Upper Tract Urothelial Carcinoma in Ectopic Pelvic Kidney

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Abstract

Upper tract urothelial carcinoma (UC) is an uncommon tumor. Ectopic kidney is also a rare entity. The combination of these two conditions is very rare. A 49-year-old male complained of right flank pain with hematuria. On CT scan he was found to have a malrotated right kidney with soft tissue seen in the upper calyceal group and a normal left kidney. Diagnostic cystoscopy was unremarkable. Radical nephroureterectomy with bladder cuff excision was performed. Pathology report revealed low grade urothelial carcinoma. Patient’s symptoms disappeared postoperatively. Follow up showed no recurrence during the first two years in the bladder and upper tract in the contralateral kidney. Isolated UC of ectopic kidney is rare disease three cases were reported in literature. Although treatment of this tumor can be challenging due to its complex blood supply and position inside the pelvis, treatment strategy is still similar as for orthotopic kidneys.

Introduction

Renal ectopia or ectopic kidney is a rare condition involving a failure of the mature kidney to reach its normal location within the renal fossa. Different types of ectopic kidneys exist according to its location: pelvic, abdominal, thoracic, iliac, contralateral or even crossed. About one in 900 people has an ectopic kidney. Urothelial carcinoma of the upper tract is rare. It constitutes less than 10% of the upper tract tumors. The incidence of ectopic kidney with UC is still rare. Here, we report a case of UC in a pelvic kidney in a patient with painful hematuria.

Case presentation

A 49-year-old male, previously well, non-smoker, presented to our clinic complaining of right flank pain associated with microscopic hematuria for few weeks duration. He recalls no previous similar episodes in the past. He was on anticoagulant medication for cardiac cause. He is a merchant. Physical examination was unremarkable and no abdominal masses were palpable.

Laboratory workup showed microscopic hematuria and normal serum creatinine. Ultrasonic examination of the kidneys and bladder showed right pelvic kidney with soft tissue mass. Computerised tomography (CT) of the abdomen and pelvis with IV contrast and urogram phase was done showing right malrotated pelvic kidney with enhancing soft tissue mass measuring (2.3 * 1.8 * 2.8 cm) in the upper calyceal group. The right pelvic kidney has two renal arteries; one supplies the upper pole including the tumor location and arising from abdominal aorta near the bifurcation, and the other supplies the lower pole away from the mass. The left kidney appeared normal. And no evidence of metastasis was found.

Diagnostic cystourethroscopy and uretero-renoscopy to left renal pelvis was performed and no abnormality was identified. Then, right open radical nephroureterectomy with bladder cuff excision was performed. Post operative period was unremarkable and he had smooth recovery. Histopathological examination of the specimen revealed low grade papillary urothelial carcinoma in the upper calyceal group limited to the mucosa and no lamina propria invasion (pTa) with negative margins.

Follow up schedule was arranged and cystoscopies, upper tract imaging studies and urine cytology were done regularly. The patient has been recurrence-free for over 2 years since tumor resection.

Discussion

The relationship between malignancy and ectopic kidneys is still unclear. Although three cases of urothelial carcinoma and few cases
of renal cell carcinoma (RCC) in ectopic kidneys have been reported in the literature, there does not appear to be an increased risk of malignancy in ectopic kidneys.3

The reason for the lack of association between these entities is still unknown. Upper tract urothelial carcinomas (UTUC) are uncommon and account for only 5–10% of UCs.4 At the time of writing this report, there were only three reported cases of urothelial carcinoma in an ectopic pelvic kidney.

Joseph Philipraj and his colleagues reported a renal mass in pelvic kidney presented with suprapubic pain difficulty voiding. It was detected on CT scan and a provisional diagnosis of RCC was made. Radical nephro-ureterectomy was carried out and revealed grade II transitional cell carcinoma (Figs. 1 and 2).5

Arisawa C and his colleagues reported a left renal pelvis mass in synchronous with bladder mass and contralateral renal mass. The left renal pelvis mass appeared to be grade II transitional cell carcinoma, the bladder mass was TCC and the contralateral renal mass was clear cell RCC.6

Terai and his colleagues reported in 1994 a transitional cell carcinoma in pelvic kidney associated with bronchogenic carcinoma in a patient presented with lower abdominal painful mass.7 The rarity of such cases in daily practice made it a challenge for the urologists. Most cases of ectopic kidneys present with atypical presentation. However, the existence and availability of modern radiological imaging techniques increased the detection of such anomaly over the years.

The treatment options for UTUCs vary from endoscopic to surgical approaches depending on the type, site, clinical stage of malignancy, co-morbid conditions and the surgeon’s experience. Radical nephro-ureterectomy with bladder cuff excision appears to be the standard treatment for localized UTUCs in ectopic kidneys. However, in all reported cases, the histopathological findings revealed low grade urothelial carcinoma and thus the question regarding the need for this radical treatment and the significance of this tumor is raised. Cystourethroscopy is advised prior to the surgery to detect any synchronous tumors. Follow up is required to detect any metachronous bladder tumors, local recurrence and distant metastases.

The ectopic kidneys often maintain the fetal blood supply from the iliac vessels or the distal aorta. Pelvic kidneys have an anomalous vascular supply and collecting systems. There may be a single or multiple renal arteries. Anomalous renal arteries may arise from the distal aorta, superior mesenteric, common iliac, internal iliac or external iliac arteries. The surgical approach to ectopic kidneys merits caution because of the uncertain vascular anatomy; vascular studies may be indicated to assist preoperative planning. Identification of the renal arteries and veins near the mass is necessary to avoid intra-operative complications.

Conclusion

Upper tract urothelial carcinoma of ectopic kidney is a rare disease with only three cases were reported in the literature. Although the treatment of this tumor can be challenging due to the complex blood supply and position inside the pelvis, treatment strategy is still similar as for orthotopic kidney.

Conflict of interest

This case report was done in the division of urology for academic purposes and was not funded by any external fund, the submission has no commercial interests, and the authors of this case report are not linked to any external agencies.

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