

Case Report

Primary squamous cell carcinoma of kidney - a case report of unusual local recurrence with literature review

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Abstract

Primary squamous cell carcinoma of the kidney is a rare clinical entity. Primary squamous cell carcinoma is more frequently reported in urinary bladder and male urethra than in kidney. Primary renal squamous cell carcinoma (RSCC) accounts for less than 5% of urothelial tumours in urinary system. Very few cases of primary squamous cell carcinoma of kidney have been reported in the world literature. Only forty-six cases over forty four years had been reported from Mayo Clinic. These neoplasms are highly aggressive and locally advanced or metastatic at the time of presentation. Here we report such a case of primary squamous cell carcinoma of kidney in a 50-year-old lady at a locally recurrent stage with fistulous tract formation. Palliative radiotherapy came out to be a good clinical modality even in locally recurrent stage in this patient.

Key words: primary squamous cell carcinoma of kidney, locally recurrent, fistulous tract.

Introduction

The common malignancies arising from kidney in adults is of clear cell carcinoma, followed by papillary carcinoma and chromophobe cell carcinoma.^{1,2} Squamous cell carcinoma is not common in kidney. Renal squamous cell carcinoma (RSCC) is known to arise from renal collecting system.³ The insidious onset of symptoms and absence of any pathognomonic feature leads to delay in diagnosis and treatment.

Case report

A 50-year-old lady presented with pain at left flank for six months. There was no history of fever, haematuria or weight loss. On per-abdominal examination, a ballotable mass was palpable at left flank. Ultrasonography of Kidney, Ureter and Bladder region (USG-KUB) was suggestive of hydronephrosis of left kidney with multiple echogenic calculi. Intravenous urogram showed left kidney to be non-functional. Serum biochemistry profile, blood counts and urine analysis - all were within normal limits. Left sided simple nephrectomy was done and a hugely distended and thinned out left kidney was found during operation. Post-operatively, the patient was lost to follow-up by Urologist. She was asymptomatic for six months, after which she had to attend the hospital again for the complaints of reappearance of flank pain, followed by discharge from incision

site and intermittent fever. Mantoux test and sputum for Acid Fast Bacilli (AFB) were negative. A Sinogram was performed and it showed sinus tract formation at left lumbar region without any obvious communication with gut. Gram stain, Ziehl-Neelsen (Z-N) stain and culture-sensitivity of swab taken from the discharging wound were negative. Discharge was persisting even with vigorous conservative management. Then, the biopsy report of the previously operated nephrectomy specimen was collected and it revealed - well differentiated squamous cell carcinoma of kidney. The ureter and perinephric fat was free of lesion, but hilar lymph nodes showed presence of tumour deposit. This patient was then referred to the department of Radiotherapy of the institute. CT Scan of lower abdomen was performed which showed scoliosis with an ill-defined poorly enhancing lesion (measuring 11.7 cm × 6 cm) with necrotic centre in left paravertebral region from just below the left dome of diaphragm to the lumbar region. It had protruded out of the abdominal cavity into the subcutaneous plane at left flank. Fine Needle Aspiration Cytology (FNAC) from left paravertebral mass was suggestive of squamous cell carcinoma. Patient's performance status was ECOG 2 (Eastern Cooperative Oncology Group) and only palliative External Beam Radiotherapy has been provided in the schedule of 30 Gy in 10 fractions over 2 weeks. Patient is symptomatically improved

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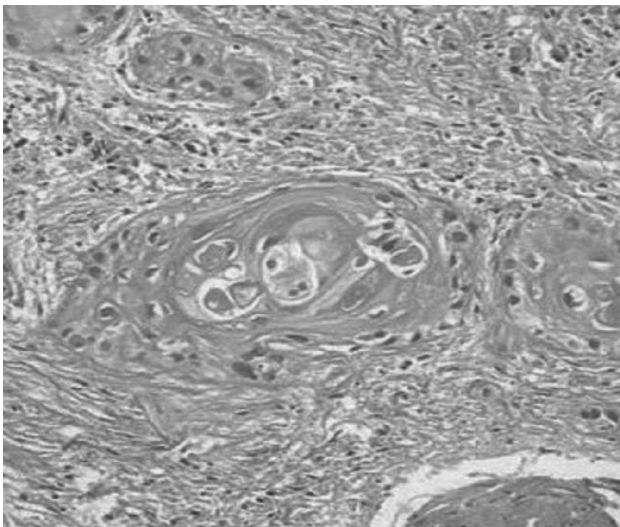
and still alive one year post-radiotherapy. This case is unique because though the case was managed primarily with surgery, but it was diagnosed at the time of recurrence in absence of any adjuvant therapy and local recurrence with fistulous tract formation has not been found in any of the previously reported cases. This may be the first one. Palliative radiotherapy came out to be a good clinical modality even in locally recurrent stage.

Discussion

Primary neoplasms of the renal collecting system account for less than 5% of urothelial tumours in urinary system.⁴ The most frequent variety is transition-

but now it is not considered true as this disease is reported equally in male. The mean age of presentation is 56 years.^{2,7} Pain and hematuria are the most common presenting symptoms. Pain is due to pelvi-ureteric junction obstruction and/or local extension.^{2,7} The reported patient was a female with similar age presenting with pain, but not haematuria.

RSCC is usually detected with extensions into the renal vein, inferior vena cava or even with distant metastasis. This patient was presented without any extension into these venous systems or any distant metastasis.



al cell type (85%-95%) and very few cases are squamous cell carcinoma and adenocarcinoma.⁵ The incidence of primary renal squamous cell carcinoma (RSCC) ranged from 0.5%-5%.^{1,2} Only forty-six cases over forty four years had been reported from Mayo Clinic suggesting its rarity.⁶ Female sex predilection was found in older reports

In a retrospective study by Lee et al, that reported 15 patients of RSCC, these tumour were classified into two groups depending on localization as central and peripheral. Central RSCC presents more with intraluminal components and lymph node metastasis is common whereas peripheral RSCC presents with prominent renal parenchymal thickening and might

invade the perirenal fat tissue without identifiable lymph node or distant metastasis. The survival of patients with central type was reported to be significantly shorter than those with peripheral RSCC.⁸ The present case with hilar nodal metastasis without perirenal fat invasion is a peripheral one as per Lee classification.

Chronic irritation of urothelium is presumed to be the cause of squamous metaplasia with subsequent malignant progression to SCC.³ Various factors have been implicated for squamous metaplasia and subsequent carcinoma. The etiological factors strongly associated with this malignancy are chronic renal calculi, pyelonephritis and phenacetin consumption.³ Among them urinary calculi is accepted as the main carcinogenic risk factor.³ The incidence of co-existing urinary stone was reported in a wide range between 18% and 100%.^{1,2} Other factors implicated include Vitamin A deficiency, hormonal imbalance, schistosomiasis and smoking. However, cases have been reported in which no apparent etiological factor could be detected.⁹ The present case also was associated with renal stone.

A solid mass, hydronephrosis and calcifications are common radiologic findings, but being nonspecific, the diagnosis could be missed before the histopathological examination. The present case also presented with renal calculi and hydronephrosis and diagnosed only by histopathology.

These tumors are highly aggressive, high grade, and locally advanced or metastatic at the time of presentation.⁶ The prognosis of renal SCC is very poor. A median survival of 5-to-11-month is reported in previous case-series.^{2,3,7} This patient has been surviving for 15 months post-surgery without any systemic therapy except palliative radiotherapy at the time of recurrence.

Radical nephroureterectomy with excision of the bladder cuff [guidelines proposed by Johansson and

Wahlqvist (1979)] appears to be the treatment of choice for patients not having distant metastasis.^{2,7,10} Unfortunately, majority of the patients have locally advanced or metastatic disease at the time of presentation and in these patients, nephrectomy with or without ureterectomy is recommended.⁷ Nephrectomy is necessary even in metastatic disease to establish a histological diagnosis, for control of symptoms such as pain, fever and hematuria or to eliminate the source of infection before systemic chemotherapy can be instituted.² Radiotherapy or systemic chemotherapy seems to have little benefit.^{1,7} Cisplatin based chemotherapy and palliative radiotherapy have been advocated only for the control of local symptoms in metastatic disease but have failed to show any survival benefit.² Nativ et al. also reported that the treatment modalities like nephrectomy, nephroureterectomy, adjuvant radiotherapy or chemotherapy, irrespective of tumor stage, did not affect the survival of these patients.⁶ As performance status of the present case was not suitable for combination chemotherapy, this patient was treated with palliative radiotherapy only.

So the conclusion is that as RSCC are strongly associated with urinary calculi existing for prolonged duration, patients with renal stone who are kept on observation or managed with extracorporeal shock wave lithotripsy or has non-functioning kidney should be carefully examined with imaging modalities; if nephrectomy has been done, histopathological examination of the specimen should always be sought for, unlike our case, to detect existing primary RSCC; in recurrent stage, palliative radiotherapy is a good treatment option.

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