). Secondary signs of inflammation were also observed, including perinephric fat stranding and thickening of Gerota's fascia (Fig. 1B). The patient rapidly developed septic shock, and a bilateral radical nephrectomy was performed. Post-operatively, the patient remains dialysis-dependent and has been readmitted on several occasions for recurrent multi-drug resistant *E. coli* (MDR *E. coli*) pyocystitis, anemia and persistent urinary retention, requiring intravenous antibiotics and red blood cell transfusions with iron supplementation. Suprapubic catheter placement allowed for drainage of the patient's defunctionalized bladder.

Pathology: On gross examination, the left kidney weighed 1,063g and measured 24.2 × 13.0 × 8.4cm; the right kidney weighed 666g and measured 21.1 × 11.0 × 6.7cm. The cut surfaces of the bilateral renal parenchyma were tan-brown and congested, with hemorrhage and necrosis comprising 50% of the right kidney and 30% of the left kidney parenchyma. Air spaces were identified in the subcapsular and perinephric spaces of both kidneys.

\* Corresponding author. Thomas Jefferson University Hospital, 132 South 10th Street, Main Building, Room 285R, Philadelphia, PA, 19107, United States.

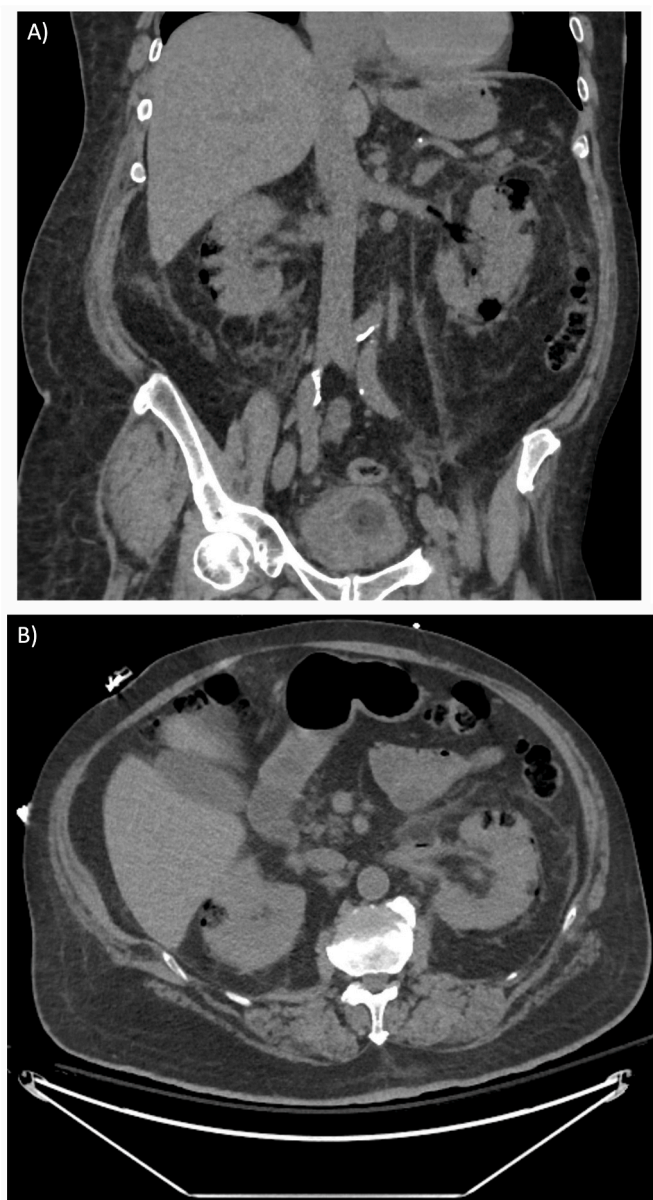
E-mail address: [li.li@jefferson.edu](mailto:li.li@jefferson.edu) (L. Li).

<https://doi.org/10.1016/j.eucr.2023.102376>

Received 1 February 2023; Accepted 2 March 2023

Available online 7 March 2023

2214-4420/© 2023 Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



**Fig. 1.** Unenhanced CT images of emphysematous pyelonephritis. A. The coronally reformatted CT images demonstrated extensive gas within the renal parenchyma bilaterally (arrows) associated with bilateral perinephric fat stranding and gas within the left renal sinus (thick arrow). A Foley catheter was noted within the bladder (dashed arrow), with mild associated perivesical fat stranding. The mural thickening was not well visualized due to the lack of distention and absence of intravenous contrast. B. The axial CT image showed gas within the periphery of the renal parenchyma bilaterally (arrows) and in the left renal pelvis (thick arrow), along with perinephric fat stranding and thickening of Gerota's fascia (dashed arrow), secondary signs of inflammation.

Microscopic examination demonstrated multifocal hemorrhage, acute pyelonephritis with abscesses and multiple foci of infarction involving the renal pelvicalyceal system, extending to the kidney cortex and perirenal fat (Fig. 2A). Abscesses were filled with abundant neutrophils, necrotic tissue and bacterial colonies. Numerous gas-filled cysts formed within the abscesses and adjacent necrotic kidney tissue (Fig. 2B and C). A large artery also showed subendothelial inflammatory infiltrate, consistent with vasculitis (Fig. 2D). A diagnosis of bilateral emphysematous pyelonephritis (EPN) was made.

Of note, the non-affected kidney cortex parenchyma exhibited

features of diabetic nephropathy, Renal Pathology Society (RPS) class IIB, showing diffuse thickening of the glomerular and tubular basement membranes and severe mesangial expansion without hypercellularity and nodular formation (Fig. 3A). Diffuse acute tubular injury/necrosis was seen, with loss of the proximal tubule brush border, simplified tubular epithelium and sloughed epithelial cells in the tubular lumen. There was mild to moderate intimal fibrosis of the arteries and thickening of the arteriolar walls (Fig. 3B).

### 3. Discussion

Up to 95% of EPN cases occur in patients with uncontrolled DM.<sup>1</sup> It is hypothesized that high levels of glucose impair blood supply, which, in combination with reduced host immunity, facilitate bacterial colonization and gas production.<sup>2,4,5</sup> Although DM is a common risk factor, this condition has not been associated with an increased risk of mortality.<sup>1,3,5</sup>

There is a greater incidence of EPN in women (female:male ratio = 6:1), most likely attributed to an increased susceptibility to urinary tract infections.<sup>1</sup> *E. coli* remains the most common causative pathogen, identified in nearly 70% of reported cases.<sup>1,2</sup> *Proteus mirabilis*, *Klebsiella pneumoniae*, Group D *Streptococcus* and coagulase-negative *Staphylococcus* infection have also been implicated.<sup>2</sup> These gas-forming microorganisms ferment glucose and lactose, leading to high levels of carbon dioxide and hydrogen, as well as nitrogen, oxygen and trace amounts of ammonia and methane.<sup>2</sup> This gas may spread beyond the site of inflammation into the perirenal space, as seen in this patient. In some cases, gas can extend all the way into the scrotal sac and spermatic cord.<sup>2</sup> The risk of developing EPN secondary to a urinary tract obstruction is 25–40%.<sup>2</sup> Bilateral EPN has been reported in 0%–38.5% of cases.<sup>3</sup>

CT imaging is the gold standard for diagnosis, confirming the presence of gas (Fig. 1).<sup>1,4</sup> Once diagnosed, patients with EPN must be aggressively and immediately treated due to potential life-threatening associated complications, including kidney failure, liver function deterioration, disseminated intravascular coagulation and septic shock.<sup>1,2,4</sup> Options include antibiotics, percutaneous drainage or nephrectomy.<sup>1,4</sup> Emergency nephrectomy may be a salvage procedure or can be performed immediately if warranted, such as in patients with several risk factors, severe clinical deterioration or poor prognosis.<sup>4</sup>

In our patient's case, bilateral emergent nephrectomy was a dramatic intervention, and the decision was made in concert with the intensive care unit, interventional radiology, the hospital's ethics team and the patient's family. The patient was critically ill, and infection source control could not be obtained by drainage; there was simply no fluid to drain. Although this decision was drastic and certainly non-standard, it was deemed lifesaving for this patient.

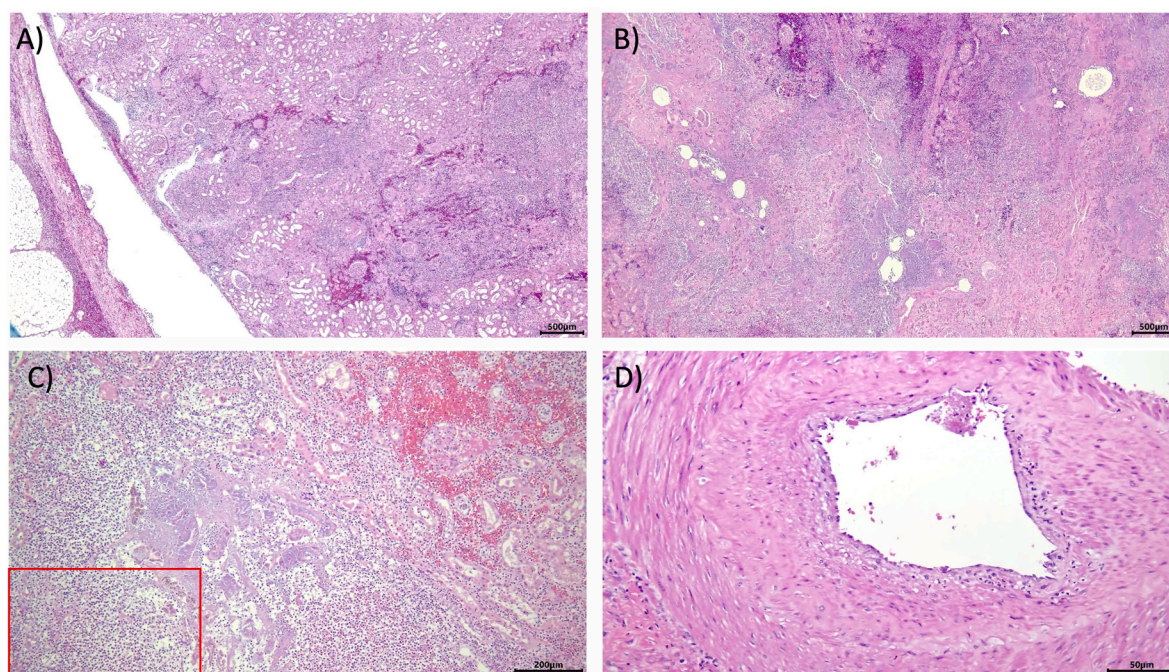
Mortality from EPN, which has declined from 78% in the 1970s to 21% as of 2018,<sup>1,3</sup> primarily arises from septic complications. Studies have shown that blood glucose level, age, sex and site of infection are not prognostic indicators.<sup>1,3</sup> Bilateral EPN, thrombocytopenia, acute renal impairment, altered consciousness and shock are associated with higher rates of mortality.<sup>3</sup>

### 4. Conclusion

Herein, we report a case of EPN necessitating emergent bilateral nephrectomy. Histologic analysis demonstrated characteristic features, including bacterial pyelonephritis with extensive abscesses, gas cysts, necrosis and foci of infarctions bilaterally. Despite rapid management, the patient has experienced several post-operative complications, including MDR *E. coli*, pyocystitis, anemia and persistent urinary retention. He remains dialysis-dependent.

### Author contributions

Christine E. Wamsley: Writing- Original Draft, Writing- Review &



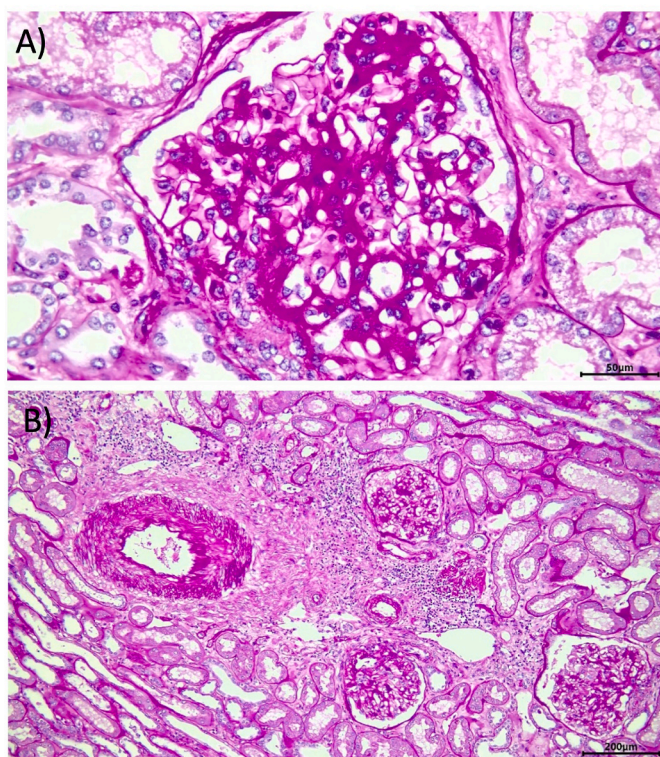
**Fig. 2.** Representative images from both kidneys.

A. Extensive abscesses filled with debris and air spaces surrounded by an intense neutrophilic inflammatory infiltrate were present in the kidney cortex extending into the perirenal fat.

B. Abundant neutrophils were present in the edematous interstitium, predominantly as polymorphonuclear (PMN) casts. Round and empty air space were present in the kidney parenchyma, with infarction and hemorrhage.

C. Bacterial colonies responsible for creating the air spaces and associated hemorrhage were identified in a focus of abscess. Bacterial colonies (inset).

D. An example of focal vasculitis showed neutrophils and mononuclear cell infiltration underneath the endothelial cells.



**Fig. 3.** Representative Periodic acid-Schiff (PAS)-stained images from the non-affected kidney parenchyma. Diffuse acute tubular injury was seen, with loss of proximal tubule brush border, simplified tubular epithelium, and sloughed epithelial cells in tubular lumen.

A. Several glomeruli exhibited diffuse thickening of the glomerular and tubular basement membranes and mesangial expansion without hypercellularity or nodular formation.

B. There was mild-to-moderate atherosclerosis and arteriosclerosis with mild-to-moderate intimal fibrosis and thickening of the arteriolar wall.

Editing, Visualization, Project Administration. Arianna Morton: Writing-Review & Editing, Visualization. Christopher G. Roth- Investigation,

Writing- Review & Editing, Visualization. Joseph K. Izes: Data Curation, Investigation, Writing- Review & Editing. Li Li: Conceptualization, Investigation, Writing- Review & Editing, Visualization, Supervision. All authors read and approved the final manuscript.

### Consent

The patient has consented to the submission of the case report to the journal.

### Data availability

All relevant data have been included in this manuscript.

### Funding statement

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

### Declaration of competing interest

Christine E. Wamsley reports compensation for consultation services provided to Innovative Optics (Sarasota, FL) for research studies outside of the submitted work. The remaining authors have no financial interests to disclose.

### References

1. Dutta P, Bhansali A, Singh SK, et al. Presentation and outcome of emphysematous renal tract disease in patients with diabetes mellitus. *Urol Int.* 2007;78(1):13–22. <https://doi.org/10.1159/000096929>.
2. Ubee SS, McGlynn L, Fordham M. Emphysematous pyelonephritis. *BJU Int.* 2011;107(9):1474–1478. <https://doi.org/10.1111/j.1464-410X.2010.09660.x>. May.
3. Falagas ME, Alexiou VG, Giannopoulos KP, Siempos II. Risk factors for mortality in patients with emphysematous pyelonephritis: a meta-analysis. *Sep J Urol.* 2007;178(3 Pt 1):880–885. <https://doi.org/10.1016/j.juro.2007.05.017>. quiz 1129.
4. Pontin AR, Barnes RD. Current management of emphysematous pyelonephritis. *Nat Rev Urol.* 2009;6(5):272–279. <https://doi.org/10.1038/nrurol.2009.51>. May.
5. Najafian B, Fogo AB, Lusco MA, Alpers CE. AJKD atlas of renal Pathology: diabetic nephropathy. *Am J Kidney Dis.* 2015;66(5):e37–e38. <https://doi.org/10.1053/j.ajkd.2015.08.010>. Nov.