Case Report

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Pathology Section

Primary Squamous Cell Carcinoma of Renal Pelvis Associated with Staghorn Calculi Masquerading as Xanthogranulomatous Pyelonephritis

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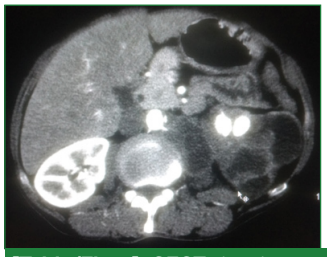
ABSTRACT

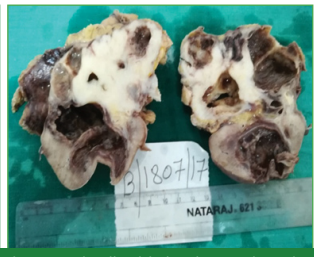
Primary squamous cell carcinoma of renal pelvis is a rare tumour associated with long standing renal calculi. We report here a case in which patient presented with pain abdomen and renal calculus with left pelvi ureteric junction obstruction and final diagnosis of primary squamous cell carcinoma of kidney was made only on histopathological examination. This case has been reported to highlight the silent and unusual presentation of common tumour at an uncommon site. Hence, a high degree of suspicion of squamous cell carcinoma should be made and considered as a differential diagnosis in patients presenting with history of renal stones.

Keywords: Common tumour, Histopathology, Renal calculi, Renal stones

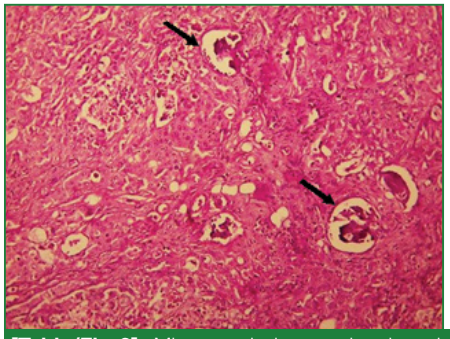
CASE REPORT

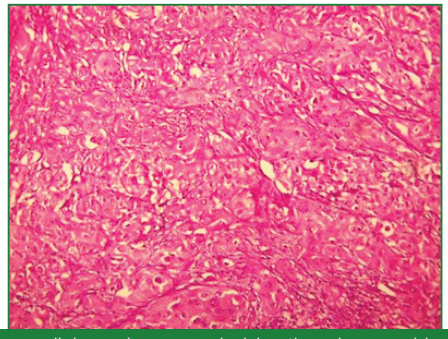
A 73-year-old male patient presented with pain in the right flank on and off since 3 months. Pain increased in intensity since 1 week associated with burning micturation. There was no history of fever, weight loss and gross haematuria. There was no history of renal calculi, urinary tract infection in the past. Had undergone surgery for peptic ulcer 10 years back. General physical examination was normal. On per abdominal examination, tenderness was present on the left loin region. Routine haematological and urine analysis was done in which complete blood count was normal and urine microscopy showed few leucocytes. Further, radiological investigations were done. In which ultrasound examination revealed a large calculi measuring 3.4 cm in the renal pelvis causing moderate nephrosis with thinning of the cortex. Contrast Enhanced Computed Tomography (CECT) of kidney and urinary bladder was performed and it showed radiodense

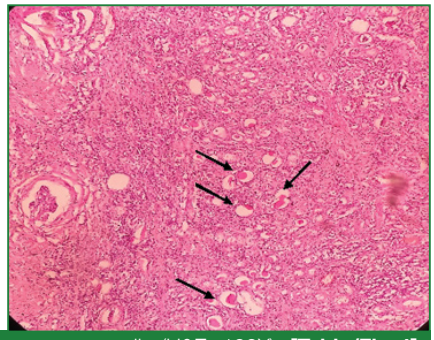




[Table/Fig-1]: CECT showing staghorn calculi with hydronephrosis and diffuse parenchymal thinning with surrounding mild enhancing soft tissue component; **[Table/Fig-2]:** Gross image of the cut section of the kidney showing grey white tumour (arrow) extending from upper pole to the hilum.







[Table/Fig-3]: Microscopic image showing glomeruli (arrow) surrounded by the pleomorphic squamous cells (H&E, 100X). **[Table/Fig-4]:** Microscopic image showing pleomorphic squamous cells in sheets (H&E, 400X). **[Table/Fig-5]:** Microscopic image showing thyroidisation of the tubules (arrow) with infiltration of lymphocytes which are features of chronic pyelonephritis (H&E, 100X).

renal pelvis with two staghorn calculi measuring 31X27 mm and 22X19 mm causing marked hydronephrosis with diffuse parenchymal thinning and surrounding mild enhancing soft tissue component and possibility of xanthogranulomatous pyelonephritis was made [Table/Fig-1].

Patient underwent left nephrectomy for left sided non functioning kidney and was sent for histopathological examination. On gross examination, kidney was enlarged and had bosselated appearance with adherent capsule. Cut section of the kidney showed thinned out cortex with a grey white tumour extending from upper pole to the hilum measuring 7X6 cms, pelvi calyceal system was dilated, cortico-medullary junction was not made out [Table/Fig-2] and the cystic cavities were filled with pus and two staghorn calculi were noted. Part of the ureter was identified in the hilum.

Extensive sampling was done and microscopy showed moderately to poorly differentiated squamous cell carcinoma infiltrating capsule [Table/Fig-3,4] and perinephric fat with Focal areas of Chronic pyelonephritis features [Table/Fig-5]. Stage pT3aNxMx was given on histopathology on histopathology. Ureter was free from the tumour. Patient had uneventful post-operative course and put on radiotherapy. Informed consent has been taken from the patient.

DISCUSSION

Tumours of kidney and renal pelvis are the ninth most common malignant cancer and 12th most common malignant cancer among all the cancers [1]. Transitional cell carcinoma of renal pelvis is the most common malignancy arising in the upper urinary tract. Primary squamous cell carcinoma of renal pelvis are very rare accounting to 0.5% to 15% of all urothelial malignancies [1,2]. It is difficult to diagnose clinically since it is an unusual tumour and early diagnosis is impossible due to its rarity, misleading clinical presentation and non specific radiological findings. Hence, most of the patients are diagnosed in advanced stage and they will have poor prognosis.

Pure squamous cell carcinoma of renal pelvis are highly aggresive tumours with poor clinical course [3]. Primary Squamous cell carcinoma has got a female preponderence with a common age of 50-70 years [4]. In present case was a 73 years old male patient. The common risk factor is renal calculi in which there is development of metaplasia due to chronic irritation leading to dysplasia and finally squamous cell carcinoma [5]. Other risk factors are infections, vitamin A deficiency, endogenous and exogenous chemicals, radiotherapy, smoking [6,7]. In present case, tumour might have arisen due to long standing renal calculi which might have caused chronic irritation.

Mode of presentation is varied in patients with renal Squamous cell carcinoma and is dull aching flank pain, fever, weight loss which overlap with symptoms of renal stones. In our patient presenting features were dull aching loin pain with burning micturation. The diagnosis of primary squamous cell carcinoma by radiological investigation is inconclusive due to its non specific features and this tumour tends to grow from urothelium directly into the renal parenchyma which may lead to a diagnostic dilemma as in our case. Therefore, surgical resection and histopathological examination is the mainstay of diagnosis [8].

Histologic examination reveals tumours with extensive squamous differentiation with keratin pearl formation and intercellular bridging. These tumours are usually moderately to poorly differentiated tumours which are likely to be invasive and presents in advanced stages [9]. Our case was also reported as moderately to poorly differentiated squamous cell carcinoma infiltrating the capsule and perinephric fat with pT3aNXMX. Thyroidisation of renal tubules and interstitial infiltration by chronic inflammatory cells which are the features of chronic pyelonephritis were also seen in this case.

The differential diagnosis include urothelial carcinoma with squamous differentiation, metastatic squamous cell carcinoma, xanthogranulomatous pyelonephritis associated squamous cell carcinoma [10]. Immunohistochemistry markers like 34bE12, uroplakin , GATA 3 and PAX8 helps to differentiate between transitional cell carcinoma from Primary squamous cell carcinoma [11] and markers like CK14 and Mac 387 helps to differentiate between urothelial carcinoma with squamous differentiation and primary squamous cell carcinoma [12].

There are no standard guidelines for management due to the rarity of this tumour. Therefore, current modality is radical nephrectomy for the localised disease and for advanced disease multidisciplinary approach is followed which includes surgery, chemotherapy and radiotherapy. Our patient underwent radical nephrectomy and is currently on radiotherapy treatment with regular follow-up. The prognosis is poor because of its advanced stage of presentation. As surgical resection and adjuvant chemotherapy and radiotherapy are ineffective the prognosis with a 5 years survival rate is less than 10% [13,14].

CONCLUSION

Primary squamous cell carcinoma of pelvicalyceal system is a very rare entity and is associated with renal stones. Due to its non specific symptoms and inconclusive findings on radiological imaging, a high degree of suspicion of squamous cell carcinoma should be made in any patient presenting with history of renal calculi. It is a masquerader which can present in any form. Because of its aggressive nature and poor prognosis, it should be considered as a differential diagnosis and the diagnosis is confirmed only by histopathological examination.

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