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Squamous Cell Carcinoma Arising in Keratinizing Desquamative Squamous Metaplasia (KDSM) of the Renal Pelvis

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ABSTRACT

Keratinizing desquamative squamous metaplasia (KDSM) of the upper urinary tract is a rare condition. We present a case of a 45-year-old male smoker of more than 20 years presenting with a 1-month history of recurrent, intermittent left flank pain of moderate intensity associated with dysuria and nausea. In this case the differential diagnosis included renal cell carcinoma, tuberculosis of the kidney, and KDSM. However, the pathological findings showed squamous cell carcinoma arising in KDSM. Hence we report this rare association of a squamous cell carcinoma of the renal pelvis arising in KDSM.

INTRODUCTION

Keratinizing desquamative squamous metaplasia (KDSM) of the upper urinary tract is a rare condition. It is often referred to by several different names, including cholesteatoma, leukoplakia, and hyperkeratosis [1]. The etiology of KDSM includes a reactive epithelial response to chronic irritation (urinary tract infection, urolithiasis, tuberculosis, or hydronephrosis) [1]; secondary to vitamin A deficiency [2]; representation of a spontaneous epithelial transformation; or representation of a congenital anomaly whereby abnormal epithelial cells from the ectoderm embryologically contaminate the primitive Wolffian duct [3]. We are presenting here a rare case of a squamous cell carcinoma of the kidney arising in KDSM.

CASE HISTORY

A 45-year-old male smoker for more than 20 years presented with a 1-month history of recurrent, intermittent left flank pain of moderate intensity associated with dysuria and nausea. He gave a history of losing 5 kg of his body weight during this period. However, there was no history of hematuria, fever, vomiting, or a history of tuberculosis, diabetes, or hypertension. There were no features suggestive of paraneoplastic syndromes.

A physical examination revealed a thin built man with normal vital signs. There was abdominal tenderness in the left flank area with no palpable mass. Investigations revealed normal CBC, coagulation, renal functions, electrolytes, bone profile LFT, and chest X-ray. HIV 1 and 2 were negative. Hepatitis screen revealed that he had previous exposure to HBV. Urinalysis, microscopy, and culture showed 500 leucocytes and 300 red blood cells per microliter but no growth. Mantoux test for tuberculosis was negative. On computed tomography (CT) scan of the abdomen and pelvis, there was a complete lack of enhancement in the enlarged left kidney. There were multiple cystic areas with peripheral calcifications. The left renal pelvis was significantly dilated with debris/hematomas seen, and the wal0-l was thickened and calcified. There were multiple left renal hilum and perinephric lymph hypodense nodes, the largest measuring 1.5 cm. The left renal artery was diffusely attenuated. The right kidney appeared normal. The urinary bladder was unremarkable. There was no significant sized retroperitoneal or mesenteric adenopathy (Figure 1).

The patient underwent radical left nephrectomy on the suspension of renal cell carcinoma. Intraoperative findings were suggestive of tuberculosis of the kidney due to the finding of an enlarged left kidney with hard cystic changes and many lymph nodes that discharged thick white material. Perinephric

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Figure 1. CT of the abdomen showing an enlarged left kidney with calcification and cystic changes (arrow).

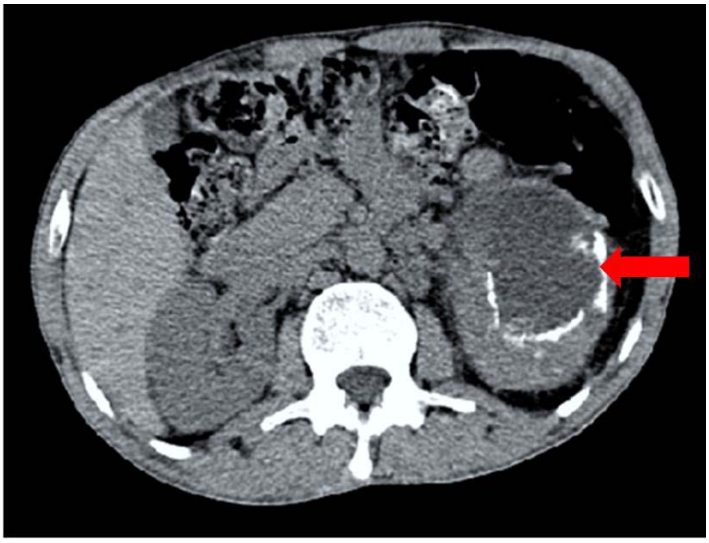


Figure 2. Left kidney with cut surface with brown necrotic material mixed with white thick keratin flakes. Note the irregularly enlarged pelvicalecyal system (white arrow) filled with brownish material (red arrow) mimicing tumor necrosis rather than cheesy caseating material of tuberculosis.

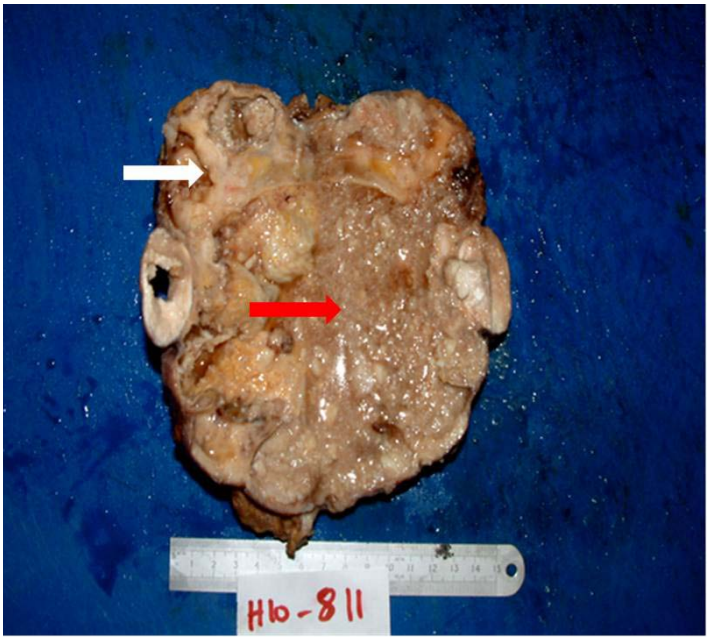


Figure 3. H&E; urothelium with adjacent thick hypekeratotic metaplastic squamous epithelium (arrow).

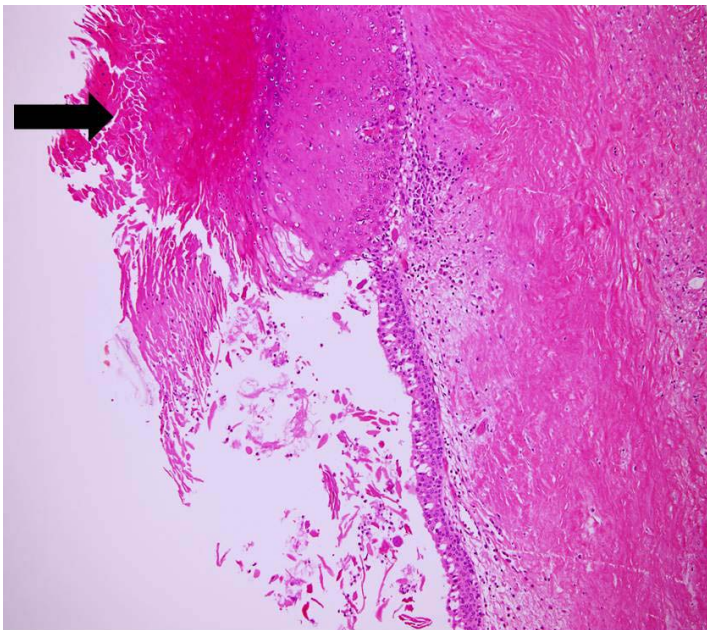
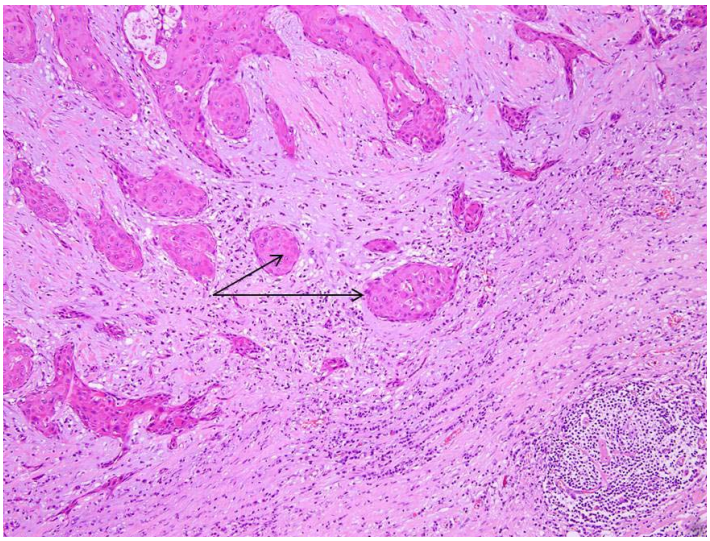


Figure 4. An area of squamous cell carcinoma (some highlighted with arrows).



fat and Gerota's fascia adhered to the kidney with an intense inflammatory reaction. The pathology revealed a cystic mass with compressed renal medullary tissue over the outer surface of the wall. The cyst lining is of squamous epithelium showing

hyperkeratosis and parakeratosis, which was focally ulcerated. At the base of the epithelium there were disorganized, dysplastic, irregular infiltrative down growths of epithelium, indicative of squamous cell carcinoma. At some foci, the tumor infiltrated the renal capsule. Four lymph nodes examined revealed extensive metastasis. As the metaplasia and the areas of squamous cells were well differentiated and the morphological diagnosis was unequivocal, immune-histochemical investigations were not performed. Well-differentiated squamous cell carcinoma arising in a desquamative keratinizing squamous metaplasia (cholesterotomatous leukoplakia) of the renal pelvis was diagnosed (Figure 2, Figure 3, Figure 4). On postoperative follow-up the patient was well and had gained weight. He was referred to oncology for further follow-up. The patient received chemotherapy. However, due to local and metastatic recurrence of the disease, the patient died after 1 year from the date of the diagnosis of his disease.

DISCUSSION

Keratinizing desquamative squamous metaplasia (KDSM) has been described coincidentally with squamous cell carcinoma (SCC) of the upper urinary tract in 8 to 12% of cases; however, it has never been demonstrated that SCC arises from KDSM [4]. Reece and Koontz, [3] reported 2 cases of concurrent renal pelvis KDSM and neoplasm (1 transitional-cell carcinoma and 1 squamous-cell carcinoma). However, they were unable to provide histological evidence that the KDSM was involved in the tumor. Sheaff et al [5] described a case of squamous cell carcinoma in association with extensive keratinising squamous metaplasia of the pelvic urothelium. The general appearance of KDSM as a space-occupying mass within the kidney prompts consideration of malignancy. It has been reported that keratinizing squamous metaplasia of the kidney may mimic transitional cell carcinoma [6]. Keratinizing desquamative squamous metaplasia of the renal pelvis without presence of malignancy has been described bilaterally [7].

In our case the differential diagnosis included renal cell carcinoma, tuberculosis of the kidney, and KDSM. However, to our surprise the pathological findings showed squamous cell carcinoma arising in KDSM. In this case we prove for the first time in the literature a squamous cell carcinoma of the kidney arising in KDSM. Historically, KDSM of the upper urinary tract is treated with extirpative surgery. However, Borofsky et al. [8] reported 2 cases of renal KDSM managed conservatively with a nephron-sparing manner with biopsy, diagnostic ureteroscopy, and imaging surveillance. They concluded that conservative management of renal KDSM is preferable as it is almost certainly a benign condition. In addition, Ganeshappa et al. [9] reported conservative management for KDSM. Our case shows that SCC is a potential complication of KDSM and we recommend that the conservative management of KDSM should be guarded and, if employed, a vigorous surveillance with renal biopsy to be undertaken regularly.

Most tumors of the renal pelvis and ureter are of urothelial origin and are, most commonly, transitional cell tumors. Squamous cell carcinomas and adenocarcinomas may be associated with chronic infection or stones, but this does not occur frequently. In those patients at risk for upper tract SCC, a high index of suspicion should be maintained to improve detection of disease at an earlier stage. In our case, which included left radical nephrectomy performed for the suspicion of RCC, the incidental finding of SCC on histological examination of the nephrectomy specimen was found. The standard surgical management of upper tract SCC is radical nephroureterectomy. However, in our case only radical left nephrectomy was done due to the initial suspicion of RCC. Subsequent completion of ureterectomy was not carried out due to the advanced disease of the patient and the need for chemotherapy.

CONCLUSION

In this case we report this rare association of squamous cell carcinoma of the renal pelvis arising in KDSM. However, squamous cell carcinoma is a potential risk of keratinizing desquamative squamous metaplasia, and conservative management of KDSM needs further support by future studies.

REFERENCES

1. Hertle, L. and P. Androulakakis (1982). "Keratinizing desquamative squamous metaplasia of the upper urinary tract: leukoplakia--cholesteatoma." *J Urol* 127(4): 631-635. PubMed

2. Liang, F. X., M. C. Bosland, et al. (2005). "Cellular basis of urothelial squamous metaplasia: roles of lineage heterogeneity and cell replacement." *J Cell Biol* 171(5): 835-844. PubMed | CrossRef

3. Reece, R. W. and W. W. Koontz, Jr. (1975). "Leukoplakia of the urinary tract: a review." *J Urol* 114(2): 165-171. PubMed

4. Denes, F. T. (1984). "Keratinizing squamous metaplasia of the upper urinary tract." *Int Urol Nephrol* 16(1): 23-28. PubMed

5. Sheaff, M., P. Fociani, et al. (1996). "Verrucous carcinoma of the renal pelvis: case presentation and review of the literature." *Virchows Arch* 428(6): 375-379. PubMed

6. Boswell, P. D., B. Fugitt, et al. (1998). "Keratinizing desquamative squamous metaplasia of the kidney mimicking transitional cell carcinoma." *Urology* 52(3): 512-513. PubMed

7. Angulo, J. C., A. Santana, et al. (1997). "Bilateral keratinizing desquamative squamous metaplasia of the upper urinary tract." *J Urol* 158(5): 1908-1909. PubMed

8. Borofsky, M., R. B. Shah, et al. (2009). "Nephron-sparing diagnosis and management of renal keratinizing desquamative squamous metaplasia." *J Endourol* 23(1): 51-55. PubMed | CrossRef

9. Ganeshappa, A., A. Krambeck, et al. (2009). "Endoscopic management of keratinizing desquamative squamous metaplasia of the upper tract: a case report and review of the literature." *J Endourol* 23(8): 1277-1279. PubMed | CrossRef