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Case Report

Renal Cholesteatoma: A Case Report of Keratinising Desquamative Squamous Metaplasia of the Upper Urinary Tract

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ABSTRACT

Metaplastic changes in the urothelium of the upper urinary tract are relatively infrequent. We report an interesting and rare case of KDSM of the kidney with the involvement of entire renal parenchyma including the major and minor calyces and upper ureter in a 38 year old lady presenting with renal staghorn calculi and end stage renal disease. We also reviewed the etiopathogenesis of this condition.

***Key words*:** cholesteatoma, desquamative, metaplasia.

INTRODUCTION

Cholesteatoma of the upper urinary tract involving the renal pelvis and upper ureter is a rare entity with only few cases being described in the literature. The characteristic histology is squamous metaplasia of the urothelium associated with exuberant keratinization and desquamation of keratinized layers, hence called as Keratinizing desquamative squamous metaplasia (KDSM). This metaplasia within the lower urinary tract is not very uncommon, but KDSM of the upper urinary tract and renal pelvis is rare [(1)]

CASE REPORT

A 38 year old lady came with the complaints of right sided loin pain and fever since one year. This was not associated with vomiting, dysuria, hematuria, weight loss or night sweats. There was no significant family or personal history. On examination, abdomen was soft. No tenderness or palpable mass was noted. Her hemoglobin was 8.6 gm%. Renal function tests were normal. Urine examination showed mild increase in epithelial cells 6-8/HPF (N 0-5/HPF). On ultrasound abdomen right kidney was not clearly visualized and appeared contracted. Plain and contrast CT scan of abdomen and pelvis showed right kidney measuring 8.6x 5.8 cm, hypoenhancing with a pelviureteric junction staghorn calculus measuring 34x 29 mm causing gross hydronephrosis and renal cortical thinning. There was no evidence of excretion of contrast on delayed phases. Left kidney was normal in size and attenuation. A diagnosis of non functioning right kidney was given (Fig 1, 2). Following this right nephrectomy was done. Per operative right kidney was found contracted with pelvic calculus and multiple dense perinephric adhesions. Grossly, specimen weighed 250 gm and measured 11x 5x 3.5 cm. On cut section the corticomedullary differentiation was not noted. Entire pelvicalyceal system was

distorted and showed multiple cystic spaces surrounded by grey white and yellow areas along with calcifications (Fig 3,4). Histology showed entire renal urothelium replaced by metaplastic keratotic squamous layer with keratin debris and luminal keratin flakes. Renal parenchyma composed of marked sclerotic glomeruli, atrophic tubules and dense chronic interstitial infiltrate. Ureter also showed this squamous metaplasia (Fig 5,6,7,8). A diagnosis of extensive keratinizing desquamative squamous metaplasia in end stage renal disease with pyelonephritis was given. Patient was on regular follow up post operatively and had no complaints.

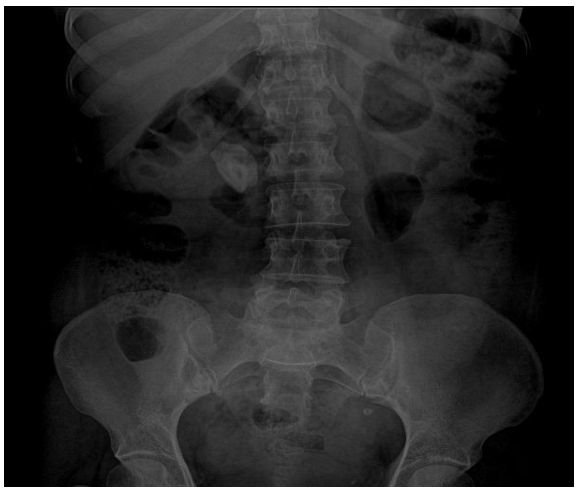

Fig 1- Pelviureteric junction staghorn calculus measuring 34x 29 mm

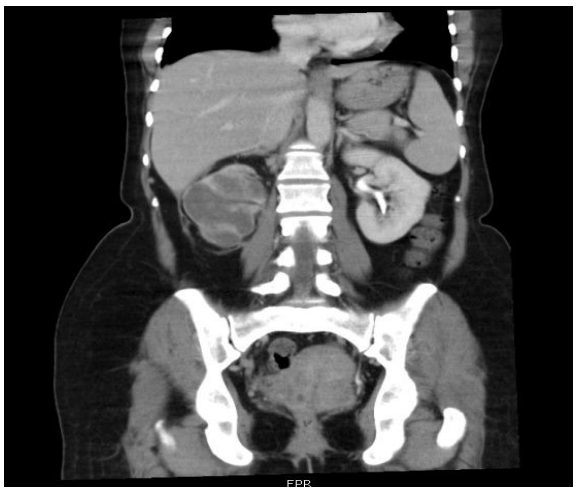

Fig 2- Right kidney measuring 8.6x 5.8 cm with gross hydronephrosis and renal cortical thinning

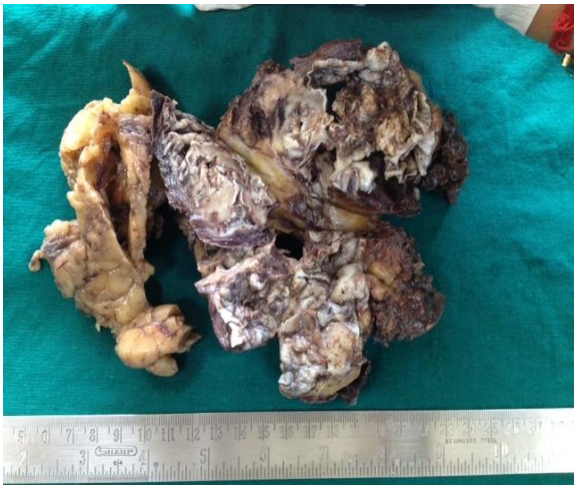

Fig 3- Grossly, on C/S no corticomedullary differentiation noted. Entire pelvicalyceal system was distorted

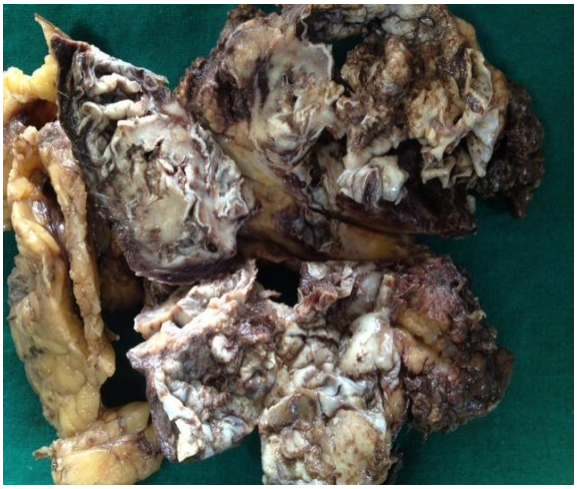

Fig 4- Grossly, multiple cystic spaces surrounded by grey white and yellow areas along with calcifications

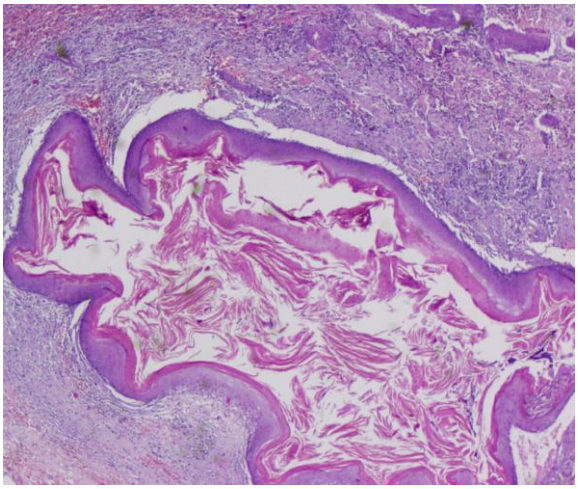

Fig 5- Entire renal urothelium replaced by metaplastic keratotic squamous layer with keratin debris and luminal keratin flakes H&E X20

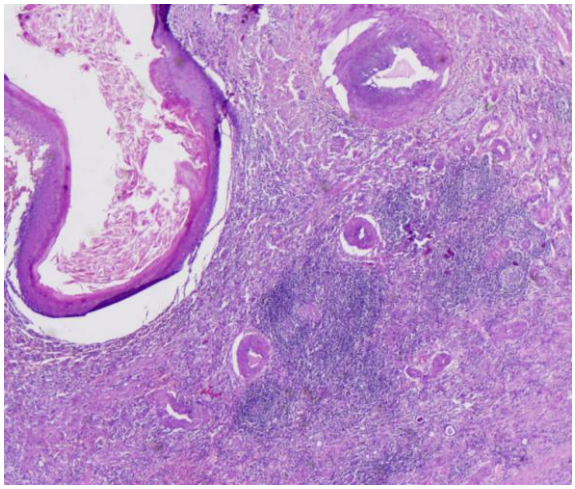

Fig 6- Metaplastic keratinized squamous layer overlying renal interstitial dense chronic inflammatory infiltrate H&E X20

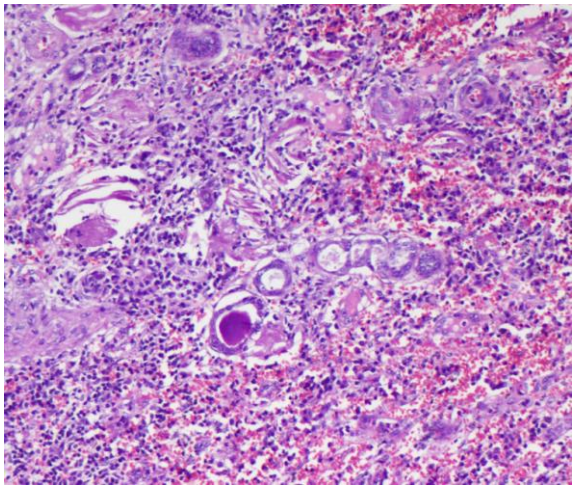

Fig 7- Renal parenchyma with marked sclerotic glomeruli, atrophic tubules and dense chronic interstitial infiltrate H&E X100

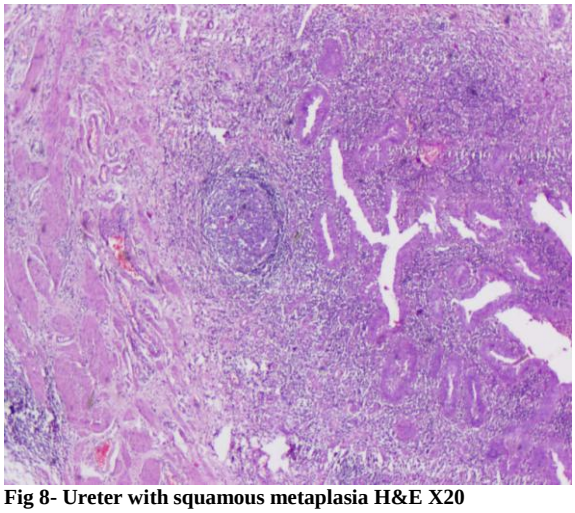

Fig 8- Ureter with squamous metaplasia H&E X20

DISCUSSION

Originally referred to as renal cholesteatoma, KDSM is an uncommon condition with an uncertain etiology. The condition is commonly believed to be reactive and has been associated with chronic irritant exposure and chronic infections. It may be seen with xanthogranulomatous pyelonephritis, renal stones, cigarette smoking, infections like syphilis and tuberculosis. Metaplasia is a protective adaptation in response to a chronic insult, converting the transitional epithelium to a more robust keratinized squamous epithelium, thereby reducing the damage to underlying stroma. [(1)] KDSM may also be secondary to vitamin A deficiency or may represent a congenital anomaly where abnormal ectodermal epithelial cells embryologically contaminate the primitive Wolffian duct. [(2)] In few cases no associated pathology has been found. [(3)] However, in this case report patient had renal calculi. KDSM is predominantly seen in adult population usually in third to sixth decades of life with a slight male predilection, although cases in young children have been documented, suggesting a possible genetic etiology. [(4,5)]

The term KDSM is descriptive of the histology seen in this condition and was first described by Hertle and Andraulakakis in the year 1982. [(6)] They found cells to be metaplastic, not dysplastic with keratinization and subsequent desquamation. Other causes of presence of squamous epithelium in the kidney include teratoid variant of Wilm's tumor, teratoma, dermoid cyst or associated with renal malignancies. [(7,8)]

To establish a clinical diagnosis of KDSM is difficult. Recurrent renal colic, characteristically passage of desquamated keratinised cells and a lamellar configuration on excretory urography suggest a possibility of KDSM, however differential diagnosis include urothelial tumor, radiolucent stone, blood clot, tuberculosis and papillary necrosis. [(3,4)] Our case had mild increase in the epithelial cells in urine examination.

It is still a matter of controversy whether KDSM should be considered as a premalignant lesion or it is more of a benign process. In few cases, KDSM have been seen in association with squamous cell carcinoma. It is stated that KDSM has chances of malignant transformation in nearly 8-12% of the cases. [9] Reece and Koontz [10] reported 2 cases of concurrent renal pelvis KDSM and malignancies, transitional-cell carcinoma and squamous-cell carcinoma. Sheaff et al. [11] have also described a case of squamous cell carcinoma in association with extensive keratinising squamous metaplasia of the pelvic urothelium. However, authors were unable to prove on histology that KDSM was involved in the tumor. Also the progression from metaplasia to neoplasia has never been demonstrated. [5] Hence, the possibility of KDSM being a premalignant lesion has been challenged and questioned. Endoscopic management and close follow up of KDSM are the preferred treatment rather than any radical procedures. Since ureteral obstruction by desquamated keratinized debris has been thought to result in hydronephrosis and pyelonephritis, alleviation of this obstruction should preserve the kidney function. [1] Borofsky et al. [12] have managed 2 cases of renal KDSM conservatively by nephron-sparing procedure with the help of diagnostic ureteroscopy and imaging. Few others like, Ganeshappa et al. [13] also reported and preferred conservative management for KDSM. However, in most cases nephroureterectomy has been done as it is difficult to distinguish this condition from other neoplastic processes preoperatively. [1] However, in the present case, the kidney was non functional, hence warranting radical surgery.

CONCLUSION

KDSM of the kidney is an uncommon condition where urothelial epithelium of the renal pelvis and ureter transforms into stronger keratinized squamous layer following chronic insult. Its malignant and recurrence potential is yet to be determined. The precise treatment protocols for management of KDSM need to be established and require further support by future studies.

REFERENCES

1. Siderits RH, Fingerman J, Hazra A, Rimmer C, Colaco M, Mikhail N, Ardeleanu C, Mazari PM. Renal Pelviceal Keratinizing Squamous Metaplasia with Sparing of Pyramidal Zones. Case Rep Urol. 2012 26;2012: 242780
2. Al-Marhoon MS.Squamous Cell Carcinoma Arising in Keratinizing Desquamative Squamous Metaplasia (KDSM) of the Renal Pelvis. Uro Today Int J. 2013 August; 6(4):art 49. http://dx.doi.org/10.3834/uij.1944-5784.2013.08.08
3. Keratinising desquamative squamous metaplasia of the upper urinary tract: A case report. Indian J Surg. 2004; 66(2):105-6
4. Lima DX, Rabelo EAS, Salles PGO. Cholesteatoma of the upper urinary tract. Int Braz J Urol. 2004;30:494-5
5. Prashant Jain, Ashwani Mishra, and Deepti Shukla Misra. Keratinizing squamous metaplasia of the upper urinary tract in a child with a solitary kidney. Indian J Urol. 2014 Apr-Jun; 30(2):230–2
6. Hertle L, Androulakakis P. Keratinising desquamative squamous metaplasia of the upper urinary tract: Leukoplakia-cholesteatoma. J Urol. 1982;127:631–5
7. IntrarenalEpidermoid Cyst Presented as an Enlarged Multicystic Kidney. Saudi J Kidney Dis Transpl. 2010;21(4):728-31
8. Huang YP, Chen B, Sun XZ, Guo Y, Yang SC, Deng CH, Huang YR. High-grade Neuroendocrine Carcinoma with Focal Squamous Metaplasia of Renal Pelvis Associated With Renal Calculus: Study of a Case. Urology Case Reports 2014;2:93-6

9. Boswell PD, Fugitt B, Kane CJ. Keratinizing desquamative squamous metaplasia of the kidney mimicking transitional cell carcinoma. Urology. 1998;52(3): 512-3
10. Reece RW, Koontz WW Jr. Leukoplakia of the urinary tract: a review. J Urol.1975;114(2): 165-71
11. SheaffM, Fociani P, Badenoch D, Baithun S. Verrucous carcinoma of the renal pelvis: case presentation and review of the literature. Virchows Arch. 1996;428 (6): 375-9
12. Borofsky M, Shah RB, Wolf JS Jr. Nephron-sparing diagnosis and management of renal keratinizing desquamative squamous metaplasia. J Endourol. 2009;23(1):51-5
13. Ganeshappa A, Krambeck A, Grignon DJ, Lingeman JE. Endoscopic management of keratinizing desquamative squamous metaplasia of the upper tract: a case report and review of the literature. J Endourol. 2009; 23(8):1277-9.

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