Original Article

Mortality in emphysematous pyelonephritis: Can we reduce it further by using a protocol-based treatment? The results of a prospective study

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Abstract

Introduction: Even though the mortality rate in emphysematous pyelonephritis (EPN) is brought down presently to 13%–25%, there is still scope for improvement. The hurdle lies in identifying those patients at risk of mortality earlier in the disease process and providing intensive management to them. In this study, we created risk groups by combining both clinical and radiological presentations and applied a protocol-based treatment to evaluate its role in reducing mortality.

Methods: We formulated a treatment protocol based on the available literature. The first step was to recruit all patients diagnosed with EPN into the treatment protocol as soon as possible without any delay. The second step was to stratify the patients into risk groups based on our clinicoradiological risk group classification. The third step was to apply the treatment protocol according to the risk group they belonged to.

Results: We treated 24 patients with EPN in the past 4 years. According to the radiological classification – four patients had Type 1 disease, five patients had Type 2A disease, six patients had Type 2B disease, four patients had Type 3A disease, two patients had Type 3B disease, two patients had Type 4A disease, and one patient had 4B disease. Following risk stratification, we have categorized seven patients into category 1, eight patients into category 2, and nine patients into category 3. All except one patient survived following the treatment protocol followed by us.

Conclusions: Early risk stratification, intensive management, and prompt treatment according to a protocol can reduce mortality even further in patients with EPN.

Keywords: Emphysematous pyelonephritis, golden day, mortality reduction, risk stratification, treatment protocol

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INTRODUCTION

Emphysematous pyelonephritis (EPN) is a life-threatening necrotizing infection of the kidney characterized by production and accumulation of gas in the kidney and surrounding tissues.[1] From being a death sentence, the outcome of this life-threatening disease gradually improved in parallel with the improvements in imaging modalities, supportive care, and better understanding of the disease pathophysiology. The mortality rate of this disease

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gradually reduced from a historic high of 50% to around 13%–25% at present.[2-6] As with other areas of urology, with the reduction in mortality rate, the focus has shifted now from radical treatments toward nephron-sparing approach.[7]

At present, no one has second thoughts regarding initial treatment protocol – resuscitation, electrolyte management, rapid control of blood sugar levels, initiation of broad-spectrum antibiotics, emergency decompression of disease by either double J (DJ) stenting or percutaneous nephrostomy (PCN) or percutaneous drainage (PCD) along with judicious usage of early (EN) or delayed nephrectomy (DN). However, despite advances in the management, there still remained a group of patients who have a rapidly declining course. Wan *et al.* hypothesized that the difference in outcomes is probably related to the differences in severity of immune compromise and the vascular insufficiency in the kidneys.[8] Hence, the last hurdle in further reduction of mortality lies in identifying this group earlier in the disease process and providing them with intensive management.

However, crossing the last hurdle is not easy as there are some problems inherent in the management of this disease. The first problem is that the available case series on EPN were comprised of few patients and were retrospective in nature. It is very difficult to undertake large well-controlled studies in an uncommon and emergency disease condition. The second problem is that even after identifying the prognostic factors for mortality, there is still confusion prevailing as each study identified a different set of prognostic factors and there is no uniformity in them.[8-17] The third problem is that even though the radiological classification (Wan's or Huang-Tseng's) used by most of the investigators gave an indication about the radiological severity of the disease, the clinical picture differed greatly among individuals with similar radiological presentation.[8,9]

In this study, we have assimilated the data and interpretations from previously available studies and have applied them prospectively by combining early risk stratification factors to evolve a protocol-based treatment and assess whether these measures would help in further reduction of mortality. We will discuss our results in this study.

METHODS

We made a protocol-based treatment on the available literature with two distinct goals: the first one was reduction of mortality and the second one was organ preservation, wherever possible.[8-17] The first step in the protocol was to recruit all the patients diagnosed with EPN into the treatment protocol as soon as possible without any delay. The principle was to treat the 1st day as Golden day just like the Golden hour in trauma. We made an observation that most of the deaths happened in patients admitted under other departments and later referred to us. The reason was that most of the patients presented with nonspecific symptoms and septic shock or multiorgan dysfunction were admitted in General Medicine or its allied specialties and by the time, the diagnosis was made, the patient would deteriorate further. The patients who consulted in the Urology Outpatient Department or referred directly to the Urology from casualty got immediate medical or surgical attention depending on their clinical and radiological picture. Hence, to reduce the delay in treatment, we had to sensitize the Casualty and our General Medicine colleagues regarding this clinical condition. In consultation with them, we made a protocol that all diabetic patients with unexplained fever and systemic symptoms should undergo noncontrast computed tomography (NCCT) kidney-ureter-bladder scan as soon as possible after initial stabilization. We have treated 104 patients with acute pyelonephritis over 4 years from January, 2016 to December, 2019, out of which, 24 patients were found to have EPN.

The second step was to classify these patients into risk groups [Table 1]. Group 1 comprised all stable patients with low-risk computed tomography (CT) findings. Group 2 comprised all stable patients with high-risk CT findings. Group 3 comprised all unstable patients irrespective of CT findings. The criteria for placing the patient in the stable/unstable category were based on the general condition and clinical picture of the patient [Table 2]. The risk factors identified were based on the previous research articles [Table 3].[8-17]

Abdominal CT scan and echography were performed in all these cases. Modified Huang and Tseng classification was used to classify the radiological findings on the CT scan[9] [Table 4]. Class 2 was subdivided into 2 groups – Class 2A and 2B in our study. 2A is with low volume gas in the kidney <3 cm in a single zone (upper, middle, or lower) and 2B is with high volume disease in the kidney with a cut-off value of 3 cm – >3 cm in a

Table 1: Risk groups

	General condition of the patient	Radiological picture (NCCT)
Group 1	Stable	Low risk
Group 2	Stable	High risk
Group 3	Unstable	Low/high risk

NCCT: Noncontrast computerized tomography

Table 2: Criteria for placing the patient in stable/unstable category based on general condition and clinical profile of the patient *Unstable – any one major or two minor criteria*

Major criteria
Shock (systolic BP <90 mm Hg)
Disturbance of consciousness in the form of confusion, delirium, stupor or coma
Severe thrombocytopenia (a marker of disseminated intravascular coagulation)-platelet count $<75\times10^9$/L
Renal function impairment (creatinine level >5 mg/dL)
Minor criteria
Hyponatremia <125 mEq/L
Renal function impairment (creatinine level >3 mg/dL)
Acute renal function impairment:
Further elevation of the serum creatinine level of more than 1 mg/dL from baseline (baseline serum creatinine level >3 mg/dL)
Further elevation of the serum creatinine level more than 0.5 mg/dL from baseline (Baseline serum creatinine level <3 mg/dL)
Serum albumin <2.5 g%
Patients with a high urinary red blood cell count
Elevated leukocyte count (>20,000/μL)

single zone or mottled gas appearance in multiple zones. This cut-off value was derived based on the renal abscess treatment protocol where we treat abscesses <3 cm with antibiotics. If there was mottling or streaky gas pattern, the whole intervening area (if it is nearby) was also included in calculating the volume. Class 4 was subdivided into 4A and 4B. Bilateral Class 1 was categorized as Class 4A. Class 1, 2A, and 4A were classified as low-risk CT findings and Class 2B, 3, and 4B were classified as high-risk CT findings. Hydronephrosis was defined as any dilatation of renal pelvis and calyces and was not accorded any specific risk.

The treatment protocol was formulated based on the various meta-analyses published previously [Table 5].[8-17] The treatment algorithm is shown in Table 6. "Unsuccessful" treatment was defined as clinical manifestations of unstable hemodynamic parameters for 48 h, persistent fever of more than 100°F, and progressive or persistent lesions on further imaging studies. If there are progressive or persistent lesions on follow-up imaging, they were treated according to the original protocol.

RESULTS

We had diagnosed 25 patients with EPN in the past 4 years (from January, 2016 to December, 2019). One patient refused treatment and left the hospital before intervention. Hence, 24 patients were included in our study, and they were classified into three groups and treatment was given according to protocol. Patient demographics and characteristics are represented in Table 7. Age of the patients ranged from 32 years to 73 years. Twenty-one patients were diabetic. Blood glucose at presentation ranged from 188 to 512 mg/dL. Eighteen of the 21 diabetic patients had blood glucose more than 300 mg/dL at presentation. Most of the patients presented with flank pain and fever. The duration of symptoms ranged from 2 days to 1 month. Five patients admitted in medicine intensive care unit had nonspecific symptoms at presentation. Nine patients had features of obstructive uropathy because of necrosed papilla and two patients had obstructive uropathy because of urolithiasis. Nine patients had DJ stent placed and four had PCN placed in them.

Treatment of patients by risk categories is shown in Table 8. One patient had an interesting presentation. He had undergone PCD elsewhere at the time of initial presentation and presented to us with recurrent low-grade fever after 1 month. There was pus drainage from the drain after initial drainage of approximately 100 ml/day for 1 week. The kidney was auto-nephrectomized on NCCT with gas limited to the kidney. During nephrectomy, there was only a nubbin of kidney tissue left. Among the nine patients who belonged to Category 3, one patient had Type 2B disease on NCCT who had rapidly deteriorated despite bedside PCD. She was treated for 5 days elsewhere initially and presented to us with shock, delirium, and respiratory compromise.

All the patients had improved renal parameters following recovery. The improvement in creatinine ranged from 0.6 to 3.8. Twenty-two out of 24 patients had elevated creatinine at admission (>2 mg%) and four patients had creatinine more than 5 mg%.

DISCUSSION

Kelly and MacCallum first described about a gas-forming renal infection in 1898.[18] Since then, a variety of terms were used to describe this clinical condition – renal emphysema, pneumonephritis, pneumonephrogram, pyelonephritis emphysematosa, etc., Schultz and Klorfein suggested the term EPN as in their opinion, this stressed the importance between kidney infection and gas formation.[19] In 1941, Gillies and Flocks laid down three essential factors for gas production in the urinary tract – diabetes mellitus, obstructive uropathy, and gas-producing organisms.[20] Since then, the criteria that there should be obstructive uropathy are refuted by multiple reports. Now, the essential criteria are diabetes mellitus, obstructive uropathy, or both along with gas-producing organisms.

The initial investigators advised early aggressive surgical treatment in the form of emergency Nephrectomy, as they thought this might reduce mortality. Michaeli *et al.* presented data of 55 patients in 1984 and echoed the same

Table 3: Risk factors identified by various studies

Authors	Number of patients	Prognosis significant	Prognosis-NS
Wan *et al*. (1996)[8]	38 patients	Platelet count 60,000/mm^3 or less Serum creatinine level >4 mg/dl Hematuria Patients with radiological Type I emphysematous pyelonephritis versus Type II (69 vs. 18%)	Age DM Blood glucose level Bacteremia Leukocyte count Presence or absence of urinary tract obstruction Modes of treatment
Huang and Tseng[9]	48 patients	Thrombocytopenia Renal function impairment Disturbance of consciousness Shock Radiological class	-
Falagas *et al*.[11]	175 patients	Conservative mode of treatment alone Bilateral EPN Type 1 EPN (Wan *et al*.) Thrombocytopenia Shock (<90 mm Hg) Disturbance in consciousness Serum creatinine >3 mg%	Age Uncontrolled DM Hematuria Proteinuria
Somani *et al*.[4]	210 patients	Reduced level of consciousness Shock Patients with a high urinary red blood cell count Serum creatinine level greater than 4 mg/dl Thrombocytopenia (platelets less than 60,000/mm^3)	Age Urinary tract obstruction Blood glucose level
Aswathaman *et al*.[7]	41 patients	Shock Altered sensorium Need for hemodialysis	Age Blood sugar Duration of symptoms Serum creatinine WBC count
Khaira *et al*.[12]	19 patients	Shock at admission Serum creatinine>5 mg% DIC	Age of patient Unilateral/bilateral Disease class Sepsis >or=2 or <2 poor prognostic factors
Kuo *et al*.[13]	16 patients	Impaired renal function Hematuria	-
Kapoor *et al*. (2010)[5]	39 patients	Altered mental status Thrombocytopenia <40,000 Renal failure Cr >2.5 mg% Severe hyponatremia <120 mEq/L	Disease class Age Delay in presentation Hydronephrosis Poor glycemic control HbA1c >7%
Ubee *et al*. (2011)[14]	-	Systolic blood pressure <90 mm Hg Disturbance of consciousness Increase in serum creatinine level Thrombocytopenia Bilateral EPN MM with antibiotics alone	Age Uncontrolled DM Nephrolithiasis
Lin *et al*.[15]	23 patients	Shock Long hospital duration Disease class	Proteinuria Age Serum albumin <2.5 g%
Olvera-Posada *et al*. (2013)[16]	18 patients	Altered consciousness Multiple organ failure (≥3) Hyperglycemia >400 mg/dl Elevated leukocyte count (>20,000 K)	One organ failure Acute renal failure Thrombocytopenia Disease class
LU *et al*.[17]	44 patients	Need for emergency hemodialysis Shock on initial presentation Altered mental status Severe hypoalbuminemia <3.0 g/dL Inappropriate empirical antibiotic treatment Polymicrobial infections	Disease class Age Serum albumin 4Hyponatraemia HbA1C >8% Thrombocytopenia (≤120,000/mL)
Aboumarzouk *et al*.[6]	628 patients	Shock at presentation	Obstructive uropathy

NS: Not significant, EPN: Emphysematous pyelonephritis, DM: Diabetes mellitus, DIC: Disseminated intravascular coagulation, WBC: White blood cell, MM: Medical management

sentiment[1] PN Klein *et al*. reviewed 66 reported cases of EPN in 1986 and found an overall mortality rate of 38%, i.e., 71% in medically treated patients and 29% in those surgically treated.[2] Others – Ahlering *et al*., Pontin *et al*., and

Table 4: Risk categorization based on Modified Huang-Tseng radiological classification

Risk category	Radiological category	Definition
Low risk	Class 1	Gas in the collecting system only (emphysematous pyelitis)
	Class 2A	Gas in the renal parenchyma without extension to the extrarenal space-low volume gas in the kidney <3 cm in a single zone (upper, middle, or lower)
	Class 4A	Bilateral class 1
High risk	Class 2B	Gas in the renal parenchyma without extension to the extrarenal space-high volume gas in the kidney >3 cm in a single zone or mottled gas appearance in multiple zones
	Class 3A	Extension of gas or abscess to the perinephric space
	Class 3B	Extension of gas or abscess to the pararenal space
	Class 4B	Bilateral EPN (2A, 2B, 3A, 3B) or solitary kidney with EPN

EPN: Emphysematous pyelonephritis

Table 5: Treatment based mortality outcome (percentage)-meta-analyses

	Medical treatment (%)	Immediate nephric (%)	Medical treatment with PCD (%)	Open drainage (%)	PCD-delayed nephrectomy (%)
Mydlos *et al*. (2003)	5/15 (33)	14/61 (23)	-		2/16 (12.5)
Falagas *et al*. (2007) 175 patients	2.85 OR 95% CI 1.19-6.81	-	-		-
Somani *et al*. (2008) 210 patients	12/24 (50)	16/64 (25)	16/118 (13.5)	0/2 (0)	1/15 (6.6)
Aboumarzouk *et al*. (2014)	25/167 (15)	42/126 (33.3)	39/283 (13.8)	1/18 (6)	5/47 (10.6)

PCD: Percutaneous drainage, OR: Odds ratio, CI: Confidence interval

Table 6: Treatment algorithm

Category	Treatment	Follow-up
Category 1 (clinically stable and low-risk CT)	Antibiotics (3rd generation cephalosporin) Supportive therapy DJ stenting PCD	Clinically stable, no flank pain or fever-NCCT KUB after 1 month and proceed with check ureteroscopy if hydroureteronephrosis was present on initial presentation If fever and flank pain persist - limited CT KUB in 3 days. Change antibiotics (culture specific) or consider another PCD (in case of Type 2A) If patient becomes unstable, escalate the treatment to Group 3
Category 2 (clinically stable with high-risk CT)	Antibiotics (3rd generation cephalosporin) Supportive therapy DJ stenting +/- PCD (preferably CT guided)	Clinically stable, no flank pain or fever- limited CT after 1 week If fever and flank pain persist- limited CT in 3 days. Change antibiotics (culture specific) or consider additional PCD or consider open drainage If patient becomes unstable, escalate the treatment to category 3
Category 3 (clinically unstable)	Antibiotics (carbapenems) Supportive therapy DJ stenting +/- PCD (bed side USG guided PCD, CT guided if possible)	Clinically stable, no flank pain or fever- limited CT after 5 days Clinically stable, fever and flank pain persist- limited CT in 3 days. Change antibiotics (culture specific) or consider additional PCD or consider open drainage Clinically unstable despite treatment - Consider emergency nephrectomy or open drainage

PCD: Percutaneous drainage, USG: Ultrasonography, NCCT: Noncontrast computerized tomography, KUB: Kidneys ureters and bladder, CT: Computerized tomography, DJ: Double J

Shokeir *et al*. – concluded that resuscitation and appropriate medical treatment should be attempted, but immediate nephrectomy should not be delayed, for the successful management of EPN.[3,21,22]

The whole scenario changed in 1986 with the report by Hudson *et al*., who first described fluoroscopically guided PCD for treating EPN, with successful clinical results.[23] However, this was slow to get accepted by the urology community. There was even a provocative article in 1993 by Koh *et al*., titled "emphysematous nephrectomy - drainage or nephrectomy" which still supported early nephrectomy (EN).[24] But with increasing reports of successful treatment of EPN with medical management and percutaneous drainage by multiple authors led many more urologists to question the role of emergency nephrectomy and adopt the nephron saving strategy in the treatment.[25-29]

With multiple authors reporting improved survival rates with PCD, the focus ultimately shifted from EN to salvaging the remaining kidney. Wan *et al*. reported their results of 38 patients and classified patients into 2 types: Type 1 – Dry type with mottled or streaky gas and Type 2 – wet type with fluid collections and bubbly or loculated gas.

The mortality in Type 1 was more than Type 2 (69% vs. 18%)[8,30] Chen *et al*. (1997) published their 10-year results of 25 patients. Twenty patients became alright with antibiotics and PCD. Three patients required delayed nephrectomy and two patients died of septic shock. Importantly, gas pattern and the response to treatment had no correlation in their study.[31] Subsequently, Huang and Tseng published their clinicoradiological classification after analyzing the results of 48 patients.[31] Twenty seven out of 41 patients who had medical management along with PCD had successful outcome while rest underwent

Table 7: Patient characteristics and demographics

Variable	Study results data	
Age (years)	32-73	
Sex		
Male	9	
Females	15	
Diabetes	21/24	
Other comorbidity	17/24	
	13/24 CAD, CVA, COPD	
	3/24 on steroids	
	1/24 neurogenic bladder	
Side		
Right	10/24	
Left	11/24	
Bilateral	3/24	
Admission		
Urology OP	7	
Casualty	12	
Medicine ICU	5	
Duration of symptoms	2 days-1 month	
Urine culture		
E. coli	17	
E. coli + *Proteus mirabilis*	4	
E. coli + *K. pneumoniae*	3	
Blood culture		
E. coli	9/24	
Sterile	15/24	
Serum creatinine (mg %)		After 2 months
<2	2/24	11/23
2-3	7/24	7/23
3-5	11/24	3/23
>5	4/24	2/23
Radiological classification	Type 1-4	
Low risk-11	Type 2A-5	
High risk-13	Type 2B-6	
	Type 3A-4	
	Type 3B-2	
	Type 4A-2	
	Type 4B-1	
Risk categories		
Category 1	7	
Category 2	8	
Category 3	9	

E. coli: *Escherichia coli*, CAD: Coronary artery disease, CVA: Cerebrovascular accidents, COPD: Chronic obstructive pulmonary disease, ICU: Intensive care unit, *K. pneumoniae: Klebsiella pneumoniae*

nephrectomy. Totally, nine out of 48 patients died with a mortality rate of 18.8%. According to CT findings, they classified patients into 4 groups and mortality rate had a direct correlation with the class. Following these reports, multiple authors validated the successful treatment with percutaneous drainage.[32-35]

Meta-analysis by Mydlo *et al.* (160 patients) in 2003, Falagas *et al.* (175 patients) in 2007, and Somani *et al.* (210 patients) in 2008 firmly established the role of PCD in the treatment of this disease.[4,10,11] Somani *et al.* even went ahead and titled their article – Is PCD the new gold standard in the treatment of EPN.[4] Aswathaman *et al.* reported that those patients who were not nephrectomized maintained an average relative renal function of 42%.[7] Therefore, now it is established that the best treatment strategy should be the one that improves the patient survival and at the same time, maximizes the renal salvage. Finally, in 2014, Omar M. Aboumarzouk *et al.* published a review article calling for a proper management strategy.[6] They reviewed results of 628 patients and reported that the overall mortality rate was 18%. They also reported that shock was associated with a high mortality rate and therefore should be managed aggressively. PCD and medical management were associated with significantly higher survival rates than EN, and therefore, EN should only be considered if the patient does not improve despite other treatments. Along with authors prescribing early PCD, multiple reports also emerged advocating medical management alone for the treatment of EPN.[36,37]

EPN can be compared to nontraumatic gas gangrene although the etiology and pathogenesis differ. The gas isolated in both conditions is similar, containing hydrogen, carbon dioxide, nitrogen in large quantities, and oxygen in small quantities – implying that the bacteria are indulging in mixed acid fermentation.[8,9,38-40] EPN is the interplay of high tissue glucose levels, impaired tissue perfusion, and weak immune system along with obstructive uropathy in some patients. At what point, acute pyelonephritis becomes EPN is not clear.

Almost all the published reports were case series and retrospective in nature. They identified a group of risk factors in the group of patients succumbing to the disease. However, different authors reported different risk factors. We selected a group of risk factors which were reported by majority of the authors and applied it together toward a prospective management plan. Many recent studies also claimed that the mortality is higher in the emergency nephrectomy group compared to the group of patients who underwent percutaneous drainage. However, the problem with this data is that the patients who underwent emergency nephrectomy were either very sick or underwent late nephrectomy. Most of these patients have multiple comorbidities and were at the extreme end of the sepsis spectrum. At what point of time, we have to resort to emergency nephrectomy which was not clear – it should not be either too early or too late. Unless we have a prospective management plan, the correct decision cannot be made. Radiologic risk stratification also alone would not do enough justice in evaluating the results as the clinical picture may not always correlate with the radiologic picture. Initial clinical presentation, clinical picture after resuscitation, comorbidities, and radiologic picture should be amalgamated to correctly stratify each patient and tailor the treatment accordingly. This will also

Table 8: Treatment of patients by risk categories

Risk group category	No of patients	Radiological category	Treatment given
Category 1 (clinically stable and low-risk CT)	7	Type 1-2	DJ stenting
		Type 2A-4	Medical treatment-2
		Type 4A-1	DJ stenting + PCD-2
			Bilateral DJ stenting
Category 2 (clinically stable with high-risk CT)	8 (3 patients had hydronephrosis and underwent DJ stenting)	Type 2B (HDN)-3	DJ stenting + PCD
		Type 2B-1	PCD alone
		Type 2B-1	PCD followed by nephrectomy
		Type 3A-2	PCD + PCN (in 1 patient)
		Type 4B-1	Bilateral PCD + PCN
Category 3 (clinically unstable)	9	Type 1-2	DJ Stenting-1/PCN-1
		Type 2A-1	PCD
		Type 2B-1	PCD-died
		Type 3A-1	PCD + PCN followed by Nephrectomy
		Type 3A-1	PCD
		Type 3 B-1	PCD followed by open drainage
		Type 3B-1	PCD followed by nephrectomy
		Type 4A-1	Bilateral DJ stenting

HDN: Hemolytic disease of the newborn, PCD: Percutaneous drainage, PCN: Percutaneous nephrostomy, CT: Computerized tomography, DJ: Double J

help in comparing the results from different institutions. We created 3 risk groups based on the clinical condition, laboratory parameters, and radiological features. Even though the previous authors tailored treatment based on the general condition of the patient, this type of prospective risk group classification and treatment was not attempted by them.

The treatment and outcome correlated fairly well with the risk groups we have created. Among the patients who were categorized as risk Group 2, patients who had obstructive uropathy rapidly improved with DJ stenting. It appears that obstructive uropathy is a good prognostic factor as these patients are diagnosed earlier, and corrective measures could be undertaken earlier in the course of the disease. Among the patients who were categorized as risk Group 3, those who could be resuscitated and stabilized had recovered even though some of them required open drainage and emergency nephrectomy. It appears that patients who could not be stabilized despite aggressive resuscitation and PCD should be taken up for emergency nephrectomy as soon as possible. The mortality rate in our study was 4% (1 in 24). Three patients had emergency nephrectomy and all of them improved after surgery. The only patient who died in our study was a risk Group 3 patient who was referred to us in a very critical condition. The patient had such a rapid deterioration that she could not be adequately stabilized for taking up for emergency nephrectomy and succumbed despite PCD. Her diagnosis was delayed because of nonspecific presentation and delayed imaging. This underscores the importance of sensitizing the fellow specialties regarding the importance of this disease. The main problem with our study is the limited number of patients. However, at the same time, we must acknowledge the fact that a single center cannot get adequate number of patients to validate study in this group of patients, and further, the study should be initiated in other centers for a safer inference to be drawn.

CONCLUSIONS

The clinical course of EPN can be changed with early identification, risk stratification, and aggressive management based on an active protocol-based treatment.

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Conflict of Interest
There are no conflicts of interest.

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