Emphysematous pyelonephritis is a rare, life endangering gas-forming suppurative infection of the renal parenchyma and perirenal spaces. Gas in the renal substance, perirenal area, bladder wall or the lumen of the collecting system may result from infection, penetrating trauma, fistulous communication with the intestinal tract and various diagnostic and surgical procedures. Pathogenic aerobic organisms such as Escherichia, Klebsiella, and Aerobacter and very rarely anaerobic organisms like Clostridium perfringens are known to cause gas-forming infections of the urinary tract. Diabetes mellitus and obstructive uropathy are the commonest predisposing factors. Radiological evidence of gas in the renal parenchyma or in the bladder wall, in a patient presenting with severe prostrating illness with or without urinary tract obstruction or Diabetes mellitus confirms the diagnosis of this entity. Whereas emphysematous pyelonephritis carries a grave prognosis, emphysematous cystitis has earned a good reputation as a benign disease.

Awareness of this disease entity and adequate management of Diabetes mellitus and urinary tract infection would help in curtailing the high mortality. Hence, we present an interesting case of emphysematous pyelonephritis and cystitis.

CASE REPORT

A 60-year-old Saudi female patient was admitted in hospital, complaining of left flank pain, nausea, vomiting, and rigors associated with suprapubic pain, increased frequency of urine, urgency, and dysuria for ten days.

Past medical history was significant for Diabetes mellitus and hypertension of 15 and 10 years duration, respectively. She was on glibenclamide, metformin, nifedepine retard and lisinopril. She was also recently prescribed a short course of co-amoxiclav by a general practitioner. The clinical examination showed an old, obese, drowsy, ill and toxic looking lady. She was febrile having temperature of 38.5°C, blood pressure of 135/75 mm Hg, and pulse rate of 126/minute. She was tachypneic with respiratory rate of 22/minute. Abdominal examination found diffuse tenderness, and hypoactive bowel sounds but no guarding, rebound tenderness or visceromegaly. Lungs were clear to auscultation, and cardiovascular examination was unremarkable.

Laboratory investigations showed white blood cells of 18000 cells/microliter, with a differential count of 89% neutrophils, and a hemoglobin of 11.6 g/dl. Renal function tests revealed serum creatinine of 226 umol/L, urea of 19.5 mmol/L, serum sodium was 129 mmol/L and serum potassium was 3.9 mmol/L. She was hypoalbumenic as well with a serum albumin of 28 g/L. Urine dipstick showed glucose++, blood++, protein++ and bacteria++. Urine microscopy revealed > 100 pus cells and 10 to 25 red blood cells/high power field.

Blood and urine samples for culture and sensitivity were obtained and patient was empirically commenced on intravenous cefotaxime along with fluids. After 72 hours, the urine culture grew Escherichia coli, which was sensitive to gentamicin, ciprofloxacin and aztreonam. As fever persisted, injection cefotaxime was replaced with aztreonam.

Ultrasound of abdomen was performed which showed an enlarged left kidney, mildly dilated pelvicalyceal system and multiple hyperechoic lesions giving dirty shadows, which were confused with renal stones.
So non-contrasted computerized tomography was done, which revealed gas in the left renal parenchyma (Figure 1) and an air-fluid level in the urinary bladder (Figure 2).

The diagnosis of emphysematous pyelonephritis, and emphysematous cystitis was made. The patient was continued on intravenous aztreonam for two weeks and later received ciprofloxacin orally for another two weeks along with anti-hypertensive agents and insulin to control her diabetes. After one week, she improved, became afebrile, and white blood cells came to normal range. She was discharged home after 30 days of admission and a repeat ultrasound of abdomen two months later was found to be normal.

**DISCUSSION**

Emphysematous pyelonephritis is defined as the presence of gas in the renal parenchyma and is often associated with perirenal gas. It is severe life-threatening necrotizing infection with mortality rates in medically managed patients reported to be as high as 70-90%. Ninety percent of cases are associated with Diabetes mellitus with mean age of 54 years and women are affected twice as often as men. Bilateral involvement in association with Diabetes has been rarely described.

The clinical presentation is often suggestive of severe acute pyelonephritis, but it may have an indolent course over several months. Nausea and vomiting (40%), abdominal pain in (55%) are common symptoms and fever is found in about 80% of the cases. A palpable mass is rare, abdominal tenderness is common and pneumaturia is not found unless there is coincidental emphysematous cystitis. Neutrophil leukocytosis is observed in the majority of cases and pyuria is found in 96%. Microbiologic investigation show *Escherichia coli* to be the most common causative organism. In one series, 68% of cases were due to this organism and 9% due to *Klebsiella* and multiple organisms were found in 14%. Other organisms are rare and many are reported as a single case: *Candida albicans*, *Candida tropicalis*, *Cryptococcus* and *Anaerobic streptococcus* have been reported rarely.

Renal emphysema is a radiological diagnosis as the symptoms and signs are little different from other renal infections. The diagnosis should be suspected in any diabetic patient with nausea, vomiting and abdominal pain, particularly when antibiotic therapy fails to improve symptoms in 3 to 4 days.

In gas-forming urinary tract infections, plain abdominal X-ray or intravenous urography reveals gas in 50-80% of cases. If gas is present on plain films, CT scanning should be performed to better define the extent of the infection and any associated obstructing lesions in the genitourinary tract.

Huang et al. classified gas-forming infections into 4 prognostic classes based upon CT scan findings as follows: Class 1: gas in the collecting system only (i.e. emphysematous pyelitis), Class 2: gas in the renal parenchyma, Class 3A: extention of gas or abscess to the peri-nephric space, Class 3B: extention of gas or abscess to the para-nephric space and Class 4: bilateral emphysematous pyelonephritis or solitary kidney with emphysematous pyelonephritis. Based upon the treatment and outcomes of 48 patients, the same authors made the following recommendations. Antibiotic plus percutaneous catheter placement are sufficient for patients with class 1 or 2. Antibiotic plus percutaneous catheter placement is the initial treatment of choice for patients with class 3A or 3B disease without organ dysfunction. Antibiotic plus immediate nephrectomy is needed for patients with class 3A or 3B with organ dysfunction. Bilateral percutaneous drainage is needed for patients with class 4 disease. All failures with percutaneous drainage should proceed to nephrectomy. The overall mortality was 18.8% (9 cases of 48). Age, gender and blood glucose level were not associated with mortality. Patients who initially presented with organ system dysfunction had worse outcomes.

Emphysematous pyelitis is characterized by gas occurring solely in the renal collecting system. Evanoff et al. found this condition to be more common in women (3:1) and 59% of subjects were diabetics, presumably due to higher proportion of patients with obstruction in this group (64% versus 37%). For reasons that are not clear the left kidney was more affected than the right one (53% versus 36%) and bilateral gas was rarely observed. The patients have similar symptoms to those in emphysematous pyelonephritis. *Escherichia coli* is again the most common organism. Radiology reveals gas in the collecting system and demonstrates associated obstruction. Gas occurs rarely in the bladder in association of emphysematous pyelitis. Antibiotics alone can be successful if no obstruction is present, and when it is present, it must be corrected, the reported mortality was 18%. The early description of pneumaturia in the absence of fistulous communication or instrumentation must have referred to this condition.

Prior to the review of Baily, gas production was arbitrarily described as primary pneumaturia (46 cases) or cystitis emphysematosa (52 cases). Baily who
considered those to be the same entity, added another 19 cases to the literature. Diabetes mellitus was recorded in 29/46 cases and 13/19 cases, or in total 51% of the combined series. Women were twice more common than men. In this series, mean age was 54 (range 25-83) years and the mean duration of diabetes was 14 years. The clinical presentation can include typical symptoms of urinary tract infection: frequency, nocturia, and dysuria, in addition to suprapubic pain, some patients will present with gross haematuria. However, the patients may present with nausea, vomiting, altered mental state, and fever, features of systemic sepsis and hypotension, sometimes with an acute abdomen. The commonest infecting organism. Enterobacter is relatively common, and there are occasional reports of Proteus species, Staphylococcus aureus, Streptococcus species and yeasts. Abdominal radiography may show a radiolucent line around the bladder wall or gas within the bladder, an air-fluid level can be seen within the bladder. Computed Tomography (CT) scans can clearly show gas within and around the bladder as well as the extent of gas collection. The treatment of this rare condition is by controlling Diabetes mellitus, systemic administration of antibiotics and adequate drainage. The prognosis is usually good. The cause of gas formation (carbon dioxide and hydrogen) is not well-understood. One proposal is that gas is formed at mixed acid fermentation of a glucose-rich substrate in patients with severe necrotizing infection, the resultant necrotic tissue may then act as a substrate for gas formation, and the gas then becomes entrapped by an obstructive process. The pathogenesis probably involves several factors: gas-forming bacteria, enhanced proliferation of microorganisms due to altered immune defences, vascular impairment with ischemia or infarcts, and a high tissue glucose concentration. The risk factors in this patient were female gender, diabetes mellitus, and the growth of gas-producing organism (E.coli). The diagnosis of emphysematous pyelonephritis and emphysematous cystitis was suspected initially by ultrasound and confirmed by CT scan of the abdomen along with the growth of E.coli. The prolonged course of antibiotics was sufficient to cure the infection in this patient.

REFERENCES