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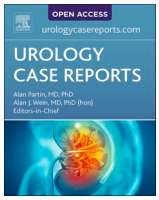

Oncology

A curious case: Concurrent collecting duct renal cell carcinoma and upper tract urothelial carcinoma

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A B S T R A C T

A 71-year-old male presented to Urology with three weeks of overt haematuria and increasing lethargy. Contrast-enhanced CT scans revealed an 8 × 6cm partially exophytic lesion in the left kidney's upper pole, extending beyond the capsule and invading the superior cortical vein, accompanied by abnormal retrocrural lymph nodes. Signs of paraneoplastic syndrome prompted a left radical nephrectomy for symptom relief. Histological analysis identified high-grade collecting duct renal carcinoma and invasive urothelial cell carcinoma. Post-surgery, he was referred for oncological treatment but passed away within two months of the initial diagnosis.

1. Introduction

Collecting duct renal cell carcinoma (CDRC) is a rare form of renal cell carcinoma (RCC) that originates from collecting duct cells of the kidney.[1] Despite CDRC representing less than 1% of all RCC cases, it is highly aggressive and approximately 32% are metastatic at diagnosis.[1]

Urothelial cancer of the renal pelvis (upper tract) is a much rarer presentation of urothelial carcinoma accounting for 5–10% of cases.[2]

The combination of the two provides similar presentation characteristics, these include abdominal pain, flank mass, haematuria, lethargy and classical systemic features.[3,4]

2. Case presentation

A 71 year old man was referred by his General Practitioner (GP) to our Urology service after 3 weeks of abdominal pain, frank haematuria, progressive lethargy and intermittent subjective fevers. Medical history included previous Gleason 3 + 4 = 7 Prostate Cancer which was treated with ADT and radiotherapy, type 2 diabetes mellitus, dyslipidaemia and an essential tremor. He was a non-smoker and a social consumer of alcohol, previously independent living with his wife and having unlimited effort tolerance.

Physical exam was unremarkable with no flank pain or palpable mass.

3. Investigations

Full blood count revealed an elevated white cell count at 23.6 × 10^9/L with a predominant neutrophilia, interestingly monocytes and eosinophils were both markedly elevated (1.48 × 10^9L and 2 × 10^9/L respectively. Urine microscopy revealed 210 leucocytes and >500 erythrocytes with no bacterial growth. Urine cytology was suspicious for high grade urothelial carcinoma, specifically it showed urothelial cells with increased nuclear to cytoplasmic ratio, moderate-to-severe hyperchromasia and irregular nuclear borders. CT Chest/abdomen/pelvis showed performed by the GP described a solid lesion in the left upper pole of the kidney with extracapsular extension and venous invasion into superior cortical vein with pathological retrocrural lymph nodes (Figs. 1 and 2). PSMA PET[5] showed an avid heterogenous mass lesion and the left upper pole of the kidney and mildly PSMA avid nodal disease in the left para-aortic and the retrocrural stations. Several osseous foci with PSMA avidity and subtle bone changes were also noted in the posterior left 1st rib, left glenoid, right sacral alar and right medial ilium.

4. Management

Initial management included a cytoreductive laparascopic simple nephrectomy was performed to alleviate symptoms of para-neoplastic syndrome, the main concern being unrelenting fevers.

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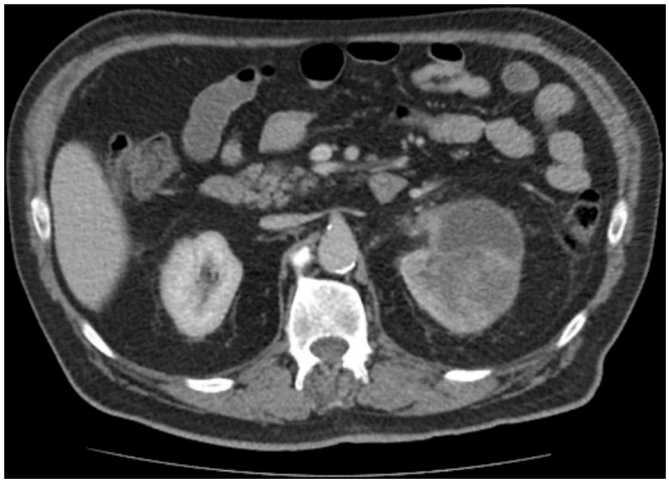

Fig. 1. Axial CT demonstrating a large heterogenous endophytic lesion in the upper pole of the left kidney with an adjacent simple cyst.

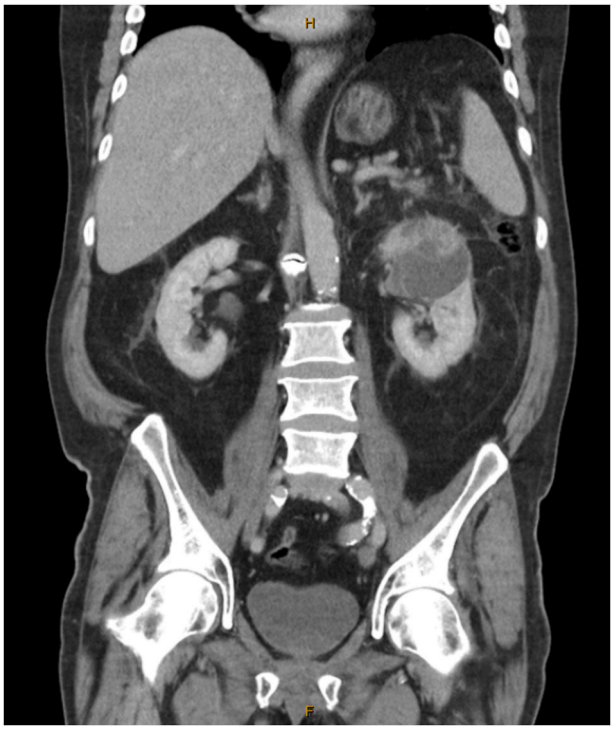

Fig. 2. Coronal CT demonstrating heterogenous lesion in the upper pole of the left kidney with an inferiorly adjacent simple cyst.

Histopathological analysis served as a foundation for tailoring subsequent chemotherapy regimens. Following an extended period of rehabilitation necessitated by significant postoperative deconditioning, the patient was discharged.

5. Diagnosis

The gross specimen was prepared into slides for analysis (Fig. 3). Histopathological interpretation of the specimen showed two distinct malignancies present (Fig. 4). Poorly differentiated rhabdoid, glandular and sarcomatoid malignancy were immunohistochemically strongly positive for PAX8, CK8/18 and 34βE12 but negative for GATA3, desmin or HMB-45; consistent with collecting duct renal carcinoma.

A high grade papillary urothelial carcinoma was also identified with

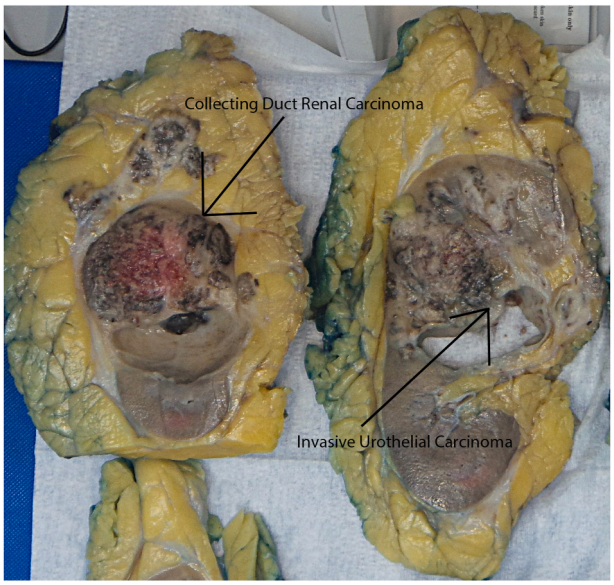

Fig. 3. Macroscopic preparation of the ex vivo left kidney, an obvious tumour is demonstrated in the upper pole.

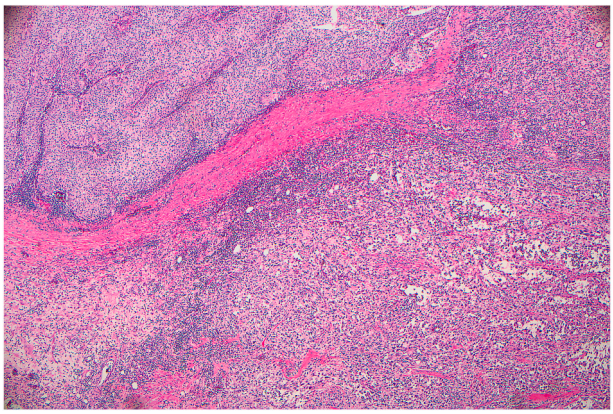

Fig. 4. In the upper pole of the left kidney, a high grade papillary and invasive urothelial carcinoma (upper left) abuts a poorly differentiated sarcomatoid malignancy (lower right).

confirmed invasion into the lamina propria but not into the muscularis propria or further. Immunostaining for this component was positive for CK8/18 and 34βE12 but negative for GATA3, desmin or HMB-45.

6. Outcome and follow up

The patient was referred to the medical oncology team for urgent follow up to determine definitive management, a multidisciplinary team meeting determined that a preliminary chemotherapy regime would include a combination of carboplatin and gemcitabine. Unfortunately the patient passed away less than two months after diagnosis from progressive decline and ultimately starvation ketoacidosis. Post surgically there was a residual post operative seroma but no complications otherwise.

7. Discussion

We present a novel case of concurrent collecting duct renal carcinoma and papillary urothelial carcinoma. Collecting duct renal carcinoma is exhibited to be aggressive in nature. Due to its low incidence, there is poor understanding of the condition. It represents <1% of all

renal malignancies and can affect ages 13–83 years old with a mean age of 55; male to female ratio of 2:1.[6] UTUC accounts for 5–7% of all renal tumours and 5–10% of all urothelial tumours.[7] A study showed that UTUC incidence has been increasing over the last 30 years[8] however, there is no comparative long term data to echo the same for CDRC.

Prognostic factors identified by Chromecki et al. namely include tumour grade, architecture and location as the most readily available measure to guide counselling, selecting neoadjuvant systemic therapy and guiding extent of lymphadenectomy during radical nephroureterectomy.[9] A more recent study also identified lymph node involvement, age at diagnosis, lymphovascular invasion and gender as a supplement to guide outcome.[10] Long term prognosis of UTUC was showed to be 90%, 76.4% and 67.7% for 1, 3 and 5 year overall survival rates.[11]

Similarly with CDC age, tumour size/grade, lymph node involvement and chemotherapy received were the main prognostic factors associated with survival. 1, 3 and 5 year survival rates were reported to be 56.4%, 32.5% and 28.7% respectively.

Despite similar symptoms and prognostic factors CDC has worse outcomes chiefly owing to its aggressive nature and 42–43.6% of patients having metastasis at presentation.[12,13]

Management of concurrent disease is not well documented in the literature however both CDC and UTUC the mainstay of initial treatment is the most radical surgical technique possible providing both survival benefit in CDC and reduction in ureteral stump recurrence (nephroureterectomy plus lymphadenectomy).[14–16]

With respect to CDC Tang et al. has shown that there is significant survival benefit in surgery patients vs non-surgery patients and between radiotherapy patients and non-radiotherapy patients (cancer-specific survival of 24 months vs 4 months, $p < 0.001$; and 8 months vs 23 months, $p < 0.001$ respectively). More importantly thee combination of surgery plus chemotherapy had significantly higher survival rates than surgery or chemotherapy alone (14 months, 5 months and 9 months respectively).[17]

In the study by Choo et al. the efficacy of lymph node dissection (LND) combined with radical nephroureterectomy in upper tract urothelial carcinoma (UTUC) management was evaluated. The results indicated a correlation between the number of lymph nodes excised and improved cancer-specific survival (CSS), with a marginal decrease in hazard ratio (HR = 0.95, 95% CI: 0.91–0.99, $p = 0.07$) as the lymph node count increased. Notably, this survival advantage was pronounced in patients without lymph node metastasis (pN0), enhancing overall survival (OS), but was not significant in patients with lymph node metastasis (pN+).[18]

8. Conclusion

Current research on the concurrent management of CDC and UTUC remains scarce. Enhanced reporting of clinical cases and treatment strategies is essential to develop a comprehensive understanding of optimal management approaches for this rare co-occurrence. Preliminary evidence suggests that in selected patients, aggressive surgical intervention might offer a survival advantage. However, further data accumulation is necessary to substantiate this hypothesis and to refine treatment protocols for these patients.

CRediT authorship contribution statement

Adib Rahman: Writing – review & editing, Writing – original draft, Investigation. **Daniel Matheson:** Visualization, Investigation. **Joanna Perry-Keene:** Investigation. **Devang Desai:** Conceptualization.

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