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Nephroureterectomy for emphysematous pyelonephritis: An aggressive approach is sometimes necessary. A case report and literature review



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ABSTRACT

INTRODUCTION: Emphysematous pyelonephritis (EPN) is a life-threatening urological emergency. A high index of suspicion is required for diagnosis as such patients may present to physicians with typical features of pyelonephritis.

PRESENTATION OF CASE: A 67 year old lady presented atypically to the Emergency Department with symptoms of renal colic. The diagnosis of emphysematous pyelonephritis was established on prompt CT scanning. She did not respond to conservative management. Due to acute, critical deterioration, she underwent a radical right nephroureterectomy. The resected kidney involved a long segment of necrotic, gangrenous ureter. The patient had a smooth post-operative recovery and was successfully discharged. She remains well on follow-up after one year.

DISCUSSION: Early radiological diagnosis is imperative for risk stratification of EPN. Current evidence recommends percutaneous catheter drainage with interval nephrectomy as the gold standard treatment. We review the literature for pathophysiology and clinical prognostic factors. This case adds onto the limited evidence base on ureteric involvement in EPN, suggesting a revision of EPN classification.

CONCLUSION: Further research on ureteric involvement and treatment outcomes in EPN is required. Even in the current era of minimally invasive surgery and renal preservation therapies, early open nephrectomy still has a role in the management of EPN.

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1. Introduction

Emphysematous pyelonephritis (EPN) is an acute, severe, necrotising infection of the kidneys and collecting system. It is characterised by the presence of inter or intramural gas within the renal parenchyma, collecting system or perinephric tissue. The term was first used by Schultz and Klorfein in 1962. This condition occurs predominantly in elderly patients with poorly controlled diabetes mellitus. The high incidence of urinary tract infections in this population group, coupled with the entity's similar presentation to pyelonephritis may lead to delayed diagnosis and intervention if patients are managed entirely by non-surgical specialties.

This case highlights the importance of clinical vigilance, prompt radiological diagnosis and pathophysiology of EPN. We review the evidence on the various management modalities based on radiological classification.

Abbreviations: EPN, emphysematous pyelonephritis; CT, computed tomography; WCC, white cell count.

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Minimally invasive interventional therapy is emerging as the gold standard treatment for EPN. Radical nephroureterectomy is an operation reserved for malignant urological disease. This case adds to the limited evidence base on its use in the management of emphysematous pyelonephritis [1,2].

2. Presentation of case

A 67 year old lady presented to the emergency department, in the early hours of the morning, with sudden onset right loin pain. She described a 12 h history of constant colicky pain radiating to the right loin. This was associated with vomiting. She denied urinary symptoms or haematuria. Her past medical history included autoimmune liver cirrhosis, atrophic gastritis and Type 2 diabetes mellitus (T2DM). Previous operations included a hysterectomy and coronary artery bypass surgery 5 years before. Her drug history included metformin and non-anaphylactic reactions to penicillin and intravenous (IV) contrast.

On general examination, the patient was pale with a temperature of 36.8 °C. Initial vital signs were BP 141/72 with pulse of 81 beats per minute. Abdominal examination revealed generalised

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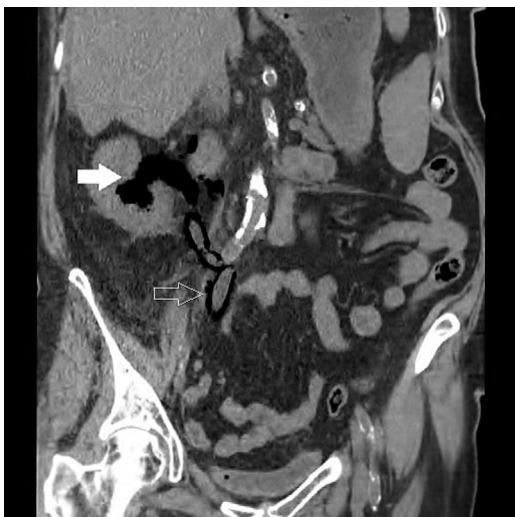


Fig. 1. EPN on CT (coronal view)—gas within the renal collecting system (thick arrow) and outlining the right ureter (thin arrow).

right sided tenderness, worse in the right upper quadrant and loin, with no crepitus.

Initial blood tests revealed anaemia (Hb 9.8 g/dL), impaired renal function (urea 13.2 mmol/L, creatinine 183 μ mol/L) and HbA1c of 5.3%. Liver function tests were normal. Prothrombin time and activated partial thromboplastin time were mildly elevated at 17.5 s and 37.4 s, respectively. The absence of a neutrophil leucocytosis (WCC 9×10^9 /L) and urinalysis displaying trace of blood prompted a Urology referral for renal colic. A non-contrast CT of the kidneys, ureters and bladder (CTKUB) to exclude urolithiasis was arranged. This showed gas outlining the pelvicalyceal system of the right kidney, with perifascial gas tracking along the right ureter down to its lower third (Fig. 1). There were no discrete fluid collections or

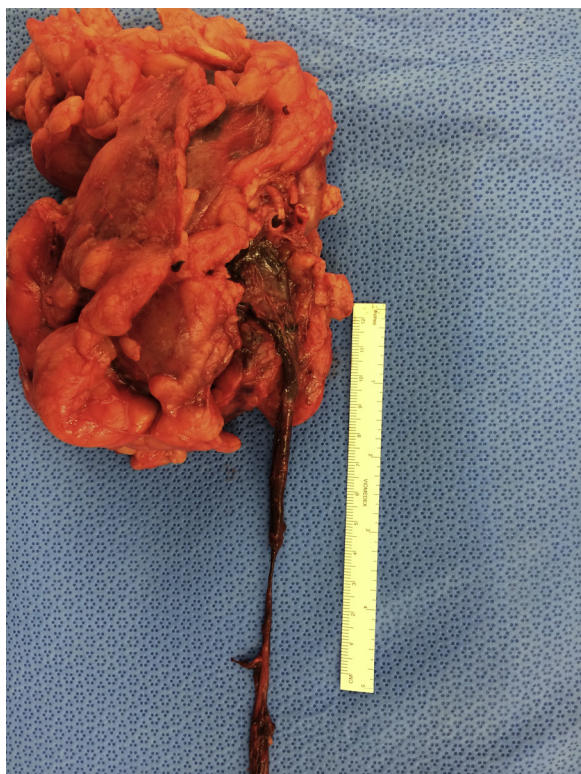


Fig. 2. Specimen: Infected kidney with necrotic ureter.

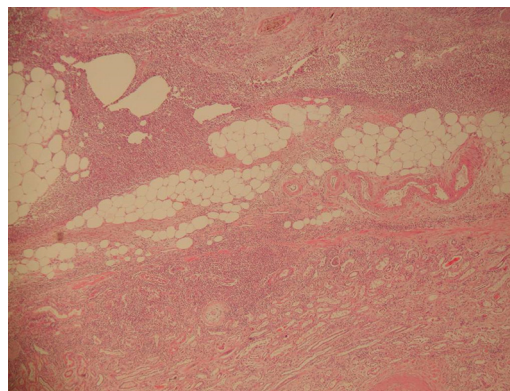


Fig. 3. Perinephric abscess.

calculi. Following blood and urine cultures, intravenous antibiotics and fluids were started.

A radiological diagnosis of emphysematous pyelonephritis was suggested. Potential differential diagnoses included retroperitoneal perforation of abdominal viscus, a psoas abscess with gas-forming bacteria and entero-pelvic or ureteric fistula.

Blood and urine specimens grew *Escherichia coli* sensitive to Meropenem. Microbiology advised the addition of Metronidazole for anaerobic cover in case of fistulous bowel contamination. The patient developed pyrexia, tachycardia, hypotension and oliguria despite copious intravenous fluid resuscitation. She was transferred to a high dependency unit (HDU). The C-reactive protein continued to rise from 8 to 109 mg/L, while she failed to mount an inflammatory response (leucocyte count of 6 g/dL). Her platelet count dropped from 198×10^9 /L to 100×10^9 /L. Her kidney function deteriorated further with creatinine rising from 183 μ mmol/L on admission to 258 μ mmol/L (baseline of 160 μ mmol/L).

There was evidence of falling platelet count, severe sepsis and end-organ failure despite optimal medical management and inotropic support on HDU. The consensus between urologists and the intensive care team was to proceed with emergency right nephroureterectomy to control sepsis.

An open approach through a right loin incision was used. The peritoneum was inadvertently opened. There was free peritoneal fluid due to cirrhosis of the liver but the abdominal viscera were otherwise normal. There was obvious necrosis of the right renal pelvis and ureter with oedema in the retroperitoneum. There was peri-ureteric gas but no perinephric abscess or collection. The ureteric necrosis extended down to the lower third of the right ureter just above the superior vesical pedicle. Following the right nephrectomy with ligation of the renal vessels, the kidney specimen together with the attached ureter was placed in a retroperitoneal space inferiorly, towards the pelvis. The main wound was closed and a second incision was made inferiorly to excise the diseased distal ureter that was removed en bloc with the kidney specimen (Fig. 2). A retroperitoneal drain and urethral catheter were left in situ and the patient was transferred to the intensive care unit following surgery.

Histopathology analysis of the specimens confirmed microscopic parenchymal abscesses within the kidney and perinephric fat (Fig. 3). There was haemorrhagic infarction of the ureteric mucosa with serositis (Fig. 4).

On post-operative day 4 the patient developed pyrexia. A CT of the abdomen and pelvis was performed. Besides small bilateral pleural effusions and normal post-operative changes, there were no significant retroperitoneal collections to explain the fever. The patient had a smooth recovery with 7 days of IV antibiotics. She was discharged 13 days after admission. At 2 weeks follow-up, there were no early complications. She had one admission within 3

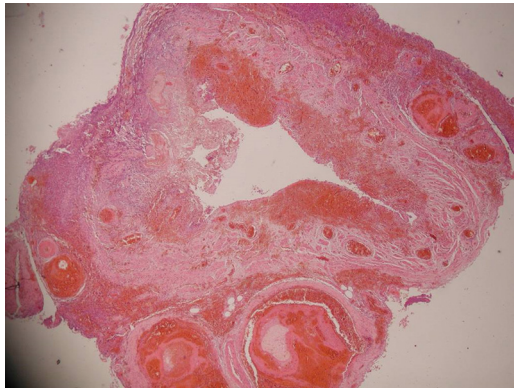


Fig. 4. Perinephric abscess.

Table 1
Classification of EPN by Huang and Tseng.

Class I	Gas in collecting system only
Class II	Parenchymal gas only
Class IIIa	Extension of gas into perinephric space
Class IIIb	Extension of gas into paranephric space
Class IV	EPN in solitary kidney or bilateral disease.

months of discharge with worsening renal function. This was managed conservatively and she remains under long term nephrology follow-up.

3. Discussion

Current evidence suggests a 3:1 female to male predominance for EPN. Poorly controlled DM, predisposing the patient to a defective immune response, is the single most common risk factor in 90% of cases. This is closely followed by urinary tract obstruction by urolithiasis [3]. High blood and tissue glucose levels, coupled with poor tissue perfusion in the diabetic kidney result in fermentation of glucose by facultative anaerobes, releasing carbondioxide. *E. coli*, *Klebsiella pneumonia* and *Proteus* spp. are the commonest micro-organisms grown in laboratory cultures [4].

This presentation of EPN is atypical given her good glycaemic control, the absence of pyrexia, crepitus and pyuria. However, renal angle tenderness was noted on clinical exam and this is the most common positive finding. In the presence of leucocytes and/or nitrites on urinalysis, this may be erroneously mismanaged as pyelonephritis delaying appropriate treatment. This illustrates the importance of a high index of suspicion in these cases.

The expedient use of CT as opposed to ultrasound scanning in this case was invaluable. CT has 100% sensitivity, is not operator dependent, can exclude other pathologies and it is useful to monitor response to treatment [5]. Two radiological classifications have been described, based on CT findings. The radiologist employed Wan et al's CT grading for EPN, a simple classification based on the absence or presence of perinephric fluid collections (Type 1 or 2, respectively), with prognostic implications [6]. An alternative is Huang et al's classification based on the anatomical distribution of gas within the renal parenchyma and perinephric tissues [7] (Table 1).

In their study, Huang et al. did not comment on ureteric involvement in the cohort managed by nephrectomy [7]. We can hypothesise that, in the absence of obstruction, ureteric necrosis would be linked to the same infective process responsible for gas formation within the renal parenchyma. The process may have been limited to the proximal ureter as the distal ureter has a better vascular supply. Scant literature exists regarding ureteric involvement in EPN and subsequent management, thus further laboratory

research and case series are needed to enhance our understanding. The degree of ureteric involvement prompted an open, as opposed to laparoscopic approach to deliver the specimens.

A recent meta-analysis quoted successful treatment rate of 90–100% with a combination of medical management and percutaneous catheter drainage [3] (PCD). This involved intravenous antibiotics, fluids and tight glycaemic control. CT-guided, percutaneous drainage tubes were placed in the retroperitoneum. A percutaneous nephrostomy may be inserted into the renal pelvis to relieve hydronephrosis. This nephron-sparing strategy is clearly beneficial if there are multiple fluid collections and abscesses [4]. In the meta-analysis, 20% of patients subsequently had elective nephrectomy after 3–6 weeks of PCD.

Transfer to a tertiary centre for percutaneous drainage was contemplated, however given the critical state and the absence of discrete fluid collections on CT, it was deemed of minimal benefit in this case. The decision to proceed to nephrectomy so early in the course of this patient's treatment was based on the presence of septic shock, rising creatinine, thrombocytopenia and Wan et al's Type 1 EPN. Along with disturbance of consciousness and bilateral EPN; these prognostic factors have been shown to be associated with poor outcomes [7]. Abourmazouk et al. found that over 54% of EPN patients with shock and end-organ failure died [5]. This group therefore warrant more aggressive management.

Finally, as the evidence base for ureteric involvement in EPN increases [1,2], further research to understand the pathophysiology and impact on survival is necessary.

4. Conclusion

Septic shock, rising creatinine, thrombocytopenia, Wan's Type 1 EPN, disturbance of consciousness and Bilateral EPN are indicators of poor prognosis in EPN. Emergency nephrectomy should be reserved for severe EPN. A revision of the established classification systems to include ureteric involvement may be required.

Conflicts of interest

None.

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None.

Ethical approval

Not applicable.

Author contribution

GRN and AB collected the data, reviewed the evidence and drafted the manuscript. WA provided clinical and critical analysis. HM and OK performed surgery and revised the article for intellectual content. All authors approve the final version of the article.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Gael R. Nana.

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