Primary renal squamous cell carcinoma in a non-functioning kidney with nephrolithiasis: A case report

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ABSTRACT

Primary squamous cell carcinoma (SCC) of renal pelvis is a rare neoplasm. A 50-year-old female presented with a history of chronic dull aching pain in left flank region and burning micturition for 8 months. Workup showed multiple calculi in non-functioning, hydronephrotic left kidney. Histopathological examination of nephrectomy specimen revealed SCC of renal pelvis; although, there was no obvious growth. The case report highlights the fact that SCC should be suspected in a patient with renal mass in a non-functioning kidney following long-standing nephrolithiasis.

Key words: Calculus, Kidney, Nephrolithiasis, Renal, Squamous cell carcinoma

Primary malignancies of the renal collecting system are rare accounting for 4-5% of all urothelial tumors [1]. Squamous cell carcinoma (SCC) of the renal pelvis is a rare tumor with incidence being 1.4% of all renal malignancy [2]. Risk factors for primary renal SCC are renal calculi, infection, endogenous and exogenous chemicals, hormonal imbalance, radiotherapy, and Vitamin A deficiency. These tumors are highly aggressive, high grade, and locally advanced, or metastatic at the time of presentation related to poor prognosis [1]. Renal SCC is strongly associated with renal stones; hence, the patients with renal stones and non-functioning kidney should be carefully examined with early histopathological diagnosis for early detection and management.

Patients of renal SCC mainly present with dull aching pain, hematuria, fever, or weight loss. It presents in late adulthood with equal incidence among men and women [3]. Radiologically renal SCC has diverse presentation as solid irregular mass, hydronephrosis, calcification, or renal infiltrative lesion without any distinct mass. Clinical and radiological non-specific features lead to diagnostic confusion. Thus, precise histopathological diagnosis after nephrectomy is important for diagnosis.

CASE REPORT

A 50-year-old female came to the surgical OPD with complaints of left flank pain, burning micturition, on and off hematuria along with on and off fever for 8 months. There was no significant history or family history. Physical examination revealed a tender lump in the left lumbar region. Urine examination showed 2+ proteinuria along with presence of pus cells and red blood cells. Routine hematological and biochemical investigations were normal with blood urea of 20 mg/dL and serum creatinine of 1.0 mg/dL. Ultrasonography

revealed left kidney staghorn calculus with gross hydronephrosis. Further computed tomography (CT) of the kidneys revealed a large 4×2 cm left renal staghorn calculus with multiple secondary calculi with gross hydronephrosis and marked parenchymal thinning. DTPA scan revealed a grossly hydronephrotic non-functioning left kidney and a normally functioning right kidney.

Left nephrectomy was performed which peroperatively showed pyonephrosis with the presence of a large staghorn calculus along with dense adhesion between renal capsule and surrounding tissue. Left nephrectomy specimen was sent to pathology department. Gross examination revealed a distorted, enlarged kidney with thickened capsule adherent to renal parenchyma. Sectioning revealed a markedly dilated pelvicalyceal system filled with a large staghorn calculus and many smaller calculi. The walls of the dilated cavities were thickened and pearly white in appearance (Fig. 1). There was no obvious tumor. Histological examination revealed nests of malignant squamous cell with large areas of tumor necrosis (Fig. 2). There were areas of compressed normal tubules and glomeruli in between. There was presence of squamous metaplasia and dysplasia in the adjacent urothelium. Foci of lymph-vascular invasion and perinephric tumor extension were also seen. However, sections from ureter were free from tumor. A diagnosis of moderately differentiated SCC of renal pelvis with extensive involvement of renal parenchyma was made. The patient is undergoing cisplatin-based chemotherapy and is doing well 6 months after surgery with no signs of metastasis or recurrence.

DISCUSSION

SCC of pelvicalyceal system with involvement of kidney is a very rare entity and should be considered while evaluating a



Figure 1: Distorted, enlarged kidney with dilated pelvicalyceal system having thickened pearly white wall

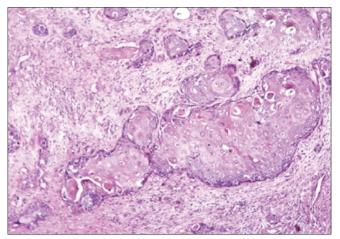


Figure 2: Histological section showing moderately differentiated invasive squamous cell carcinoma (H and E \times 400)

non-functioning renal mass associated with renal calculi. The predisposing factors such as renal calculi, infections, exogenous, and endogenous chemicals (e.g., arsenic), prior history of renal stone surgery, analgesic abuse, radiotherapy, and vitamin A deficiency cause chronic irritation which leads to development of urothelial metaplasia further progressing to dysplasia's, and finally to an SCC. The strongest association has been reported with nephrolithiasis, especially with impacted staghorn calculi of a long duration as seen in our case. Li and Cheung in their study reported an incidence of renal SCC in 2% cases of recurrent renal stones [4]. Renal pelvic tumor is mostly non-palpable clinically; however, they may lead to palpable hydronephrosis due to urinary outflow obstruction as was seen in our case. In our case, we have speculated that the tumor has arisen in a chronically inflamed hydronephrotic pelvis due to long-term irritation by large calculi.

Many studies in literature presented similar findings of primary renal SCC arising in a hydronephrotic kidney with

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renal calculi [5,6]. However, Badruddoza et al., in their case report of primary renal SCC did not find any etiological factor for the same [7]. These tumors are highly aggressive and are usually high grade at the time of diagnosis. Extensive local and retroperitoneal soft tissue infiltration are very common [8]. Conventional ultrasonography which is the choice of imaging modality for renal diseases lacks specific echoic pattern in renal SCC. Diagnosis of SCC is difficult by available imaging modalities; therefore, the diagnosis of the renal SCC is usually made only after histological examination of resected specimen as was seen in present case. Surgical resection (nephrectomy or nephroureterectomy) is considered as the mainstay of treatment for renal SCC. However, in advanced disease, surgery along with adjuvant chemoradiotherapy has been tried with not much survival benefit, emplacing the need for early diagnosis.

CONCLUSION

SCC of urothelial tract arises following metaplasia and dysplasia of urothelium. Any patient who presents with a renal mass following a long history of renal calculus in non-functioning kidney should undergo careful investigation with early surgery and thorough histopathological examination.

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