A Rare Case of Emphysematous Pyelonephritis within a Horseshoe Kidney

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INTRODUCTION

Emphysematous pyelonephritis is an acute severe necrotizing infection of the renal parenchyma; it causes gas formation within the collecting system, renal parenchyma, and/or perirenal tissues. It is caused by gas forming organisms E coli (68%), Klebsiella spp (9%) and Proteus mirabilis most commonly. It typically occurs in immunocompromised patients especially diabetics (85–90%) and patients with obstructive uropathy (3). Diagnosis is made radiologically and clinically. It is bilateral in approximately 10% of cases (8). Radiologically, it is recognized by the presence of gas either within the renal parenchyma, the collecting system or perinephric tissues. This is seen as lucencies on plain radiographs, high amplitude echoes on ultrasound, mottled areas of low attenuation on CT and signal void on T1WI and T2WI on magnetic resonance imaging (MRI).

Historically, diagnosis was made based on plain abdominal Xrays and intravenous drip infusion urograms (4).

This report is of a case of a 47-year old woman with extensive emphysematous pyelonephritis within an incidentally de-tected horseshoe kidney.

CASE REPORT

A 47-year-old diabetic and hypertensive woman presented to the emergency room with right flank pain, fever, chills, nausea, vomiting and abdominal distension. Physical examination revealed right upper quadrant and flank tenderness with bibasal lungs crepitations.

Vitals signs including her temperature (T 36° C) were normal, however her random blood glucose was elevated at 254 mg/dL. Laboratory investigations revealed a white blood cell count of 18.1 X 10⁹/L (normal 4–11 X10⁹), haemoglobin of 5.9 (normal 15.5–18.8) g/dL), platelets of 44 X 10⁹/L (normal 150–400 X 10⁹/L) with features of renal failure: urea 79 mg/dL and creatinine 4.4.mg/dL Urinalysis showed 3⁺ glucose, no ketones, 2⁺ nitrites, no blood red cells, no white blood cells.

Transabdominal sonography done one week prior to presentation only showed an echogenic focus in the lower pole of the left kidney which was suggested to be a calculus; no hydronephrosis and chronic renal parenchymal changes were seen. Plain abdominal radiographs were not requested.

From: Department of Radiology, Eric Williams Medical Sciences Complex, Trinidad and Tobago, West Indies. A non-contrast enhanced computed tomography (CT) scan using the renal calculi protocol was done of the abdomen and pelvis and revealed multiple air pockets that had almost replaced the normal renal parenchyma of a horsehoe kidney (right moiety > left) with no calculi visualized. Gas was noted to track into the perirenal tissues with perinephric fat stranding also noted. A diagnosis of emphysematous pyelonephritis in a horseshoe kidney was made and intravenous antibiotics were commenced namely ceftazidime, metronidazole and levofloxacin.

Supportive care was also provided with insulin for blood glucose control, transfusion of platelets, fresh frozen plasma and packed red blood cells to correct the thrombocytopenia and anaemia and she had dialysis for deteriorating renal function.

She improved gradually over two weeks and was discharged on oral antibiotics.



Fig. 1: Scout image from the initial CT shows gas densities in the region of the right renal outline. This is difficult to differentiate from the normal intestinal gas pattern.

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Fig. 2: Ultrasound images (a) Transverse view of the right kidney and (b) longitudinal view of the right kidney show irregular areas of increased echogenicity likely representing the pockets of air seen on CT imaging.



Fig. 3: Axial CT images of the abdomen at the level of fusion of the renal moieties in the horseshoe configuration. Renal emphysema is seen with pockets of air in the perinephric tissues and right perirenal fat stranding, findings in keeping with Type 2 emphysematous pyelonephritis.



Fig. 4: Coronal CT images of the abdomen (a) at the level of renal fusion showing renal emphysema with perirenal pockets of air (b) shows the right renal parenchyma almost completely replaced by gas, with suprarenal extension of the emphysematous pyelonephritis.

DISCUSSION

The first case of pneumaturia was reported in 1898 by Kelly and MacCullum. Since then approximately 200 cases of emphysematous pyelonephritis have been reported in the United States of America (3). Clinically, patients with emphysematous pyelonephritis present with chills, fever, flank pain, lethargy, confusion and not responding to antibiotics. A palpable mass was reported in 45% of cases (1). Positive blood and urine cultures, urosepsis, septic shock and pyrexia of unknown origin are also features.

The majority of patients are diabetic (85–95%) and may present with uncontrolled hyperglycaemia, acidosis, dehydration and electrolyte imbalances. In those without diabetes, ureteral obstruction is common and normally due to a stricture or aberrant blood vessel.

In this index case, the patient was diabetic and also had the congenital anomaly of a horseshoe kidney with the inferior poles fused by parenchyma. Despite the abnormal renal configuration, the two ureters were noted to course normally inferiorly toward the urinary bladder with no sites of obstruction.

A horseshoe kidney is the most common congenital fusion abnormality of the kidney but is however still quite rare occurring in approximately 1 in 400–800 live births. It is even much less common to see emphysematous pyelone-phritis in this anomaly, with only one documented case being published (2). On review of the literature, a horseshoe kidney is associated in 50% with caudal ectopia, vesico-ureteral reflux and hydronephrosis secondary to pelvi-ureteric obstruction. Hydronephrosis or caudal ectopia was not seen on the images acquired and no further testing such as voiding cysto-urethrography or radionuclide cystography was done to determine the presence of vesico-ureteric reflux which would predispose to a urinary tract infection.

A horseshoe kidney is typically fused at the lower poles (90%) as seen in the index case and less commonly at the upper poles (10%). The renal long axis is medially oriented with the isthmus at the L4–L5 level and the renal pelves and ureters situated anteriorly. This is also associated with multiple renal arteries including the isthmus artery and with a number of other abnormalities such as genitourinary (hypospadia, undescended testis, bicornuate uterus), cardiovascular, skeletal and central nervous system anomalies and Turner's syndrome. It is often complicated by infection, as was seen in the index case, and by renal calculi. There are two types of emphysematous pyelonephritis; Type 1 involves the cortical and medullary interstitium of the kidney with no fluid collections and Type 2 involve the renal parenchyma with gas in the collecting system and renal/ perinephric fluid collections.

The pathogenesis of emphysematous pyelonephritis has not been clearly established. Because it occurs most frequently in diabetics, it is thought that the high levels of glucose provides a suitable medium for organisms to ferment the glucose into carbon dioxide. Gas production may also result from the fermentation of necrotic tissue (1).

Emphysematous pyelonephritis is associated with a very poor prognosis, Type 1 (33%) and Type 2 (66%), and prompt radiological diagnosis is necessary to allow treatment. Treatment options classically include nephrectomy with antibiotic therapy, though current trends are towards less invasive percutaneous drainage procedures and relief of precipitating factors. In the index case, medical treatment with antibiotic coverage alone was opted for as the patient had a

horseshoe kidney which would have resulted in excess blood loss because of the fused kidneys and also she was a poor surgical candidate: severely anaemic, thrombocytopenic and in renal failure. Typically, patients do poorly on this type of conservative management with a mortality of 60% with antibiotic therapy alone. This decreases to 30–50% when surgical intervention occurs as well. Extension into the perinephric space as in the index case carries an even worse prognosis at 80% typically (8).

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