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(CASE REPORT)

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# A case series on emphysematous pyelonephritis of north Kerala: Case series and review of literature

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## Abstract

Background-Emphysematous pyelonephritis (EPN) is an acute necrotizing infection of the renal parenchyma and surrounding tissues resulting in gas formation in the renal parenchyma, collecting system and perinephric tissue. It's a life- threatening infection if not detected and treated promptly. High mortality rate in EPN is due to its septic complications. The most common predisposing factors to this entity are uncontrolled diabetes mellitus, immune incompetence, and urinary tract obstruction. While clinically difficult to detect, radiological confirmation can help clinch the diagnosis. A plain x-ray can raise suspicion of abnormal gas shadow in the renal fossa, based on which ultrasonography or computed tomography can be performed. The latter confirms the presence of intrarenal gas and supports the diagnosis of EPN. As the most common gas-forming organisms are E.coli and Klebsiella, treatment can be initiated after a radiological suspicion to prevent sepsis and morbidity. Here, we present 5 cases of EPN diagnosed by various radiological modalities and brief review of existing literature.

**Keywords:** Renal abscess; Necrotising infection; Klebsiella

## 1. Introduction

Emphysematous pyelonephritis is a gas- producing acute infection of the renal parenchyma collecting system and perinephric area. EPN is commonly seen in patients with uncontrolled diabetes. It's a life-threatening infection and carries a mortality rate of ~ 80%. It is caused by gas-forming bacteria, most commonly associated with E. coli while infection with klebsiella, clostridium, cryptococci, aspergillus and candida can also lead to EPN. Early detection and aggressive medical and interventional approach are advised for this condition.

## 2. Case 1

A 63-year-old female patient with history of hypertension and newly detected diabetes came with complaints of right flank pain and nausea for 1 week with no associated fever. There was renal derangement, with serum creatinine of 3.4 mg/dl and uric acid values of 10.7 mg/dl. Ultrasound imaging of the patient revealed a globular, bulky right kidney with altered echotexture, mild to moderate hydroureteronephrosis and a right-sided mid pole renal echogenicity with dirty shadowing of ~ 15mm with perinephric inflammation, suspicious of emphysematous pyelonephritis (EPN). The CT findings showed a bulky right kidney with ill-defined hypodense areas showing multiple pockets of air and air-fluid levels within the upper and lower pole (figure 1&2). The air foci were seen extending outside the renal outlines into the perinephric and paracaval regions and adjacent to the upper pole, upto the sub-diaphragmatic region. Along with this, there was minimal perinephric and proximal periureteric fat stranding.

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The patient underwent DJ stenting and right percutaneous nephrostomy on the same day. The post-operative period was uneventful. She improved rapidly with this intervention and she was discharged after a week.

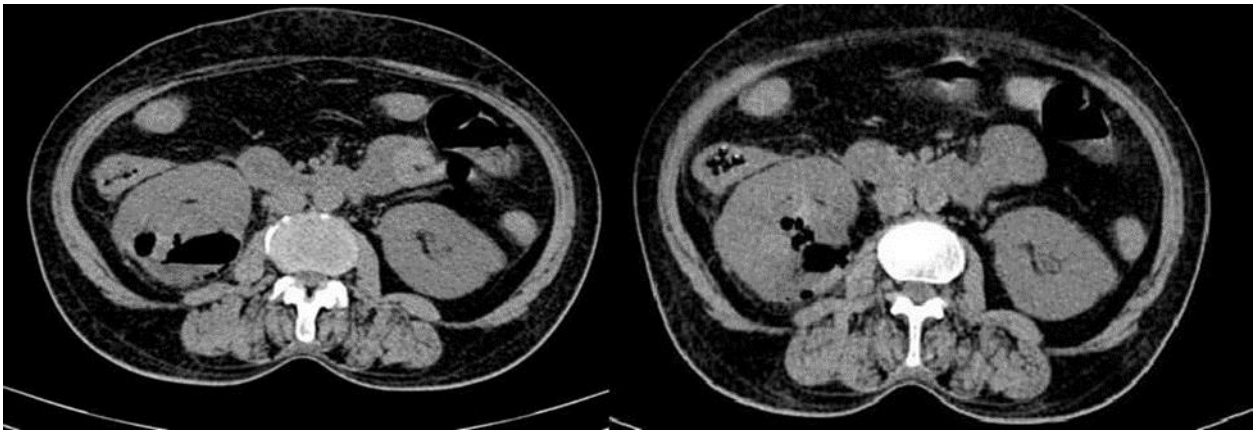

**Figure 1 a and b** NCCT KUB showing bulky right kidney with ill-defined hypodense areas with air foci and air-fluid levels within the renal calyces.

Diagnosis: Emphysematous pyelonephritis with abscess (Class 3B).

### 2.1. Case 2

**Figure 2** NCCT KUB showing few pockets of air foci and significant surrounding fat stranding in the left kidney. Right kidney is bulky with multiple hypodense areas within.

A 68-year-old female, a known case of chronic kidney disease with sub-optimally treated diabetes mellitus presented with increased tiredness and increased urine frequency for 1 week associated with mild fever and burning micturition. Patient also gives a history of recurrent urinary tract infections, uremic gastritis, and multiple hospital admissions. The patient's serum creatinine was 5.6 mg/dl and serum potassium levels were 5.1mmol/l, with haemoglobin of 7.6 gm/dl. Urine culture was ordered with revealed extended spectrum beta-lactamase (ESBL) klebsiella. The patient also underwent a bilateral DJ stenting 3 months back.

Computed tomography revealed that the right kidney is bulky in size, globular in contour with reduced parenchymal density. Right mild to moderate hydroureteronephrosis is noted with abrupt tapering of the ureter just distal to the iliac

vessel crossing. The left kidney also showed reduced parenchymal density with few pockets of air foci in the upper calyceal system (Figure 3). Minimal hydroureteronephrosis is seen on the left side. Bilateral perinephric and periureteric fat stranding with few enlarged para-aortic lymph nodes were also noted. The patient was later referred to a higher center for further evaluation.

Diagnosis: Emphysematous pyelonephritis (Class 2).

## 3. Case 3

A 61-year-old lady with history of poorly controlled diabetes mellitus and acute kidney injury (AKI) came to medicine OPD with complaints of excessive vomiting, right sided flank pain and reduced urine output since 3 days. Patient gives history of urolithiasis since 5 years. Investigation showed her totals count around 12510 cells/mm3 and serum creatinine 3.4 mg /dl. On urine routine examination numerous pus cells were found, urine culture yielded heavy growth of ESBL eschericia coli. Emergency computed tomography (CT) showed an enlarged right kidney with reduced parenchymal density. Moderate hydroureteronephrosis was noted with pockets of air foci within the collecting system (Figure 4). A calculus with central air foci is seen at the right pelviureteric junction (PUJ) (Figure 5). Peri ureteric and perinephric fat stranding is also noted along with thickening of Gerotas fascia and the lateroconal fascia. Left sided mild – moderate hydroureteronephrosis noted with air foci within the collecting system. Mild perinephric fat stranding was also seen.

The urinary bladder wall is mildly thickened. Multiple intraluminal air foci were seen along the wall of the urinary bladder. Few intramural air foci are seen at places. Mild peri vesical fat stranding was noted. Air foci are also seen within the urethra. Patient was immediately started on broad-spectrum antibiotics on the basis of her culture sensitivity reports.

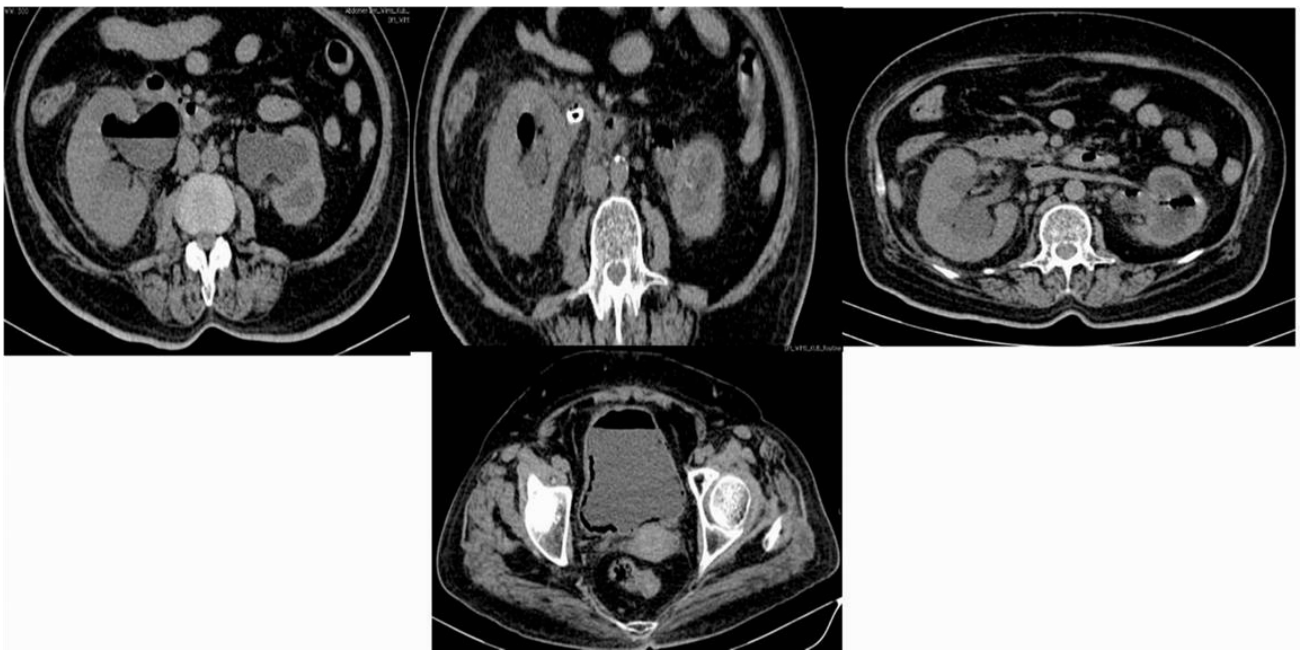

**Figure 3 a** NCCT KUB showing an enlarged right kidney with moderate hydronephrosis and large pockets of air foci within the collecting system. **Figure 3 b**: NCCT KUB showing an enlarged right kidney with a PUJ calculus showing a central air focus. **Figure 3 c:** NCCT KUB showing an air focus within the dilated calyceal system of the left kidney with mild perinephric fat stranding. **Figure 3 d:** Mildly thickened urinary bladder wall with intraluminal air foci along the wall of the urinary bladder

Diagnosis: Emphysematous pyelonephritis with emphysematous cystitis (Class 4)

## 4. Case 4

A 56-year-old male patient with history of diabetes mellitus on insulin since last 8 years and bilateral urolithiasis came to medicine OPD with complaints of left flank pain, radiating from loin to groin, associated with reduced urine output

and vomiting since 1 week. Investigation showed his total counts 15650 cells/mm$^3$ ,serum creatinine 6.2 mg/dl and serum sodium level 122 mmol/l. Urine culture revealed the presence of citrobacter koseri (CC>10^5 CFU/Ml).

On emergency computed tomography (CT), the left kidney was bulky with reduced attenuation density. There were perinephric and periureteric fluid and fat stranding with thickening and stranding of left anterior and posterior pararenal fascia and lateral conal fascia (Figure 8). Multiple renal and ureteric air foci were seen. Retroperitoneal air was seen in the perinephric and paranephric space.

The patient underwent left sided DJ stenting in the next day and tolerated the surgery well. Patient was discharged with IV antibiotics and was advised for follow-up.

**Figure 4** NCCT KUB showing an enlarged left kidney with perinephric and peri ureteric fat stranding, perinephric fluid, proximal ureteric calculi, perinephric air foci and thickening of adjacent fascia. Diagnosis: Emphysematous pyelonephritis (class 3B)

## 5. Case 5

A 70-year-old female patient with history of poorly treated type 2 diabetes mellitus, hypertension and K/C/O carcinoma breast status post unilateral mastectomy came to emergency department complaining of dysuria, pyuria left flank pain and high-grade fever for 1 week. Patient's serum creatinine was 1.5 mg/dL and total WBC counts were 11660 cells/ mm$^3$. Her urine routine showed presence of numerous bacteria. On urine culture, she was found to have growth of multidrug-resistant organism, metallo-beta-lactamase (MBL) E coli (CC>10^5 CFU/Ml)

On emergency CT KUB, the left kidney showed reduced parenchymal attenuation with multiple pockets of air foci within the pelvicalyceal system and in the renal parenchyma (Figure 9). An ill-defined hypodense collection noted in the mid pole of left kidney. The collection was seen extending outside the renal outlines forming an ill- defined pararenal collection. Mild perinephric fat stranding, perinephric fluid and thickening of perirenal fascia was noted. (Figure 9)

The patient underwent left sided DJ stenting in the next day and tolerated the surgery well. The patient was discharged with IV antibiotics and was advised for follow-up.

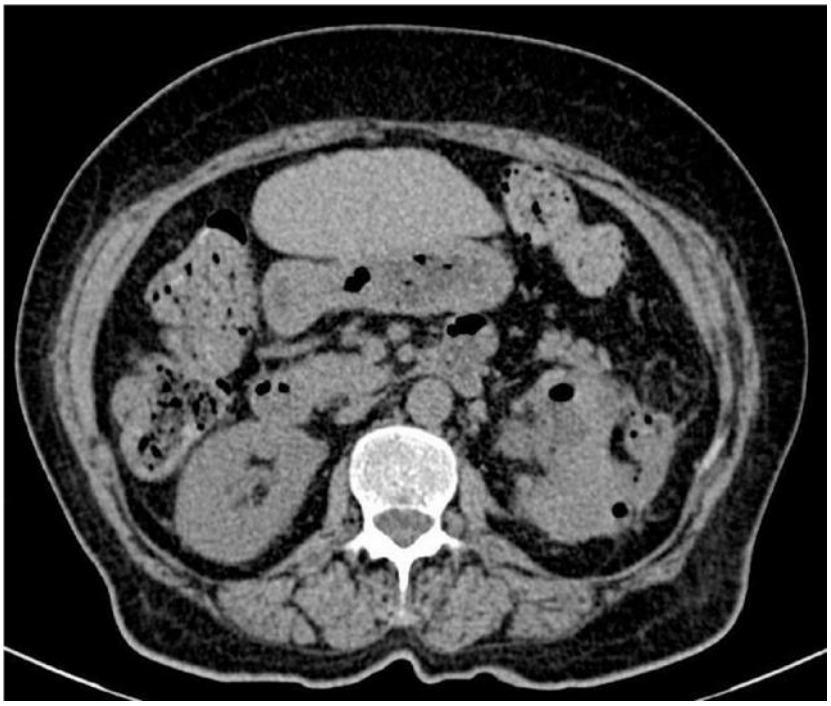

**Figure 5** NCCT KUB showing an enlarged left kidney with few pockets of air foci within the calyceal system and in the renal parenchyma.

Final Diagnosis: Emphysematous pyelonephritis (Class 2)

## 6. Discussion

Emphysematous pyelonephritis is a severe life-threatening necrotizing infection of the renal parenchyma that causes gas accumulation within or around the kidneys. It's caused by infection of the collecting system by gas forming bacteria. These gas forming microorganism's produce carbon dioxide and hydrogen. This gas can be trapped within the collecting system and that could be trapped by urinary calculi (1). This is a medical emergency which needs prompt medical intervention.

There is a degree of ambiguity surrounding the terminology used to identify the relevant involving renal gas. Emphysematous pyelonephritis is a term that should only be used for infections of the renal tract that are accompanied by intraparenchymal renal gas. Emphysematous pyelitis is the correct name for gas that is limited to the renal pelvis, and perinephric emphysema is the correct name for gas that is confined to the space surrounding the kidney.

Kelly and Maccullum described a case of gas forming necrotizing renal infection in the year 1898(2).Multiple names were used for this infection as renal emphysema, pnuemonephritis and emphysematous pyelonephritis.

We have done a study on 5 patients who were diagnosed by EPN in our department.

All of our patients presented with fever and flank pain initially. Ashok kumar sokhal et al conducted a study among 74 patients and found that fever followed by flank pain was the most common initial presentation of EPN (3).

In our study we included 5 patients in which four of them were above 60 years and were women which is in accordance by a study by raiz ahammed et al. In their study they found that nearly all of the patients were women and e coli was the most frequently isolated pathogen and nearly third of the patients had bilateral disease. (2)

In a study which included 48 patients of EPN jj huang et al found that acute renal infection with e coli or klebsiella in patients with diabetes mellitus is the main reason for developments of EPN(4). In our study all the 5 patients had increased total counts, creatinine values and were having uncontrolled diabetes mellitus. Out of the 5 patients 3 of them showed growth of e-coli on urine culture and one had klebsiella and the other showed citrobacter koseri.

In our study approximately 20 percent of the patients only had bilateral disease (1 out of 5).

Emphysematous pyelonephritis is diagnosed using plain abdomen radiography because it delineates air in the renal collecting system better than ultrasonography. Ultrasonography is often the primary radiological investigation. [17, 18, 19, 20, 21, 22] Plain radiographs can show air in the renal collecting system, but superimposed intestinal gas can obscure visibility and reduce accuracy. Gas in the retroperitoneum and renal or perinephric abscesses can also mimic emphysematous pyelonephritis. Ultrasonography is limited because gas in the kidney and/or renal pelvis resembles renal calculi and causes reverberation echoes and shadowing. [23, 24, 25]

CT scanning detects emphysematous pyelonephritis best. Non-enhanced CT scans show intraparenchymal, intracalyceal, and intrapelvic gas and perinephric space extension, mottled areas of low attenuation radially around the pyramids, and pus in the renal pelvicalyceal system. [8, 19, 26, 27, 28, 29] CT scans detect air in the renal tract and show renal and perirenal structure and infection spread to perinephric tissues. [19, 8, 30, 31].Reflux from the bladder and bronchorenal, enterorenal, and cutaneorenal fistulae are not always visible on CT images. Xanthogranulomatous pyelonephritis and localised renal abscesses can also mimic these findings [32, 33, 34]

In emphysematous pyelonephritis, renal function is diminished or absent on the affected side, and radionuclide scan is a good way to detect differential function before nephrectomy. Antimicrobial therapy responses are assessed using scintigraphy. Intravenous urography may be needed for renal intervention.

Emphysematous pyelonephritis evaluation is restricted by radionuclide studies' non-specificity. Moreover, radionuclide imaging is scarce. However, it does not cause false-positive or false negative diagnosis.

**Table 1** Classification of EPN

| CLASSIFICATION | YEAR | EPN CLASSIFICATION | EXAM |
|---|---|---|---|
| Langston and Pfister | 1970 | **-Diffuse mottling of the renal parenchyma**<br>**-Bubbly renal parenchyma surrounded by crescent-shaped gas in the perinephric space**<br>**-Extension of gas through the Gerota fascia** | X ray and pyelogram |
| Michaeli | 1984 | **Stage I** - Gas within the renal parenchyma or the perinephric tissue<br>**Stage II** - Presence of gas in the kidney and its surroundings<br>**Stage III** - Extension of gas through Gerota's fascia or bilateral EPN | X-Ray<br>IVP |
| Wan | 1996 | **Type I** - parenchymal destruction, with streaky or mottled parenchymal gas and absence of fluid collection<br>**Type II** - renal or perirenal fluid collection, with bubbly gas collection in the perinephric space or collecting system | Plain radiography, computed tomography, ultrasonography |
| Huang and Tseng | 2000 | **Class 1** - Gas confined to the collecting system<br>**Class 2** - Gas confined to the renal parenchyma alone<br>**Class 3A** - Perinephric extension of gas or abscess<br>**Class 3B** - Extension of gas beyond the Gerota fascia<br>**Class 4** - Bilateral EPN or EPN in a solitary kidney | CT and Ultrasound |
| Al-Geizawi | 2010 | **Stage 1**: Gas in the collecting system<br>**Stage 2**: Gas replacing <50% of renal parenchyma, with minimum spread to the surrounding tissues; Sepsis rapidly controlled<br>**Stage 3**: Gas replacing >50% of renal parenchyma; or Extensive spread of infection in the perinephric area; or Patient with evidence of multiple organ failure, uncontrolled sepsis, or shock not responding to medical management | Ultrasound + CT |

MRI is not used to diagnose emphysematous pyelonephritis. MRI findings are signal-free on T1- and T2-weighted images. Nevertheless, renal calculi or rapidly flowing blood might cause signal voids on MRI scans, resulting in false positives. Perinephric and intraparenchymal fluid accumulation are seen on MRI. [19, 20]

On the basis of radiological findings on CT scan jj huang et al classified EPN into 4 classes,

Huang and Tseng staging protocols: [7]

- Class 1: Gas in the collecting system
- Class 2: Gas restricted to renal parenchyma
- Class 3A: Perinephric gas or abscess extension

- Class 3B: Gas extension beyond the Gerota's fascia
- Class 4: Bilateral or single-kidney EPN

### 6.1. Distinctive diagnosis

Emphysematous pyelitis (EP) is distinguished from pyelonephritis by renal pelvis gas. These illnesses have similar pathophysiologies and are linked to diabetes mellitus. [35]

Emphysematous and xanthogranulomatous pyelonephritis (XP) are both septic conditions that involve stones in a non-functioning kidney with a severe gram-negative infection. Xanthogranulomatous pyelonephritis may also create gas in the renal parenchyma and perinephric space, but not as much as EPN. Co-occurrence of xanthogranulomatous and emphysematous pyelonephritis has been documented. [36, 37]

#### *6.1.1. Differential diagnosis*

- Abdominal viscous retroperitoneal perforation
- Psoas abscess due to gas-forming microbes
- Reflux of bladder air
- Bronchorenal, enterorenal, or cutaneorenal fistulas may arise with xanthogranulomatous pyelonephritis.
- Air in a localised renal abscess—not fatal.

Gas in the renal parenchyma may be visible on CT scanning in circumstances other than emphysematous pyelonephritis. Intraparenchymal renal gas may develop from urologic intervention like nephrostomy insertion or a gastrointestinal-renal fistula. Like emphysematous pyelonephritis, these conditions are not life-threatening. Radiologists, especially in emergencies, should be aware of this unusual finding and its differential diagnosis as abdominal CT scanning becomes more common.

Emphysematous pyelonephritis can show as a urinary bladder adenocarcinoma in a middle-aged, nondiabetic patient. [38]One of our case showed a rare finding of an air foci within the calculus which is in accordance with a study done by Peter et al were they found an air foci within a staghorn calculus(6).

A study done by MA Badaway et al over 16 years from 2000 to 2016 in 54 patients found that the presence of air locule volume >54 cc and hydronephrosis were associated with 12 and 5.5 folds of risk of failure of conservative treatment(7).

Most recent literature advocates aggressive medical management along with percutaneous drainage and partial or complete nephrectomy as the treatment for EPN. Hudson et al. [40] were the first to demonstrate that PCD might be used as a therapeutic option or EPN due to significant advancements in the percutaneous catheters employed. In further case studies, PCD was successfully employed in conjunction with MM to treat patients, significantly lowering mortality rates [41,42]. In roughly 70% of cases, PCD aids in maintaining the afflicted kidney's functionality. Patients with localised regions of gas and those who have functioning renal tissue should undergo PCD. The placement of a pigtail drain should be at least 14 Fr in size, ideally under CT guidance because it has a higher success rate than ultrasonography. PCD is not contraindicated in the presence of an abscess with loculations or several abscesses because multiple catheters can be used to drain each loculation [43]. The abscess should be the first place that PCD is directed because it is technically simpler to reach and would greatly relieve strain on the viable renal tissue.

The drainage catheters should be left in place until a follow-up CT demonstrates that the EPN characteristics have resolved; during this time, the tubes can be flushed with antibiotic treatments if necessary.

## 7. Conclusion

In our study we found that EPN is a life threatening disease which has to be detected as early as possible.

EPN commonly seen in aged women and the major organism causing EPN is Eschericia Coli.

Patients usually presents with fever and flank pain with underlying history of uncontrolled diabetes or renal calculi and with deranged renal function test values. In such patients, early imaging can be essential in guiding the clinicians towards the diagnosis and initiation of empirical treatment.

Early aggressive management of EPN is advised as it can save the patient's life.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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