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Case report: Bilateral emphysematous pyelonephritis with pneumatosis intestinalis

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ABSTRACT

Emphysematous pyelonephritis is a severe and rare gas producing infection in the renal parenchyma or collecting system. Diabetes mellitus is an established risk factor. Treatment options include antibiotics, minimally invasive management such as percutaneous drainage, or nephrectomy. This case report describes a 40-year-old female with newly diagnosed diabetes and cirrhosis who presented with left sided emphysematous pyelonephritis. After initial management with intravenous antibiotics and drainage of the collecting system, this patient's clinical condition worsened, and subsequent repeat imaging revealed bilateral emphysematous pyelonephritis as well as pneumatosis intestinalis.

1. Introduction

Emphysematous pyelonephritis (EPN) is a life-threatening urologic emergency characterized by a gas producing microbial renal infection. Symptoms include acute cystitis, an elevated white blood cell (WBC) count and positive blood cultures.[1]

EPN is associated with severe morbidity and mortality, and the most common predisposing risk factors are diabetes and urinary tract obstruction. Patients presenting with thrombocytopenia, altered mental status, and shock are associated with a higher mortality rate.[1]

Typically, EPN has been associated with a mortality rate up to 78%, however over the past several decades changes in detection and management have reduced mortality to 21%.[2] Treatment options include medical therapy alone with antibiotics, antibiotics plus minimally invasive management (MIM) such as percutaneous drainage or insertion of double J stents, or nephrectomy. There has been a gradual shift towards a nephron preserving approach.[2]

Here, we present a case of bilateral EPN in a patient with newly diagnosed diabetes and cirrhosis.

2. Case report

A 40-year-old female with history of obesity presented to an outside hospital with confusion and one week of dysuria. She had a CT scan showing left EPN and was transferred to our tertiary care center for evaluation.

Upon admission to the Medical Intensive Care Unit (ICU), the patient was awake and oriented. She denied hematuria, flank pain, chest pain, shortness of breath, or a history of nephrolithiasis.

The patient was afebrile, tachycardic, normotensive, tachypneic, and saturating well on room air. Significant lab findings included a WBC of 34, platelet count 66, lactate 5.8, BUN 56, creatinine 3.22, glucose 485, alkaline phosphatase 498, AST 39, ALT 43. On presentation, Hemoglobin A1C was 7.6. Urine and blood cultures grew *E. coli*. The physical exam was notable for suprapubic tenderness. CT imaging studies revealed gas within the parenchyma of the upper left kidney and a 4 mm stone within the proximal left ureter without hydronephrosis. There was air in the collecting systems of both kidneys, without definite parenchymal gas on the right. CT also revealed a heterogenous liver with cirrhotic nodules, and a moderate amount of abdominal ascites (Fig. 1). There was no drainable collection.

The patient was diagnosed with Child Class C cirrhosis and started on rifaximin and lactulose. Workup was not completed; however, it was felt nonalcoholic fatty liver disease seemed most likely given the patient's body mass index and history. Her Model for End-Stage Liver Disease (MELD-Na) score was 31,which was likely skewed by her renal function in the setting of EPN, though even assuming normal renal function her MELD-Na would be 19.[3] Regardless, the patient had a guarded prognosis.

Initial clinical management included intravenous fluids, insulin drip, and Zosyn. That afternoon, the patient was taken to the operating room for insertion of bilateral double J stents. Retrograde pyelogram showed

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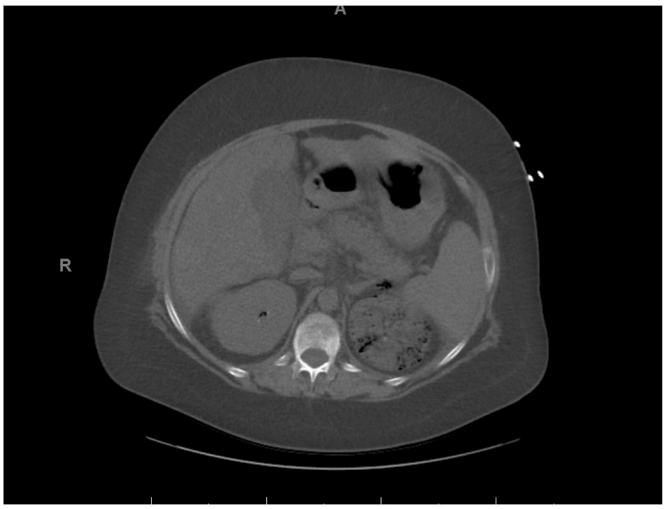

Fig. 1. Gas within the parenchyma of the upper left kidney.

no evidence of hydroureteronephrosis. A foley was left in place for maximal drainage. During the immediate perioperative course, the patient had worsening acidosis and required continued intubation. The patient returned to the ICU in stable condition.

Despite initial improvement, on hospital day three, the patient required dialysis, bicarbonate drip, and pressors. Tachypnea and low volumes prevented extubation. The family was updated, who wished to proceed with surgery only if percutaneous drainage was not possible.

A repeat CT scan was obtained to assess for a drainable collection. CT showed worsening left EPN with near complete necrosis of the left renal upper pole, new right EPN, and new right colon pneumatosis and portal venous gas (Figs. 2 and 3). Planning began for definitive surgical intervention for emergent left nephrectomy. General surgery was consulted regarding necessity of bowel resection. In light of the patient's comorbidities there was concern that surgery would be fatal. In discussion with urology, general surgery, medical ICU, and the patient's family, the decision was made that comfort measures were appropriate.

That evening the patient was terminally extubated and died soon after.

3. Discussion

Given the overall rarity of EPN, there is a paucity of management guidelines.

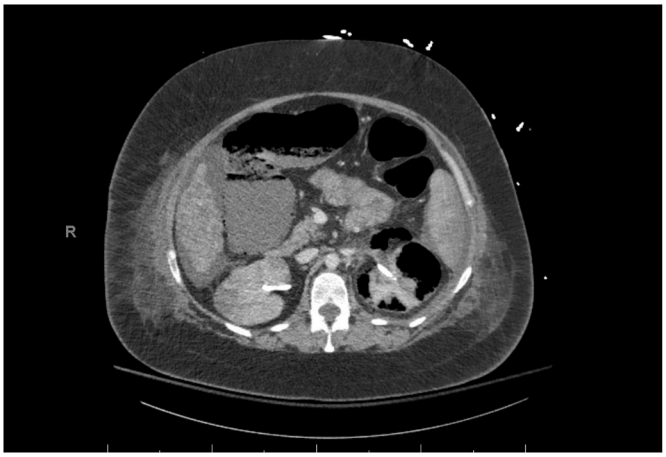

Fig. 2. Worsening left EPN and new right colon pneumatosis and portal venous gas.

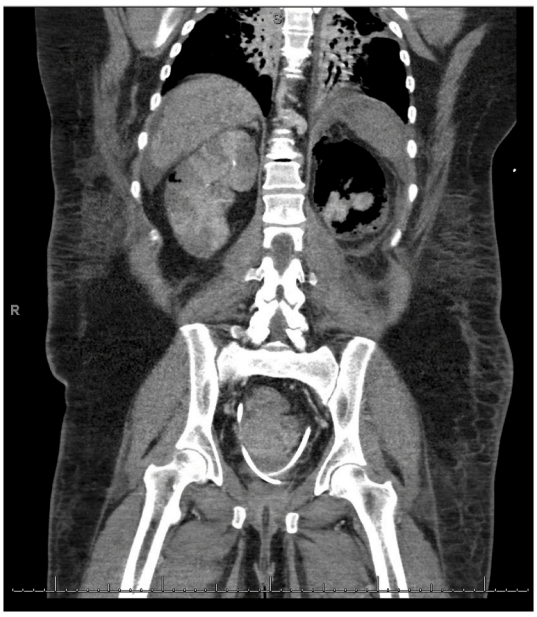

Fig. 3. Progressive left emphysematous pyelonephritis, new gas in the parenchyma of the right kidney as well as wedge shaped infarcts.

With a trend towards a nephron sparing approach, the difficult question for urologists involves when a nephrectomy is warranted. A meta-analysis by Somani et al. showed that mortality from MIM alone (13.5%) and MIM followed by elective nephrectomy (6.6%), were significantly lower than medical management with antibiotics alone (50%) or emergent nephrectomy alone (25%), $p < 0.001$. Nephrectomy after MIM was performed for prolonged sepsis or fever, but they did not report time-specific clinical guidelines.[2] Based on these findings, antibiotics and MIM are recommended for the treatment of choice for a majority of patients. They did not directly compare ureteral stent and percutaneous drainage. With deterioration after that point, they suggest consideration of nephrectomy.

Similarly, Huang et al. recommends percutaneous drainage for class one or two EPN and/or relief of urinary tract obstruction along with antibiotics. For class three or four, for patients with <2 risk factors, they recommend attempt of percutaneous drainage along with antibiotics, however nephrectomy should be promptly considered for EPN with a fulminant course or unsuccessful minimally invasive management.[1]

In this case, CT showed Huang's class two EPN and multi-organ dysfunction ensued, which progressed to Huang's class four bilateral EPN and new pneumatosis intestinalis. Of the risk factors for prognosis (thrombocytopenia, renal dysfunction, shock, or confusion), our patient had all four.[1]

A unique aspect of our case is the availability of sequential CT imaging two days apart. While the patient initially improved with aggressive medical therapy and drainage of the collecting system, this improvement was most likely in response to supportive care and may have masked a progressive necrotizing infection. An earlier second CT may have prompted nephrectomy sooner, though given our patient's severe comorbidities it is difficult to predict if she would have survived surgery. Many reviews suggest clinical response as the trigger for emergent nephrectomy.[4] One algorithm by Adapala et al. suggests repeat imaging after three days,[5] which would have been too late in our patient's case.

It is clear larger prospective studies are needed to define a clear management algorithm for EPN, incorporating serial CT imaging, perhaps even daily in selected cases, to evaluate a response to treatment.

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None.

Statement of ethics

The patient's next of kin provided an informed consent for this case report and agreed to the publication of details and figures related to the case.

Declaration of competing interest

"The authors have no conflicts of interest to declare."

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