Primary Squamous Cell Carcinoma Of Kidney - A Case Report And Review Of Literature.

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Abstract
Primary squamous cell carcinoma of the kidney is a very rare clinical entity. The lack of characteristic presentation like hematuria, pain and palpable mass causes delay in diagnosis results in locally advanced or metastatic disease at presentation. We report a case of this rare tumor present in right kidney with calculi and hydronephrosis, treated with right nephrectomy. Adjuvant treatment has been tried in form of combination chemotherapy. However, due to aggressive nature of this rare tumor, the prognosis is poor and most of patients develop recurrence or disseminated metastatic disease.

INTRODUCTION
Primary malignancies of the renal collecting system are rare accounting for 4-5% of all urothelial tumors. Most frequently diagnosed cases are the transitional cell carcinoma 85-94% followed by squamous cell carcinoma 6% to 15% adenocarcinoma 7%. Squamous cell carcinoma of the renal collecting system is a very rare clinical entity. The incidence of primary renal squamous cell carcinoma of kidney ranged from 0.5%-8%. These tumors are highly aggressive, high grade, and locally advanced or metastatic at the time of presentation related to poor prognosis.

CASE REPORT
A 47 year old male chronic smoker, farmer by occupation patient presented with history of intermittent pain in right side of abdomen radiating to right side scapular region, and intermittent hematuria of one month duration. There was also history of on and off fever. Fever was associated with chills and rigors. There was no other significant past medical and family history. General and physical examination of the patient were within normal limits. On local examination revealed a slight tenderness on right costovertebral angle. Complete blood count, erythrocyte sedimentation rate, biochemical analysis and were all within normal limits. Urine examination showed albumin and full of pus cell and red blood cells. Chest x ray was also within normal limits. Plain X ray abdomen showed multiple radiopaque shadows seen in the right kidney opposite L3 L4 vertebrae. (Fig 1)
Ultrasound examination of abdomen and pelvis showed right renal calculi with moderate hydronephrosis. Renal scintigraphy with isotope 99mTc-DTPA showed nonfunctioning right kidney with split function 05%, no viable cortex, GFR <10 ml/min. The patient was diagnosed to have right nonfunctioning kidney with hydronephrosis and planned for right nephrectomy. Per operative findings were right hydronephrotic kidney enlarged in size contain two large stones. Cortex thinned out except upper pole, adhesion was present between kidney capsule and surrounding tissues. Histopathological examination of the specimen revealed squamous cell carcinoma measuring 4x3 cm in size, moderate to poorly differentiated involving the resection margins focally. Stones were staghorn type of stones. (Fig 3&4)

The patient was kept on follow up. The patient remained disease free up to three months post surgery. After three months of follow up, patients complaints pain abdomen. Ultrasound examination abdomen and pelvis showed multiple lymph nodes in portal, pericaval and preaortic regions, which later confirmed metastatic from squamous cell carcinoma ultrasound guided FNAC. Then patient planned for adjuvant combination chemotherapy Inj. Paclitaxel, Inj. carboplatin and Inj 5FU-1gm three weekly. The patient has received only two courses of chemotherapy and then lost to follow up.

**DISCUSSION**

Squamous cell carcinoma of the urinary tract is more
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frequently reported in urinary bladder and male urethra, in it is rarely encountered in renal pelvis. The etiological factors which play in the genesis of this rare malignancy are strongly associated with phenacetin consumption, chronic renal calculi, pyelonephritis and squamous metaplasia. Li MK et al in their study reported incidence of coexisting renal stone in 100% cases. Urinary calculi was accepted as a main carcinogenic risk factor squamous cell carcinoma. Chronic irritation and infection are believed to induce reactive changes in the urothelium and leads to neoplasia via metaplasia and lecoplakia. Staghorn calculi are most frequently associated with renal pelvis squamous cell carcinoma. In present case renal squamous cell carcinoma associated with staghorn calculi, correlated with data given in literature.

Lee et al in their study classified these tumors into two groups, according to localisation of the tumors as central and 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. Based on these criteria the present case classified as peripheral renal squamous cell carcinoma.

The survival of patients with central renal squamous cell carcinoma was reported to be significantly shorter than those with peripheral renal squamous cell carcinoma. Diagnosis of renal SCC is difficult as characteristic features usually not associated with renal SCC, added by imaging techniques which reveals only calculi and hydronephrosis. Therefore, initial diagnosis of SCC is mostly based on histological analysis as was seen in present case.

Extensive review of the available medical literature on this rare malignant entity revealed a poor prognosis. 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.

Review of literature suggested current primary treatment of renal squamous cell carcinoma is nephrectomy. Adjuvant chemotherapy or radiotherapy indicated in metastatic disease. Corral et al in their study used a combination chemotherapy included cisplatin, methotrexate, bleomycin if the sigh of metastatic disease are revealed. The patient has been given combination chemotherapy in form of Paclitaxel, carboplatin and 5 FU. The patient received only two courses and then lost to follow up.

Primary squamous cell carcinoma is an aggressive tumor, throughout medical literature; authors have underlined the poor prognosis when tumor recurrence occurs; as was seen in present case; warranting aggressive management as soon as metastasis develops. The patient in present case developed lymphnodes metastasis after three months of primary surgical treatment. As these tumors are strongly associated with renal stones, the patients with renal stones and non-functioning kidney should be carefully examined with newer imaging modalities for early detection of the tumor, and warrants aggressive treatment with surgery followed by adjuvant aggressive combination chemotherapy that may provide a better outcome.

Conclusion: Primary squamous cell carcinoma is an aggressive tumor, throughout medical literature; authors have underlined the poor prognosis when tumor recurrence occurs; as was seen in present case; warranting aggressive management as soon as metastasis develops. The patient in present case developed lymphnodes metastasis after three months of primary surgical treatment. As these tumors are strongly associated with renal stones, the patients with renal stones and non-functioning kidney should be carefully examined with newer imaging modalities for early detection of the tumor, and warrants aggressive treatment with surgery followed by adjuvant aggressive combination chemotherapy that may provide better results.

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