Emphysematous Pyelonephritis—a Rare Surgical Emergency Presenting to the Physician: A Case Report and Literature Review

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Abstract Emphysematous pyelonephritis is an acute necrotizing parenchymal and perirenal infection caused by gas-forming uropathogens. It is a rare condition, usually occurring in diabetic patients. Mortality rates in medically managed patients are as high as 70–90 %. It should be suspected in diabetic patients with urinary tract infections and worsening of renal function. CT scan is diagnostic and is the method of choice for diagnosis and follow-up. Both physicians and surgeons should be aware of this rare condition which might present to the physician as fulminant urinary tract infection in an uncontrolled diabetic patient, but which might warrant urgent surgical intervention by way of an emergency nephrectomy. We report a 60-year-old diabetic woman who presented with urinary infection and sepsis. Initially she was managed conservatively, but had to be taken up for emergency nephrectomy in view of her worsening condition. We review the clinical presentation, radiological diagnosis with characteristic CT scan pictures, and the management of this rare condition.

Keywords Diabetes complications · Emphysematous pyelonephritis · Nephrectomy · Urinary infections

Introduction

Emphysematous pyelonephritis is a rare acute necrotizing parenchymal and perirenal infection caused by gas-forming

uropathogens. Occurring usually in diabetic patients, mortality rates in medically managed patients are as high as 90 % [1]. Both physicians and surgeons should be aware of this rare condition which might present to the physician as fulminant urinary tract infection in an uncontrolled diabetic patient, but which might warrant urgent surgical intervention by way of an emergency nephrectomy.

Case Report

A 60-year-old woman, a known diabetic for 12 years on irregular treatment with oral hypoglycemics, presented to the emergency department with pain in the left hypochondrium for 4 days, associated with high-grade fever, dysuria, and pyuria. She was an obese, disoriented and febrile, with tachycardia, facial puffiness, and bilateral pedal edema. Her abdomen was distended with tenderness in the left lumbar region and renal angle. Her investigations revealed a random blood sugar of 278 mg%, blood urea 85 mg%, serum creatinine 2.6 mg%, total leukocyte count 14,200/mm³, and a platelet count of 18,000/mm³. Urine examination was positive for sugar and ketones, and it showed proteinuria, RBCs, pus cells, and casts. An ultrasound of the abdomen showed increased cortical echogenicity of the left kidney, with a heterogenous area with gas seen in the upper and middle portions suggestive of emphysematous pyelonephritis. There was no hydronephrosis or pyonephrosis, or free fluid in the peritoneal cavity. A CT scan of the abdomen revealed parenchymal destruction in the left kidney with presence of streaky or mottled gas radiating from the medulla to the cortex, characteristic of emphysematous pyelonephritis type 1. There was no evidence of obstruction or hydronephrosis (Fig. 1).

With a diagnosis of diabetic ketoacidosis with left emphysematous pyelonephritis, the patient was resuscitated with antibiotics, intravenous fluids, insulin, packed cells, and platelet-

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Fig. 1 Transverse and coronal sections of the CT scan showing parenchymal destruction of the left kidney with presence of gas characteristic of emphysematous pyelonephritis

rich plasma. She was initially managed conservatively in view of the ketoacidosis, deranged renal functions, and low platelets. She showed progressive signs of recovery, and by day 7, she was fully conscious and oriented, ambulatory, and accepting orally. Her hematological and renal parameters returned to normal limits. A renal scan showed a non-functioning left kidney and normal renal function on the right side.

On the eighth day, however, the patient's condition started to deteriorate rapidly. She became febrile, drowsy, disoriented, with tachycardia and tachypnea. She also had left flank pain and tenderness. Her blood sugar rose to 378 mg%, and her total leukocyte count rose to 18,000/mm³; however, her platelets and renal parameters remained within normal limits. The patient was subjected to an emergency left nephrectomy. Her postoperative recovery was uneventful, and she was discharged on the eighth postoperative day with normal blood and renal parameters.

Discussion

Emphysematous pyelonephritis (EPN) is a rare acute necrotizing parenchymal and perirenal infection caused by gas-

forming uropathogens occurring in adult diabetic patients. Of such patients, 96 % have diabetes mellitus and 29 % have urinary tract obstruction. Juvenile diabetic patients do not appear to be at risk. Women are affected more often than men in the ratio of 4:1 [2].

The usual clinical presentation is severe, acute pyelonephritis; although sometimes, a chronic infection precedes the acute attack. Patients invariably display the classic triad of fever, vomiting, and flank pain [3]. Pneumaturia is absent unless the infection involves the collecting system. Urine cultures are invariably positive with *Escherichia coli* or *Klebsiella* [2].

The diagnosis is established radiographically. On abdominal radiographs, tissue gas in the renal parenchyma may appear as mottled gas shadows over the involved kidney. This finding is often mistaken for bowel gas. A crescentic collection of gas over the upper pole of the kidney is more distinctive. As the infection progresses, gas extends to the perinephric space and retroperitoneum. This distribution of gas should not be confused with "emphysematous pyelitis" in which air is in the collecting system of the kidney secondary to a gas-forming bacterial UTI, often occurring in nondiabetic patients, is less serious, and usually responds to antimicrobial therapy.

Excretory urography is rarely of value in emphysematous pyelonephritis because the affected kidney is usually non-functioning or poorly functioning. Because of the significant risk of contrast nephropathy in critically ill, dehydrated diabetic patients with abnormal renal function, retrograde pyelography rather than excretory urography is advisable to demonstrate obstruction. Obstruction is demonstrated in approximately 25 % of the cases. Ultrasonography usually demonstrates strong focal echoes suggesting the presence of intraparenchymal gas [4].

CT is the imaging procedure of choice in defining the extent of the emphysematous process and guiding management [5, 6]. Two subtypes of EPN based on CT appearances have been described. Type I EPN (33 % of patients) is characterized by parenchymal destruction with either absence of fluid collection or presence of streaky or mottled gas radiating from the medulla to the cortex. A crescent of subcapsular or perinephric gas may be present. The absence of fluid collection implies a poor immune response. The mortality rate is high, at 66 %. Type II EPN (66 % of patients) typically has a confined, bubbly, intrarenal gas pattern—probably within abscesses associated with renal and perinephric fluid collection—and gas within the renal pelvis. The mortality rate in type II is 18 % [7].

A nuclear renal scan should be performed to assess the degree of renal function impairment in both kidneys.

In a meta-analysis representing 175 patients, conservative treatment alone, bilateral emphysematous pyelonephritis, type I emphysematous pyelonephritis, and thrombocytopenia were associated with increased mortality, as also were systolic

blood pressure less than 90 mmHg, serum creatinine greater than 2.5 mg/dl, and disturbance of consciousness [8].

In another analysis of 10 retrospective studies on 210 patients, the mortality from medical management alone was 50 %, medical management combined with emergency nephrectomy was 25 %, and medical management combined with percutaneous drainage was 13.5 % [2].

Emphysematous pyelonephritis is a surgical emergency. Most patients are septic, and fluid resuscitation and broad-spectrum antimicrobial therapy are essential. If the kidney is functioning, medical therapy can be considered [5, 6]. Nephrectomy is recommended for patients who do not improve after a few days of therapy [8]. If the affected kidney is not functioning and not obstructed, nephrectomy should be performed because medical treatment alone is usually lethal. If a kidney is obstructed, catheter drainage must be instituted. If the patient's condition improves, nephrectomy may be deferred pending a complete urologic evaluation. Although there are isolated case reports of retention of renal function after medical therapy combined with relief of obstruction, most patients require nephrectomy [2].

Growth of the diabetic population warrants heightened attention to these potentially fatal infections which might require urgent surgical intervention. They should be suspected in diabetic patients with urinary tract infections and worsening of renal function. CT scan is diagnostic and is the method of choice for diagnosis and follow-up. Both physicians and surgeons should be aware of this rare condition

which might present to the physician as fulminant urinary tract infection in an uncontrolled diabetic patient, but which might warrant urgent surgical intervention by way of an emergency nephrectomy.

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