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PRIMARY SQUAMOUS CELL CARCINOMA OF KIDNEY: REPORT OF TWO CASES

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ABSTRACT

Primary squamous cell carcinoma of renal pelvis is rare clinical entity with only few cases have been reported in the literature. It is usually associated with long standing renal calculi. Insidious onset of symptom and inconclusive clinical and radiological features leads to locally advanced or metastatic disease at presentation; resulting in poor prognosis. Here we are reporting two cases of squamous cell carcinoma of kidney having renal calculi to highlight its clinical presentation and to document the association of squamous cell carcinoma in longstanding nephrolithiasis due to its rarity.

Keywords: *Carcinoma, Kidney, Renal stone, Squamous cell*

INTRODUCTION

Primary carcinoma of the renal pelvis accounts for only 4-5%of all the urothelial tumor.[1]Transitional cell carcinoma is the most common histopathological type followed by squamous cell carcinoma and adenocarcinoma.[2]
Primary squamous cell carcinoma of renal pelvis is a rare clinical entity that constitutes only about 0.5-8% of renal tumors.[3]The lack of definite clinical presentation and inconclusive imaging features result in advanced stage of presentation. Solid mass, hydronephrosis, and calcification are common but nonspecific radiological finding that explain why this tumor is not diagnosed before histopathological examination of resected surgical specimen.[4] These tumors are high grade, highly aggressive tumors with poor prognosis. Very few cases of primary squamous cell carcinoma of kidney have been reported in the literature. Here we present two cases of squamous cell carcinoma of renal pelvis due to its rarity and also to highlights the silent presentations of these tumors and the need to keep in mind the association of malignancies in patients having nephrolithiasis. This report highlights the rarity and aggressiveness of squamous cell carcinoma.

CASE SERIES

CASE: 1

A 65 years old male clinically presented in the Department of Radiation oncology of our institute with lose of weight, fever, vomiting on and off and intermittent dull aching pain in right flank of last six months. There was no significant past medical history as well as family history. Physical examination of the patient revealed mild tenderness in right renal angle. A lump of about 12×8cm was palpable in the right lumbar and hypochondrial region, which was firm, and moving with respiration. Routine haematology revealed leucocytosis and thrombocytosis. Biochemical test and chest radiograph were normal. Plain x-ray abdomen revealed multiple radiopaque shadows in the right kidney (Figure-1). Ultrasonography of whole abdomen showed hydronephrosis of right kidney with multiple renal calculi and multiple hepatic metastases. Normal architecture of right kidney was lost. Patient had undergone right nephrectomy. On gross examination right kidney was enlarged measuring 14×8×5.5cm. Cut surface revealed loss of architecture of right kidney. Medulla and cortex could not be differentiated.
Whole kidney was converted into multiple lobules with thickened septa. Multiple black hard stones were present (figure-2). Histopathology of the renal pelvis revealed moderately differentiated squamous cell carcinoma (figure-3). Patient was in low general condition and was treated symptomatically. He died due to disease 6 months after.

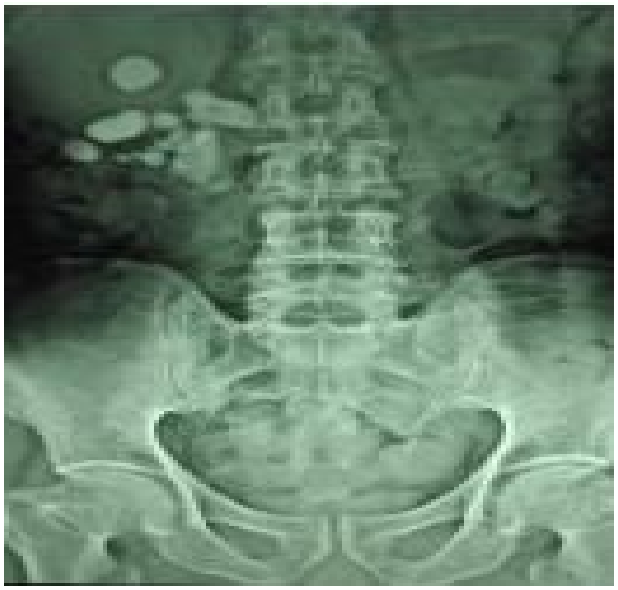

Fig 1: Plain x-ray abdomen showing multiple radiopaque shadows in the right kidney

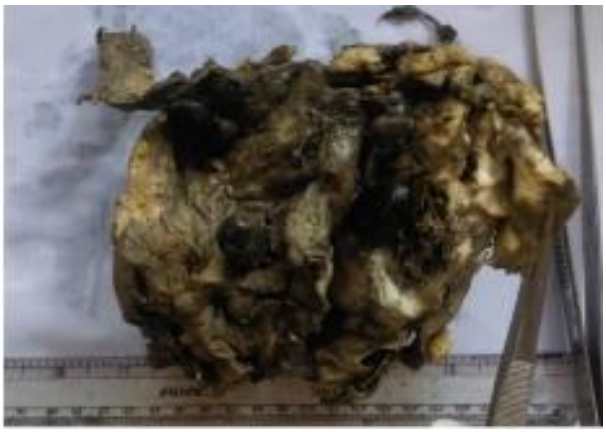

Fig 2: Gross specimen of nephrectomy

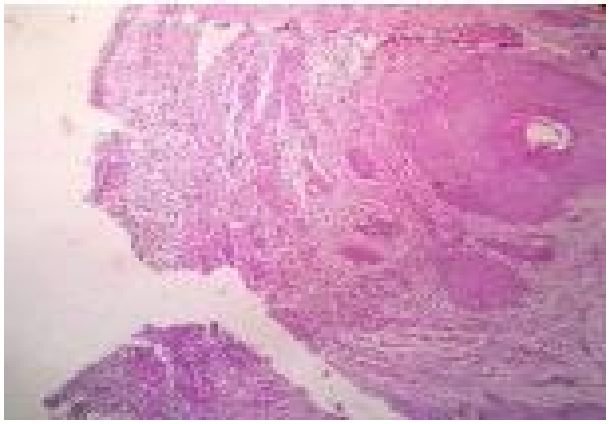

Fig 3:Photomicrograph showing Transitional epithelium of renal pelvis and underlying stroma with tumour tissue showing moderately differentiated squamous carcinoma cells.(H & E ,×10x]

CASE: 2

A 67 years old male patient presented in Radiation oncology Department of our institute with history of intermittent right flank pain of 6 months duration. There was no history of hematuria, fever or dysuria. Clinical examination showed mild tenderness in right renal angle. A mass of about 16×9 cm was palpable in right hypochondrium. It was firm and moving with respiration. Rest of the clinical examination was insignificant. Routine haematological and biochemical investigations were within normal limits. Intravenous pyelograpy showed non-functioning right kidney with a calculi in the right renal pelvis. Ultrasonography of abdomen and pelvis showed large calculi in right renal pelvis, hydronephrosis and cortical thinning of right kidney. Left kidney was normal. He underwent right nephrectomy. Grossly the right kidney was irregular, brownish, and oval shaped of 18×9×7 cm in size. Cut section showed a large brownish calculus in pelvis with surrounding whitish tissue of size 2×2×1 cm. Rest of kidney appeared cystic and distorted (figure-4). Histopathology revealed well differentiated squamous cell carcinoma and thyroidisation of renal tubules (figure-5) Patient was in regular follow up without any evidence of disease since 1 year.

Fig 4: Gross specimen of nephrectomy showing large brownish calculi in pelvis and surrounding whitish tissue.

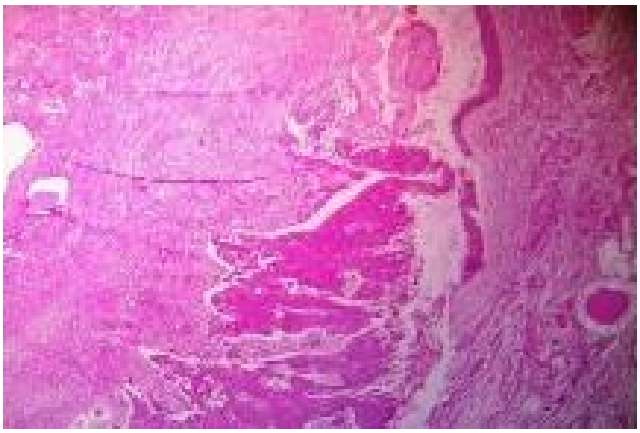

Fig 5: Photomicrograph showing squamous metaplasia, squamous carcinoma in situ and foci of invasion.stroma shows inflammation and thyroidisation of renal tubules. (H & E 100)

DISCUSSION

Squamous cell carcinoma of urinary tract more frequently reported in urinary bladder and urethra. It rarely occurs in renal pelvis. Transitional cell carcinoma is the most common tumor originating from renal pelvis followed by squamous cell carcinoma and adenocarcinoma. The median age of presentation is 57, with slight female preponderance.[5]In the present case one patient was 65years,another was 67years, and both are male. Due to lack of pathognomonic sign they present in advanced

stage resulting in poor prognosis. Common etiologic factors are renal stone, infection, chemicals; hormone imbalance, vitamin A deficiency, schistosomiasis and smoking.[6]Chronic irritation and infections are thought to induce metaplasia of urothelium which subsequently leads to squamous cell carcinoma. Li MK et. al in their study found coexisting renal stone in 100% cases.[4] Staghorn calculi being the most frequent variant. In both the reported cases patient had calculi in renal pelvis producing chronic irritation leading to squamous cell carcinoma which correlated with data given in literature. Hypercalcemia, leukocytosis and thrombocytosis have been reported as Para neoplastic syndrome in renal squamous cell carcinoma. One of our patients also had leucocytosis and thrombocytosis.

Histopathology is the hallmark of diagnosis because of lack of characteristic clinical and imaging feature. Squamous cell carcinoma of renal pelvis is diagnosed histopathologically by extensive squamous differentiation. The histologic hallmark of pearl formation, intercellular bridges and keratotic cellular debris are like those of squamous cell carcinoma at any site. Most of these carcinomas are moderately differentiated or poorly differentiated and more deeply invasive than the transitional cell carcinoma.[7]Detail work up to exclude secondary renal squamous cell carcinoma should be done. Lee et. al in their study classified squamous cell carcinoma into two groups based on the location of the tumor, central and peripheral.[8]Central one has more intraluminal component with lymph node metastasis. Peripheral variant has prominent renal parenchymal thickening invading perirenal fat tissue before lymph node or distant metastasis. Central variant has worse prognosis. One of our cases was moderately differentiated central type who had hepatic metastasis. However another patient had peripheral variant of squamous cell carcinoma.

Nephroureterocystectomy is considered the primary treatment. Surgery is the standard of care even in the faces of metastasis to establish a histological diagnosis, to control symptoms or to eliminate the source of infection.[9]In one of our case nephrectomy was done even in the presence of liver metastasis. Adjuvant treatment is considered in case of metastasis. Cisplatinum based chemotherapy and radiotherapy are usually given in advanced cases but failed to show any survival benefit. However, because of less number of cases no fixed treatment guideline is present.

It is highly aggressive tumor with unfavorable outcome. In one series 84% of tumor was locally advanced or had metastasis at the time of operation.[10] In the present study one patient had hepatic metastasis at diagnosis due to central variety. He died 6 months after the surgery. The other patient had peripheral type of lesion and surviving for last one year without any evidence of disease. Nativet. al in their study reported locally invasive renal squamous cell carcinoma had 1 and 2 year survival rate of 33% and 22% respectively.[11]They also found that treatment modalities like nephrectomy, nephrourectomy, adjuvant chemotherapy or radiotherapy did not affect survival of patients irrespective of tumor stage.

CONCLUSION

Primary squamous cell carcinoma of renal pelvis is a rare aggressive tumor with poor prognosis. Due to no definite pathognomic sign and symptoms, most of the patients presented with advanced stage. As these tumors are strongly associated with renal stones, patient with renal stones and non-functioning kidney should be evaluated with newer imaging technologies for early detection of the tumor that may lead to a better outcome for the patients.

Conflict of Interest: The authors declare that there is no conflict of interests

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