CASE REPORTS

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Emphysematous pyelonephritis – case report and review of literature

Emfizematozni pijelonefritis

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Abstract

Introduction. Emphysematous pyelonephritis (EPN) is a severe, potentially fatal necrotizing infection of the kidney with the clinical picture ranging from the mild abdominal pain and discomfort to the septic shock and multiorgan failure. We presented here a case of EPN in a poorly controlled diabetic patient that was the first registered case of EPN in our clinic for more than ten years. **Case report.** A 78-year-old diabetic male patient was referred to the Clinic for Nephrology and Clinical Immunology of the Clinical Center of Vojvodina, Novi Sad, Serbia, with weakness, malaise, abdominal discomfort and reduced daily urine volume. After complete physical exam, laboratory work up, echosonographic and computed tomography scanning we diagnosed the patient with EPN class IV, according to the Huang and Tseng classification, with the presence of 5 risk factors for mortality (systolic blood pressure below 90 mmHg, altered consciousness, thrombocytopenia, elevated serum creatinine level, bilateral disease). Treatment with conservative therapy and percutaneous drainage was not successful, further deteorioration of the patient status ensued so the patient passed away on the 8th day of hospitalization due to the development of septic shock with multiorgan failure that was refractory to all measures that were instituted. **Conclusion.** EPN is a severe, potentially fatal necrotizing inflammation of the kidney and surrounding tissue. Management and prognosis of this disease depends on the clinical status, risk factors and radiological classification of the disease.

Key words:
pyelonephritis; emphysema; diabetes mellitus, type 1; drainage; treatment outcome.

Apstrakt

Uvod. Emfizematozni pijelonefritis je teška, potencijalno fatalna, nekrotizirajuća infekcija bubrega sa kliničkom slikom koja može varirati od blagih abdominalnih bolova do septičkog šoka sa multiorganskom insuficijencijom. Prikazan je slučaj emfizematoznog pijelonefritisa kod višegodišnjeg dijabetičara, što je prvi registrovan slučaj ovog oboljenja na našoj klinici u periodu dužem od deset godina. **Prikaz bolesnika.** Muškarac star 78 godina, sa višegodišnjim, loše regulisanim dijabetesom, hospitalizovan je na Klinici za nefrologiju i kliničku imunologiju Kliničkog centra Vojvodine u Novom Sadu, sa tegobama u vidu slabosti, malaksalosti, bolova u stomaku i smanjene količine mokraće. Nakon fizikalnog pregleda, laboratorijskih analiza, ehosonografskog pregleda i kompjuterizovane tomografije abdomena postavljena je dijagnoza emfizematoznog pijelonefritisa klase IV po klasifikaciji Huanga i Tsenga. Kod prikazanog bolesnika verifikovano je postojanje pet faktora loše prognoze (sistolni krvni pritisak ispod 90 mmHg, poremećaj svesti, trombocitopenija, povišen nivo serumskog kreatinina, bilateralna bolest). Primenjena konzervativna terapija i perkutana drenaža nisu rezultirale povoljnim ishodom i došlo je do daljeg pogoršanja stanja bolesnika. Bolesnik je preminuo osmog dana hospitalizacije usled razvoja septičkog šoka i multiorganske insuficijencije koja je bila refrakterna na sve primenjene mere. **Zaključak.** Emfizematozni pijelonefritis predstavlja tešku, potencijalno fatalnu nekrotizujuću inflamaciju bubrega i okolnog tkiva. Terapija i prognoza ove bolesti zavise od kliničkog statusa, prisutnih faktora rizika i radiološke klasifikacije bolesti.

Ključne reči:
pijelonefritis; emfizem; dijabetes melitus, insulin zavisni; drenaža; lečenje, ishod.

Introduction

The first case of kidney infection with gas accumulation in kidneys and the surrounding tissues was described by Kelly and MacCallum [1] in 1898. The term emphysematous pyelonephritis (EPN) was introduced in clinical practice in 1962, when Schultz and Klorfein [2] used it to point to the connection between kidney tissue infection and formation of gas accumulation.

EPN is a severe, potentially fatal necrotizing kidney infection with the clinical picture ranging from the mild abdo-

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minal pain and discomfort to septic shock and multiorgan failure. Most cases of EPN occur in poorly controlled diabetic patients, while fewer cases of these potentially fatal infections occur in patients with urinary tract obstruction or some other malformation of the kidneys, like polycystic kidneys [3, 4]. Treatment of EPN consists of a combination of conservative measures among which the most important is an antibiotic therapy, and invasive, surgical procedures ranging from percutaneous drainage to nephrectomy. We presented a case of EPN in a patient with poorly controlled diabetes. It was the first registered case of EPN in our clinic for more than ten years.

Case report

A male patient, aged 78 years, was admitted to the Clinic for Nephrology and Clinical Immunology of Clinical Center of Vojvodina, Novi Sad, Serbia, with complaints of fatigue, malaise, abdominal discomfort and reduced daily urine volume. The symptoms began two days prior to admission, and worsened on the day of admission. The patient had diabetes for decades and had been receiving insulin therapy for the past 6 years. He had a poorly controlled primary illness with the hemoglobin A1c (HbA1c) values of 10.2%. The patient`s vital signs showed normal body temperature (36.9 Cº), sinus tachycardia of 110 beats per minute (bpm), arterial hypotension with blood pressure of 90/60 mmHg and number of respirations of 21/min. Physical examination found extensive pain on succusion of left lumbar region as well as on palpation of left paracolic region. The patient`s general condition was poor, he was somnolent, with Karnofsky performance status of 30, Glasgow Coma Scale of 12, and he was oligoanuric in the previous 12 hours.

Laboratory analyses done upon admission showed the existence of leukocytosis with leukocytes measuring 21.8 × 10^9/L [normal range (nr) 4.5–10.0 × 10^9/L] and 83% of granulocytes (nr 50–70%), thrombocytopenia measuring 42 × 10^9/L (nr 150–450 × 10^9/L), C-reactive protein level was 224 mg/L (nr < 10 mg/L), serum procalcitonin level was 84.52 pg/mL (nr less than 50 pg/mL), as well as elevated serum creatinine level measuring 418 μmol/L (nr 70–120 nmol/L) and elevated blood urea level measuring 28.3 mmol/L (nr 2.5–7.1 nmol/L). Urine analysis showed the presence of a pyuria and bacteriuria, and the urine sample was sent for a microbiological analyses. Echosonography of the abdomen found suspected presence of gas accumulation on both kidneys, but predominantly on the left side with the presence of free fluid in the perisplenic and hepatorenal recessus. The patient was hospitalized and started on a dual antibiotic therapy of ciprofloxacin and ceftazidime, with other supportive measures. Considering the high serum levels of urea and creatinine as well as oligoanuria, a dual lumen dialysis catheter was inserted into the inner jugular vein on the patient`s right side. This was followed by a computed tomography (CT) scan of the abdomen which confirmed the presence of extensive gas accumulation in the left kidney parenchyma, calyceal system on the same side as well as in the left pararenal space. Gas collection was found in a right kidney as well, but the collection was limited to the parenchyma (Figure 1).

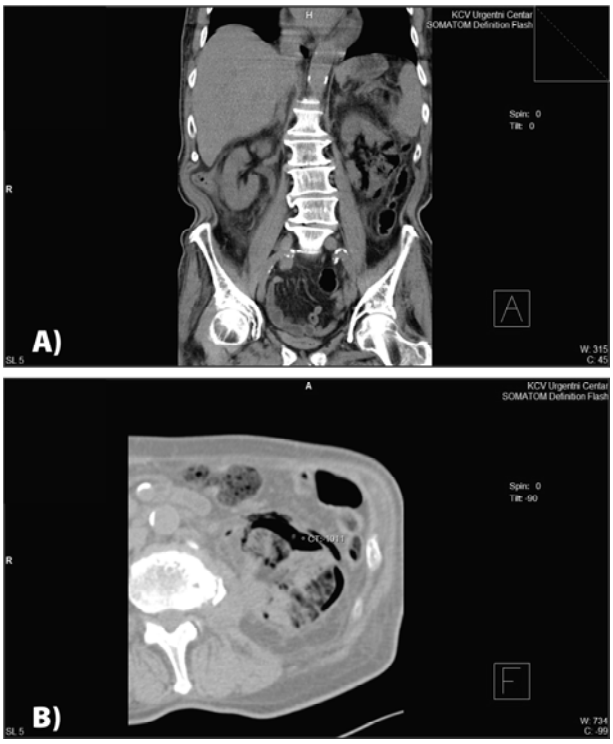

Fig. 1 – Computed tomography (CT) scan of abdomen: A) longituidinal view, showing minimal gas accumulation in the right kidney and extensive gas accumulation in the left kidney, collecting system on the left side and in the left pararenal tissue; B) transversal view, showing extensive gas accumulations in the left kidney, collecting system on the left side and in the left pararenal tissue.

Based to the CT findings and clinical status of the patient, left side lumbotomy with the intention of performing nephrectomy, was performed on the second day of hospitalization. Due to the patient's extremely poor condition accompanied by hypotension and profuse bleeding caused by thrombocytopenia, nephrectomy was abandoned, and only percutaneous drainage of left pararenal space was performed. As the patient's condition continued to deteriorate, he was transferred to an Intensive Care Unit. Measures of advanced life support, assisted ventilation and supportive measures among which infusion of platelets, crystalloid solutions and plasma expanders as well as positive inotropic drugs were instituted. On the third day of hospitalization and upon receipt of the results of the urine cultures which showed the presence of *Escherichia coli* resistant to then available antibiotics, piperacilin-tazobactam was introduced in the therapy. On a sixth day of hospitalization the patient started having diuresis in a range of 500–1,000 mL, but we continually performed renal replacement therapy (RRT) with continuous veno-venous hemodiafiltration (CVVHDF), due to the inadequate kidney function and volume status of the patient. In the course of further treatment, the patient`s status deteriorated and he developed septic shock with multiorgan failure, refractory to therapeutic measures, which caused a fatal outcome on the 8th day of hospitalization.

Discussion

EPN is a necrotizing infection of the kidney and surounding tissue where gas is formed in kidney parenchyma, collecting system and perinephric tissue. More than 90% of cases of EPN occur in patients with poorly controlled diabetes [5]. Other predisposing factors for EPN are urinary tract obstruction, polycystic kidney disease, immunosuppresive therapy and end stage renal disease [3, 4]. This disease has a higher predilection for a female patients [4], which is in accordance with the higher incidence of urinary tract infections in females, in general.

The pathogenesis of EPN depends on several factors which include the existence of bacteria capable of producing gas, high level of tissue glucose favorable for the fast growth of bacteria, disruption in tissue perfusion and inadequate immune response of the patient due to disrupted vascularization. The most frequent causes of EPN are *Escherichia coli, Klebsiella pneumoniae, Proteus mirabilis* and *Pseudomonas aeruginosa* [6, 7], the same bacteria that most frequently causes urinary infections in general. Anaerobic bacteria very rarely cause EPN. The gas is formed because of the fermentation of glucose and lactate which produces large amounts of carbon dioxide and hydrogen, which are then accumulated in places where inflammation occurred. Clinical manifestations of EPN are similar to cases of "ordinary" pyelonephritis, most frequently accompanied by elevated body temperature and pain in the loins and abdomen. Depending on the severity of the clinical picture, nausea and vomiting may occur, as well as different levels of altered state of consciousness. Laboratory analyses are dominated by leukocytosis with neutrophilia, pyuria and parameters pointing to an infection such as elevated levels of C-reactive protein, erythrocyte sedimentation rate and procalcitonin level. Impaired kidney function may occur, or an existing poor kidney function may deteriorate.

A number of different methods are available for the visualization of the changes in kidney parenchyma. Although echosonography is not a preferred method of proving the presence of gas accumulation in kidneys and surrounding tissue, it is used to initially examine the upper urinary tract if an infection is suspected. It can, to a large extent, confirm or rule out the existence of a urinary tract obstruction. CT is the method of choice for diagnosing EPN [3, 6]. There are several staging systems based on the radiological findings that can have a great significance in the prognosis and treatment of EPN [3, 8, 9]. According to the classification by Huang and Tseng [3] (Table 1), which is the one most widely in use, the patient treated at our clinic had class IV EPN. Aside from the radiological classification of EPN, other important prognostic factors, i.e. risk factors, that are associated with outcome of EPN are systolic blood pressure below 90 mmHg, altered state of consciousness and initially elevated sera creatinine level (> 221 μmol/L or 2.5 mg/dL). In addition, the presence of thrombocytopenia (< 100×10^9/L) and the existence of bilateral EPN also lead to a less favorable outcome [5].

The treatment of EPN depends on the evaluation of the patient's general condition and the morphological report based on the CT scan. A decision on the treatment of each individual patient is made on the basis of evaluation of the condition including risk factors for a less favorable prognosis and the outcome of the illness, as well as on morphological diagnostics by Huang and Tseng [3]. Conservative treatment with antibiotics and measures aimed at stabilizing the patient is justified in all cases [10]. It is combined with surgical procedures ranging from percutaneous drainage [3, 7, 11, 12] to nephron-sparing surgery [13] and nephrectomy [14], depending on the patient's condition and the extent of changes visualized by CT scan. One of the first study on EPN, found an average mortality rate of 31% in those patients. Mortality rate was as high as 80% for patients who received conservative therapy only, 60% for patients treated with percutaneous drainage, and 20% in nephrectomized patients [15]. Shokeir et al. [4] registered a mortality rate of 20% in their series of nephrectomized patients. Procedures that are less invasive and have a goal to preserve kidney mass are justifiable, but only for a less extensive cases of EPN. In a meta-analysis of the different treatment strategies, the most successful one was combination of medical therapy and percutaneous drainage, with the mortality rate of around 13% [12]. In cases where advanced changes are evidenced by a CT scan (class III and IV by Huang and Tseng [3]), and especially if there are two or more risk factors, nephrectomy is recommended as the combination of conservative therapy and percutaneous drainage does not lead to very good outcomes, resulting in mortality in over 92% of patients [7].

Table 1

Classification system for emphysematous pyelonephritis (EPN) by Huang and Tseng [3]

Radiological test	Class of the disease
Computed tomography	I: Gas in collecting system only
	II: Parenchymal gas only, without extension to extrarenal space
	III: Extension of gas or abscess to pararenal space
	IV: EPN in solitary kidney or bilateral disease

Conclusion

EPN is a very serious necrotizing infection of the kidneys and the surrounding tissue which, in a number of cases, can result in a fatal outcome. This is especially true in cases of extensive gas accumulation in the kidney and surrounding tissue, as well as when two or more factors of unfavorable prognosis are present. Therapy for EPN consists of combination of conservative measures and surgery, and the decision for the every patient is individual, based upon radiological findings and clinical status.

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