Case Reports

A Young Diabetic Lady with Emphysematous Pyelonephritis

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Abstract:

A rare and potentially life-threatening condition is reported. A young diabetic lady presented with septic shock and features of paralytic ileus. A plain abdominal radiograph suggested "megacolon with gas-fluid levels". But high degree of clinical suspicion persuaded us to perform a CT scan of abdomen, which revealed emphysematous pyelonephritis. This enabled us to start early effective treatment and eventually save the patient.

Keyword: Emphysematous Pyelonephritis, Diabetes mellitus

Introduction:

The first case of gas forming renal infection was reported in 1898. Since then, many names, such as "renal emphysema", "pyelonephritis emphysematousa" and "pneumonephritis", have been used to describe this disease until 1962, when Schultz and Klorfein suggested the term "Emphysematous pyelonephritis" as the preferred designation name, because it stresses the relationship between acute infectious process and gas formation.¹

Emphysematous pyelonephritis is an acute severe necrotizing infection of the renal parenchyma and its surrounding tissues that results in the presence of gas in the renal parenchyma, collecting system or perinephric tissue. It deserves special attention because of its lifethreatening potential.² A high index of suspicion is required as the usual presenting features resemble pyelonephritis.^{1,2} We report such a case, who presented with multiple adverse prognostic factors.

Case report:

In June, 2011, a 20-year-old diabetic lady presented to the emergency department with shock, preceded by fever, vomiting, abdominal pain, and constipation for a week and oliguria for one day. On admission, she had a low Glasgow Coma Scale (12/15), fever (38·3°C), and was hypotensive (80/40 mmHg) with tachycardia (114 beats/min). Examination of cardiovascular and respiratory systems was unremarkable. Abdomen was distended, tender over the right loin and renal

angle, crepitus was felt over the right lumber region, and bowel sound was not audible. Random blood glucose was 23.6 mmol/L and urine dipstick showed leukocytes and glucose, but no nitrates or ketones. She was resuscitated with intravenous fluids, and intravenous insulin was administered. Intravenous antibiotics (meropenem and metronidazole) were commenced after collecting blood and urine for routine labs and culture. Despite all efforts, she failed to respond, and was admitted to the Intensive Care Unit. Noradrenalin was required to raise her blood pressure. A clinical diagnosis of acute pyelonephritis with sepsis with paralytic ileus was made.

Important laboratory tests showed leukocytosis (20.8 K/iL) with neutrophilia (91.9%), platelet count $214 \text{ K/}\mu\text{L}$, C-reactive protein >270 mg/L, serum creatinine 159.1 imol/L, sodium 130 mmol/L, potassium 2.9 mmol/L, chloride 97 mmol/L, TCO₂ 21 mmol/L, and HbA1C 10.8%. A plain x-ray of abdomen in erect posture showed gas shadow resembling "megacolon" with gas-fluid levels (Figure A).

But the presence of crepitus on the renal area persuaded us for a CT-scan of abdomen, and it revealed right sided emphysematous pyelonephritis of class 3B (Huang and Tseng¹) (Figure B). Urgent percutaneous incision and drainage was performed, releasing "foul smelling" gas and pus.

Blood, urine, and pus cultures were positive for *E. coli*. Antibiotics were adjusted according to sensitivity. Glycaemic control achieved with subcutaneous insulin. Right sided

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simple nephrectomy was performed a week later, to remove the remaining pus and necrosed tissues, including whole right kidney, perinephric fat, and part of right adrenal. After two weeks of hospital stay and completion of antibiotics course, she was discharged with advice on strict glycaemic control with subcutaneous insulin. On her first follow-up visit, two weeks later, she was well with no further complaint.



Fig.-A: Plain X-ray of abdomen in erect posture showing gas shadow resembling "megacolon" with gas-fluid level and gas under the skin and within subcutaneous and intramuscular planes on the right side. There are multiple small gas-fluid levels on the left also.



Fig.-B: *CT* scan of abdomen showing architecturally distorted right kidney largely replaced by gas. Gas is also seen in right perinephric and intra-peritoneal spaces, subcutaneous and intramuscular planes.

Discussion:

Emphysematous pyelonephritis (EPN), a rare, acute necrotizing, gas forming infection of the renal parenchyma and surrounding tissues, occurs almost exclusively in diabetics (H"95%).^{1,2} Females suffer approximately six times more, mostly in their fourth or fifth decade; from this potentially life-threatening condition.² Commonest agent reported is *E. coli*, usually isolated from urine or pus.¹⁻³ *Proteus mirabilis, Klebsiella pneumoniae*, Group D *Steptococcus* and coagulase-negative *Staphylococcus*, and in rare cases, anaerobic micro-organisms including *Clostridium septicum, Candida albicans, Cryptococcus neoformans* and *Pneumocystis jirovecii* have also been reported as the causative pathogens for EPN.²

Various factors in the pathogenesis of this condition have been suggested. They include high levels of glucose within the tissues, presence of gas-forming micro-organisms, impaired vascular blood supply, reduced host immunity and the presence of obstruction within the urinary tract. A high level of tissue glucose in association with impaired blood supply to the kidneys, which is prevalent in patients with diabetes, facilitates the process of anaerobic metabolism.²

EPN requires a high index of suspicion as the usual presenting features (fever, rigours, nausea, vomiting, flank pain, and dysuria) resemble pyelonephritis. Acute renal dysfunction, acid-base disturbances, impaired consciousness, or rapid progression to septic shock, also can be the initial presentation.^{1,2} Unusual presentations with subcutaneous emphysema, pneumoperitoneum and pneumomediastinum have also been reported.^{3,4} Tender loin and crepitus around the renal area may be the only physical finding.²

EPN is a radiological diagnosis. A plain radiograph usually shows an abnormal gas shadow in renal bed, whereas an ultrasonography or CT will confirm the presence of intrarenal gas. CT-scan is more sensitive and defines the extent of parenchymal destruction.²

Huang and Tseng classified EPN according to the radiological findings on CT scan, into the following classes: (1) class 1: gas in the collecting system only; (2) class 2: gas in the renal parenchyma without extension to extra-renal space; (3) class 3A: extension of gas or abscess to perinephric space; class 3B: extension of gas or abscess to pararenal space; and (4) class 4: bilateral EPN or solitary kidney with EPN.¹ It is important for the treating physician to note the classification, as it helps in planning the management, which could be either conservative or surgical.

EPN was associated with a mortality rate of up to 78% until the late 1970s but, over the last two decades, improvement in management techniques has reduced the mortality rate to 21%.² Mortality from EPN is primarily attributable to septic complications. Systolic BP <90 mmHg, impaired consciousness, and raised serum creatinine level, thrombocytopenia, and bilateral EPN are associated with higher mortality. Medical management only is also associated with poor outcome.^{2,3}

Correction of fluids and acid-base imbalance, commencement of antibiotics, and good glycaemic control are the mainstay of resuscitation. Current treatment strategy is a nephronsparing approach with percutaneous drainage. But, in patients with extensive EPN, signs of organ dysfunction or with two or more risk factors, early nephrectomy along with medical management reduce the mortality rate and shorten the recovery period.^{2,3,5} Early diagnosis and successful management improved outcome in our patient despite multiple poor prognostic factors.

Conclusion:

High level of clinical suspicion is important to clinch diagnosis of rare, potentially life-threatening conditions. Early and correct recognition is essential for appropriate management of such diseases, like emphysematous pyelonephritis.

Conflict of Interest: None declared.

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