

*Prikazi bolesnika/
Case reports*

Correspondence to:

Mr sci. dr Slobodan Radulović, Beograd,
KBC „Zvezdara“, Kliničko odeljenje urologije,
Dimitrija Tucovića 161,
Tel: ++381 3810-298; 064-167-42-10,

E-mail: uroboban@ptt.rs

Ključne reči/Key words

emfizematozni pijelonefritis,
ureterolitijaza, dijabetes, sepsa,
trombocitopenija, ureteroskopija,
nefrektomija.

Emphysematous pyelonephritis;
ureterolithiasis, diabetes mellitus, sepsis,
thrombocytopenia, ureteroscopy, nephrec-
tomy.

COMBINED – ENDOUROLOGICAL AND
SURGICAL TREATMENT OF EMPHYSEMA-
TOUS PYELONEPHRITIS CAUSED BY
SILENT URETEROLITHIASIS IN DIABETIC
PATIENT

KOMBINOVANO – ENDOUROLOŠKO I
HIRURŠKO LEČENJE EMFIZEMATOZNOG
PIJELONEFRITISA IZAZVANOG
LATENTNOM URETEROLITIJAZOM KOD
DIJABETIČNE PACIJENTKINJE

Slobodan Radulović¹, Božo Vavić, Saša Patrnogić and
Zorica Miljuš²

¹Clinical department of Urology, Clinical Hospital "Zvezdara",
Belgrade, Serbia;

²Center of Anesthesiology and Reanimatology, Clinical Hospital
"Zvezdara", Belgrade, Serbia

Apstrakt

UVOD: Emfizematozni pijelonefritis je akutna, nekrotizirajuća infekcija bubrežnog parenhima i perirenijuma izazvana sa uropatogenim mikroorganizmima koji koristeći nekrotično tkivo kao substrat produkuju gas. Javlja se isključivo kod odraslih osoba, pretežno kod dijabetičara. Mortalitet oboljenja se kreće do 43%. Drugi mogući kontributivni faktori u nastanku ovog oboljenja predstavljaju imunosupresivna stanja i opstrukcija uretera ili ureteropijeličnog segmenta izazvana urolitijazom ili nekrotičnom papilom. Emfizematozni pijelonefritis se klinički najčešće prezentuje kao težak oblik akutnog pijelonefritisa koji se ni na intenzivnu antibiotiku terapiju ne smiruje unutar tri dana. Ponekad je klinička slika oboljenja atipična pa je dijagnostika i diferencijalna dijagnostika emfizematoznog pijelonefritisa teška i na njega se isprva, pri prijemu pacijenta najčešće ne posumnja. Dijagnoza se uspostavlja na osnovu anamneze, podataka o komorbiditetu, kliničke slike, ehosonograma, nativnog snimka abdomena i urotakta i CT-a abdomena. Lečenje ovog oboljenja se sastoji najpre u intenzivnom tretmanu septičko-endotoksinskog šoka, najčešće dekompenzovanog dijabetesa, te drenaži urinarnе opstrukcije. Nakon drenaže i sprovedene pripreme je u skoro svim slučajevima neophodna nefrektomija.

PRIKAZ SLUČAJA: Pacijentkinja stara 45 godina, prevedena sa Instituta za endokrinologiju naše ustanove, s neregulisanim dijabetesom, u prostraciji i trombocitopeniji (KKS: Le=5,5, 98,7% granulocita i 16.000 trombocita), u "hladnom" - endotoksičkom šoku. EHO pregledom viđen uvećan levi bubreg u stazi gr. IV i kolekcijama gasa u kaliksima i perirenijumu. Rđ nativnim snimkom abdomena i urotakta videna krećna senka u projekciji levog processusa transversusa L3 veličine oko 9 x 11mm i hidroaerične senke. CT-om abdomena videne kolekcije gasa u perirenijumu i potpuna destrukcija bubrežnog parenhima. Hitno je urađena ureteroskopija kojom je konstatovan inklaviran kalkulus lumbalnog dela uretera, ali endoskopska litotripsija nije bila izvodljiva zbog jake krivine uretera distalno od kalkulusa. Urađena je deblokada bubrega repozicionom Rđ nativnom snimku urotakta se vidi pravilno plasiran "double J" stent i kalkulus koji se sada nalazi u projekciji primarnog donjeg kaliksa. Potom je pacijentkinja prevedena u intenzivnu negu. Nakon sedam dana je izvedena nefrektomija. Patohistološkim pregledom je postavljena dijagnoza: "Pyelonephritis acuta abscondens et gangrenosa". Pacijentkinja je otpuštena kući zdrava.

ZAKLJUČAK: emfizematozni pijelonefritis je teška forma infekcije bubrežnog parenhima koja se u ogromnoj većini slučajeva ne može izlečiti bez nefrektomije. Oboljenje je udruženo sa sepsom i stoga neretko s trombocitopenijom ili poremećajem koagulacionog statusa koji predstavljaju kontraindikaciju za neposrednu nefrektomiju i perkutanu ugradnju nefrostomskog katetera. Retrogradnim ureterskopskim pristupom se može uspešno izvršiti drenaža urina iz bubrega i otkloniti kalkulusna opstrukcija.

INTRODUCTION

Emphysematous pyelonephritis is acute, necrotizing infection of renal parenchyma and perirenal space caused by gas producing uropatogens that use necrotizing tissue as a food (1). It occurs only in adults (2). Bilateral process is registered in approximately 10% of cases and mortality is recorded in almost 43% of cases (3). Pathogenesis is not completely understood. Because of predominant occurrence in diabetic patients (in 85% cases), it is hypothesized that high level of glucose in tissue represents substrate for *E. coli* and other microorganisms (Gram negative uropatogens, anaerobes and even fungi) that produce gas by glucose splitting (4), but this cannot be explained in non-diabetic patients. Thus, emphysematous pyelonephritis has to be comprehended as complication of severe pyelonephritis rather than separate entity (1). Other possible contributing factors in genesis of this disease are immunosuppressing conditions (hepatic cirrhosis for example) and ureteric or ureteropelvic junction obstruction caused by urolithiasis or necrotic papilla (1).

In vast majority of cases emphysematous pyelonephritis is clinically presented as severe form of acute pyelonephritis with high fever, vomiting and flank pain without resolution in spite of intensive antibiotic treatment for more than three days (1). Sometimes clinical appearance can be atypical – afebrile patient or patient in hypothermia caused by endotoxic shock or it can appear in form of acute abdomen or in form of sepsis with worsened glycoregulation, acute renal failure, thrombocytopenia and disseminated intravascular coagulation. Because of rarity, severity of disease and limited diagnostic possibilities (presence of gas as disimproving factor in echosonographic and radiographic findings interpretation, impossible application of contrast because uremia or afunctional and by infection severely destructed renal parenchyma), diagnosis and differential diagnosis of emphysematous pyelonephritis is complicated and disease can be easily misdiagnosed.

Diagnosis can be established by anamnesis, comorbidity data (diabetes mellitus), clinical appearance, echosonography, KUB and abdominal computed tomography (presence of gas collections in necrotic renal and perinephritic tissue and renal collection system and ureter, with signs of pneumaturia in non-obstructed ureter cases).

Treatment firstly consists of intensive treatment of septic/endotoxic shock and usually worsened diabetes and urgent urinary drainage (by retrograde, or percutaneous approach). In spite of that, nephrectomy is usually necessary. In rare cases affected kidney can be preserved only with drainage and conservative treatment (5).

CASE REPORT

45 years old female patient was hospitalized on endocrinology department for reasons of worsened diabetes. She was referred to urologist in prostration with no fever because of developed endotoxic shock with 5.500 Leucocytes, but 98,7% granulocytes and only 16.000 plateletes in blood count. Symptoms appeared 7 days before hospitalisation as severe left paraumbilical pain without

propagation and provocation, vomiting and diarrhea and no macroscopic haematuria. She also denied other previous diseases and surgery except of 8 years long non-insulin dependent diabetes mellitus and cholecystectomy.

Echosonography revealed 180mm long left kidney with 8mm thick and hyperechoic parenchyma, marked (grade IV) hydronephrosis and caliceal and perirenal gas collections that disimproved examination. Right kidney was 125mm long, with good parenchyma. KUB and plane abdominal radiogram revealed gas collections around kidney silhouette, with appearance of spontaneous pneumoretroperitoneum

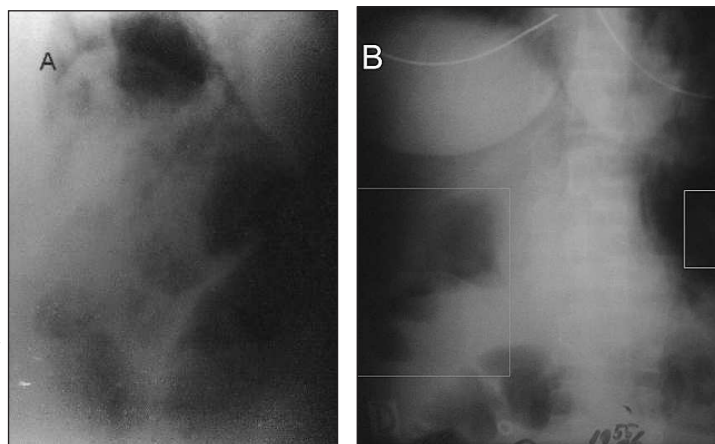


Figure 1 KUB: A – spontaneous pneumoretroperitoneum (kidney silhouette surrounded with gas collections) and B – 9 x 11mm big proximal ureteral stone near third left vertebral process transversus and hydroechoic shadows over small bowel and ascending part of colon (framed areas)

(figure 1a), 9 x 11mm calcium-dense shadow in the level of proximal ureter and third left vertebral process transversus and hydroaeric shadows over small bowel and ascending part of colon (figure 1b).

Computed tomography demonstrated crescent gas collections in perirenum, complete destruction of kidney parenchyma and unclear parenchyma-sinus boundary

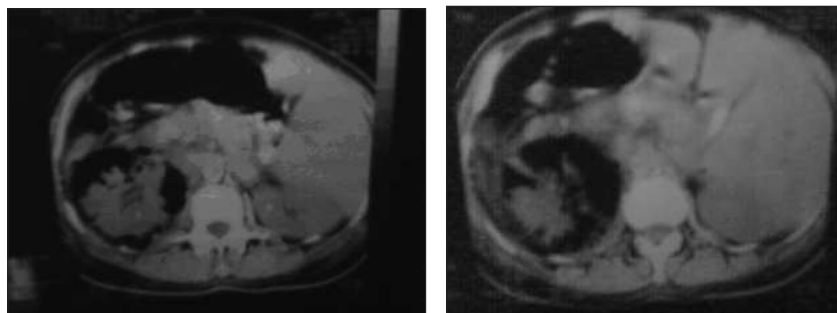


Figure 2 Computed abdominal tomography: crescent gas collections around the kidney with destructed renal parenchyma

After being transferred from Endocrinology department the urgent ureteroscopy was performed with no "Swiss Lithoclast" ballistic lithotripsy success because due to sharp ureteric curve proximal of stone. After the stone has been pushed back with ureteral catheter and purulent urine appeared, culture specimens have been taken. After procedure "double J" stent has been inserted. Control KUB demonstrated good position of "double J" stent.

After ureteroscopic urine drainage, patient has been transferred to intensive care where she has been treated empirically with three antibiotics and intravenous rehydration; blood, plasma and platelet transfusions, oxygen and

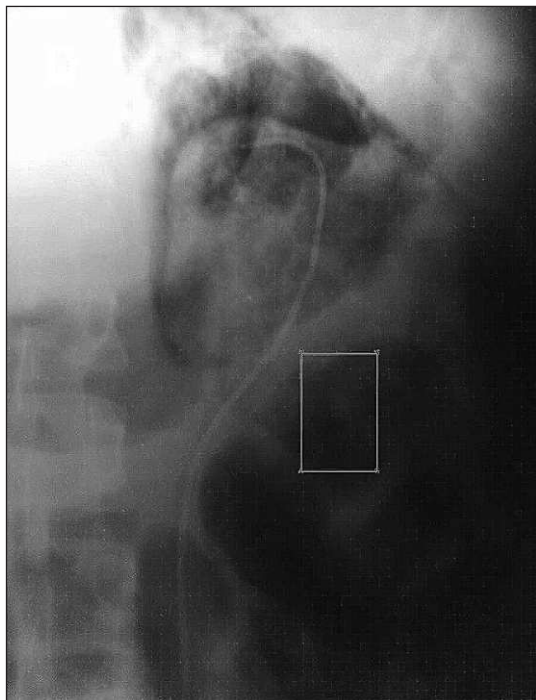


Figure 3 KUB after "double J" left ureteral stenting: evidence of good "double J" stent position, with stone pushed back from proximal ureter to primary lower calix (framed zone on radiogram). Small diameter between "double J" stent and kidney silhouette denotes severe degree of kidney parenchymal atrophy

insulino-therapy. After seven days of urgent ureteroscopy elective nephrectomy has been performed. During kidney preparation perinephritic abscess was found, and its pus taken for culture. Kidney was bulky, so its preparation was very complicated. In order to make debulking of the kidney, multiple punctures (protected with purse string sutures) has been performed, but only small amount of pus which was also microbiologically analysed has been aspirated. After removal, kidney was assessed pathohistologically.

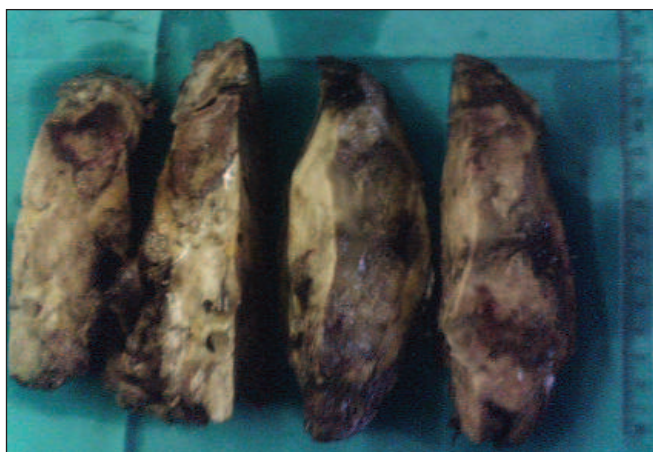


Figure 4 Pathoanatomy: completely destroyed renal parenchyma, replaced with solid tissue without recognisable renal cavities and cortex-sinus boundary

Figure 4 demonstrates completely destroyed renal parenchyma, no clear differentiation of cortex-sinus boundary, without recognisable renal cavities which explains presence of very little amount of pus taken by multiple punctures and aspirations. Pathohistological examination revealed final diagnosis: "Pyelonephritis acuta abscednens et gangrenosa". Due to antibiotic administration few days before hospitalisa-

tion in our hospital all microbiological findings were falsely sterile. Postoperative course passed without any complication that could be anticipated regarding disease nature and comorbidity, so patient was dismissed from our hospital urology department completely recovered.

DISCUSSION

Emphysematous pyelonephritis as a rare form of pyelonephritis is described in literature in form of case reports or descriptive and retrospective studies based on small number of patients conducted in a period of more than one year in vast majority of cases. Emphysematous pyelonephritis is disease oftenly accompanied with massive, severe and necrotising infection of renal parenchyma so it is usually equalised with gangrenous pyelonephritis as our case which is also pathohistologically confirmed in that manner. Consequently, we share opinion that therapy of emphysematous pyelonephritis impossible without nephrectomy. Symptomatology and diagnostic signs of sepsis are persisting even after surgical, percutaneous or ureteroscopic drainage of renal collecting system with mortality rate up to 75% in cases of late nephrectomy, (6, 7, 8, 9) which in rare cases can be performed laparoscopically (10). In some cases of nonmassive necrosis of renal tissue, emphysematous pyelonephritis can be completely resolved conservatively, with kidney preservation (11). Nielsen and al. (12) even described a case of emphysematous pyelonephritis accompanied with nephrolithiasis with successful kidney preservation after PCNL. It is still questionable if immediate nephrectomy, or delayed nephrectomy, after kidney drainage and patient reanimation better therapeutic choice. Septic shock is oftenly complicated with thrombocytopenia (as in our case) or DIC (13) that represent a form contraindication for urgent open surgery. Kidney drainage can be minimally invasive, performed in a form of percutaneous nephrostomy or in a form of retrograde (ureteroscopic) catheterisation of collecting system. Opposite of Chen M.T. (14) and Yamaguchii S. (15) we share attitude of Matsuda D. (16) and Okamoto T. (17) et al. who advocate retrograde-transurethral drainage. Pathoanatomical finding of our case described in figure 4 confirms Matsuda's D. (16) and Okamoto's T. (17) attitude. Percutaneous nephrectomy in our case could provoke dissemination of infection or hemorrhage, having in mind presence of severe thrombocytopenia. On the contrary, ureteroscopy allows not only less invasive approach, but also can permit ureteral or ureteropelvic junction stone removal by its intentional retrograde propulsion or lithotripsy in cases of emphysematous pyelonephritis caused with calculous obstruction of ureter or ureteropelvic junction.

CONCLUSION

Emphysematous pyelonephritis is severe and destructive infection of renal parenchyma, that cannot be resolved without nephrectomy in vast majority of cases, accompanied with septic shock and thus not rarely complicated with thrombocytopenia and impaired blood coagulation that contraindicates immediate nephrectomy or percutaneous nephrostomy. By retrograde, especially ureteroscopic approach, drainage of renal collecting system or calculous obstruction removal (by lithotripsy or retrograde stone propulsion) can be successfully performed.

Abstract

INTRODUCTION: Emphysematous pyelonephritis is acute, necrotizing infection of renal parenchyma and perirenal space caused by gas producing uropatogens. It occurs only in adults, predominantly in diabetic patients with mortality in almost 43% of cases. Other possible contributing factors in pathogenesis are immunosupriming conditions and ureteric or ureteropelvic junction obstruction caused by urolithiasis or necrotic papilla. In vast majority of cases emphysematous pyelonephritis is clinically presented as severe form of acute pyelonephritis without resolution in spite of intensive antibiotic treatment for more than three days. Sometimes clinical appearance is atypical, so diagnosis is complicated and disease can be misdiagnosed. Diagnosis can be established by anamnesis, comorbidity data, clinical appearance, echosonography, KUB and abdominal computerized tomography. Treatment consists of intensive treatment of sepsis, usually worsened diabetes and urinary drainage, but nephrectomy is necessary in almost all cases.

CASE REPORT: 45 years old female patient hospitalized with worsened diabetes, afebrile, with 5.500 leucocytes, but 98,7% granulocytes and 16.000 platelets in blood count. Echosonography revealed bulked left kidney with marked hydronephrosis and caliceal and perirenal gas collections. KUB and plane abdominal radiogram revealed appearance of spontaneous pneumoretroperitoneum, 9 x 11mm big left lumbar ureteral stone and bowel hydroaeric shadows. Computed abdominal tomography showed crescentic gas collections in perirenum, and complete kidney parenchyma destruction. Urgent ureteroscopy was performed, but lithotripsy was unsuccessful due to sharp ureteric curve. Retrograde stone propulsion and "double J" stent indwelling was done. Control KUB demonstrated good position of "double J" stent. After ureteroscopic urine drainage, patient has been transferred to intensive care. After seven days nephrectomy has been performed. Pathohistology revealed: "Pyelonephritis acuta abscondens et gangrenosa". Patient was dismissed from our hospital completely recovered.

CONCLUSION: Emphysematous pyelonephritis is severe and destructive infection of renal parenchyma that cannot be resolved without nephrectomy. It is accompanied with sepsis and thus not rarely with thrombocytopenia or impaired blood coagulation that contraindicates immediate nephrectomy or percutaneous nephrostomy. By retrograde, especially ureteroscopic approach, drainage of renal collecting system or calculus removal can be successfully performed.

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