Papillary Renal Cell Carcinoma Presenting As Pyonephrosis In A Young Adult.

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ABSTRACT

Papillary Renal Cell Carcinoma is the second most common type of renal malignancy in adults. It presents in fifth to seventh decade of life and is commoner in patients with End Stage Renal Disease and acquired renal cystic diseases. The presentation is similar to the clear cell type of Renal Cell Carcinoma and is asymptomatic usually. We report a case of Papillary Renal Cell Carcinoma in a young adult who presented with pyonephrosis.

Key words: Pyonephrosis, Papillary Renal Cell Carcinoma, Young Adult.

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INTRODUCTION

Papillary Renal Cell Carcinoma (RCC), the second most common histologic subtype of RCC, represents 10% to 15% of all the RCCs [1]. It is more commonly seen in patients with End Stage Renal Disease (ESRD) and in patients with acquired renal cystic disease [1]. The clinical presentation is the same as for clear cell RCC. Its presentation as pyonephrosis is rare. We describe such a presentation in a young adult.

CASE REPORT

A 23 year male was referred with history of painless hematuria not associated with clots, right hypochondriac and right lumbar pain associated with lump since 6 months. There was history of pigtailing and drainage of about 2 litres of pus elsewhere for right pyonephrosis. There was no history suggestive of Urinary Tract Infection. At admission, he was stable; his abdomen was soft, non tender, bladder was not palpable and pigtail was in situ in the right hypochondrium. Blood haematological investigations, renal function and liver function tests were normal. Contrast Computed Tomography (Figures 1a and 1b) suggested normal left kidney; pyonephrotic right kidney with pigtail in situ with extension of the pyonephrotic collection outside the Gerota's fascia; parenchymal loss of the Right kidney and loss of plane between the Right Kidney and surrounding structures. Right open nephrectomy was done. Intra-operatively, there was evidence of a hard mass at the middle and lower pole of the right

kidney (Figure 2 and 3). Histopathology revealed type I Papillary RCC of Fuhrman Grade III with capsule and perirenal infiltration.

Figure 1 (a and b): Contrast Computed Tomography showing normal Left Kidney; Pyonephrotic Right Kidney with pigtail in situ with extension of the pyonephrotic collection outside the Gerota's fascia; Parenchymal loss of the Right kidney and loss of plane between the Right Kidney and surrounding structures.

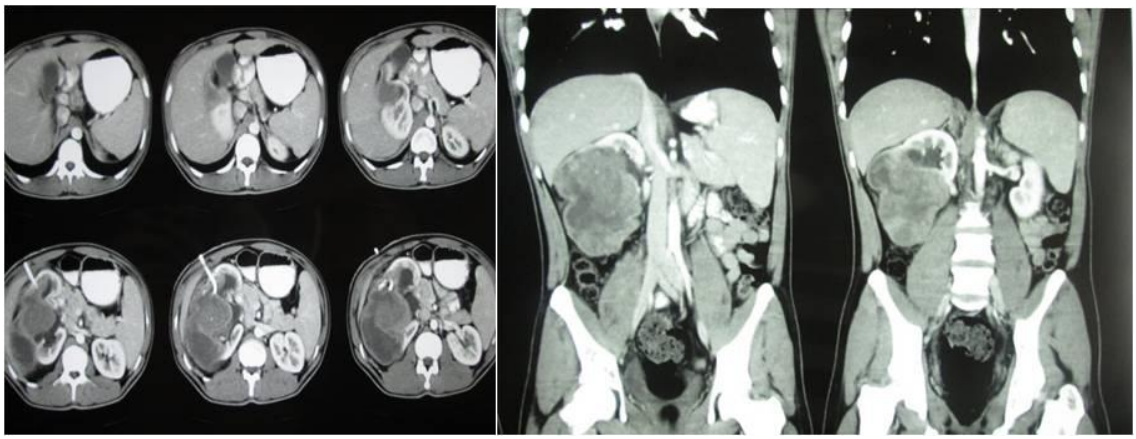


Figure 2: Intra-operative image showing pyonephrotic Right kidney with inflamed peri-nephric fat and Gerota's fascia.

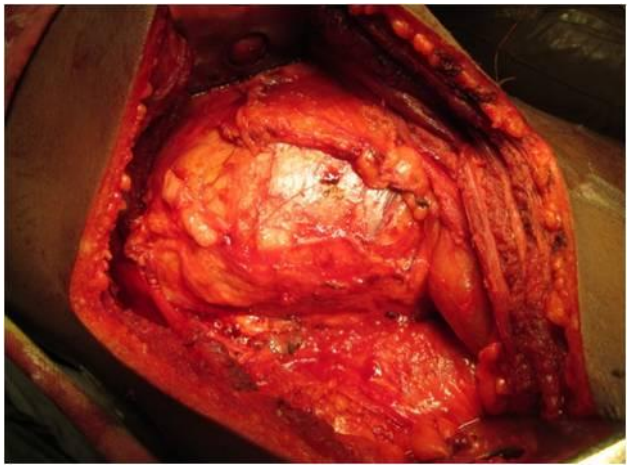
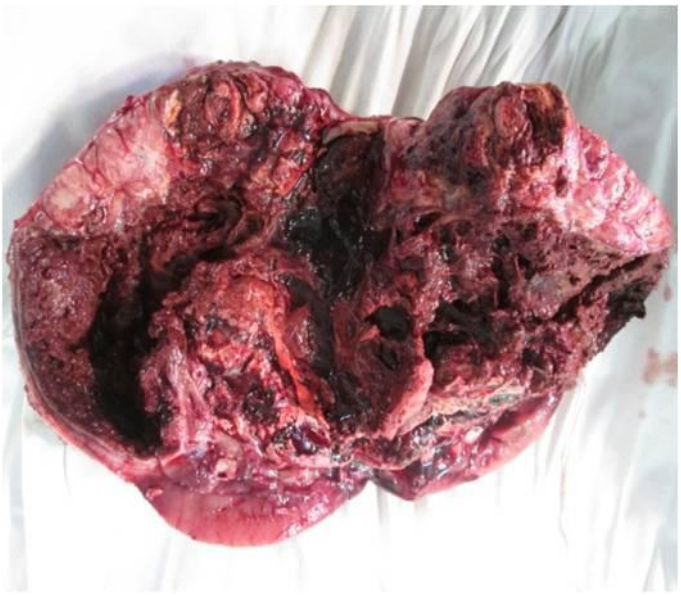


Figure 3: Cut-section of the excised specimen showing parenchymal destruction with inflamed, haemorrhagic and necrotic tissue with a mass at lower pole of the right kidney.



DISCUSSION

Malignant neoplasms of the kidney are common tumors and represent approximately 2% of all the human tumours. Renal cell carcinoma is the most common malignancy of the kidney in adults[2]. Papillary renal cell carcinoma represents the second most common histological subtype of RCC and comprises 2 subtypes [3]. Prognosis for type 1 Papillary RCC is relatively good, whereas type 2 Papillary RCC is associated with poor clinical outcomes [3].

The age of presentation is 50 to 70 years for type I Papillary RCC and approximately a decade earlier for type II Papillary RCC [4]. Approximately 50 percent of papillary carcinomas remain asymptomatic and are detected at advanced age or incidentally during non-invasive radiological investigations [1]. The symptoms are usually due to local tumour growth, haemorrhage, paraneoplastic syndromes or metastases [1]. The classic triad of flank pain, gross haematuria and palpable abdominal mass are rarely seen in the present days and always denote advanced disease [1]. The other indicators of advanced disease include constitutional symptoms like weight loss, fever and night sweats and findings include palpable cervical lymphadenopathy, non-reducing varicocoele, and bilateral lower limb edema because of venous congestion [1]. But presentation of papillary carcinoma as pyonephrosis is rare as seen in our case.

The modalities of investigation are Ultrasound and Computed Tomography (Plain and Contrast). Multiple gene mutations have been identified in Papillary RCC which may provide

clues regarding its tumorigenesis and serve as a basis for future developments of targeted therapies against type 1 and 2 Papillary RCC [3]. S100A11 and ferritin light chain proteins have been identified as the proteins abundant in papillary RCC regions compared to normal tissue [5]. Radical nephrectomy is the treatment of choice. The prognosis is better for type I Papillary RCC [4].

CONCLUSION

Papillary Renal Cell Carcinoma is the second most common histologic subtype of RCC which is more commonly seen in elderly patients with End Stage Renal Disease (ESRD) and in patients with acquired renal cystic disease. Its presentation in a young adult as pyonephrosis is rare.

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