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CASE REPORT

Emphysematous Cystitis: An unusual disease of the bladder diagnosed on imaging

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Abstract
Emphysematous cystitis (EC) is defined by the presence of gas in the urinary bladder wall. This rare disease entity is caused by gas fermenting bacterial and fungal pathogens. It complicates urinary tract infections especially in diabetic patients but other disabling general medical conditions may be present. Because the clinical symptoms are nonspecific, the diagnosis is often made incidentally on X-rays. Early diagnosis and treatment improve the outcome.
We report a case of 83-year-old diabetic female who presented with fever, dysuria and gross hematuria. Ultrasonography and computed tomography revealed gas accumulation in the wall and lumen of the bladder leading to the diagnosis of emphysematous cystitis. She was treated with antibiotics, insulin, rehydratation and a Foley catheter placement.
Diabetic patients with urinary tract infections who are severely ill should have radiological investigations to rule out the prsence of emphysematous complications.

Key-words: Emphysematous cystitis - Diabetes mellitus - Urinary tract infections - Klebsiella pneumoniae.

Introduction

The prevalence of Urinary Tract Infections (UTI) is high in diabetic patients. They are also at a greater risk of complications than non-diabetics (1).
Emphysematous cystitis is an uncommon, but severe manifestation of infection of the urinary bladder. It is characterized by air within the bladder wall and lumen produced by gas forming organisms. The presentation may be atypical and contrary to the degree of inflammation, patients may present with subtle clinical findings (2, 3). The diagnosis is always radiological (4).
There has been a recent increase in reported cases due to a wider use of abdominal imaging and a greater awareness of this unusual disease. Although there are numerous published case reports, EC remains poorly understood. The paucity of clinical information on diagnosis, treatment and outcomes warrants a review of this condition (4).
Herein we present a new case with late diagnosis in a diabetic patient.

Case Report

An 83-year-old diabetic woman was admitted to our department because of confusion, fever, abdominal pain

and hematuria which was evaluated for 15 days prior to her transfer. She rapidly progressed to septic shock. Her past medical history was unremarkable.

Physical examination revealed a regular pulse with a rate of 90/min. Her temperature was 38.8°C, blood pressure 132/80 mmHg and respiratory rate 24/min. Cardiac and pulmonary examinations were unremarkable and there was no hepatosplenomegaly on abdominal examination. The lower abdomen was painful on palpation. She had a palpable bladder with mild suprapubic tenderness. There were no peritoneal signs and the bowel sounds were normal.

Laboratory tests showed pancytopenia with blood cell count 4300/mm^3, hemoglobin 7.7 g/dl and platelet count 75000/mm^3. Prothrombin time was 62% and hepatic enzymes were elevated.

Initial blood glucose level was 43.5 mmol/l with Ketonuria. Serum creatinine was 107 μmol/l, blood urea nitrogen was 44 mg/dl and inflammatory markers were increased (C reactive protein 108.3 mg/l and fibrinogen 4.7 g/l). Nutritional proteins were decreased (albumin 26 g/l).

Chest X-ray was normal. KUB radiography was unremarkable but abdominal ultrasound showed normal kidneys and hyperechoeic thickened bladder wall (Fig. 1).

Computed tomography scan of the abdomen demonstrated a thickened, trabeculated bladder wall containing pockets of gas and lumen with air-fluid level, which indicated EC (Fig. 2,3). There were no findings that suggested pyelonephritis. The diagnosis of emphysematous cystitis was made.

Urine contained 5.10^4 leukocytes/mm3 and culture grew a highly resistant Klebsiella Pneumonia 10^7 Colony Forming Unit/ml. Blood cultures, which were performed three times, were all negative.

A urinary catheter was inserted, yielding 800 ml of macroscopic hematuria containing pus and air.

She was treated with antibiotics (ceftriaxone® 2 g intravenously once daily and gentamicin 80 mg intatravenously every 12 hours, amoxicilline + clavulanic acid and ofloxacine®) and rehydratation. She was started on insulin for newly diagnosed diabetes mellitus. Because of the hematuria, she underwent a cystoscopic exploration. Cystoscopy revealed multiple submucosal bullous lesions. Bladder mucosa was studded with vesicles varying in size and arranged in clumps (Fig. 4). Bladder mucosal biopsy showed mild non-specific active inflammation, with marked oedema and congestion of the lamina propria.

The investigations were all consistent with a diagnosis of emphysematous cystitis in a patient with a newly-diagnosed diabetes mellitus.

Despite intravenous antibiotics and bladder catheterization, the patient died two days after admission.

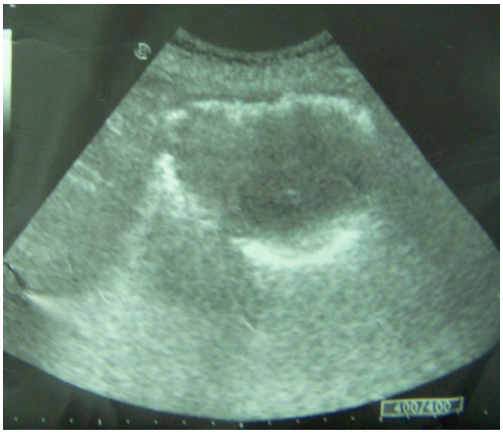

Figure 1: Abdominal ultrasound showing hyperechoeic thickened bladder wall

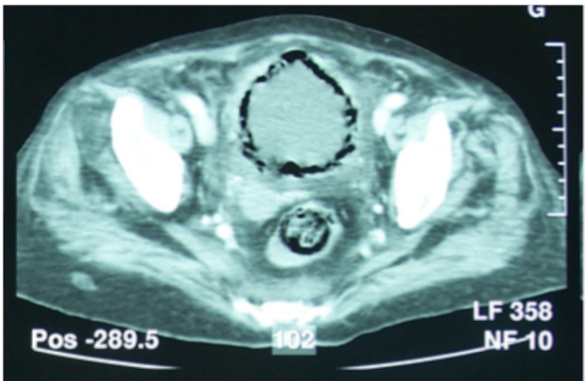

Figure 2: Abdominal CT showing trabeculated bladder wall

Discussion

Emphysematous cystitis was first identified in 1882 by Keyes (5). Bailey (6) further characterized the condition by defining the link between pneumaturia and this condition, and defined it in 1961 as 'cystitis emphysematosa' (6, 7).

EC is a rare, serious entity relatively unknown to physicians (3, 8). It is characterized by pockets of gas, produced by bacterial or fungal fermentation, in and around the bladder wall (1, 6, 7, 9).

Emphysematous conditions are typically diagnosed

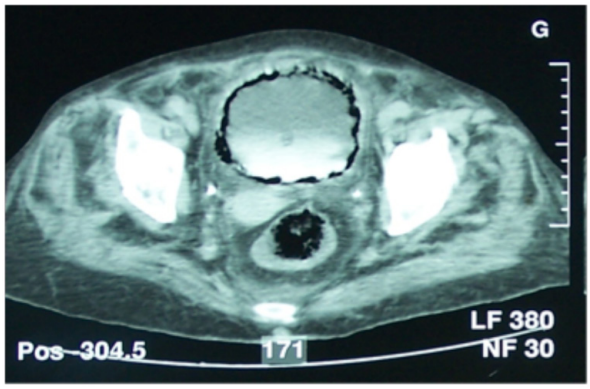

Figure 3: Abdominal CT showing air fluid level in the baldder

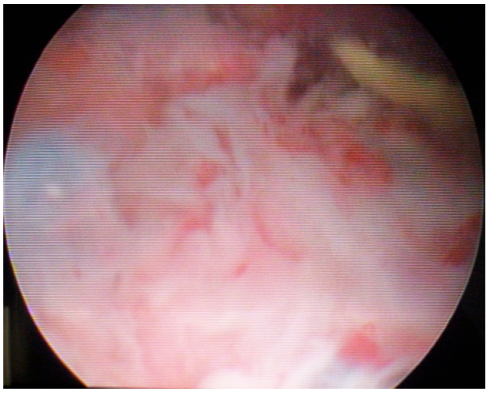

Figure 4: Cystoscopy picture showing the bladder lesions

radiologically, thus the number of emphysematous infections might be underreported and under-diagnosed (4). The incidence of reported cases is increasing with the increased use of abdomino-pelvic imaging and a greater awareness of such diseases.

The disease is most commonly seen in middle-aged women (3, 4, 10), immunocompromised patients(3, 8), diabetes mellitus (8-10), recurrent urinary tract infections (3, 8), urinary stasis, bladder outlet obstruction (8, 10), neurogenic bladder (3, 8, 10), indwelling urinary catheters (8, 10), and in transplant recipients (11). We noticed three risk factors in our case. It may complicate UTI in other pre-existing conditions such as alcoholism (9, 12), undernutrition (9, 13) or disabling general medical conditions (9, 14).

Overall, two-thirds of the reported cases were diabetic and 64% were women (4). There are several theories on the pathogenesis of these rare gas-forming infections. The pathogenesis is not yet fully understood, but a multifactorial aetiology of impaired host responses with transluminal dissection of gas or true infection of the bladder wall seems to be a plausible explanation for the production of gas within the affected tissues (3,4). It is thought that the gas collections are carbon dioxide produced by the anaerobic (15) fermentation of glucose or albumin by micro-organisms infecting the bladder (7, 16). The most common causative organisms are E. coli accounting for 60-70% of cases (6,9,17), but other organisms reported to produce emphysematous cystitis include Enterobacter aerogenes, Klebsiella pneumonia as in our case, Proteus mirabilis, Staphylococcus aureus, streptococci, Clostridium perfringens (18), and Candida albicans (19). Aerobic and anaerobic cultures are required for identification of the etiologic agents (17).

The clinical presentation of EC is nonspecific and ranges from incidental diagnosis on abdominal imaging to severe sepsis (4) as in our patient's case. Patients may complain of irritative symptoms, abdominal discomfort or pneumaturia. A history of pneumaturia is highly suggestive, but is rarely offered by the patient (3).

The radiographic findings provided the first and the only diagnostic clue (3) and all patients with diabetes and UTIs should have an abdominal X-ray screen to detect the presence of gas within the bladder wall or other complications (17).

Conventional imaging shows usually a curvilinear area of radiolucency delineating the bladder wall with or without intraluminal air. In our cese, KUB film was almost normal.

Ultrasonography (as in our case) might suspect the diagnosis (9), it commonly demonsastrates diffuse bladder wall thickening. Focal regions of high-amplitude echoes with posterior dirty acoustic shadowing into the lumen may be seen in extensive cases of emphysematous cystitis and these echogenic shadows may be misinterpreted as bladder calculi with ultrasound (20).

Nevertheless, computed tomography is considered to be the preferred method because of its high sensitivity and specificity in the detection of abnormal gas and its anatomic extension (21).

The radiological findings include small gas-filled vesicles in the bladder mucosa, producing a cobblestone appearance, and can go on to form a thin zone of gas outlining the

perimeter of the bladder. The bladder wall is thickened. Gas-filled blebs are seen in the bladder wall, which, when ruptured, allow gas into the bladder lumen (8).

Diagnostic entities associated with gas in the genitourinary tract include emphysematous pyelonephritis, emphysematous pyelitis, and gas-forming renal abcess (3).

Patients with EC are not as acutely ill as those with emphysematous pyelonephritis. It is important to differentiate emphysematous cystitis from emphysematous pyelonephritis, in which gas involves the renal parenchyma, since the latter has an increased mortality and generally requires nephrectomy. In contrast, surgical intervention is rarely needed in EC except when an anatomical abnormality like an obstruction or stone is present (22).

Other sources of gas within the urinary tract were reported as trauma, vesico-enteric fistulas from radiation therapy, rectal carcinoma, diverticular disease or Crohn's disease and iatrogenic causes, such as diagnostic or surgical instrumentation. History, physical examination and imaging are the best modalities to differentiate the above etiologic causes. Fistulous tracts, abscess, can be excluded on CT scan (3).

Cystoscopic examination is not essential for the diagnosis, although it can add information about bladder outlet obstruction or the origin of hematuria. It might show bladder wall thickening with vesicles of varying size, and microscopically there are multiple gas-filled vesicles predominantly within the bladder mucosa, lined by necrotic tissu and fibrin deposit (6, 23).

The prognosis for emphysematous cystitis is favorable if it is diagnosed promptly and treated properly (10). It usually responds well to treatment, which includes glycaemic control, appropriate systemic antibiotics and good urinary drainage (1, 8, 24). Some authors had proposed a continuous bladder irrigation with amikacin as adjuvant treatment for EC to reduce hospital stay and accelerate the patient's healing process, as well as an earlier negativisation of urine cultures (2).

Delayed diagnosis may lead to unfavorable outcomes including overwhelming infection, extension to ureters and renal parenchyma, bladder rupture and death (3).

Patients with few comorbidities and presenting with EC should respond favourably to conservative management with close monitoring, whereas patients with necrotizing infections will require more aggressive treatment that includes surgery (4).

About 1 in 10 patients (7%) who are diagnosed with emphysematous cystitis will die from the condition (9, 25, 26). Cases of EC associated with another emphysematous infection of the urinary tract had a higher death rate of 14%, but the overall death rate for EC indicates that it might not be as severe as once thought, and can be successfully treated with medical management (27), reserving surgical intervention for severe cases. Improved outcomes may be achieved by early recognition of the infection, by clinical and radiological assessment, and by prolonged course of appropriate antibiotherapy (3-6 weeks) (3, 4).

Conclusion

Emphysematous cystitis is a relatively uncommon disease. It is defined by the presence of gas in the urinary bladder wall and often in the bladder lumen. It complicates UTI especially in diabetic patients but other pre-existing conditions.

This clinical entity may present with fairly nonspecific findings. It is most often not diagnosed by routine or systematic approach and imaging is essential for the diagnosis.

The physician should be cautious and a high degree of suspicion should be maintained with diabetics and at-risk patients for complicated UTIs that do not respond to standard therapy.

They should have an abdominal ultrasonography as a screening tool to detect the presence of gas within the bladder wall. CT scan define the severity and extent of the disease and allows an assessment of ascending infection and other complications.

The treatment of this entity involves management of diabetes, asystemic antibiotherapy and external drainage of the bladder. Early diagnosis and treatment are mandatory to improve the outcome.

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