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CASE REPORT

Emphysematous pyelonephritis in a diabetic leading to renal destruction: pathological aspects of a rare case

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Abstract

Emphysematous pyelonephritis is a severe, 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. Urine culture was positive for *Escherichia coli* with a significant colony count. Radiological examination gave a diagnosis of Left Type 1 Emphysematous Pyelonephritis. Inspite 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, but also to focus on the histopathological findings of the same, documentation of which is limited in literature.

INTRODUCTION

Emphysematous pyelonephritis (EPN), a rare renal infection in the form of a necrotizing pyelonephritis, deserves special attention because of its life threatening potential and occurrence in diabetics. However with the extensive use of ultrasound and computed tomography (CT) in the evaluation of patients with symptoms and signs of complicated urinary tract infections, more cases of EPN are being recognized and reported.[1] It is important to distinguish this condition from emphysematous pyelitis (gas limited to the renal excretory system), as the management and the prognosis of the two are different.[2]

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 with a focus on the microscopical findings.

CASE REPORT

A 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 – 18.98 mmol/l, haemoglobin - 9.5 g/dl, platelet count – 160 x 10^9/l, leukocyte count – 24.3 x 10^9/l 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 showed a significant growth of *Escherichia. coli*.

Ultrasound: The left kidney showed loss of corticomedullary differentiation, in the upper 2/3rd. Anechoic focus with multiple discrete hyperechoic foci within it was seen. The right kidney and other organs were unremarkable. *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

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parenchyma (more than 2/3rds) of the enlarged left kidney (Figure 1). Air pockets were also detected in the left perinephric region, anterior pararenal space, subhepatic space and in the retroperitoneum. 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. Serum electrolytes and other renal parameters showed dramatic improvement post-surgery. The patient was discharged after his condition was stable.

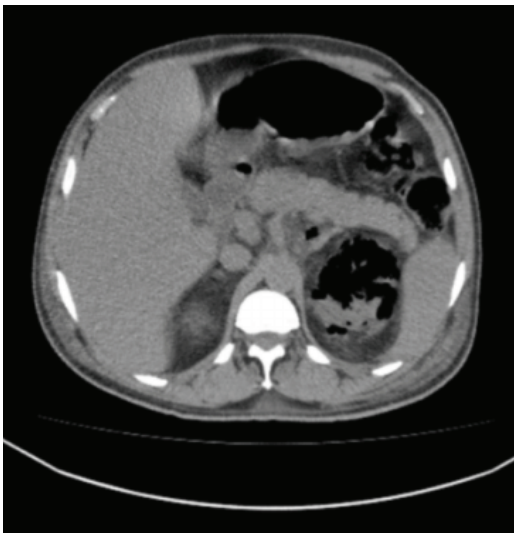

FIG. 1: CT abdomen showing multiple air pockets in the renal parenchyma, perinephric and anterior paranephric spaces.

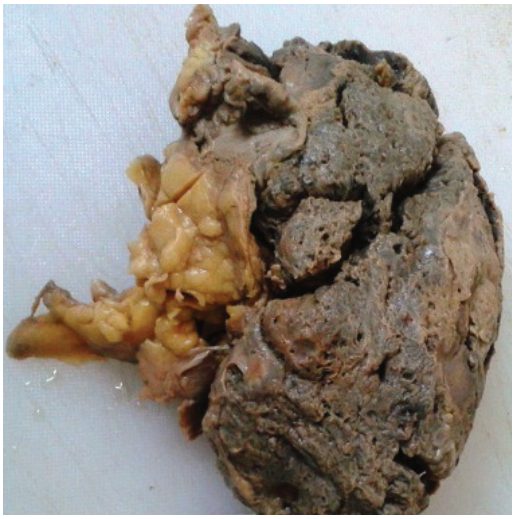

FIG. 2: Nephrectomy specimen showing honeycomb-like air spaces

Pathology

Gross: The nephrectomy specimen measured 10 x 6 x 3 cm. The external surface showed tiny honeycomb-like air-filled spaces (Figure 2). Cut sections showed soft necrotic areas with intact corticomedullary differentiation**.** *Histopathology* of the nephrectomy specimen showed features of: (1) emphysematous pyelonephritis: areas of coagulative necrosis, infarction (Figure 3a), loose degenerating areas of renal tissue, with emphysematous spaces showing no definite lining, surrounded by destroyed renal parenchyma (Figure 3b), and microabscesses, (2) diabetic nephropathy: glomerular basement membrane thickening (Figure 4a), expansion of the glomeruli with mild mesangial hypercellularity in nodular pattern (Kimmelstiel-Wilson lesions) (Figure 4b) and periglomerular fibrosis, (3) Non-diabetic nephropathy: membranous glomerulonephritis (Figure 5a) with epimembranous spikes (arrow) highlighted on silver stain (Figure 5b), features of chronic pyelonephritis and wall thickening of large vessels.

DISCUSSION

The first case of gas forming renal infections was reported in 1898 by Kelly and McCallum. In 1962 Scultz and Klorfein proposed emphysematous nephritis as the preferred designation stressing the relationship between acute renal infection and gas formation. The majority of the cases occured in diabetics with poor glycemic control.[1]

The mean age of patients with EPN is 55 years, with females being six times more commonly affected. The typical symptoms are fever (97%), flank pain (71%), nausea and vomiting (17%), acute renal impairment (35%) and thrombocytopenia (46%), most of which were present in the present case.[3]

Factors in the pathogenesis of EPN includes presence of gas forming bacteria, high levels of glucose in the tissues, impaired tissue perfusion, reduced host immunity and urinary tract obstruction. The causative organism is Escherichia coli in the majority of the cases (as in our case), followed by *Klebsiella pneumoniae*, *Proteus* and *Pseudomonas*.[4]

CT is considered the best imaging technique for the diagnosis and identification of gas (within the renal parenchyma, collecting system, urinary bladder, peri and paranephric space). In 1998 Wan *et* al classified EPN into two types on CT findings. Type 1 was the classical form with

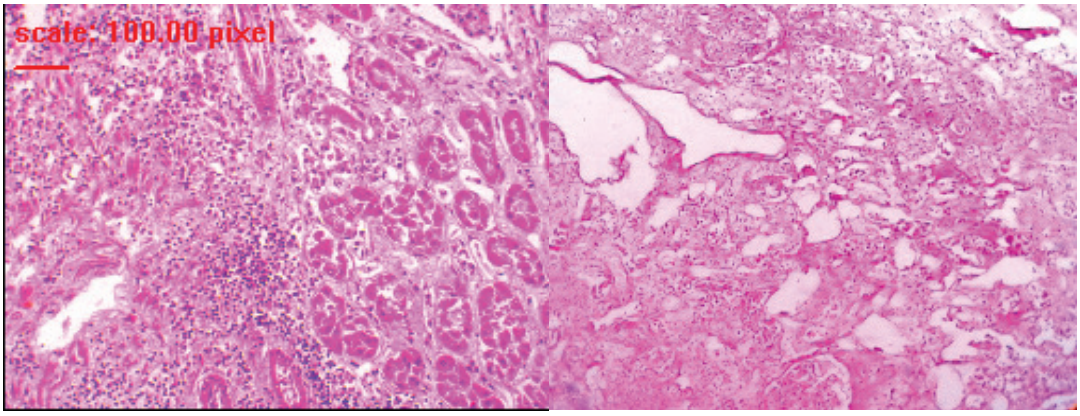

FIG. 3: Histomorphology of emphysematous pyelonephritis. H&E. (A) coagulative necrosis, infarction and microabscess formation, (B) empty emphysematous spaces with adjacent destroyed renal tissue

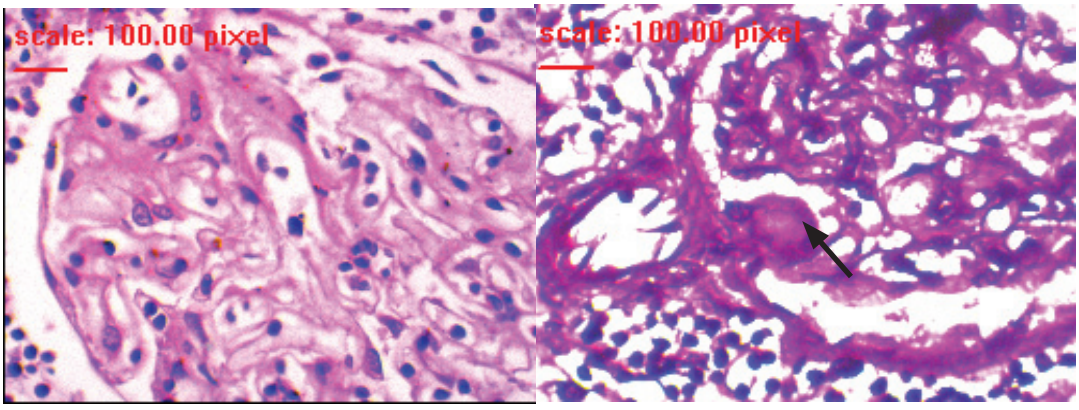

FIG. 4. Diabetic nephropathy (A) thickening of glomerular basement membrane. H&E. (B) Kimmelstiel-Wilson lesion (arrow). PAS.

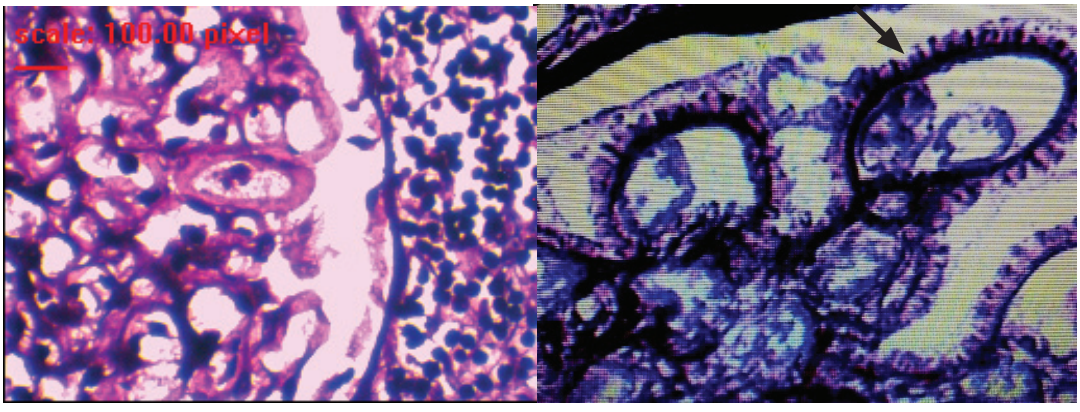

FIG. 5: Non-diabetic renal lesions. (A) membranous glomerulonephritis with thick capillary walls, PAS (B) Epimembranous spiking of the glomerular capillary basement membrane (arrow) on Silver stain – oil immersion

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 1 has a higher mortality rate than type II.[5] Our case had Type I EPN.

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 1 EPN include necrosis, hemorrhagic infarction, and fragile spongy kidney with honeycomb-like gas containing spaces. Microscopically, vasculitis, extensive necrosis, microabscesses and infarcts, as in our case, are seen. Type II EPN is characterized by infiltration of acute and chronic inflammatory cells, abscess formation and necrosis. The mottled gas pattern and absence of exudative response in patients with type 1 EPN may reflect a defective immune response in the host, while the presence of exudates in patients with type II EPN suggests better host immunity and a more favorable prognosis.[6,7]

In addition, glomerulosclerosis, Kimmelstiel-Wilson nodules, hyalinised arteriolosclerosis, suggestive of diabetic nephropathy (DN) have been documented in nephrectomy specimens of emphysematous pyelonephritis.[8] However, DN is not the only renal abnormality found in diabetics. Microscopic features like membranous glomerulonephritis, chronic pyelonephritis, infarcts, thrombosis (as in our case) can also be seen in diabetics.[9] The complete histopathological work up in our case documents diabetic, non 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%.[5] *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 non-specific, but are seen to develop in a background of diabetic nephropathy, supporting the association between the two.

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