Case report

Massive bilateral grade IV emphysematous pyelonephritis: Case report

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Abstract

Emphysematous pyelonephritis (EPN) is a severe disease of the renal system in which gas formed by facultative anaerobe microorganisms accumulates, being Escherichia coli the most representative causative agent. A series of conditions foster its development including uncontrolled diabetes mellitus, diabetic nephropathy and obstructive uropathy. Abdominal CT scan continues to be the gold standard for diagnosis. Currently, nephrectomy is avoided as much as possible, and more conservative treatments are given. Mortality is still as high as 21% despite new therapeutic options, all of which are mostly surgical. The rarest cases are those classified as grade IV, which affect both kidneys; these are considered the most lethal of the clinical presentations and they are also of particular interest, since a multidisciplinary team must be in charge of treatment. Herein, we present the case of an elderly woman with chronic diseases and a presentation of massive emphysematous pyelonephritis concurrent with emphysematous cystitis and pneumoperitoneum, who required bilateral radical nephrectomy despite efforts of preserving at least one of the kidneys.

Introduction

Urinary tract infections (UTI) in patients living with type 2 diabetes mellitus pose an important public health issue, with a prevalence of 8.2% in developed countries [1]. Within the spectrum of UTI, infections that involve gas production stand out, including emphysematous pyelitis (gas in the renal pelvis), perinephric emphysema (gas in the perinephric space) and emphysematous pyelonephritis (intraparenchymal gas) [2].

Emphysematous pyelonephritis (EPN) is an infection in which gas forms in the renal parenchyma and, sometimes, outside from it. It was first documented in 1898 by Kelly and MacCullum, which they called “pneumaturia” [3]. Later, in 1962, it was Schultz and Klorfein who coined the term “emphysematous pyelonephritis” [4].

Initially, the only accepted management option was radical nephrectomy, with mortality rates being as high as 80%; however, therapeutic strategies are currently diverse depending on the severity of the presentation, and mortality in general has decreased to 21% [5].

We present the case of a Mexican female patient with fulminant, massive bilateral EPN. Due to the severity of the presentation, we decided to perform the most radical surgical treatment available.

Case presentation

A 54-year-old female was admitted to our emergency department (ED) due to urinary symptoms, nausea and diffuse abdominal pain. Her relevant past medical history includes a diagnosis of type 2 diabetes mellitus nine years prior, currently on treatment with basal insulin 20 IU per day. Home-based capillary blood glucose monitoring was 300 mg/dL on average, on a daily basis. The patient denied follow-up appointments with her primary care physician. She denied prior hospitalizations secondary to poor glycemic control. She reported recurrent UTI in the last 4 months that were treated with empirical antibiotics. The rest of...
Fig. 1. Abdominal CT scan. Axial cut showing abundant intraparenchymal gas and loss of renal anatomy.

Fig. 2. Abdominal CT scan. Coronal cut showing intraparenchymal gas, as well as gas within the bladder.

her past medical history was non-contributory.

She began her symptoms three days prior to admission with fatigue, increased urinary frequency, urgency and acute urinary retention, reason why she consulted with a private physician who placed a Foley catheter, successfully achieving diuresis, while giving non-specific management. After 24 h from symptom onset, she developed fevers of 39 °C, as well as nausea and vomiting, which prompted her to seek medical attention at our hospital.

Vital signs at admission were as follows: blood pressure 58/36 mmHg, pulse 77 beats per minute, respiration rate 26 breaths per minute, body temperature 36 °C, room-air oxygen saturation 92 %, capillary blood glucose 337 mg/dL. The patient had altered mental status with a Glasgow Coma Scale (GCS) of 12 points. Physical examination revealed capillary refill of 3 s, normal lung examination and abdomen without signs of peritoneal irritation. Vesical globus and bilateral costovertebral angle tenderness were noted. A Foley catheter was placed, revealing hematuria, and resuscitation was initiated with IV fluids, vasopressor agents and third-generation cephalosporins.

Lab tests revealed leukocytes $35.7 \times 10^3/\mu$L, total neutrophils $33.4 \times 10^3/\mu$L, hemoglobin 11.7 g/dL, platelets $138 \times 10^3/\mu$L, fibrinogen 981 mg/dL, creatinine 4.3 mg/dL, glucose 334 mg/dL, urea 132 mg/dL, corrected sodium 136 mEq/L, potassium 5 mEq/L, chloride 94 mEq/L and calculated serum osmolality 282 mOsm/L. Venous blood gases revealed pH 7.31, PO2 25 mmHg, PO2 37 mmHg, HCO3 12.37 mmol/L and base excess (BE) – 13.7 mmol/L. Urinalysis showed a positive leukocyte esterase test, negative nitrite test, 500 leukocytes per high-power field (HPF) and presence of ketones. Urine culture was obtained, which came back positive to extended-spectrum beta-lactamase (ESBL)-producing E. coli, which was meropenem-sensitive and levofloxacin-resistant.

A non-contrast abdominal CT scan was ordered under the clinical suspicion of a kidney and abdomen infection, which revealed both kidneys with loss of anatomy and presence of gas through the entire perirenal space that extended over the course of the ureters and bladder, as well as pneumoperitoneum. A diagnosis of septic shock due to grade IV emphysematous pyelonephritis, mild diabetic ketoacidosis and stage 3 acute kidney injury was made.

The patient was hospitalized and wide spectrum carbapenem antibiotics were given at full doses due to septic shock, coupled with fluid resuscitation and IV insulin for treatment of uncontrolled glycemia. A joint management with the Department of Urology was set who, given the patient’s compromise and severity of disease, proposed a total bilateral nephrectomy.

During surgery, the patient’s appendix was also removed. The specimens were revised by the Surgical Pathology Department, who observed both kidneys with necrosis greater than 90 %, with areas of parenchymal abscesses, as well as renal vein and arteries with septic thrombi without signs of malignancy. The appendix showed signs of acute appendicitis.

The patient was admitted to the Intensive Care Unit (ICU) and underwent continuous veno-venous hemofiltration (CVVH) renal replacement therapy. During her stay, she progressed poorly until her death 11 days after symptom onset and 8 days after admission.

Discussion

EPN is an infection which mostly affects women, at a ratio of 6:1 [6]; this can be explained by the higher prevalence of UTI in this population. Mean age of onset is 55–60 years, and the left kidney is the most affected in up to 67 % of cases [6].

There are several risk factors of the host that predispose to developing this infection such as: long-standing, uncontrolled diabetes mellitus, uncontrolled hypertension, obstructive uropathy and chronic kidney disease (CKD) [7]. The causative agents involved in most of the cases include: Escherichia coli (60 %), Klebsiella (15 %), Proteus mirabilis (10 %) and Pseudomonas (7 %) [8]. These bacteria are facultative anaerobes, that is, under hypoxic conditions (such as in diabetic microangiopathy), they ferment glucose and lactate, resulting in the production of carbon dioxide (CO2), hydrogen, nitrogen, ammonia, methane and carbon monoxide [9], which accumulate in the renal parenchyma and spread from there to other underlying structures.

The prevalence of ESBL-producing E. coli is still unknown in this disease, and the treatment of choice are carbapenems. The use of fluoroquinolones as part of the initial empirical treatment of UTIs is discouraged, given their high resistance rate deemed at more than 20 % [10].

The clinical manifestations of EPN are usually very non-specific due to immunosuppression caused by uncontrolled diabetes. However, in a considerable proportion of patients there is a characteristic triad: fever, flank pain and nausea/vomiting, though renal colic and hematuria have also been reported [11]. Of note, most of the findings were present in our case presentation.

There are four factors of severity that complicate the progression of EPN: thrombocytopenia less than 120,000 × 10^3/μL, altered mental status, acute kidney injury and septic shock [6]. Mortality rates in the presence of 1, 2 and 3 of these factors are 27 %, 75 % and 100 %, respectively[12]. These factors explain the patient’s fatal outcome.

Urineanalysis is a diagnostic test useful in any UTI, however, even
though it usually reveals leukocytosis and gross hematuria, up to 30 % of them may show negative nitrites test [8] just as in our case presentation. On the other hand, urine cultures are positive in 90 % of the cases [7].

According to Huang and Tseng [6], EPN may be classified into four grades depending on the localization of gas in the renal system:
- Class I: Gas in the collecting system only (10.5 %).
- Class II: Gas in the renal parenchyma (23 %).
- Class IIIa: Gas or abscess in the perirenal space (14.5 %).
- Class IIIb: Gas or abscess in the para-renal space (43 %).
- Class IV: Bilateral EPN or solitary kidney with EPN (8.3 %).

This classification guides the clinical in deciding the treatment: classes I and II usually respond well to medical management with antibiotics alone (as long as there is no obstruction), class IIIa can be managed with antibiotics and percutaneous drainage with a survival of up to 80 %[11], and classes IIIb and IV very frequently end up requiring urgent nephrectomy, which has a mortality rate of 42 % [6].

Thereby, there are several treatment approaches [13]:
- Conservative management alone.
- Conservative management with percutaneous drainage.
- Emergency nephrectomy.
- Delayed nephrectomy after failure of conservative management.

In the literature, less than 10 % of cases belong to a class IV EPN, and the few that are reported show bilateral involvement with moderate gas that allows for medical management and percutaneous drainage. Our patient showed an intense destruction of the renal parenchyma of both kidneys, with concurrent emphysematous cystitis, pneumoperitoneum and appendicitis due to intraperitoneal dissemination of the infection. Sepsis was so severe that it was decided to perform both a bilateral radical nephrectomy and an appendectomy, despite a significant metabolic decompensation.

Some physicians choose to perform a renal scintigraphy before opting for nephrectomy to confirm there is functional renal tissue, thereby delivering a more conservative management, however, it has been shown that up to 47.4 % of patients with kidney preservation present recurrent UTIs[7], putting them again at risk for EPN. On the other hand, both conservative and minimally invasive treatment have managed to decrease mortality to 21 %[14].

Conclusions

UTIs are the second most frequent cause of sepsis in patients living with diabetes [15], and their occurrence is determined by several modifiable factors, including duration and control of disease. The most severe of presentations is EPN, which can harbor high mortality rates. When facing a compatible clinical presentation in high-risk patients, an abdominal CT scan is the test of choice since early identification of EPN can be decisive in terms of disease prognosis. Fig. 1–4.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.
Author contribution

Carlos Adrian Pérez Martínez: Care of patient, writing, editing, revising and submit of manuscript. Ixchel Guadalupe Rodríguez Barajas: Care of patient and writing manuscript. Maria Fernanda Carballar Jerez: Care of patient and writing manuscript. Brenda Lara Gonzalez, Jonathan Ramirez Porras, Luis Alberto García Fierro: Care of patient and revising manuscript. Edmundo Rivero Sanchez: Care of patient and revising manuscript.

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Declaration of Competing Interest

We declare that there are no conflicts of interest amongst the authors.

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