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Squamous Cell Carcinoma of the Renal Pelvis Presented as Post-Obstruction Pyonephrosis: A Commonly Known Malignancy in a Rare Location

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Abstract

Squamous Cell Carcinoma (SCC) of the renal pelvis is an extremely rare clinical entity, constituting less than 1% of all urinary tract neoplasms. The diagnosis is usually not possible due to inconclusive clinical and radiological features. Chronic urothelial irritation due to recurrent infection and long-standing nephrolithiasis would encourage squamous metaplasia of the renal collecting system and predispose to such aggressive nature of the tumor. A typical manifestations and consequent delay in diagnosis may result in locally advanced or metastatic disease upon presentation and poor prognosis and outcomes. We report a case of incidentally diagnosed squamous cell carcinoma of the renal pelvis in a 47-year-old man, presented with asymptomatic bilateral renal stones. Post-right percutaneous nephrolithotomy, the patient developed severe intermittent left hypochondrial pain with spikes of fever with rigidity extending from below the left costal margin to the groin and crossing the middle line. Urethral catheter drained pure pus while the right nephrostomy tube drained clear urine. Left nephrectomy performed with intra operative drainage of ~2200 cc of pus from such a huge kidney containing multiple variable sized impacted calculi. Histopathology revealed moderately differentiated T4NxMx squamous cell arising from the renal pelvis. The case highlights the importance of careful gross and histopathological assessment of any specimen when associated with hydro or pyonephrosis secondary to a long-standing history of nephrolithiasis and an associated poor or non-functioning kidney. In the current report, a case will be presented with its clinical and morphological features together with a review of the relevant current literature.

Keywords: Squamous cell carcinoma; Kidney; Renal pelvis; Calculi; Pyonephrosis

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Introduction

Primary malignant tumors of the renal pelvis constitute 8% to 14% of all the renal neoplasms. Transitional Cell Carcinoma (TCC) is the most common malignancy arising from the renal collecting system, occurs in more than 90% of such cases, whereas Squamous Cell Carcinoma (SCC) has a reported incidence of only 0.5% to 0.8% [1,2]. A typical presentation and consequent delay in diagnosis may result in locally advanced or metastatic disease with a poor prognosis and outcomes of such a highly aggressive neoplasm, compared to TCC of the upper urinary tract [3]. SCC of the renal pelvis thought to be secondary to metaplasia of the urothelium due to chronic irritation, inflammation and infection from renal calculi, even when they are non-obstructive by nature, although it may occur without apparent etiological factors [4]. If neglected or inadequately treated, renal calculi may lead to recurrent infection, deterioration of renal function and risk of developing urosepsis. When tumor is complicated with kidney stones, symptoms and even diagnostic work-up may flop to differentiate between renal stone and upper tract SCC. Surgery is the mainstay of therapy in SCC of renal pelvis, with no established benefits from chemotherapy and the management still individualized, depending on sparse case reports. As only few cases have been reported to date, the current report will present the clinical and morphological features of this tumor with a review of the relevant available literature.

Case Presentation

A 47-year-old man presented with asymptomatic bilateral renal stones. He is a stone-passer since 25-years and has a history of hypertension and familial Parkinsonism, with no history of smoking. BMI was 21.13 kg/m^2 with unremarkable laboratory work-up, apart from high serum

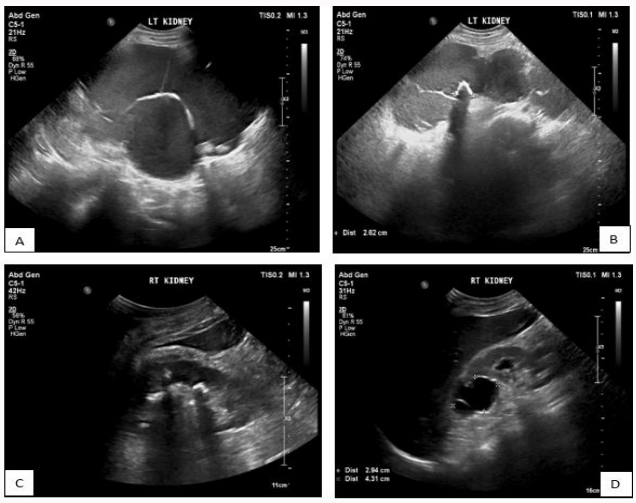

Figure 1: Abdominal ultra sonography reveals multiple left renal calculi with diffuse loss of parenchyma [A&B], while the right kidney contains multiple obstructing renal calculi and large upper pole cyst measuring 3 cm × 4 cm [C&D].

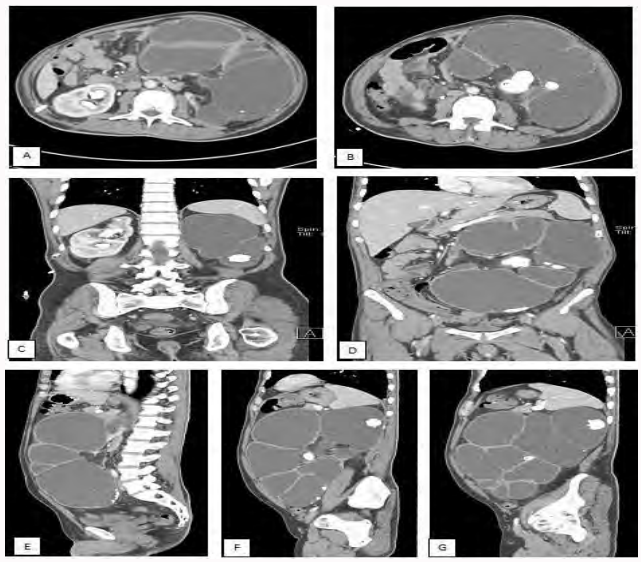

Figure 2: Multiple axial computed tomography after intravenous contrast injection, including sagittal (C-E) and coronal sections (F&G) showing multiple stones both kidneys. The left kidney measures 25-cm in CC dimension with severe cortical thinning and marked hydronephrosis.

uric acid (8 mg/dL), with a baseline serum creatinine of 1.25 mg/dL. Abdominal ultrasonography revealed multiple bilateral obstructing renal calculi with diffuse loss of left renal parenchyma and 3 cm × 4 cm upper pole cyst in the right kidney (Figure 1). Renal DMSA scan sowed scintigraphic evidence of inhomogeneous cortical tracer distribution, consistent with cortical scarring at the lateral border of the right kidney while the left kidney was not visualized and did not contribute to the total renal function. Abdominal CT revealed showing bilateral multiple obstructive staghorn renal calculi. The left kidney measures 25 cm in CC dimension with severe cortical thinning and marked hydronephrosis. An abnormal worrisome left upper pole renal hypodensity measures about 1.6 cm × 3.6 cm was noticed with small residual medially located left upper pole cortex. An enlarged left para-aortic lymph node measures 1.3 cm was also observed (Figure 2).

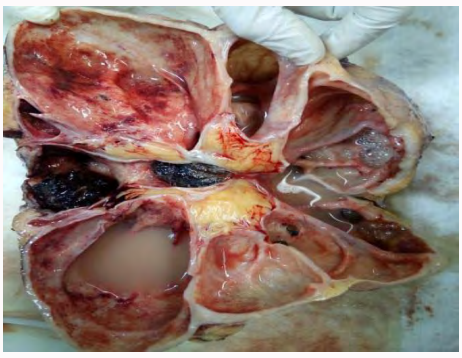

Figure 3: Left kidney specimen weighed 1130 gm and consisted of a single cystic enlarged kidney measured 16 cm × 12 cm × 6.0 cm, with intact capsule. On opening, a yellowish color fluid oozed out with multiple variable sized, black-colored impacted calculi, and the largest of them measured 4.5 cm × 3.5 cm × 1.0 cm. The collecting system was hugely dilated with thin parenchyma.

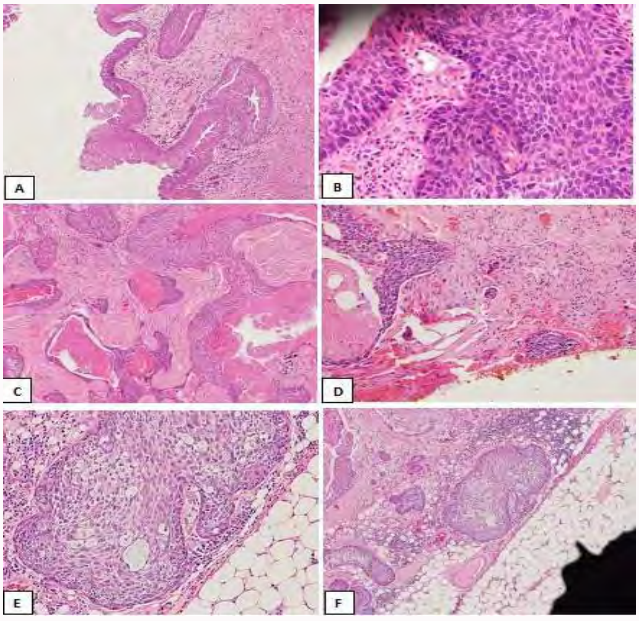

Figure 4: Histopathological examination revealed squamous metaplasia [A] and dysplasia (40x) [B], squamous cell carcinoma [C], near the resection margin [D] and invading the perinephric fat [E&F].

Patient underwent right percutaneous nephrolithotomy, followed by flexible ureteroscopy for residual multiple right kidney stones, with bilateral ureteral stenting. Postoperatively, the patient developed severe intermittent left hypochondrial pain with spikes of fever up to 39°C. Clinically, there was right hypochondrial tenderness and rigidity extending from below the costal margin to the groin and crossing the middle line. Urethral catheter drained pure pus while the right nephrostomy tube drained clear concentrated urine. Serum creatinine elevate up to 1.9 mg/dL and gradually descent to average values. Last lab chemistry was unremarkable apart from decreased total protein (5.4 g/dL) with hypoalbuminemia (1.81 g/dL), high alkaline phosphatase (140 U/L), with normal serum creatinine (1.22 mg/dL) and complete blood count.

The patient underwent left nephrectomy through a flank incision and resection of the 12th rib, with intraoperative drainage of ~2200 cc of pus from the huge kidney, which was opened inadvertently. Specimen weighed 1130 gm and consisted of a single cystic enlarged

kidney measured 16 cm × 12 cm × 6.0 cm, with intact capsule. On opening, a yellowish color fluid oozed out with multiple variable sized, black-colored impacted calculi, the largest of them measured 4.5 cm × 3.5 cm × 1.0 cm (Figure 3). Histopathological examination revealed moderately differentiated p4NxMx squamous cell carcinoma (3.5 cm × 2.0 cm × 1.7 cm) arising from the renal pelvis, reaching up to the renal capsule, in the background of chronic pyelonephritis and squamous metaplasia. Tumor was 2-mm away from the Gerota's fascia and free resected ureteral margin (Figure 4).

Discussion

Primary SCC of the renal collecting system represent rare of rarity condition, where it represents a rare presentation of malignant tumors of the renal pelvis, which are relatively rare, with urothelial carcinoma occurs in more than 90% of cases [5]. At presentation, SCC of the renal collecting system tends to be sessile, ulcerated, infiltrative and at a higher clinical stage, representing more aggressive disease when compared to other histologies [6]. SCC of the urothelial tract seems to arise from metaplasia of the urothelium due to chronic irritation of the urothelium mostly by nephrolithiasis and recurrent infection, and less commonly by exogenous or endogenous chemicals, hormonal imbalance, schistosomiasis, smoking and vitamin A deficiency. However, some cases have been reported with no apparent etiological factor [7]. Staghorn stone has been reported to be a leading cause of SCC of the renal pelvis with a grave prognosis [8].

Early detection of the neoplasm by imaging studies may be difficult due to non-specific finding of the tumor, such as calcification or hydronephrosis [9].

The present index case was presented by huge hydronephrosis with multiple renal calculi, followed by extensive pyonephrosis with almost complete loss of renal parenchyma. Similar presentation has been previously reported with pyonephrosis and peritoneal abscess formation [10]. However, in the present case the diagnosis of SCC was incidentally discovered after nephrectomy, where the diagnosis was difficult to explain preoperatively, with no specific radiological features that allow for preoperative diagnosis. Beside the chronic obstruction caused by long-standing stones and recurrent infection, necrotic materials sloughed tumor tissue might have obstructed the ureter and had caused hydronephrosis. Consequently, the kidney was transformed into a bag of pus with neglected treatment and persistent obstruction. This would highlight the importance of meticulous screening for malignancy in patients with long-standing history of renal calculi, even non-obstructing, in association with infection and poor functioning kidney.

Patients with SCC histology fared worse, with a median overall survival time of 10 months compared to 63 months for TCC. After correction for age, gender, prior treatment, race and decade of diagnosis, the SCC had a hazard risk of 3.7 (95%CI 3.0-4.5) compared to TCC [6]. However, the authors found a comparable survival of TCC and SCC stage by stage, when appropriate staging done according to lymph node status. In the present case, the disease was localized with no involvement of perirenal tissue and no evidence of distant metastasis or regional lymph node disease. Therefore, surgery might be curative in this patient with no need for systemic chemotherapy, which has only marginal benefit in these cases. The currently used combination of cisplatin, methotrexate, and bleomycin fails to show any survival benefit. It seems that more case reports and pathological studies are still needed to establish the risk factors for primary SCC of the renal collecting system.

Conclusion

SCC of the renal pelvis may have unpredicted presentation, such as hydronephrosis or pyonephrosis secondary to a long-standing history of nephrolithiasis and an associated poor or non-functioning kidney. Careful gross and histopathological assessment of any suspicious area should be considered, especially in cases with predisposing conditions such as recurrent infection and obstructing renal calculi.

References

1. Odabas O, Karakok M, Yilmaz Y, Atilla MK, Akman E, Aydin S. Squamous cell carcinoma of the kidney. East J Med. 2000;5(1):35-6.
2. Kalayci OT, Bozdag Z, Sonmezgoz F, Sahin N. Squamous cell carcinoma of the renal pelvis associated with kidney stones: radiologic imaging features with gross and histopathological correlation. J Clin Imaging Sci. 2013;3:14.
3. Berz D, Rizack T, Weitzen S, Mega A, Renzulli J, Colvin G. Survival of patients with squamous cell malignancies of the upper urinary tract. Clin Med Insights Oncol. 2012;6:11-8.
4. Talwar N, Dargan P, Arora MP, Sharma A, Sen AK. Primary squamous cell carcinoma of the renal pelvis masquerading as pyonephrosis: a case report. Indian J Pathol Microbiol. 2006;49(3):418-20.
5. Mizusawa H, Komiyama I, Ueno Y, Maejima T, Kato H. Squamous cell carcinoma in the renal pelvis of a horseshoe kidney. Int J Urol. 2004;11(9):782-4.
6. Pham A, Colvin G, Berz D. Squamous cell malignancies of the upper urinary tract: a survival study and a systematic review of the literature. J Cancer Sci Ther. 2011;S4:001.
7. Talwar N, Dargan P, Arora MP, Sharma S, Sen AK. Primary squamous cell carcinoma of the renal pelvis masquerading as pyonephrosis: A case report. Indian J Pathol Microbiol. 2006;49(3):418-20.
8. Jongyotha K, Sriphrapradang C. Squamous Cell Carcinoma of the Renal Pelvis as a Result of Long-Standing Staghorn Calculi. Case Rep Oncol. 2015;8(3):399-404.
9. Holmäng S, Lele SM, Johansson SL. Squamous cell carcinoma of the renal pelvis and ureter: incidence, symptoms, treatment and outcome. J Urol. 2007;178(1):51-6.
10. Chung SD, Lai MK, Chueh SC, Wang SM, Yu HJ. An unusual cause of pyonephrosis and intra-peritoneal abscess: Ureteral urothelial carcinoma. Int J Infect Dis. 2009;13(1):e39-40.