

CASE REPORT**INCIDENTAL SQUAMOUS CELL CARCINOMA OF THE RENAL PELVIS IN A NON FUNCTIONING KIDNEY THAT WAS MISSED ON TWO NON-CONTRAST CT-SCANS****Masroor Hassan, Asim Qureshi**

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Although the second most common malignancy after urothelial carcinoma, squamous cell carcinoma (SCC) of the renal pelvis is a rare entity. It has strong association with nephrolithiasis thus emphasizing prompt treatment of renal calculi. Because of rarity and nonspecific clinical and radiological findings, it mostly presents at pT3 or higher stage. We report SCC of renal pelvis that was missed two times on non-contrast CT scans and was diagnosed incidentally in a nephrectomy specimen. Its prognosis is similar to that of urothelial carcinoma of comparable stage. Owing to its rarity, no standard treatment guidelines are available; however radical nephrectomy with lymph node dissection is the initial treatment which can be curative in an early stage disease. Adjuvant chemotherapy and radiotherapy are usually ineffective.

Keywords: Squamous cell carcinoma, Imaging, Schistosoma haematobium

J Ayub Med Coll Abbottabad 2017;29(3):489–92

INTRODUCTION

Squamous cell carcinoma (SCC) rarely involves renal pelvis and is mostly associated with nephrolithiasis and pyelonephritis but can occur in the absence of any causative factor. It is mostly diagnosed at higher stage and has poor prognosis. Surgical management is required and usually curative for early stage disease but adjuvant chemotherapy and radiotherapy are usually ineffective.

CASE REPORT

A 45-year male patient, known case of bilateral kidney stones since childhood, who previously undergone pyelolithotomy and extra corporal shock wave lithotripsy (ESWL), came to emergency department with severe right flank pain with no fever, vomiting, burning micturition or weight loss. He also underwent bilateral double J stenting one week ago for renal decompression. Dipstick urine test showed 3+ leukocytes and 1+ blood. Total leukocyte count was 11500/mm³. Serum creatinine was 1.22 mg/dl and blood urea 28 mg/dl. Serum calcium (9.8 mg/dl) and phosphate (4.2 mg/dl) were also normal. The non-contrast CT scan abdomen and pelvis revealed small sized right kidney with fragmented staghorn calculus, severe cortical scarring more pronounced at the upper pole, impacted calculus in right proximal ureter, moderate hydronephrosis, perinephric and proximal periureteric strandy changes and thickened Gerota's fascia. Mild hydronephrosis and a calculus at the lower pole of left kidney was also seen along with few prominent precaval and aortocaval lymph nodes. The liver, spleen, gallbladder, seminal vesicles and prostate appeared unremarkable. A non-contrast CT performed six months back showed almost similar

findings. Urine culture and sensitivity revealed no growth after 48 hours of incubation. He underwent right nephrectomy for non-functioning kidney and the specimen was sent for histopathological examination.

We received 309 grams nephrectomy specimen measuring 9.8×7.3×6.0 cm. A small attached portion of ureter measured 1.0 cm in length and 0.5 cm in diameter. No renal vein was identified. The kidney was firm with intact outer capsule having multiple small notches. Upon opening the specimen along anti-hilar border, a large well circumscribed tan white tumour (7.0×5.0×4.5 cm) was seen which had almost entirely replaced the renal parenchyma (figure1). Grossly it was 0.2 cm away from the renal capsule. A fragmented staghorn calculus (4.0×3.9 cm) was seen in the pelvis along with multiple small calculi in the extensively dilated calyces. Histopathological examination revealed a tumor comprised of polygonal cells with prominent intercellular bridges, exhibiting pleomorphism and hyperchromatism (Figure-2)

Multiple keratin pearls and areas of calcification were seen. The tumour invaded peri-pelvic fat and renal parenchyma (figure3) and was 1mm away from renal capsular margin. The ureteric margin was free of tumour. Squamous metaplasia of renal pelvis was seen (Figure-4) while the background kidney showed marked glomerulosclerosis, tubular atrophy and interstitial fibrosis. It was diagnosed as moderately differentiated squamous cell carcinoma of stage pT3 Nx (as no lymph nodes were submitted). The tumour revealed only squamous differentiation and no transitional component was identified on extensive sampling.

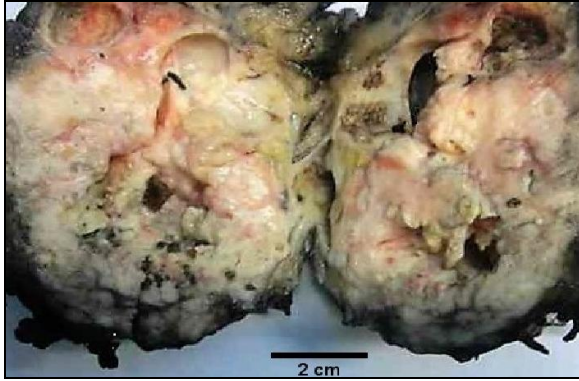


Figure-1: Gross appearance of tumour

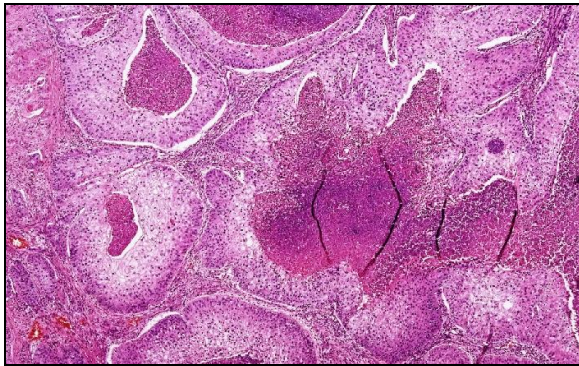


Figure-2: H and E section of SCC of renal pelvis

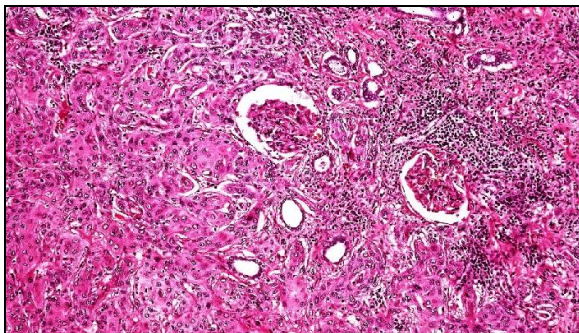


Figure-3: H and E section of tumour invading into the renal parenchyma

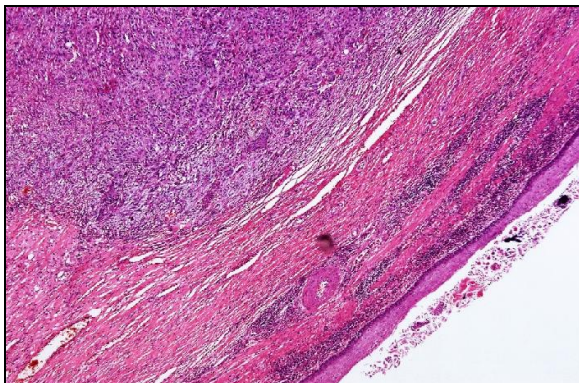


Figure-4: H and E section showing squamous metaplasia in the lower right corner. Tumour is present in the upper left portion.

DISCUSSION

Renal pelvis is a rare site for a commonly known Squamous Cell Carcinoma (SCC). It accounts for 6.1% of upper urinary tract tumours.¹ In the middle east where schistosomal infection is endemic, it is the commonest one.² So far approximately 300 cases of SCC of renal pelvis have been reported with a strong association with nephrolithiasis.¹ Females are more commonly affected than males mostly in the 5–7th decade.³

Nephrolithiasis and chronic pyelonephritis are the two commonly reported etiological factors for SCC of renal pelvis³, the former found in our case also. Others include schistosomiasis, vitamin A deficiency, smoking and exogenous and endogenous chemicals, but it may occur without any predisposing factors.³ Our patient was non-smoker and no schistosomal eggs were identified on histopathological examination.

Microscopically, SCC of renal pelvis is similar to SCC at other sites, showing polygonal cells with intercellular bridges, keratin pearl formation and keratotic cellular debris³ but only those tumours should be diagnosed as SCC which predominantly shows keratin formation.² The term 'urothelial carcinoma with squamous differentiation' should be reserved for those urothelial tumours which show variable squamous differentiation.² In contrast to urothelial carcinoma, most SCC shows deeper invasion at the time of diagnosis. Squamous metaplasia of adjacent urothelium is present in majority of patients showing that chronic irritation, inflammation and infection progress through metaplastic process to dysplasia and carcinoma.⁴ Our case also showed squamous metaplasia of adjacent urothelium.

Imaging findings of SCC of renal pelvis includes either a distinct solid mass with hydronephrosis and calcifications or an infiltrative lesion, hence difficult to differentiate it from xanthogranulomatous pyelonephritis (XGP) or other malignant renal neoplasms.⁵ Rarely XGP can coexist with SCC.⁶ In our case, there was moderate hydronephrosis and no lesion was identified on two non-contrast CT scans performed six months apart. Moreover, there was no clinical suspicion of carcinoma. According to Lee *et al*, although presence of enhancing extraluminal and exophytic mass and, in some cases, an intraluminal component are helpful radiological (CT) features of renal SCC, but because of impracticability of performing CT scan in every patient with renal calculi, IVU will be preferred option.⁷ On IVU, the presence of filling defects, delay in appearance

of dye, or thickening of renal parenchyma should raise the suspicion of renal tumour despite the absence of mass effects and preserved renal contour, which should then be followed by CT scan or renal pelvis biopsy.⁷ Raghavendran M *et al* concluded that a screening CT scan should be performed in all patients having long history of renal stones with a poor functioning kidney on urogram or haematuria.⁸ Thus there may be no evidence of SCC both clinically as well as radiologically,⁷ the possibility of SCC should always be kept in mind while operating a patient who has long history of renal calculi or infections. As humoral hypercalcemia of malignancy caused by parathyroid hormone related protein (PTH-rP) is also a rare finding in SCC of renal pelvis, so serum calcium is also important while investigating patients of chronic renal calculi.⁹

So far, only two cases of SCC of renal pelvis diagnosed with fine needle aspiration cytology (FNAC) have been reported in literature. The first one was initially diagnosed with urine cytology and FNAC and later on confirmed with histopathology of nephrectomy specimen¹⁰ while the second one was initially diagnosed with FNAC and then confirmed with core biopsy and nephrectomy.¹¹ Thus FNAC of renal pelvis is also extremely helpful in accurately diagnosing SCC especially when performed in a multidisciplinary setting.¹⁰ Because of immunosuppression, renal transplant patients are more prone to malignancies, hence a single case report of renal SCC of native kidney in a transplanted patient has also been described in literature.¹² Bones are rare sites (three case reports so far) of metastasis in SCC of renal pelvis.¹³ Furthermore, both SCC and renal cell carcinoma can coexist in the single kidney.¹⁴

More than 90% cases of SCC of renal pelvis are initially diagnosed at pT3 or higher stage¹⁴ however their prognosis is similar to that of urothelial carcinoma of comparable stage.^{2,15} Berz *et al* concludes that although SCC of renal pelvis present at higher stage with inferior overall survival, stage by stage survival is similar to that of urothelial carcinoma.¹⁶ Our case was also pT3 as the tumour had invaded into the peripelvic fat and renal parenchyma.

Patients with SCC of the renal pelvis have a median survival of 7.25 months with a 5-year survival rate of 18%.¹⁷ No standard treatment guidelines are available owing to its rarity however radical nephrectomy with lymph node dissection is the initial treatment.¹⁵ Adjuvant chemotherapy and radio-chemotherapy can be beneficial in selected patients.¹⁷ Because longstanding staghorn calculi also cause life threatening sepsis and renal

impairment in addition to squamous metaplasia and SCC, so prompt treatment of these calculi is required.⁹ Biochemical analysis of stone, metabolic evaluation along with achieving urine output of greater than 2.5 litter per day will decrease recurrence of renal calculi.⁹

In summary, we report a case of incidental squamous cell carcinoma in a non-functioning kidney that was missed on two non-contrast CT scans. Because of non-specific clinical and radiological features, its possibility should always be kept in mind by nephrologists, urologists and pathologists.

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Received: 28 December, 2016

Revised: 21 March, 2017

Accepted: 24 May, 2017

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