

## PRIMARY SQUAMOUS CELL CARCINOMA OF RENAL PELVIS ASSOCIATED WITH RENAL CALCULUS AND RECURRENT PYONEPHROSIS

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### ABSTRACT

**INTRODUCTION:** Primary Squamous Cell Carcinoma in the kidney is a rare malignant neoplasm associated with nephrolithiasis, typically monobacterial pyonephrosis and rarely Xanthogranulomatous pyelonephritis. It is an aggressive disease with a poor prognosis mostly due to lack of presenting clinical features like a palpable mass, gross haematuria and pain. We report a case presenting with renal calculus and pyonephrosis managed initially with percutaneous nephrostomy followed by nephrectomy due to complete loss of renal function. Histopathological evaluation revealed poorly differentiated squamous cell carcinoma which is managed by chemotherapy, although initially beneficial, patients later develop disseminated metastatic disease which holds a poor prognosis.

**KEYWORDS:** Carcinoma, Squamous Cell; Pyelonephritis, Nephrolithiasis.

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**INTRODUCTION:** Primary neoplasms of the renal collecting system are uncommon and account for about 5% of all urothelial tumors;<sup>[1]</sup> and squamous cell carcinomas are seen only in 0.7%-7%;<sup>[2]</sup> it is a rare malignancy of the upper urinary tract and it occurs six times more frequently in the renal pelvis than in the ureter. These are moderately to poorly differentiated tumours which are more likely to be invasive and in advanced stages at the time of their diagnosis.<sup>[3,4]</sup> A very few number of cases have been reported in literature and we report a case of primary squamous cell carcinoma of the renal pelvis.

**CASE REPORT:** A 30-year-old female presented to the outpatient clinic with the chief complaints of pain in the right flank for 5 months and fever from the past 1 week. The pain was continuous and dull aching in nature, and fever was continuous in nature and relieved on medications. She complained of burning micturition and anorexia for the past 1 week. There were no complaints of gross haematuria. She had history of renal calculus disease in the past but did not undergo surgery. She had history of similar symptoms 6 months back for which she was managed conservatively in a secondary health centre.

On general physical examination, she was lethargic and running a temperature of 102 degrees Fahrenheit, heart rate of 110/min, blood pressure of 100/60 mm Hg and a respiratory rate of 16/min., Pallor was present. Per abdominal examination did not reveal any significant findings.

In view of past history and general condition of the patient she underwent routine haematological, urine and radiological analysis. Her complete blood count revealed a Hemoglobin of 9.1, TLC count of  $33.69 \times 10^3/\mu\text{L}$ ; neutrophilic leucocytosis with toxic granules on a peripheral blood smear; serum electrolytes analysis revealed potassium levels at 2.50 mMol/L, chloride at 99 mMol/L and sodium at 139.00 mMol/L. Renal function test revealed blood urea and serum Creatinine 67.00mg% and 3.2mg% respectively. Ultrasonography of the abdomen revealed renal calculus and an ill-defined heterogeneous mass lesion with internal echos in the right renal fossa and a poorly visualised right kidney with internal echoes and paper thin cortex and few echogenic foci were noted along in an enlarged left kidney (141x59mm), revealing a picture of pyelonephritis. Intravenous Pyelogram depicted a dilated PCS with staghorn calculus in the right kidney with no flow and the visualised left collection system revealed an enlarged kidney without any defect in ureteral and bladder filling. Contrast-enhanced Computed Tomography (CECT) of abdomen revealed a poorly identified right kidney with intra and peri-renal collection and necrotic areas showing heterogeneously moderate enhancement, noted arising from renal pelvis.

The patient was taken up for percutaneous nephrostomy and frank pus was drained; she was subsequently managed on IV antibiotics and analgesic support. Culture revealed monobacterial infection by *Staphylococcus aureus* and her tissue biopsy was inconclusive. She was discharged on request after 15 days and was reviewed in the outpatient clinic for review and further management in view of nephrostomy drainage which was found blocked. Thus a repeat nephrostomy was performed and due to long standing status of the disease a <sup>99m</sup>Tc Diethylene-Triamine Pentaacetic acid test (DTPA) was scheduled, it revealed a very poorly visualized right kidney in dynamic and delayed images, a flat renogram curve and GFR at 5.45 ml/min and

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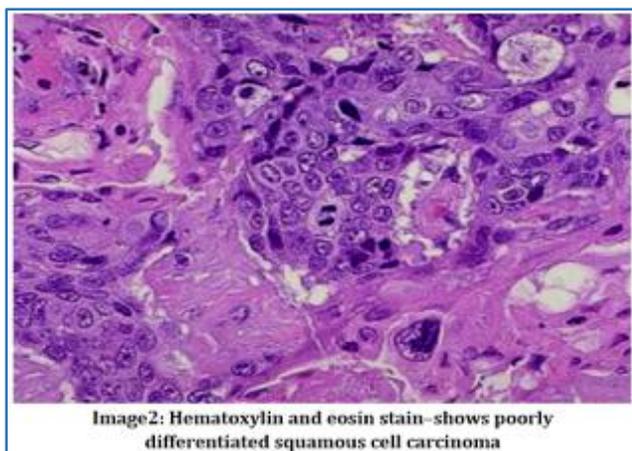
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spilt function of 6.33%.

She was electively planned for right nephrectomy and was managed uneventfully post operatively. Histopathological analysis revealed poorly differentiated squamous cell carcinoma of the renal pelvis. Patient is undergoing platinum based chemotherapy and is on regular follow up.



**Image 1: Echogenic Parenchyma, hydronephrosis with internal echoes, paper thin cortex**



**Image 2: Hematoxylin and eosin stain - shows poorly differentiated squamous cell carcinoma**

**DISCUSSION:** Renal Squamous Cell Carcinoma (RSCC) of the renal collecting system is a rare malignancy and holds a poor prognosis. It has a variable incidence of about 7-8% of all Urothelial cancers.<sup>[5]</sup> Squamous Cell Carcinoma (SCC) of the urinary tract is more frequently reported in urinary bladder and male urethra, it is rarely encountered in renal pelvis.<sup>[1,6]</sup> Pure squamous cell carcinoma is very rare though it is the second most common tumour in the renal pelvis.<sup>[1]</sup> Women are affected more frequently than men, predominant age group being 50-70 years.<sup>[7]</sup>

It is presumed that long standing renal calculi (usually staghorn calculus) causes inflammation and infection of the foci that leads to development of pyelonephritis inevitably developing squamous metaplasia and leucoplakia.<sup>[6]</sup> Other factors are exogenous and endogenous chemicals, vitamin A deficiency, schistosomiasis, smoking;<sup>[8]</sup> chronic analgesic abuse<sup>[4]</sup> and phenacetin consumption.<sup>[6]</sup> Hypercalcemia, leukocytosis and thrombocytosis have been reported as a part of paraneoplastic syndromes in RSCC cases.<sup>[9,10]</sup>

As per previous studies and existing literatures, these tumors have been classified as per their localization of the tumors as central or peripheral. Central renal cell carcinoma presents more Intraluminal components and is usually associated with lymph node metastasis whereas peripheral renal squamous cell carcinoma presents with prominent renal parenchymal thickening and might invade the perirenal fat tissue before lymph node or distant metastasis could be identified.<sup>[5]</sup>

With the presenting symptoms of the patient diagnosis of RSCC it is difficult to define the symptoms as non-specific and imaging analysis may only reveal nephrolithiasis and hydronephrosis and occasionally PUJ obstruction. Patient may present with recurrent pyonephrosis as seen in our case, even after undergoing percutaneous nephrostomy, thus considering the possibility of underlying cellular metaplasia. Thus, diagnosis is more based on histopathological features rather than signs and symptoms of the disease, as in our case only a nephrectomy revealed the underlying pathology as the cause of recurrent pyonephrosis.

Histologic presentation reveals tumors with extensive squamous differentiation, and the landmarks are pearl formation, intercellular bridges, and keratotic cellular debris and the conspicuous presence of keratinizing squamous metaplasia of the adjacent flattened urothelium, especially if associated with dysplasia, supports a diagnosis of primary SCC of the renal pelvis which is rare.<sup>[6,7]</sup>

Existing literature and previous studies reveal that metastasis is sporadic and is usually a sign of end stage disease with a poor prognosis as Nativ, et al in their study have divided renal SCC in three groups, reported 1 and 2 year survival rates of locally invasive renal SCC 33% and 22% respectively.<sup>[8]</sup>

The current modality of treatment of RSCC is primarily by Nephrectomy,<sup>[1,8,9]</sup> to rephrase Nephrectomy with or without ureterectomy even in the presence of distant metastasis.<sup>[10]</sup> Adjuvant chemotherapy or radiotherapy indicated in metastatic disease,<sup>[1]</sup> our patient had pathological staging as T3N1M0 in which radical nephrectomy with ureterectomy was done. This is the treatment of choice in these patients, even in the face of metastatic disease. There is a lack of evidence regarding survival benefit with chemo-radiation following surgery. Corral, et al in their study used a combination chemotherapy included cisplatin, methotrexate, bleomycin if the sign of metastatic disease is revealed.<sup>[11]</sup> The survival of patients with central renal squamous cell carcinoma was reported to be significantly shorter than those with peripheral renal squamous cell carcinoma.<sup>[5]</sup> However no long term studies exist in regard to patient response to chemotherapy regimens, thus no protocol exists.

Radiotherapy has been used in management of renal pelvis cancer, clinical target volume should include the renal fossa, the course of ureter to bladder, the entire bladder and the para-caval, paraaortic lymph nodes to the dose of 45-50 Gy at 1.8 to 2 Gy per day for subclinical and microscopic disease, a boost of 5-10 Gy for more extensive disease,

multiple positive nodes, microscopic positive margins, macroscopic residual margin. Our patient preoperatively has undergone nephrectomy and has received the first course of cisplatin based chemotherapy.

Furthermore, the prognosis of renal SCC is so poor that the 5-year survival rate is no more than 10% and most patients die within 1 year of surgery.

**CONCLUSION:** Primary Renal Squamous Cell Carcinoma is an aggressive tumor and literature and studies warrant a poor prognosis, thus requiring the need for an aggressive management after reviewing histopathological reports and as soon as metastasis develops. As the disease is concealed by non-specific symptoms and associated with renal stones, it is necessary to examine patients with appropriate imaging modalities for early detection and management. Thus patients presenting with an enhancing extra luminal exophytic mass or in some cases, an intraluminal component<sup>[5]</sup> should be worked up for long standing stones, filling defects and renal parenchymal thickening in intra venous pyelogram.

It is thus important to understand that careful examination of symptoms, critical imaging analysis and keeping in mind the possibility of malignancy and a confirmatory histopathological report warrants for an immediate and aggressive treatment modality by nephrectomy/radical nephrectomy followed by chemotherapy. Although the prognosis is inevitably poor it may marginally provide better results.

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