Emphysematous Pyelonephritis with Diabetic Nephropathy: A Rare Case Report

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ABSTRACT

Emphysematous pyelonephritis is a severe, uncommon, potentially fatal necrotizing pyelonephritis with a variable clinical presentation, ranging from mild abdominal pain to septic shock. The majority of cases occur in diabetics with poor glycemic control while a small percentage may be due to urinary tract obstruction. We present a case of a 57 year old male patient, diabetic on treatment, presenting with left flank pain and poor stream of urine since one week. Laboratory tests revealed that the patient had electrolyte imbalance, ketoacidosis and high blood sugar. Radiological examination gave a diagnosis of Left Type 1 Emphysematous Pyelonephritis. In spite of giving vigorous resuscitation and antibiotics with nephrostomy, the patient had to undergo nephrectomy due to extensive renal parenchymal destruction. The nephrectomy specimen was studied in detail to know the histopathological findings in a case of diabetic patient with emphysematous pyelonephritis. We present this case not only because of it being a rare complication of diabetes as well rarely seen in male but also to focus on the histopathological findings of the same, documentation of which is limited in literature.

Keywords: Emphysematous Pyelonephritis, Diabetic Nephropathy, Hyperglycemia.

INTRODUCTION

The first case of gas-forming renal infection was reported in 1898 by Kelly and MacCallum.¹ Since then many names have been used to describe emphysematous pyelonephritis (EPN) such as renal emphysema, pyelonephritis emphysematousa and pneumonephritis.² In 1962 Schultz and Klorfein proposed emphysematous pyelonephritis as the preferred designation name because it stresses the relationship between acute renal infection and gas formation³.

Emphysematous pyelonephritis is a severe, potentially fatal, necrotizing pyelonephritis with a variable clinical picture ranging from mild abdominal pain to septic shock. The majority of cases occur in diabetics with poor glycemic control.⁴⁵ Previous researchers have postulated that vigorous resuscitation and appropriate medical treatment should be followed by immediate nephrectomy.⁶ However
current advances in treatment, allow patients to be treated with Percutaneous drainage in combination with broad spectrum antibiotics.\textsuperscript{[4,7,8]}

Nevertheless, not many articles have focused on the histopathological findings in a nephrectomy specimen with a clinical diagnosis of EPN. The main objective of our case report is to present this rare clinical condition in a male patient with a focus on the histopathological findings.

**CASE HISTORY**

57-year-old male patient, a known diabetic on treatment for the previous 5 years presented with complaints of general weakness, left flank pain and poor stream of passing urine for a week associated with fever and dysuria. Physical examination revealed a conscious patient with pallor and tender left renal angle. Laboratory tests revealed diabetic ketoacidosis (confirmed by arterial blood gas values), blood sugar – 206 mg\%, hemoglobin – 9.5 g/dl, platelet count – 123 x 10\(^3\)/ul, leukocyte count – 24.3 x10\(^9\)/ul with a neutrophilic predominance, serum creatinine – 1.8 mg/dl, sodium- 119 mmol/l, potassium- 2.4 mmol/l.

Aggressive resuscitation was started. Ketoacidosis was controlled but the loin pain persisted. Urine culture of the patient revealed collection contaminated sample.

**Figure 1:** Multiple air pockets were detected in the renal parenchyma of the enlarged left kidney.

**Figure 2:** The nephrectomy specimen measured 16 x 10 x 5 cm. The external surface showed tiny honeycomb-like air-filled spaces.

**Figure 3:** Cut surface shows air filled spaces between areas of necrosis and hemorrhage with blurred cortico-medullary junction.

**Figure 4:** Diabetic nephropathy changes in glomeruli with micro-abscess in right side.
CT KUB: The left kidney was enlarged with perirenal fat stranding and thickened fascias around suggesting pyelonephritis. CT abdomen: Multiple air pockets were detected in the renal parenchyma (more than 2/3rd) of the enlarged left kidney. Air pockets were also detected in the left perinephric region, anterior pararenal space, subhepatic space and in the retroperitoneum (Fig 1). Perinephric fat stranding was noted. The Urology department was consulted and nephrectomy was recommended after DTPA scan showed a left renal function of < 10% and normal right renal function. The patient succumbed to death after 2 days of nephrectomy due to septic shock.

Pathology
Gross: The nephrectomy specimen measured 16 x 10 x 5 cm. The external surface showed tiny honeycomb-like air-filled spaces (Fig. 2). Cut sections showed necrotic, haemorrhagic areas with blurred cortico-medullary differentiation (Fig 3).

Histopathology of the nephrectomy specimen showed features of: (1) emphysematous pyelonephritis: areas of coagulative necrosis and degenerating areas of renal tissue with emphysematous spaces showing no definite lining, surrounded by destroyed renal parenchyma, and micro abscesses (2) diabetic nephropathy: glomerular basement membrane thickening, expansion of the glomeruli with mild mesangial hypercellularity in nodular pattern (Kimmelstiel-Wilson lesions) and periglomerular fibrosis (Fig 4,5,6).

DISCUSSION
Emphysematous pyelonephritis 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. More than 90% of cases occur in diabetics with poor glycemic control. Other predisposing factors include urinary tract obstruction, polycystic kidneys, end stage renal disease and immunosupression. The pathogenesis of EPN remains unclear however four factors have been implicated, including gas-forming bacteria, high tissue glucose level (favoring rapid bacterial growth), impaired tissue perfusion (diabetic nephropathy leads to further compromise regional oxygen delivery in the kidney resulting in tissue ischemia and necrosis; nitrogen released during tissue necrosis) and a defective immune response due to impaired vascular supply. The commonest organism causing EPN is
Escherichia coli. Other bacteria include Klebsiella pneumoniae, Proteus mirabilis and Pseudomonas aeruginosa.\[^{4-7}\]

The mean patient age is 55 years old. Women outnumbered men probably due to their increased susceptibility to urinary tract infections. The left kidney was more frequently involved than the right one.\[^4\] The clinical manifestations according to Huang and Tseng\[^4\] fever was encountered in 79% of the patients, abdominal or back pain in 71%, dyspnoea in 13% and shock in 29%. Laboratory testing revealed leukocytosis in 67%, thrombocytopenia in 46%. This data comes to agreement with those generally reported in the literature.\[^5-7\] Non-contrast CT scan remains the diagnostic method of choice. In addition to showing the presence of gas, it defines the extent of the infection and can diagnose any obstruction.\[^4,5\]

In 1998 Wan et al classified EPN into two types on CT findings. Type I was the classical form with renal parenchymal destruction and presence of diffuse gas in the parenchyma in a streaked or mottled pattern with little or no fluid. Type II was characterized by the presence of fluid (renal or perirenal) with a loculated gas pattern, or gas in the collecting system with acute bacterial nephritis or with perirenal fluid containing abscesses. Type I has a higher mortality rate than type II. Our case had Type I EPN.\[^9\]

The treatment of EPN involves antibiotic therapy followed by surgical intervention (nephrectomy), if the renal parenchyma is extensively destroyed. Grossly the pathological findings of Type I EPN include necrosis, hemorrhagic infarction, and fragile spongy kidney with honeycomb-like gas containing spaces. Microscopically, vasculitis, extensive necrosis, micro abscesses and infarcts, as in our case, are seen. In addition, glomerulosclerosis, Kimmelstiel Wilson nodules, hyalinised arteriolosclerosis, suggestive of diabetic nephropathy (DN) have been documented in nephrectomy specimens of emphysematous pyelonephritis.\[^{10-13}\]

The complete histopathological work up in our case documents diabetic and emphysematous microscopic features in a case of EPN. The mortality in untreated EPN is 100%. With medical treatment alone it decreases to 70%, whereas with combined medical and surgical intervention mortality can be reduced to 30%.\[^9\]

**CONCLUSION**

EPN requires early diagnosis, for which a high index of suspicion is essential in poorly controlled diabetics. CT plays a pivotal role in the diagnosis and staging. Nephrectomy is indicated when medical measures fail. The histopathological findings in emphysematous pyelonephritis are seen to develop in a background of diabetic nephropathy, supporting the association between the two diseases.

**REFERENCES**

3. Huang Kelly HA, MacCallum WG. Pneumaturia. JAMA. 1898; 31:375–381.


