

Emphysematous pyelonephritis: Our experience with conservative management in 14 cases

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Abstract

Context: Emphysematous pyelonephritis (EPN) is a rare, severe, acute, necrotizing infection of the kidney. In this study, we present the clinical details, the management strategies, and the outcome of fourteen patients of EPN managed at our center.

Materials and Methods: A retrospective analysis of the hospital records was done. A total of fourteen patients with EPN were admitted in our hospital from August 2007 to February 2011. All the patients were managed conservatively. Follow-up ranged from six months to one year.

Results: Of the fourteen patients, four belonged to class I, five to class II, four to class IIIA and one to class IIIB. All the patients had history of fever, 43% had localized flank pain while 36% had vague abdominal discomfort. Renal angle tenderness was the most common sign, seen in 86% of the patients. *E. coli* was the most common bacteria, which was isolated from urine in 57% of the patients. On the risk factor stratification, three patients had simultaneous presence of 2 or more risk factors (thrombocytopenia-2 patients; renal function impairment-7 patients; shock-1 patient). All the patients were initially managed with aggressive fluid and electrolyte resuscitation, control of blood sugar levels, and broad spectrum antibiotics. Intervention, in the form of percutaneous drainage or DJ stenting, was done in six patients. One patient failed to respond to this minimally invasive modality of treatment and had to undergo an open drainage. Thus, the acute episode was managed with conservative management strategies in all the patients; however, three patients underwent nephrectomy due to poorly-functioning kidney during follow-up.

Conclusions: EPN is now being more readily diagnosed, at an early stage, making conservative management of EPN a safe, effective, and feasible option.

Key Words: Emphysematous pyelonephritis, nephrectomy, renal infection

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INTRODUCTION

Emphysematous pyelonephritis (EPN) is a rare, severe, acute

necrotizing infection of the kidney characterized by the presence of gas within the renal parenchyma, collecting system and peri-renal tissue.^[1] EPN is common in diabetics, especially in females. Non-diabetic patients can also develop EPN, albeit rarely, with a less severe clinical course as compared to diabetics.^[2] The term Emphysematous Pyelonephritis was first used by Schultz and Klorfein.^[3] In the yesterdays, this condition was associated with high morbidity and mortality but now, with the availability of better modalities, the natural history seems to have changed. We, in this study, present the clinical details and outcome of 14 patients of

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EPN managed at our center and discuss their management and outcomes.

MATERIALS AND METHODS

Between August 2007 and February 2011, a total of 14 patients were admitted in our hospital with emphysematous pyelonephritis. All the patients were studied with respect to the clinical features at presentation. Computerized tomography was done in 13 cases to confirm the diagnosis and for classification while 1 patient was referred to us with an MR Urogram. All the patients were thoroughly investigated, and the risk factors (as proposed by Huang and Tseng) were evaluated. All the patients were initially managed by aggressive diabetic control, correction, and maintenance of fluid and hemodynamic status, and antibiotics. The patients were followed up for 6-12 months.

RESULTS

Out of the fourteen patients with EPN, eleven were females while three were males (Male:female; 1:3.66). Age range was 35-58 years with a mean age of 51 years. Twelve patients (86%) were diabetic, out of which, nine were known diabetics while three were diagnosed following admission. seven of the known diabetics were on regular treatment while two were non-compliant; however, all the diabetic patients had raised blood sugar levels at the time of admission. Both the non-diabetic patients had ureteral obstruction due to stone disease. One of the diabetic patients too had hydronephrosis due to stone in the ureter. The clinical presentation of the patients is given in Table I. All the fourteen patients (100%) had fever at the time of presentation while localized flank pain was present in six (43%) of the patients. Five (36%) patients had vague abdominal discomfort while nausea with or without vomiting was present in six (43%) patients. Dysuria and increased urinary frequency was seen in four

(29%) patients. None of the patient had altered sensorium or pneumaturia.

On examination, renal angle tenderness was present in twelve (86%) patients while abdominal mass was found in four (29%) patients. Two (14%) patients had diffuse abdominal tenderness while only one had hypotension at the time of presentation. None of our patient had crepitations in the flank.

Pyuria was found in all the patients while leucocytosis was found in twelve (86%) patients. Two (14%) had thrombocytopenia while seven had deranged renal parameters at the time of admission. The urine of all the patients was submitted for culture and sensitivity testing. Out of the fourteen patients, *E. coli* was isolated from eight (57%) patients, *Klebsiella* from two (14%) and *Proteus* from one (7%) patient. In three (21%) patients, no bacteria could be isolated from urine. CT scan was performed for confirmation of the diagnosis as well as for classification while one patient was referred to us with an MRI. The distribution of the patients into various classes based on radiological investigation is given in Table 2. The representative images of patients with EPN are shown in Figures 1-4.

On risk factor stratification, as per the criteria proposed by Huang and Tseng,^[4] three patients had simultaneous presence of two or more risk factors (Thrombocytopenia-2 patients; Renal Function Impairment-7 patients; Shock-1 patient).

Management strategies adopted

After admission, all the patients were initially managed by aggressive diabetic control, correction, and maintenance of fluid and hemodynamic status, and antibiotics. Initially, broad spectrum antibiotics were used. A combination of piperacillin and tazobactam was used as the first line antibiotic. Aminoglycosides were added in patients who had normal renal parameters while a quinolone was added in patients with deranged renal parameters (with dose adjustment). Antibiotics were changed in accordance with the sensitivity report when it was available. Percutaneous drainage under CT guidance was done in three patients who had more than two risk factors. One patient of class IIIB EPN did

Table 1: Clinical Features at presentation

	No. of patients	Percentage
Sex		
Male	3	21
Female	11	79
Clinical presentation		
Fever	14	100
Flank pain	6	43
Vague abdominal discomfort	5	36
Nausea and vomiting	6	43
Dysuria and frequency	4	29
Depressed level of consciousness	None	
Pneumaturia	None	
Signs		
Abdominal mass	4	29
Renal angle tenderness	12	86
Abdominal tenderness	2	14
Hypotension	1	7
Crepitus in flank region	None	

Table 2: Radiological classification of patients (n=14)

	No. of patients	Percentage
According to the classification by Wan et al.		
Type I	4	29
Type II	10	71
According to the classification by Huang and Tseng		
Class I	4	29
Class II	5	36
Class IIIA	4	29
Class IIIB	1	6
Class IV	0	0

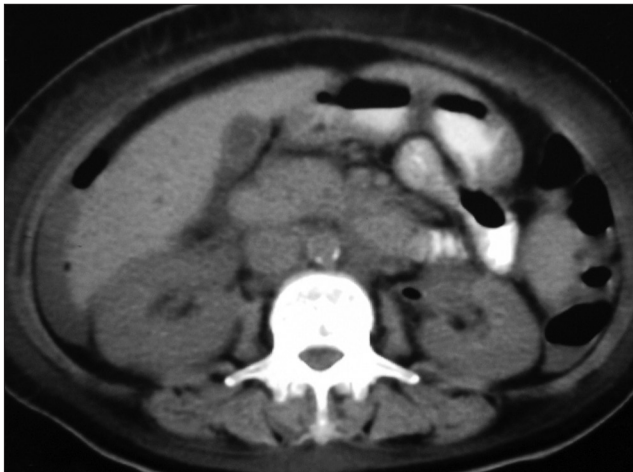


Figure 1: CT scan showing gas within the pelvis of left kidney (Class I EPN)

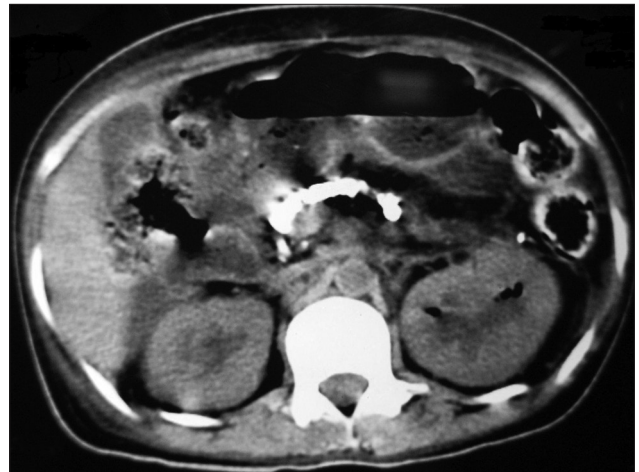


Figure 2: CT scan showing gas within the parenchyma of the left kidney (Class II EPN)

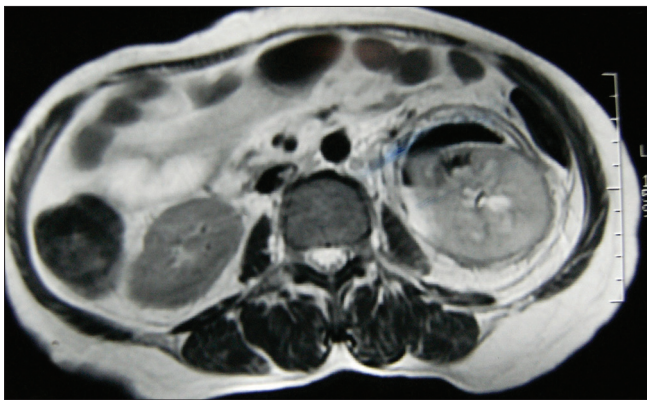


Figure 3: MRI showing gas in the perinephric space (Class IIIA EPN)



Figure 4: CT scan showing gas within the renal parenchyma extending into the perinephric space and pararenal space (Class IIIB EPN)

not respond to this management and had to undergo a surgical exploration. On exploration, the kidney was found to be severely inflamed and densely adherent to the surrounding structures. The nephrectomy could not be performed, but the necrotic material was evacuated, and drains were placed. The patient responded well to the treatment and had an uneventful recovery. DJ stenting was done in three patients who had upper tract obstruction, including the two non-diabetics.

Out of the fourteen patients, five patients were lost to follow-up while the remaining nine were followed up for a period ranging from 6 months to 1 year. On follow-up, two patients had poorly-functioning kidney on dynamic renogram and underwent nephrectomy. One patient developed nephro-cutaneous fistula and had to undergo a nephrectomy. No patient had any episode of recurrent pyelonephritis. Thus, in thirteen patients, the acute episode could be managed with conservative management strategies alone while in one patient, open surgical drainage was done. Only three patients underwent nephrectomy, that too during follow-up.

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.^[4] This condition was first described by Kelly and MacCullum^[5] while the term EPN was first used by Schultz and Klorfein.^[3] EPN is much more common in females, with various studies reporting the female to male ratio ranging from 3:1 to 43:3.^[2,6] In our study too, majority of the patients were females. EPN is common in patients with diabetes mellitus (DM), with up to 90% of them being diabetic.^[6] Occasionally, patients without DM but with obstruction of the corresponding reno-ureteral unit may also develop EPN; however, the disease is less extensive in them.^[2]

The most characteristic feature of EPN is presence of gas in the renal parenchyma. Microbial infection and rapid catabolism

have been proposed as the cause for increased gas formation, which is trapped in the tissues due to impaired transport of gas as there is vascular compromise in the pyelonephritic kidney. Huang and Tseng have postulated that 4 factors are involved in the pathogenesis of EPN, which were gas-forming bacteria, high tissue glucose level, impaired tissue perfusion, and a defective immune response.^[4] Leukocyte dysfunction seen in diabetics may contribute to the pathogenesis of EPN. Gas production in the renal parenchyma in the absence of infection has also been described following traumatic renal infarction.^[7] The infecting organisms are usually glucose-fermenting bacteria. *E. coli* is the most common bacteria implicated in EPN, others are *Klebsiella* and *Proteus*. EPN caused by *Streptococcus* and *Candida* have also been reported.^[8]

The clinical presentation is often suggestive of severe acute pyelonephritis, with fever, flank pain, and pyuria being the most common clinical manifestations. However, these are non-specific and may be seen in other forms of upper urinary tract infections. Other clinical features of EPN include non-specific abdominal pain, nausea, vomiting, depressed levels of consciousness, shock, renal angle tenderness, dysuria, crepitations in the flanks, and pneumaturia.^[2-4,6,9] In our study too, fever and flank pain were the most common symptom. Pyuria was also seen in all the cases. Huang and Tseng^[4] in their study had found that thrombocytopenia (46%), acute renal function impairment (35%), disturbance of consciousness (19%), and shock (29%) can be the initial presentations. Shokier *et al.*^[6] in their study found renal functional impairment in 80% of their patients and shock and coma in 15% of patients. In our study, only 14% of the patients had thrombocytopenia, 50% had deranged renal functions, and only 1 patient had hypotension.

Diagnosis of EPN is made radiologically, with CT being the most definitive modality. EPN can also be diagnosed by abdominal X-ray and ultrasonography. CT images are useful to define the presence, extent, and position of gas within the renal parenchyma, beside any other associated renal pathology, like calculi, and/or presence of obstruction may also be evident on CT scan. Contrast-enhanced CT scan is better as it give an idea about the function status of the renal units as well as it facilitates the description of the intraparenchymal gas (streaky, mottled, bubbly, rimlike, crescent shaped, locular, and so on). However, in patients with deranged renal parameters, a non-contrast CT scan may suffice. In addition to diagnosis and staging of EPN, CT scan is also helpful in monitoring the response to treatment. It might show resolution of the gas and abscesses or the development of new lesions.

Abdominal X-ray may reveal mottled gas shadow in the renal

region followed by development of a crescent of gas surrounding parenchyma. In the absence of CT facilities, Intravenous Urography (IVU) can be used; however, quite a few EPN patients are in shock or have elevated serum creatinine levels; therefore, IVU cannot be used in such patients. The sensitivity of IVU has been reported as 85% in a study by Paivansalos *et al.*^[10] and 100% by Ahlering and colleagues.^[11] Ultrasound reveals strong focal echoes, typically described as 'dirty shadows'. The sensitivity and specificity of ultrasonography in EPN is low. Obesity and bowel gas interfere with the interpretation of plain X-Ray, IVU as well as ultrasonography.

Staging of EPN is done radiologically based on the extent of gas in the renal parenchyma and surrounding tissues. It might be useful for decision making and prognostication. Langston and Pfister^[12] suggested a classification on the basis of abdominal X-ray and an intravenous pyelography, which was later modified by Michaeli *et al.*^[13] They classified EPN into 3 classes:

- Class I - Gas in renal parenchyma or perinephric tissue
- Class II - Gas in the kidney and its surroundings
- Class III - Extension of gas through fascia, or bilateral disease.

Wan *et al.*^[1] classified the gas collection as type I or type II, on the basis of CT scans.

- Type I: Renal necrosis with presence of gas but no fluid
- Type II: Parenchymal gas associated with fluid in renal parenchyma, perinephric space, or collecting system.

Mortality was 69% in patients with type I EPN and only 18% in patients with type II EPN. Similar mortality rates for type I and type II EPN were observed by Chen *et al.*^[14]

Huang and Tseng^[4] also used CT to classify patients with EPN as follows:

- 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 pararenal space
- Class IV: EPN in solitary kidney, or bilateral disease.

The classification by Huang and Tseng is a superior due to the better prognostic value and is also helpful in selecting a management protocol. In their study, class I and II patients, all survived following treatment with percutaneous procedures and medical therapy. While in patients belonging to class III or IV, those with fewer than two risk factors (i.e. thrombocytopenia, acute renal function impairment, disturbance of consciousness and shock) had an 85% survival rate with percutaneous drainage and medical therapy, whereas patients of class III or IV EPN and two or more risk factors had a 92% failure rate with percutaneous drainage and medical therapy. In their study,

33.33% of the patients belonged to class I or II while 66.66% of the patients belonged to class III or IV; while in our study, majority of the patients (64%) belonged to either class I or II while 36% of the patients belonged to class III or IV.

Despite the morbidity and mortality associated with EPN, there is still controversy regarding its proper management. Shokier *et al.*^[6] and Ahlering *et al.*^[11] proposed immediate nephrectomy following resuscitation of the patient. Huang and Tseng^[4] proposed the management protocol based on the radiological classification and presence of risk factors. They managed class I and II EPN with antibiotics along with percutaneous drainage or relief of obstruction while those with class III or IV EPN were given a trial of conservative management, and nephrectomy was done in patients who had more than 2 risk factors for poor prognosis or in patients in whom conservative management failed. There has been a recent trend towards conservative treatment strategies for EPN. A number of reports of successful conservative management of EPN have come up.^[14-22]

Initial treatment of patients with EPN should start with vigorous resuscitation. Fluid and electrolyte imbalances should be corrected, Diabetes should be controlled, and antibiotics should be started. Broad spectrum antibiotics targeting gram-negative bacteria should be started. Aminoglycosides should be used with caution as quite a few patients may have deranged renal functions. Patients who have obstruction of the renal drainage system should have their obstruction relieved. DJ stenting may be tried in such patients. Percutaneous drainage under CT/USG guidance is done in patients who have well-formed collections within the renal parenchyma or the surrounding tissue. Nephrectomy should be considered in a select group of patients who have gross destruction of renal parenchyma, have class IIIA or class IIIB gas distribution when there is simultaneous presence of 2 or more risk factors or when the involved kidney is non-functioning.^[2] Jayesh V Dhabalia *et al.*^[23] in their study similarly found that EPN can be successfully managed with conservative treatment modalities, and the such treatment strategies are associated with lower mortalities than emergency nephrectomies.

In our study, we started with conservative management, and reserved nephrectomy for patients who did not respond to conservative management. One patient in our study failed conservative management and was planned for nephrectomy; however, only open drainage could be done due marked inflammation and adhesion. The patient responded well to open drainage. Thus, minimally invasive modalities were successful for emergency management of most of the patients.

CONCLUSION

EPN is a rare and severe infection of the renal parenchyma. However, with increasing availability and decrease in the threshold for using imaging modalities like ultrasonography and CT scan in patients with severe urinary tract infections and sepsis, more number of patients are now diagnosed with emphysematous pyelonephritis and at an early stage. With early diagnosis, availability of more potent antibiotics and advances in critical care support systems, an increasing number of patients can be managed with conservative approaches, and nephrectomy should be reserved for patients who fail conservative management, especially those belonging to higher radiological class with simultaneous presence of two or more risk factors.

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