CASE REPORT

Emphysematous pyelonephritis: a case report and review of the literature

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We present a case of fulminant emphysematous pyelonephritis in a 44-year-old diabetic woman culminating in emergent nephrectomy. Current management strategies and their outcomes are reviewed.

Key Words: emphysematous pyelonephritis, necrotizing infections, nephrectomy

Introduction

Emphysematous pyelonephritis (EPN) is a rare, acute, life-threatening, necrotizing renal infection characterized by bacterial gas production within the renal parenchyma and surrounding perirenal tissues. Mortality rates have been reported as high as 70%. We report a case of emphysematous pyelonephritis in a diabetic woman who subsequently underwent an emergent nephrectomy.

Case report

A 44-year-old type II diabetic woman presented to her community hospital with a 3-day history of nausea, vomiting and fever. She denied urinary symptomatology, flank or abdominal pain initially. Her medical history was significant for a 1-year history of poorly controlled type II diabetes mellitus, asthma and depression. The initial clinical impression was that the source of her fever was respiratory and radiographically a left lower lobe pneumonia was suspected. She was treated with a third generation cephalosporin. Despite this, she deteriorated clinically developing left-sided abdominal pain and a fever of 40°C. Her blood glucose rose to 44 mmol/L. Her creatinine was 141 μmol/L and white blood cell count was 15.0 x 10^9/L. She was given fluid resuscitation and started on broad spectrum antibiotic treatment with intravenous gentamicin and metronidazole. Plain films of the abdomen demonstrated extensive gas accumulation, involving the entire left kidney, Figure 1. A contrast enhanced computed tomography scan of the abdomen and pelvis revealed gas in the left kidney and retroperitoneum to the level of the iliac bifurcation, consistent with a diagnosis of

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Figure 1. Preoperative plain film of the abdomen and pelvis demonstrating gas accumulation within and surrounding the left kidney.

emphysematous pyelonephritis (EPN), Figure 2. Only a small rim of non-enhancing cortex was seen. Intraluminal gas was seen in the left renal vein, Figure 3. No focal fluid collections, hydronephrosis or renal calculi were identified. The right kidney appeared normal. Urine and blood cultures grew Escherichia coli sensitive to all tested antimicrobials.

She was transferred to our tertiary care center, where initial evaluation revealed her to be slightly tachycardic (pulse 110) with a systolic blood pressure consistently greater than 100 mmHg. She had a temperature of 38°C. She was drowsy but otherwise had normal mentation. Laboratory investigations revealed a white blood cell count of 11.0 x 10^9/L, hemoglobin of 101 g/L and creatinine of 113 μmol/L. With the radiologic evidence of near complete destruction of the left kidney, emergent open nephrectomy was performed. Intraoperative observations included the findings of necrotic renal parenchyma and marked edema of the perinephric tissues. Intraluminal gas in the left renal vein was not appreciated upon surgical ligation. A closed suction drain was left. The patient tolerated the procedure well. She was monitored in the intensive care unit for 48 hours post-op and was discharged home 1 week following surgery. Her serum creatinine was 80 μmol/L at the time of discharge.

Figure 2. Preoperative contrast enhanced computerized tomography scan of the abdomen and pelvis revealing gas accumulation in the left kidney and retroperitoneum, as well as destruction of left renal parenchyma.

At pathologic examination, the kidney measured 13.0 cm x 7.0 cm x 5.0 cm, and on gross inspection was coated with purulent fibrinous adhesions. The majority of the parenchyma was necrotic with little preservation of normal renal architecture. There was disruption of the corticomedullary interface and papillary necrosis. Microscopically, acute and chronic inflammatory cells extending from the proximal portion of the ureter through the renal parenchyma, cortical surface and surrounding adipose tissue were detected.

Figure 3. Preoperative contrast enhanced computerized tomography scan showing intraluminal gas in the left renal vein (arrow).

Discussion

Emphysematous pyelonephritis is a rare, but potentially life-threatening infection. EPN must be differentiated from other complicated urinary tract infections such as emphysematous pyelitis and pyelonephritis as the clinical course of these infections may not be as ominous as EPN. This differentiation maybe difficult to make clinically as these infections may also present with septic shock. EPN typically presents with a triad of vomiting, abdominal or flank pain and fever.[1] Pneumaturia is generally associated with emphysematous cystitis and not EPN. Patients may present with leukocytosis, pyuria, elevated blood glucose, thrombocytopenia, elevated creatinine and urea, as well as positive urine and blood cultures.

There are several populations at risk of developing EPN. Ninety percent of patients are diabetic, most with poorly controlled blood glucose levels.[2] Immunosuppression, urinary obstruction and renal calculus disease also increase the risk of EPN. Reports have been documented of patients with EPN who possess none of the known risk factors.[3]

Microbial proliferation and glucose fermentation are thought to progress in the presence of hyperglycemia, poor vascular supply and urinary obstruction. Several types of bacteria have been associated with EPN. Escherichia coli (69%), Klebsiella pneumoniae (29%) and other gram negative enterobacteriae are most commonly detected.[2] Candida albicans,[4] Aspergillus[5] and the protozoan Entamoeba histolytica[6] have also been implicated in EPN.

A strong index of suspicion is required to make a prompt and accurate diagnosis of EPN. The populations at risk should undergo imaging studies once hemodynamically stable to investigate for intraparenchymal gas. In a retrospective chart review Shokeir et al[7] found kidney, ureter, bladder radiographs confirmatory for gas distribution in 11/20 cases tested, while the remaining 9/20 were highly suspicious. Often mistaken for bowel gas, a mottled, non-specific distribution of gas may be seen. Renal ultrasound is useful to diagnose obstruction of the urinary tract but was imprecise when identifying gas in the renal parenchyma as only 2/20 cases were diagnosed with ultrasound in Shokeir's series. Computed tomography scan of the abdomen was the most sensitive test to diagnose EPN, confirming 16/16 cases for the presence and extent of renal parenchymal gas. CT does not necessarily require contrast dye to delineate the extent of EPN, which is an advantage over studies such as intravenous pyelography.[7]

Several radiological classifications of EPN have been described. In 1996 Wan et al[8] described two types of EPN based on computed tomography scan results of 38 cases. Type 1 showed renal parenchymal destruction in the absence of a fluid collection with the presence of streaky, mottled gas. A fulminant course from onset of symptoms to death was associated with type 1 EPN. Type 2 EPN revealed renal or perirenal fluid collections with bubbly or loculated gas. More recently, Huang et al[2] described a classification system based on the CT imaging of 46 cases. Derived from Wan et al[8] and Huang et al,[2] Table 1 describes both classifications and the associated mortality risk.

Both authors discussed prognostic factors associated with unfavorable outcomes of EPN. These

TABLE 1. **Classification of emphysematous pyelonephritis (Wan et al,[8] Huang et al[2])**

	Class	Description	Mortality (%)
Wan et al[8]	Type 1	Renal parenchymal destruction in the absence of a fluid collection with the presence of streaky, mottled gas	69
	Type 2	Renal or perirenal fluid collections with bubbly or loculated gas	18
Huang et al[2]	1	Gas within the collecting system only (emphysematous pyelitis)	0
	2	Gas within the renal parenchyma without extension to the extrarenal space	10
	3A	Gas or abscess extension to the perinephric space[1]	29
	3B	Gas or abscess extension to the pararenal space[2]	19
	4	Bilateral EPN or solitary kidney with EPN	50

[1]Perinephric space defined as the area between the fibrous capsule and renal fascia
[2]Pararenal space defined as space beyond the renal fascia and adjacent tissues

TABLE 2. **Negative prognostic factors of EPN (Wan et al,[9] Huang et al[2])**

Wan et al[9]	**Huang et al[2]**
serum creatinine > 123.8 μmol/L (1.4mg/dL)	acute renal failure*
	platelets < 120 x 10^9/L
platelets < 60 x 10^9/L	SBP < 90 mmHg
	disturbed level of consciousness

*Defined as an increase in creatinine greater than 88.4 μmol/L if baseline creatinine was greater than 265.2 μmol/L or an increase greater than 44.2 μmol/L if baseline creatinine was less than 265.2 μmol/L

criteria are described in Table 2. In a 1998 retrospective analysis Wan et al[9] concluded that the probability of mortality for patients with Type 1 and Type 2 EPN rose from 69% to 92% and 18% to 53%, respectively, if both criteria were met. Huang et al[2] reported that the presence of two or more risk factors had worse outcomes in terms of failed treatment (percutaneous drainage and antibiotics) and mortality for class 3A, 3B and 4 EPN.

Initial management of EPN begins with attention to stabilizing the patient's vital signs including fluid resuscitation, broad spectrum antibiotic coverage and insulin if required. Medical management alone has been reported with a successful outcome by Punnose et al,[10] Best et al[11] and Flores et al,[12] who reported a case of bilateral EPN. Surgical management options include relief of obstruction via stent placement,[13] percutaneous drainage (PCD) under CT or U/S guidance[14] and nephrectomy.[6]

Due to the relative rarity of EPN, large prospective trials to assess treatment outcomes have not been conducted and there are no concrete guidelines to govern treatment. The literature is controversial. Several retrospective reviews of multiple cases have reviewed outcomes of various treatment modalities. The results in terms of survival are summarized in Table 3.

In 1997 Shokeir et al[7] reviewed 20 cases (from 1980-1995) of EPN treated with immediate nephrectomy. A survival rate of 80% led the author to recommend immediate nephrectomy as the appropriate management of EPN. There was no attempt to stratify these cases based on extent of gas production or other risk factors.

Recent literature also supports the safe and effective use of PCD with antibiotic treatment in the initial treatment of select cases of EPN. In 1997 Chen et al[15] conducted a retrospective analysis of 25 patients with EPN. All 25 patients underwent PCD following fluid resuscitation, blood glucose control and antibiotic treatment. There were two mortalities and three patients underwent delayed nephrectomy 3 weeks following PCD as the renal unit was non-functioning (n=2) or due to infected, obstructed calculus disease (n=1). This study found no significant difference in survival outcomes for PCD with antibiotic treatment of type 1 and type 2 EPN.

Percutaneous drainage of EPN may be considered in those cases where an abscess collection or renal obstruction is identifiable in a renal unit worthy of salvage. This may be necessary in patients with bilateral EPN or a solitary kidney. PCD may allow stabilization of the patient, decreasing the operative risk prior to nephrectomy.

TABLE 3. **Treatment modalities and EPN survival (Shokeir et al,[7] Chen et al,[15] Huang et al[2])**

	Shokeir et al[7] (n=20) % (n*)	**Chen et al[15] (n=25) % (n*)**	**Huang et al[2] (n=48) % (n*)**
Antibiotics alone	N/A (0)	N/A (0)	60 (3/5)
PCD + antibiotics	N/A (0)	80 (20/25)	66 (27/41)
Delayed nephrectomy following failed PCD + antibiotics	N/A (0)	100 (3/3)	13 (1/8)
Immediate nephrectomy	80 (16/20)	N/A (0)	100 (2/2)

*Surviving patients/total no. patients undergoing treatment
PCD = percutaneous drainage

Huang et al[2] reviewed four treatment modalities as summarized in Table 3. Class 1 through 4 EPN could be successfully managed with PCD and antibiotics. However, in class 3A, 3B and 4 EPN the presence of two or more prognostic factors (acute renal failure, thrombocytopenia, shock, altered level of consciousness) led to an increase in mortality. In these cases, nephrectomy is clearly considered the treatment of first choice, as the definitive way of removing the source of infection and with the greatest chance of survival. In 2003 Mydlo et al[16] conducted a literature review and concluded that initial treatment should be chosen based on the patient's condition and severity determined by CT imaging.

Clearly, rapid operative intervention retains an important role in the management of severe cases of EPN, and maybe life-saving. □

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