Pneumaturia, Can Be A Clue to A Rare Disease; An Emphysematous Cystitis Case Presentation and Review of Literatures

Dileep Sugathan Kovilazhikam, Fakhriya Alalawi*, Thouqeer Ahmed, Mohammed J. Railey, Amna Alhadari
Department of Nephrology, Dubai Hospital, Dubai Health Authority, Dubai, United Arab Emirates

*Corresponding author: Dr. Fakhriya Alalawi, Department of Nephrology, Dubai Hospital, Dubai Health Authority, Dubai, United Arab Emirates. E-mail: fjalalawi08@yahoo.co.uk


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Abstract

Emphysematous cystitis is a rare type of infection of the bladder wall by gas forming bacteria, commonly E. coli or fungi. Pneumaturia which tends to be ignored by most physicians should raise the possibility of emphysematous cystitis, particularly in elderly, diabetics and immunocompromised individuals.

In this article, we present a diabetic patient who presented only with pneumaturia and was diagnosed to have an Emphysematous cystitis.

Keywords: Pneumaturia, Emphysematous Cystitis (EC), diabetes mellitus (DM), CKD (chronic kidney disease), UTI (urinary tract infections).

Introduction

Emphysematous cystitis is a rare type of infection of the bladder wall by gas-forming bacteria or fungi. Despite disease rarity, it remains the most common type of all gas-forming bladder infections. The disease most commonly affects elderly diabetics and immunocompromised patients.

Case Report

64-year-old male with history of type-2 diabetes mellitus for more than 10 years and hypertension for more than 5 years. Patient had history of recurrent urinary tract infection, mostly with E-coli, which was treated with appropriate antibiotics. He was on follow up with urologist for benign prostatic hypertrophy and renal stones and underwent shock wave lithotripsy on several occasions. Additionally, he is a chronic smoker.

Patient was diagnosed to have chronic kidney disease (CKD) stage 3 secondary to diabetic nephropathy in 2012. He had gradual decline in renal functions till he reached CKD stage 4 in 2018.

In Jan 2019 was admitted electively for further evaluation of kidney status as his serum creatinine had increased from 3.6mg/dl to 5.7mg/dl over 1-month period. He had no fever; however, he gave history of dysuria with passing air while voiding. His lab tests were shown in the table.

In view of worsening chemistry, patient underwent urgent ultrasound KUB (image 1), reported as normal size kidneys with increased parenchymal echogenicity. Corticomedullary differentiation is partially maintained with normal Parenchymal thickness on both sides; however, there were numerous non-shadowing echogenic foci within the renal sinuses on both side and a lower calyx calculus on the left side, measuring 7.3 x 7 mm, with no hydronephrosis. Urinary bladder shows thickened heterogeneous wall with multiple echogenic foci showing posterior dirty shadowing suggestive of Emphysematous cystitis. The prostate is border line in size with Post void residue of 138 cc.

His lab investigations are shown in the table:

<table>
<thead>
<tr>
<th></th>
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<th>Ref. Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calcium</td>
<td>9.4</td>
<td>Ref Range: 8.9 - 10.2 mg/dL</td>
</tr>
<tr>
<td>Chloride</td>
<td>103</td>
<td>Ref Range: 98 - 108 mmol/L</td>
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<tr>
<td>Creatinine</td>
<td>5.2 (HH)</td>
<td>Ref Range: 0.7 - 1.2 mg/dL</td>
</tr>
<tr>
<td>eGFR (CKD-EPI)</td>
<td>10.8 (L)</td>
<td>Ref Range: &gt;60 mL/min/1.73m (2)</td>
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<tr>
<td>Phosphate</td>
<td>4.0</td>
<td>Ref Range: 2.7 - 4.5 mg/dL</td>
</tr>
<tr>
<td>Potassium</td>
<td>3.7</td>
<td>Ref Range: 3.3 - 4.8 mmol/L</td>
</tr>
<tr>
<td>Urea</td>
<td>134 (H)</td>
<td>Ref Range: 12 - 40 mg/dL.</td>
</tr>
<tr>
<td>Haemoglobin, Blood</td>
<td>11.2 (L)</td>
<td>Ref Range: 13.0 - 17.0 g/dL</td>
</tr>
<tr>
<td>HEMATOCRIT</td>
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<td>Ref Range: 40.0 - 50.0 %</td>
</tr>
<tr>
<td>PLATELETS COUNT</td>
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<td>Ref Range: 150 - 410 10^3/uL</td>
</tr>
<tr>
<td>WBC COUNT</td>
<td>10.1</td>
<td>Ref Range: 3.6 - 11.0 10^3/uL</td>
</tr>
</tbody>
</table>

His urine culture grew ESBL positive E-coli; he received appropriate antibiotics according to the culture sensitivity. He was seen by the urology team and underwent Foley's catheter insertion.

Ultrasound KUB:

Image 1: Urinary bladder shows thickened heterogeneous wall with multiple echogenic foci, showing posterior dirty shadowing, suggestive of? Emphysematous cystitis.

Image 2 (A-C): Kidneys are normal in size with mild to moderate perinephric fat stranding and non-obstructive left renal calculus. However, there are multiple foci of air in the lumen and wall of the urinary bladder, suggestive of emphysematous cystitis.
His urinary symptoms had improved, though his renal functions remained stable during his hospital stay with serum creatinine of 5.2mg/dl and eGFR of 10.8 mL/min/1.73m². Patient was discharged from the hospital with antibiotics and Foley’s catheter in situ with follow-up appointment.

**Discussion**

Emphysematous cystitis is a rare type of infection of the bladder wall by gas-forming bacteria or fungi. The most frequent offending organism is *E. coli*. Other gram-negative bacteria, including *Klebsiella*, *Proteus* and *Citrobacter* are also commonly isolated. *Enterococci* and Fungi, such as *Candida*, have also been reported as causative organisms.

Although it is a rare type of bladder infection, it is the most common type of all gas-forming bladder infections. The first case was identified in a post-mortem examination in 1888 [1-3], though the first description to such cases in literatures (Gas within the urinary tract) goes back to 1671, was described in a man who presented with pneumaturia [4].

The condition is characterized by the formation of air bubbles in and around the bladder wall. The pathogenesis of emphysematous cystitis is poorly understood. Elevated tissue glucose levels in diabetic patients may provide an encouraging microenvironment for gas-forming microbes. However, bacterial gas production does not fully explain the pathological and clinical manifestations of emphysematous UTIs [5]. Various theories have been suggested, including fermentation of glucose in urine, with emphasis on disequilibrium between gas formation and clearance, other suggested either a transluminal dissection of gas or a true infection of bladder wall with pathology. The gas found in the bladder consists of nitrogen, hydrogen, oxygen and carbon dioxide [6].

**Clinical presentation and lab testing’s:**

- Clinical manifestations can be non-specific with minimum symptoms of urinary tract infection. Generally, patients with EC are not acutely ill as the case with Emphysematous pyelonephritis/pyelitis, however, Cases of Emphysematous Cystitis in a clinical study have shown to progress quickly if not treated and can be fatal due to inflammation caused by gas forming organisms [1].
- Abdominal pain was the most common symptom of emphysematous cystitis, occurring in up to 80% of cases. By comparison, the classic symptoms of acute cystitis (fever, chills, dysuria, urinary frequency, urinary urgency, haematuria and supra-pubic pain) occurred in only about 40-50% of patients [6-8].
- Pneumaturia after bladder catheterization occurred in 7 of 10 patients in one series.
- Laboratory testing usually revealed pyuria and haematuria with positive urine cultures. The two most common pathogens were *E. coli* and *K. pneumoniae*, which accounted for 75 to 80 percent of cases. Other isolates have included *Enterococcus*, *Candida* and polymicrobes.
- Bacteremia was present in approximately one-half of the cases [6,8].

**Risk Factors**

Risk factors include indwelling urethral catheters, or recent instrumentation of the urinary tract, chronic urinary tract infections, diabetes mellitus, neurogenic bladder, and being in an immunocompromised state. In 50%- 67% of cases, patients are elderly (age over 60) and diabetic. Obstruction of the urinary tract as well as urinary stasis, often brought on by paralysis of the urinary tract, are also major risk factors in addition to diabetes. Transplant recipients as being immunocompromised individuals; have been found to be at risk. In addition, most patients were women (67% in one series), similar to the female predominance with acute cystitis and pyelonephritis [5,6,8-11].

Differential diagnosis: Air within the bladder wall is usually due to emphysematous cystitis; however, intra-luminal air can occur in the setting of an enterovesical, colovesical, or rectovesical fistula (which, in turn, may be due to diverticulitis, inflammatory bowel disease or colorectal carcinoma). Intra-luminal air can also occur following cystoscopy or bladder catheterization [1,7,8].

**Diagnosis**

Due to atypical presentation and rarity of the infection, it takes a physician longer period to diagnose than more common types of bladder infections. Diagnosis requires a personalized investigation with consideration to risk factors and symptoms. Plain abdominal X-ray of the abdominal or pubic region has proven to be an important tool in reaching a definitive diagnosis of conditions causing gas in the urinary tract with high sensitive (97.4%) [6].

Computer tomography (CT scans) are of most help due to their high sensitivity in detecting gas and air bubbles. It can detect cases that are not apparent on plain radiography, can accurately define the extent and severity of the disease, and can help in differentiating emphysematous cystitis from colovesical fistula, intra-abdominal abscesses, neoplastic disease or emphysematous pyelonephritis.

Gas in the bladder wall will often have the appearance of cobblestone or a “beaded necklace” with the use of conventional radiography. Delayed diagnosis can lead to a severe infection, bladder rupture and death. Emphysematous cystitis has an overall mortality rate of 7% [8,12-14].
Treatment

Surgical intervention is rarely needed in emphysematous cystitis compared to emphysematous pyelonephritis, except in cases with anatomical abnormalities such as obstruction or presence of stones. Emphysematous cystitis can usually be treated with medical therapy alone. However, bladder irrigation may be needed if blood clots are present, and catheter placement is often required if the patient cannot adequately void. Rarely, bladder debridement and partial or total cystectomy are necessary. In a review of 135 published cases, 10% had required combined medical and surgical therapy and the overall mortality rate in this group of patients was 7 percent.

Parenteral antibiotic selection is same as for the management of acute pyelonephritis. Duration of antimicrobial therapy depends upon the clinical response. A review of 20 cases of emphysematous cystitis for which there were data on duration of treatment reported a median length of antibiotic treatment of 10 days. Hyperbaric oxygen had cured one case in a single report in less than 48 hours.

Conclusion

Pneumaturia should always raise doubt of emphysematous cystitis, particularly in elderly, diabetics and immunocompromised individuals. Early diagnosis is essential for a favorable outcome, while delayed diagnosis can result in high mortality rate.

References