Squamous cell carcinoma of renal pelvis presenting as pyelo-entero-cutaneous fistula: case report and review of literature

Arif Akhtar¹, Parveen Kumar Pandey¹, Raman Tanwar¹, Arvind Ahuja², Rajeev Sood¹

ABSTRACT

Objective: Squamous cell carcinoma (SCC) of renal pelvis is a subtle tumour constituting only about 0.7% to 7% of all renal malignancies and is considered to be highly truculent [1]. It is believed to arise through a process of metaplasia of the urothelium. Out of various aetologies proposed, the most pertinent is long standing irritation due to calculus and infection [2]. Such patients usually present with colicky pain, urinary tract infection and haematuria. Pain is mainly caused by obstruction at pelvi-ureteric junction or local extension. Haematuria is usually by calculus and renal mass. Other unfamiliar presentation documented in literature includes isolated hydronephrosis, pyonephrosis or local extension. Haematuria is mainly caused by obstruction at pelvi-ureteric junction or local extension. Haematuria is usually by calculus and renal mass. Other unfamiliar presentation documented in literature includes isolated hydronephrosis, pyonephrosis or local extension.

Method: We report a rare case of squamous cell carcinoma of the renal pelvis presenting as pyelo-entero-cutaneous fistula in a patient with long standing staghorn calculus. Result: Renal SCC can present without evidence of a distinct mass and it may closely mimic chronic inflammatory conditions like xanthogranulomatous pyelonephritis. Inconclusive clinical and radiological traits confounded by rarity, often delay the diagnosis. Majority of patients present with locally advanced or metastatic disease leading to poor survival. The diagnosis of SCC of renal pelvis was made on post surgery pathological examination. Conclusion: Renal SCC must be kept in mind as an important differential diagnosis in patient with non-resolving fistulas associated with chronic stone disease. These patients should be scrutinized with strong clinical suspicion with newer imaging techniques to aid in timely diagnosis in order to deter poor survival in these subset of patients.

KEY WORDS: Squamous cell carcinoma; Renal pelvis, Pyelo-Colo-duodeno-Cutaneous Fistula

INTRODUCTION

Squamous cell carcinoma (SCC) of renal pelvis is a subtle tumour constituting only about 0.7% to 7% of all renal malignancies and is considered to be highly truculent [1]. It is believed to arise through a process of metaplasia of the urothelium. Out of various aetologies proposed, the most pertinent is long standing irritation due to calculus and infection [2]. Such patients usually present with colicky pain, urinary tract infection and haematuria. Pain is mainly caused by obstruction at pelvi-ureteric junction or local extension. Haematuria is usually by calculus and renal mass. Other unfamiliar presentation documented in literature includes isolated hydronephrosis, pyonephrosis with peritoneal abscess and paraneoplastic symptoms [3]. We report a rare case of SCC of the renal pelvis presenting as Pyelo-entero-cutaneous Fistula.

CASE REPORT

A 53 years smoker, non-diabetic, non-hypertensive male patient presented to us with feco-purulent discharging sinus in the right flank for the past 3 months. He also complained of fever off and on, right flank and upper abdominal pain, generalised weakness, weight loss and loose stools since 3 months. Patient gave history of percutaneous right renal surgery in some other hospital for renal calculus two years back. According to the patient, postoperative period after surgery was uneventful and nephrostomy tube was removed on post operative day 2. As per him, complete stone clearance was not achieved and no post operative imaging was done after that. There was no documents available related to intra-operative details of surgery as well.

On examination 0.5 x 0.5 cm fistulous opening was present over the right flank just posterior to posterior axillary line with indurated margins with sero-purulent discharge. Biochemical evaluations revealed anaemia (Hb-8.0 gm/dl), high total leucocyte count (=15400) and hypercalcemia (S.Ca=11.4). Urine analysis showed pyuria with microscopic haematuria. His renal function was moderately deranged (blood urea=52, S.Creatinine =1.7). Both urine culture and pus culture from the fistulous tract showed mixed growth with evidence of food particles. Subsequently, a conventional contrast-enhanced CT (CECT) scan of abdomen was done which showed right hydronephrosis with multiple inferior and superior calyceal calculi (figure 1). In addition, there was a heterogeneous mass/collection involving the pelvis with thickening of the adjoining duodenum with a fistulous tract extending to the ascending colon (figure 2). Diethylenetriaminepentaaecetic acid (DTPA) renogram revealed a non-functioning right kidney. Subsequently, open right simple nephrectomy was planned.

On surgical exploration, there were dense adhesions between right kidney, colon and duodenum. Necrosis of the postero-lateral wall of the ascending colon was observed.
Further, second and third part of duodenum was plastered with adjoining the renal pelvis. The contour of the kidney was completely lost. The lower pole was necrotic, filled with pus and calculi. Dense adhesions were present with psoas muscle posteriorly. Right nephrectomy with excision of fistulous tract was performed. In addition, right hemicolecetomy with end ileostomy and mucous fistula creation was done. There was dense adhesion present near the second part of duodenum and during adhenolysis a 0.5×0.5 cm tear occurred in 2nd part of duodenum which was repaired primarily. Along with this, gasrojejunostomy with antral exclusion and a feeding jejunostomy was also performed. The patient developed a controlled duodenal fistula on the 8th post-operative day which was managed conservatively. But, eventually he succumbed to his illness two weeks later.
On gross examination, renal specimen was greyish white with fistulous opening at one pole containing multiple stones (largest of size stone 4 × 2.5 cm). Histopathology revealed well differentiated squamous cell carcinoma of kidney (figure 3) with features of chronic pyelonephritis with focal epithelioid cell granuloma and giant cells (figure 4). Resected part of caecum showed fistulous opening on the serosal surface with transmural infiltration by SCC. Colonic mucosa also showed focal squamous metaplasia and full thickness dysplasia (figure 5). The excised duodenal margins revealed tumour tissue with features of well differentiated SCC.

**DISCUSSION**

Pyeloenterocutaneous fistula is a rare but serious condition. Hippocrates first reported a case of reno-alimentary fistula way back in 460 BC [4]. Most often the cause is iatrogenic occurring secondary to per-cutaneous renal surgeries [5]. Other etiological factors include blunt or penetrating trauma, renal or colonic transitional cell carcinomas and foreign body ingestion. Apart from this, inflammatory processes (secondary to stones, tuberculosis, xanthogranulomatous pyelonephritis) and diverticular diseases are known to cause this entity [6,7]. SCC of the renal pelvis is a rare cause of pyeloenteric and pyelocutaneous fistula with only one case of each reported in literature. Poon et al. reported a case of Pyelo-duodenocolic fistula in such a patient who presented with torrential bleeding and shock [8]. Another case of a loin sinus secondary to renal SCC was described by Giannopoulos et al [4].

Renal SCC can present without evidence of a distinct mass and it may closely mimic chronic inflammatory conditions like xanthogranulomatous pyelonephritis [1]. It must be kept in mind as an important differential diagnosis in patient with non-resolving fistulas associated with chronic stone disease. Otherwise, CT often reveals a hypodense mass with associated calculi. In our case, the mass was heterogeneous and a possibility of thick organized pus resulting from recurrent pyonephrosis with external fistula was kept as a differential diagnosis. There was no substantial evidence to support malignant pathology in preoperative workup.

Radical nephrectomy is the mainstay of therapy in SCC of renal pelvis. Inconclusive clinical and radiological traits confounded by rarity, often delay the diagnosis. Majority of patients present with locally advanced or metastatic disease leading to poor survival as compared to their transitional cell (TCC) counterpart with median survival of 3.5 months [7].

**CONCLUSION**

Our case of Pyeloenterocutaneous fistula is an unreported presentation of pelvic SCC. It may be masked by presence of associated calculus and purulent material. SCC should strongly be suspected while dealing with chronic inflammatory renal pathology with calculus in non-functioning kidney. Upper tract fistulas should be scrutinized with strong clinical suspicion with newer imaging techniques to aid in timely diagnosis in order to deter poor survival in these subset of patients.

**CONFLICT OF INTEREST STATEMENT**

There is no conflict of interest of authors.

**REFERENCES**