Case Report

Primary squamous cell carcinoma of kidney: A rare case report

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ABSTRACT

Neoplasms of the kidney are most commonly adenocarcinomas. Carcinomas of the renal pelvis and ureter are rare, accounting for only 4% of all urothelial cancers. Squamous cell carcinoma (SCC) of renal pelvis and ureter are very rare and account for 6-15% of all renal tumors of which transitional carcinomas constitute the majority followed by adenocarcinoma and SCC. Renal SCC (RSCC) is a rare malignancy of the upper urinary tract characteristically presenting with advanced stage, the reported incidence being 1.4% of all renal malignancies. Very few cases have been reported in the literature. Most patients have a history of chronic urolithiasis, renal infection or abuse of analgesics. This tumor is aggressive in nature and usually has a poor prognosis. We report two cases of RSCC one with characteristic history of chronic pyelonephritis and one without characteristic etiologic factors. These cases have been reported due to their extreme rarity and also highlight the silent presentation of these tumors and the need to keep it as a differential diagnosis while evaluating cases of nephrolithiasis and chronic inflammations.

Keywords: Chronic pyelonephritis, renal pelvis, squamous cell carcinoma

INTRODUCTION

Cancers of the kidney amounts to 2% of all the total human cancer.1 Primary tumors of the renal pelvis are quite rare accounting for only about 4-5% of all urothelial tumors and include transitional cell carcinomas, squamous cell carcinomas (SCC), and adenocarcinomas.² Renal SCC (RSCC) of pelvis is a rare malignancy of the upper urinary tract characteristically presenting with advanced stage. Of these 90% are of transitional cell origin while SCCs constitute about 6-15%.³ Among malignant renal tumors, SCC are decidedly rare neoplasms and form only about 0.5-8%. These are rare and very few cases have been reported in the literature. We report two cases of primary RSCC of the renal pelvis.

CASE REPORTS

Case 1

A 44-year-old lady presented to our Outdoor Patients Department (OPD) with a history of complaints of pain in the right flank since around 6-7 months. The pain was of dull aching type, continuous in nature. There was no history of weight loss, loss of appetite, hematuria or any other urinary complaints. The patient also had complaints of a lump in the same region since 6 months. There was no history of any renal calculus disease or calculus surgery in the past. Her general examination was normal. Per abdominal examination preoperatively had revealed, a lump of 10.0 cm × 8.0 cm was palpable in the right lumbar and hypochondriac regions. On examination, it was firm, moving with respiration and non-ballotable. The surface was bosselated, with well-defined medial, lower, and lateral borders. The upper border could not be palpated.

Hence, with a clinical diagnosis of a renal lump, most probably renal cell carcinoma (RCC), the patient was investigated. All routine hematological investigations were normal. Ultrasonography (USG) of the abdomen showed a solid mass at the upper pole of the right kidney abutting the liver segments VI and VII of liver show a hyperechoic lesion, likely to be a metastasis. Contrast-enhanced computed tomography (CECT) of abdomen revealed an ill-defined solid mass with necrotic areas showing heterogeneously moderate enhancement, noted arising from renal pelvis and superior pole of right kidney measuring 8.8 cm × 8.8 cm × 6.4 cm. The mass showed well-defined rounded calcification and has breached the perinephric fat to involve segment VI and VII of liver. Both renal veins and inferior vena cava were normal. USG guided tru-cut biopsy was inconclusive.
showing only dysplastic cells. Rest of the metastatic workup was normal.

Hence, with a diagnosis of RCC, a right nephroureterectomy with wedge resection of liver was done (Figures 1 and 2). Histopathology was suggestive of well-differentiated SCC of the renal pelvis extending into the upper pole of kidney and liver. Patient was planned for platinum-based chemotherapy after wound healing indicating being hepatic invasion. However, the patient was lost for follow-up and on enquiry patient passed away 3 months after surgical management.

**Case 2**

A 60-year-old female presented to our OPD with complaints of on and off fever since 14 months and right-sided flank pain and passage of clots in urine since 2 months, pain was dull and intermittent in nature. There was no history of weight loss, loss of appetite or any other urinary complaints. There was no history of any renal calculus disease or calculus surgery in the past. Her general examination was normal. Per abdominal examination preoperatively had revealed, a bosselated lump of 8.0 cm × 8.0 cm was palpable in the right lumbar and hypochondriac regions. On examination, it was firm, moving with respiration and non-ballotable.

With clinical diagnosis of renal mass patient was evaluated. All routine hematological investigations were normal. Urine culture and sensitivity was positive for *Escherichia coli*. Urine cytology was negative for malignant cells. USG abdomen and pelvis showed mass noted in the upper pole of the right kidney measuring 5.1 cm × 4.7 cm × 3.3 cm with mild vascularity and heterogeneous echotexture. CECT abdomen and pelvis revealed well-defined moderately enhancing mass lesion of size 5.9 cm × 5.4 cm × 6.3 cm in transverse, AP and CC axis noted arising from upper pole of right kidney with reniform shape of kidney well maintained with few non-enhancing areas noted s/o necrosis (Figures 3-5). There is thickening of Gerota's fascia with perinephric fat stranding. Right renal vein and right suprarenal gland appear normal. Chest X-ray and electrocardiogram were normal.

With a diagnosis of RCC, right radical nephrectomy was done (Figure 6). Histopathology report though confirmed diagnosis of SCC of kidney with chronic pyelonephritis with section from tumor with pelvis showing squamous metaplasia of pelvis with full thickness dysplasia (Figures 7-9).

Post-operative CECT Abdomen was within normal limits with confirming postoperative status of the right kidney (Figure 10). In accordance with that patient was confirmed to be staged as p(T3N0M0) and is kept on keen follow-up. As per, limited experience in the past chemoradiotherapy is indicated in metastatic setup.
DISCUSSION

SCC of the renal collecting system is a rare malignancy with poor prognosis accounting for about 10% of renal pelvic tumors and 0.5% of all renal tumors. Primary RSCC of the renal pelvis is a rare malignancy with a variable incidence of about 7-8% of all urothelial cancers. Pure SCC is very rare, though it is the second most common tumor in the renal pelvis. Women are affected more frequently than men, predominant age group being 50-70 years.

Figure 5: C2 - Coronal section showing right kidney renal fossa with no significant LN pathy

Figure 6: C2 - Gross specimen of radical nephrectomy

Figure 7: C2 - A: Thyroidization of glomeruli, B: Keratotic cellular debris

Figure 8: C2 - A: Normal glomeruli, B: Malignant squamous cells

Figure 9: Hyperchromic nuclei of squamous cells

Figure 10: C2 - Post-operative scan showing empty right mass
Chronic irritation of urothelium is presumed to be a cause of squamous metaplasia with subsequent malignant progression to SCC. Common causes of chronic irritation are renal calculi disease for long duration, previous history of renal calculi surgery, chronic analgesic abuse or radiotherapy. Li et al. reported incidence of co-existing renal stone in 100% of cases.8 Smoking or tobacco chewing was also observed in 60% of the patients as a known predisposing factor. Hypercalcemia, leukocytosis, and thrombocytosis have been reported as a part of paraneoplastic syndromes in RSCC cases.7,8

Overall median survival for renal pelvis SCC was 7.25 months.9 Using the current 7th edition AJCC staging, the 5-year renal pelvis and ureter cancer survival is as follows: Stage 0a - 72.3%, Stage 0 - 70%, Stage I - 63.9%, Stage II - 56.7%, Stage III - 38.5%, Stage IV - 10.2%. Their generally poor prognosis can be attributed to the typically advanced stage at diagnosis with high local recurrence rates, but the stage for stage, prognosis is similar for squamous, and usual urothelial carcinoma.10

As per literature, there are two entities when it comes to SCC of kidney, one being intraparenchymal SCC which is much rarer and pathognomonic sign of, which is normal histopathological features of renal pelvis which was not the case with our patients. Second, being primary renal SCC of pelvis, which may or may not be associated with squamous metaplasia/dysplasia. Hence, both of the reported cases are in primary renal SCC of the renal pelvis.

The diagnosis of SCC of the renal pelvis is restricted to tumors showing extensive squamous differentiation. The histologic hallmarks of pearl formation, intercellular bridges, and keratotic cellular debris are those of squamous carcinoma at any site. However, 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 rare11,12 which were characteristically seen in our case report. Thyroidization of renal glomeruli, which is the hallmark of chronic pyelonephritis, was also one of the findings of our case report.

Hence, a tumor with squamous cell morphology in the kidney in a middle aged to the elderly patient should be meticulously sampled to differentiate among the primary urothelial carcinoma with squamous differentiation, primary SCC of the renal pelvis, metastatic SCC to kidney and primary intraparenchymal SCC of the kidney. Both of our cases were negative for metastatic work up, hence excluding above differential diagnosis.

Lee et al. found that the specific feature in CT of RSCC was presence of enhancing extra luminal exophytic mass or in some cases, an intraluminal component.13 They further suggested that intravenous urography (IVU) should be carried out periodically, especially, in patients with long-standing stones. Because the filling defects, delay in appearance of pyelogram, or renal parenchymal thickening in IVU may indicate a renal tumor despite the absence of mass effect and preservation of renal contour, warranting further studies.14 One of the cases showed mass arising from upper pole of right kidney with reniform shape of kidney well-maintained.

Extensive review of the available medical literature on this rare malignant entity revealed a poor prognosis.9 Nativ et al. in their study divided renal SCC in three groups, reported 1- and 2-year survival rates of locally invasive renal SCC 33% and 22%, respectively.9 Review of literature suggested current primary treatment of RSCC being nephrectomy.9,10,12 Adjuvant chemotherapy or radiotherapy indicated in metastatic disease.15 First case reported had pathological staging as T4N1M0 (Stage IV) in which radical nephrectomy with hepatic wedge resection was done as special mention. Second case had pathological staging as T3N1M0 (Stage IV) in which radical nephrectomy was done. Nephrectomy with ureterectomy 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.

Adjuvant treatment in the form of 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, para-aortic 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.

In the setting of metastatic disease, adjuvant chemotherapy has been used which was platinum-based but definite chemotherapy protocol do not exist due to being rare clinical entity with poor prognosis. The possibility of anti-epidermal growth-factor receptor therapy in renal SCCs expressing the receptor needs further evaluation in a research setting.

CONCLUSION

Primary RSCC of pelvis has silent presentation, often detected at an advanced stage hence cases of nephrolithiasis should be kept on follow-up with IVU.

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PEER REVIEW

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CONFLICTS OF INTEREST

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REFERENCES


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