

**CASE REPORT**

Primary Squamous Cell Carcinoma of The Renal Pelvis Associated with Staghorn Calculi

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Abstract

Tumors of the renal pelvis account for approximately 10% of all renal tumors of which urothelial carcinomas constitute the majority followed by adenocarcinoma and squamous cell carcinoma. The latter are rarer and very few cases have been reported in English literature. We report a case of primary renal squamous cell carcinoma in the left renal pelvis associated with staghorn calculi and pyonephritis. This case has been reported due to its extreme rarity and also highlights the silent presentation of these tumors and the need to keep it as a differential diagnosis while evaluating cases of nephrolithiasis and chronic inflammations.

Key Words

Kidney, Squamous Cell Carcinoma, Renal Pelvis, Staghorn

Introduction

Cancers of the kidney amounts to 2% of all the total human cancer (1). Primary tumors of the renal pelvis are quiet rare accounting for only about 4-5% of all urothelial tumors and include transitional cell carcinomas, squamous cell carcinomas (scc), and adenocarcinomas (2). Of these 90% are of urothelial origin while squamous cell carcinomas constitute about 6-15% (3). Among malignant renal tumors, SCC are decidedly rare neoplasms and form only about 0.5-8% (4,5). These tumors are usually associated with a poor prognosis, have a silent presentation and tend to be sessile, deeply invasive at presentation. Squamous cell carcinomas have been associated with antecedent stag horn calculi or chronic infection (6).

Case Report

A 62 year old female patient presented with dull aching pain in the right flank, off and on for three months. Examination of the abdomen was unremarkable. Her blood pressure was 130/80 and a regular pulse rate of 80 per minute. Her hemoglobin was 8.5 g/dL and she had total leucocyte count of 8,300/mm³. Urine examination revealed mild hematuria and 2+ proteinuria. The blood

urea was 44mg/dl and serum creatinine 1.2 mg/dl. The total glomerular filtration rate (GFR) was 81% with 12.2% on right side and 90% on left side. X-ray of the kidney, ureter and bladder (KUB) showed presence of staghorn calculi in right kidney (Fig 1). Ultrasound evaluation showed grade IV right renal hydronephrosis with right renal calculi largest measuring 23mm suggestive of astaghorn calculi. CT scan was not done. The patient underwent a right nephrectomy for right sided non-functioning kidney.

Pathology

On gross examination the kidney was enlarged and showed a dilated pelvi-calyceal system, presence of staghorn calculi in the lumen, and thinning of the renal parenchyma to a narrow peripheral rim (Fig 2). The renal pelvis was diffusely thickened. The resected end of ureter showed presence of necrotic material. Sections from the thickened pelvis were taken microscopic examination of which showed an incidental finding of a well differentiated squamous cell carcinoma (Fig 3), infiltrating the perirenal fat. Tumor was staged- Stage III (pT3aN0). The patient had an uneventful postoperative course.

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Fig. 1 KUB Showing Right Staghorn Calculi



Fig. 2 Kidney with Dilated Pelvi-Calyceal System & Thinning of Renal Pelvis

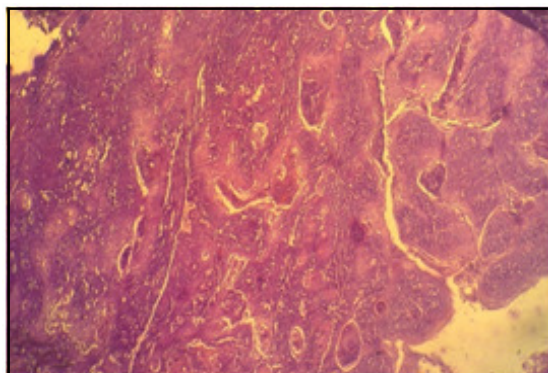


Fig. 3 Squamous Cell Carcinoma of Renal Pelvis Showing Keratin Pearls (100 X)

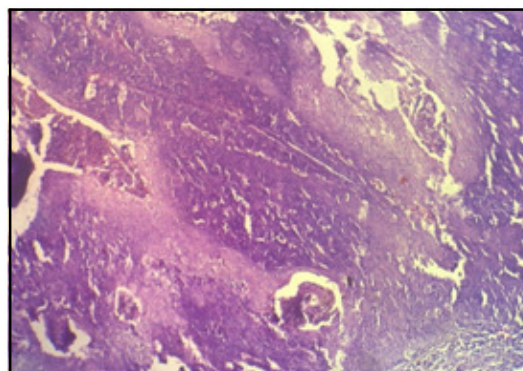


Fig. 4 Squamous Cell Carcinoma of Kidney Showing Aborted Glomeruli (100 X)

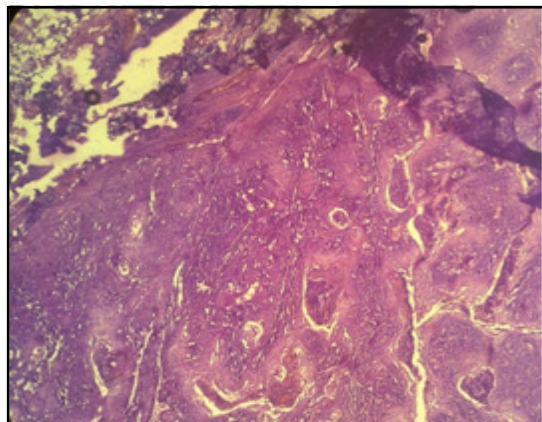


Fig. 5 Tumor Infiltrating Peri- Renal Fat (400 X)

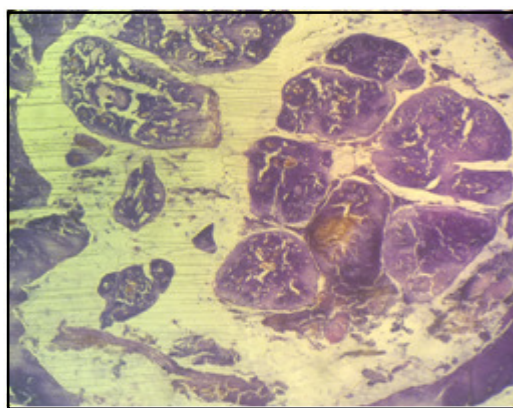


Fig. 6 Tumor Involving Upper Ureter (400 X)

Discussion

Squamous cell carcinomas of the renal pelvis are believed to develop following chronic irritation superimposed with infection leading to metaplasia and consequent development of invasive carcinoma (7).

Chronic irritation may be due to in descending order of frequency because of : renal calculi (mostly staghorn type) , following surgery for renal stones, analgesic abuse, radiotherapy, exogenous and endogenous chemicals, vitamin A deficiency, hormonal imbalance, schistosomiasis



and smoking (8). Few cases are, however, not associated with any apparent aetiological factor (9). The mean age of presentation is 56 years with no predilection for side or sex (10). The tumor is mostly unilateral. Presentation is usually silent with most patients presenting with non-specific features such as pain and hematuria. Pain may be due to pelviureteric junction obstruction and/or local extension while as hematuria may be due to primary tumor mass or calculi (10). Hypercalcaemia, leucocytosis and thrombocytosis are the paraneoplastic syndromes associated with renal squamous cell carcinoma (11).

The lack of characteristic presentation, radiological findings and insidious onset of pain and is the main cause of diagnostic dilemma (10). Lee *et al* (12) found that the specific feature in CT of Renal Squamous cell carcinoma was presence of enhancing extraluminal exophytic mass or in some cases, an intraluminal component. Most of the detected tumors are histologically high grade with 84% being locally advanced or metastatic (13). Lee *et al.*, (12) classified primary renal squamous cell carcinomas based on the location of the tumour into two categories- the central and the peripheral types.

The central type had higher rates of lymph nodal metastasis and the peripheral type showed parenchymal thickening with perirenal infiltration. Poor survival rates were associated with the central type. Our case was classified as a peripheral type of primary renal squamous cell carcinoma as per Lee *et al* (12). Nephrectomy with or without ureterectomy is the recommended treatment in primary renal squamous cell carcinomas even in the presence of distant metastasis (10) which require Cisplatin based chemotherapy and palliative radiotherapy as adjuvants (10,14) but have shown no survival benefit, highlighting the need for early diagnosis (7).

Conclusion

Squamous cell carcinoma of the renal pelvis is a very rare tumour commonly associated with staghorn calculi and infection. This tumour is highly aggressive and often detected at advanced stage with poor outcome. We describe a case report of a 62-year-old female patient who was diagnosed with right nephrolithiasis with non-functioning kidney. Histopathology revealed an unexpected co-existing SCC in renal pelvis. The present case highlights the need for use of newer imaging modalities and the role of biopsy from the renal pelvis in cases of long-standing renal calculi as these patients are susceptible of harbouring co-existing hidden malignancy.

References

1. Lipworth L, Tarone RE, McLaughlin JK. The epidemiology of renal cell carcinoma. *J Urol* 2006; 176: 235.
2. Murphy DM, Zincke H, Furlow WL. Primary grade I transitional cell carcinoma of the renal pelvis and ureter. *J Urol* 1980;123:629-31.
3. Latham HS, Kay S. Malignant tumors of the renal pelvis. *Surg Gynecol Obstet* 1974;138:613-22.
4. Li MK, Cheung WL. Squamous cell carcinoma of the kidney. *J Urol* 1987; 138:269-71.
5. Lynch CF, West MM, Davile JA, Pletz CE (2007) Cancers of the kidney and renal pelvis. In: Ries LAG, Young JL, Keel GE, Eisner MP, Lin YD, Horner M-J (ed) SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988-2001, Patient and Tumor Characteristics. National Cancer Institute, SEER Program, NIH Pub. No. 07-6215, Bethesda, MD.
6. Paonessa J, Beck H, Cook S. Squamous cell carcinoma of the renal pelvis associated with kidney stones: a case report. *Med Oncol* 2011; 28 (Suppl 1) : S392.
7. Singh V, Sinha RJ, Sankhwar SN, *et al.* Squamous Cell Carcinoma of the Kidney - Rarity Redefined: Case Series with Review of Literature. *J Cancer Sci Ther* 2010; 2: 82-85.
8. Mathur SK, Rana P, Singh S, Goyal V, Sangwan M. An incidentally detected squamous cell carcinoma in a non-functioning kidney, presenting as a multi-cystic mass. *J Surg Case Reports* 2011; 9:1-4.
9. Talwar N, Dargan P, Arora MP, Sharma S, Sen AK. Primary squamous cell carcinoma of the renal pelvis masquerading as pyonephrosis: A case report. *Ind J Pathol Microbiol* 2006;49:418-20.
10. Holmang S, Lele SM, Johansson SL. Squamous cell carcinoma of the renal pelvis and ureter: incidence, symptoms, treatment and outcome. *J Urol* 2007;178:515-6.
11. Er O, Coskun HS, Altinbas M, *et al.* Rapidly relapsing squamous cell carcinoma of the renal pelvis associated with paraneoplastic syndromes of leukocytosis, thrombocytosis and hypercalcemia. *Urol Int* 2001;67:175-77.
12. Lee TY, Ko SF, Wan YL, *et al.* Renal squamous cell carcinoma: CT findings and clinical significance. *Abdom Imaging* 1998; 23:203-08.
13. Nativ O, Reiman HM, Lieber MM, *et al.* Treatment of primary squamous cell carcinoma of the upper urinary tract. *Cancer* 1991;68:2575-78.
14. Blacher EJ, Johnson DE, Abdul-Karim FW, Ayala AG. Squamous cell carcinoma of renal pelvis. *Urology* 1985; 25:124-26.