Oncology

Squamous Cell Carcinoma of the Renal Pelvis: Atypical Presentation of a Rare Malignancy

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ABSTRACT

Primary squamous cell carcinoma (SCC) of the renal pelvis is exceedingly rare and is associated with advanced stage at diagnosis and a poor prognosis. Diagnosis is often difficult due to vague presenting symptoms and nonspecific radiologic findings. The mainstay of treatment is nephrectomy, with cisplatin based chemotherapy and palliative radiation reserved for metastatic disease. We report a SCC of the renal pelvis in a 72 year-old female who presented with abdominal pain. To date, this is the largest SCC originating from the renal pelvis reported in the literature, made even more exceptional by the lack of antecedent risk factors.

Introduction

Upper urinary tract malignancies are rare, accounting for 8% of all renal malignancies, with urothelial carcinoma comprising the vast majority of histological type. Even less common is squamous cell carcinoma (SCC) of the upper tracts, accounting for 10% of all renal pelvic tumors and 0.5% of all renal malignancies.1 SCC of the upper tracts appears to be more aggressive and is diagnosed at a later clinical stage than other more common histologic types.2 Diagnosis is challenging, as presentation is often non-specific and locally advanced or metastatic disease is common upon presentation. Radical surgical resection is the foremost treatment option, as alternative treatments are of limited efficacy. Despite aggressive surgical efforts, prognosis remains poor as most patients die within one year of surgery.3 There have been few case reports in the literature describing renal pelvic SCC; we thus describe a case of a 19 cm SCC occurring in the renal pelvis of a 72 year-old female.

Case presentation

A 72 year-old female presented to the ED with 4 weeks of gradually worsening left upper and lower quadrant pain. She has a history of hypertension, congestive heart failure, hyperlipidemia, atrial fibrillation (rate controlled; not on anticoagulation), diverticulitis, and underwent TAHBSO with 8 rounds of vaginal brachytherapy for endometrial cancer 13 years prior; she was rendered disease free since. She reported no smoking history. She endorses a 30-lb weight loss over the prior two months due to decreased appetite. Her family history is significant for a brother with multiple myeloma and colon cancer. Colonoscopy and endoscopy 1-month prior were unremarkable.

She underwent a CT scan of the abdomen and pelvis with contrast (Fig. 1) which demonstrated a 16.4 cm × 12.3 cm left renal neoplasm occupying most of the left hemipelvis encasing the distal abdominal aorta, left common iliac artery and proximal left external and internal iliac arteries, without evidence of vascular invasion. Also noted was severe left hydronephrosis with obliteration of the left ureter distal to the renal pelvis.

She underwent a biopsy of the retroperitoneal mass which revealed SCC, moderately differentiated, keratinizing type. Due to her symptomatology, she opted to proceed with left nephrectomy and resection of the retroperitoneal mass. With the involvement of urology, surgical oncology, and vascular surgery, the patient underwent an exploratory laparotomy, bowel resection, with en block resection of the retroperitoneal mass and the left kidney.

Histopathological examination (Fig. 2) revealed a 19 cm SCC arising from the renal pelvis, moderately differentiated, keratinizing type involving the left kidney, retroperitoneal fibroadipose
tissue, and serosa of the small bowel. Lymphovascular invasion was identified, and margins were negative, pT4N0Mx.

Discussion

The mean age at presentation for renal pelvic SCC is 56 years, with no predilection for sex or laterality.3 It is more common in the urinary bladder and in the male urethra and rarely presents in the renal pelvis. Paraneoplastic syndromes such as hypercalcemia, leukocytosis, and thrombocytosis have been observed. Prognosis is often poor, and most tumors have been reported to be high grade with 84% being locally advanced or metastatic.4 Nativ et al reported 1 and 2 year survival rates of locally invasive renal SCC to be 33% and 22%, respectively.4 5-year survival is under 10%, and most patients die within one year of surgery.7

The majority (94%) of renal SCC’s present at an advanced stage.3 Presentation is obscure in these patients, as they often present with vague abdominal pain and hematuria, and weight loss in advanced stages.3 Specific findings of an enhancing extraluminal mass or an exophytic mass has been previously described; however there appears to be no specific radiological features that allow for preoperative diagnosis in most cases. Imaging is often not as impressive as a 19 cm mass seen in our patient, and adds to the diagnostic dilemma as imaging may only demonstrate stones, hydrenephrosis, a filling defect on urography or a ureteropelvic junction obstruction. Lee et al2 classified SCC’s of the upper tracts into two main categories based on tumor location: central and peripheral. Central type tumors were associated with higher rates of nodal metastasis and lower survival rates, and peripheral type tumors showed parenchymal thickening with peri-renal infiltration. The present case was a central type malignancy as per this classification.

Different etiological factors including renal calculi, hydrenephrosis, chemical use/abuse, vitamin A deficiency, and hormonal imbalances have been implicated in the pathogenesis of upper tract SCC.5 The strongest association has been reported with stone disease; up to 100% of previously reported cases of upper tract SCC had concomitant nephrolithiasis.5 The proposed mechanism for the development of SCC appears to be chronic inflammation, irritation and infection that lead to the squamous metaplasia of the urothelium. Interestingly, our patient had none of the aforementioned risk factors, with the exception of hydrenephrosis which was likely

Figure 1. CT abdomen/pelvis showing A) markedly thin renal cortex and severe left hydrenephrosis, B) tumor encasing the abdominal aorta, C) tumor encasing the iliac vessels, D) coronal view of the tumor, E) sagittal view of the tumor.

Figure 2. A) Low power (4× magnification) showing tumor in kidney, B) high power (10× magnification) showing keratinizing squamous cell carcinoma.
secondary to local tumor compression. Few similar cases of upper tract SCC have been reported in the absence of the proposed risk factors.

The foremost treatment of renal pelvic SCC is nephrectomy with or without urethrectomy. Platinum based chemotherapy and radiotherapy is reserved for patients with metastatic disease, however nephrectomy is still indicated in the presence of metastasis in order to establish a histopathologic diagnosis, control symptoms, and eliminate a potential source of infection before initiating chemotherapy. Currently, a combination of cisplatin, methotrexate, and bleomycin is employed, but fails to show any survival benefit.

Conclusion

Renal pelvic SCC is exceedingly rare. The insidious onset and nonspecific radiologic findings permit for late detection, often when the tumor becomes locally advanced or metastatic. These tumors are treated with aggressive surgical resection, with chemoradiation in the metastatic setting. The possibility of SCC should be recognized when an upper tract tumor is suspected in the setting of the aforementioned risk factors, therefore accurate history taking in conjunction with proper imaging is vital. We also highlight that this malignancy may occur even in the absence of these risk factors. The present case is unique in that this is the largest SCC originating from the renal pelvis reported in the literature, made even more exceptional by the lack of antecedent risk factors.

Conflicts of interest

None.

References

1. Busby JE, Brown GA, Tamboli P, et al. Upper urinary tract tumors with non-transitional histology: A single-center experience. Urology. 2006;67:518–523.
2. Lee TY, Ko SF, Wan YL, et al. Renal squamous cell carcinoma: CT findings and clinical significance. Abom Imaging. 1998;23:203–208.
3. Holmang S, Lele SM, Johansson SL. Squamous cell carcinoma of the pelvis and ureter: Incidence, symptoms, treatment and outcome. J Urol. 2007;178:51–56.
4. Nativ O, Reiman HM, Leiber MM, et al. Treatment of primary squamous cell carcinoma of the upper urinary tract. Cancer. 1991;68:2575–2578.
5. Tyagi N, Sharma S, Tyagi SP, et al. A histomorphologic and ultrastructural study of the malignant tumors of the renal pelvis. J Postgrad Med. 1993;39:197–201.