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CASE REPORT

A renal mas(s)querader

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This is a case report of an unusual presentation of suspected gynaecological malignancy that turned out to be an upper tract urothelial carcinoma (UTUC). The patient was referred to our department after a staging laparotomy for suspected ovarian cancer revealed a massive right retroperitoneal mass. The final diagnosis was made after a relook laparotomy and radical nephrectomy.

Keywords: UTUC, renal mass, radical nephrectomy, radical nephroureterectomy, palliative chemotherapy

Case presentation

A 39-year-old previously well female from Cape Town had an atypical presentation of upper tract urothelial carcinoma (UTUC).

She initially presented to the department of obstetrics and gynaecology with a two-month history of abdominal and back pain, and no other symptoms. Notably, she had a five pack-year smoking history. She was Gravida 2 Para 2 and had two previous vaginal deliveries. There was no other relevant medical, surgical, occupational, travel or family history.

Initial ultrasonographic imaging and an elevated beta-human chorionic gonadotropin (B-hCG) of 1 744 IU/L suggested ovarian cancer and a staging laparotomy (there was no preoperative cross-sectional imaging done) revealed two normal ovaries and a uterus with a large retroperitoneal mass. A postoperative computed tomography (CT) scan demonstrated a large cystic-solid right renal mass with complete distortion of normal renal anatomy (Figure 1) and a normal left kidney, which prompted referral to our department. Clinically, she had a firm mass extending from the right upper quadrant to the pelvis, which crossed the midline. Her urine dipstix was normal and microscopy, culture and sensitivity revealed no leukocytes or erythrocytes with a mixed growth. Urine cytology was negative for high-grade urothelial carcinoma. She had a normal serum creatinine and HIV and hydatid serology were negative. She had a normal Papanicolau smear.

A relook midline laparotomy and right radical nephrectomy were performed (Figure 2). Ureterectomy and cuff of bladder were not done as UTUC was not the main differential diagnosis.

Macroscopic examination of the nephrectomy specimen (largest diameter of 26.5 cm) showed a mass lesion in the renal pelvis with extension into the adjacent renal parenchyma. The tumour communicated with a large unilocular cyst that distorted the renal anatomy. Histology showed a high-grade invasive urothelial carcinoma arising from surface dysplasia in the renal pelvis. The tumour comprised nests and cords of high-grade malignant epithelioid cells infiltrating into the adjacent renal parenchyma and present within the renal vein (pT3). The renal vein resection margin was involved by invasive carcinoma. Microscopy of the associated cyst wall confirmed the presence of a large subcapsular haematoma. Immunohistochemical staining confirmed the diagnosis of urothelial carcinoma (positive GATA3, negative PAX8, INI1 retained) and markers for mismatch repair proteins showed intact nuclear staining (negative screen for Lynch syndrome).[1,2] (Figure 3) This case demonstrated extensive infiltration into the adjacent renal parenchyma and renal vessels with haemorrhage into the mass resulting in a large subcapsular haematoma which confounded the clinical and radiological findings. Although there

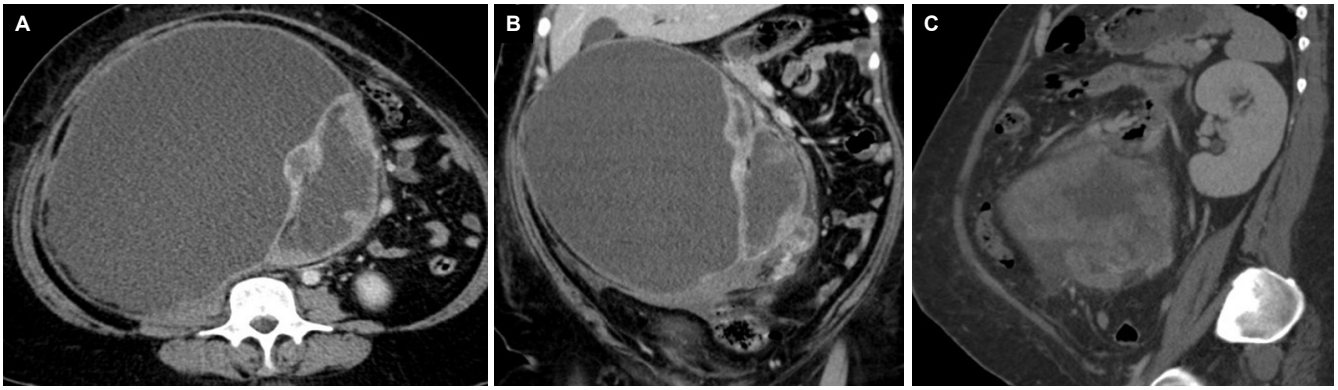

Figure 1: A) axial; B) coronal; C) sagittal contrast-enhanced abdominal CT images demonstrating a large, cystic-solid right renal mass with a solid enhancing component and complete distortion of normal renal anatomy

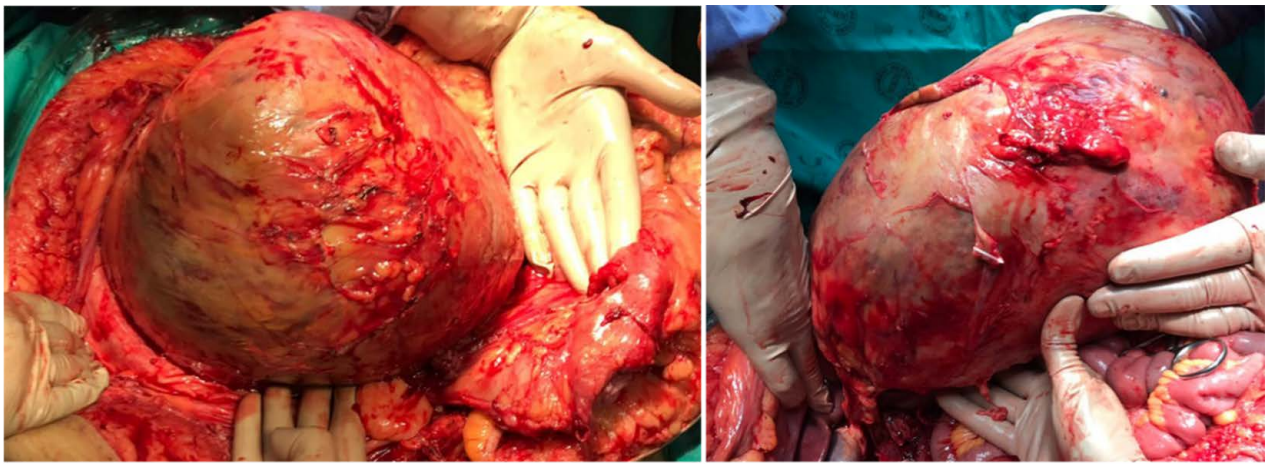

Figure 2: Dissection of renal mass during radical nephrectomy

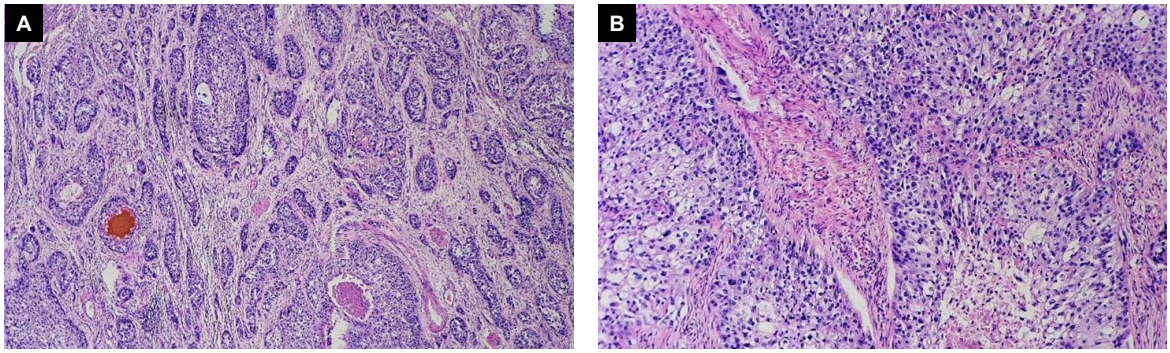

Figure 3: Haemotoxylin & eosin (H&E) stained histopathological photomicrographs; A) 40x magnification; B) 100x magnification. Nests of invasive carcinoma infiltrate into the adjacent renal parenchyma; the cells are pleomorphic with areas of necrosis and abundant mitoses; the cells stained positive for GATA3 and were negative for PAX 8, confirming their urothelial nature

was a degree of hydronephrosis, the large cystic component was mainly caused by the subcapsular haematoma.

Follow-up cystoscopy was normal, and a staging CT scan performed one month postoperatively revealed de-novo liver and lung metastases. The patient subsequently received palliative carboplatin and gemcitabine,[3] but she unfortunately demised five months later.

Discussion

UTUC accounts for 5–10% of all urothelial carcinomas. Patients may present with haematuria (micro- or macroscopic), flank pain or mass, constitutional symptoms or an incidental finding on radiological investigations.[4] Risk factors include smoking, exposure to aromatic amines, aristolochic acid and an association with Lynch syndrome, among other causes.[4-6]

CT urography is the imaging modality of choice, with a pooled sensitivity of 92% and pooled specificity of 95%.[4,8] The most useful CT signs to identify intra-renal urothelial carcinoma (compared to centrally-located renal cell carcinoma) are tumour centre in any part of the collecting system, a focal filling defect in the renal pelvis, preservation of renal shape, absence of cystic or necrotic change, homogenous enhancement of the tumour and extension of the tumour toward the pelvi-ureteric junction.[7] Imaging findings might be non-specific and there are numerous mimickers of UTUC. Benign lesions include a hypertrophied papilla, blood clots, inflammatory pseudotumours, suburothelial haemorrhage, papillary necrosis, cystic pyeloureteritis urogenital tuberculosis, mycetomas, malakoplakia, xanthogranulomatous pyelonephritis, retroperitoneal fibrosis and fibroepithelial polyps.[8-10] Non-benign lesions include renal cell carcinoma and renal lymphoma.[8] Other diagnositc investigations include urine cytology, retrograde pyelography and ureterorenoscopy.

Interestingly, B-hCG, a confounder in this case, has been shown to be elevated in urothelial cancer, especially in muscle-invasive disease.[11] There is increasing evidence that the expression of B-hCG in urothelial cancer points to dedifferentiation towards a gestational trophoblastic phenotype and these tumours histologically resemble choriocarcinoma.[11] B-hCG-secreting tumours have been associated with advanced disease, early haematogenous spread, decreased survival and decreased sensitivity to cisplatin-based chemotherapy.[11,12]

Management depends on the anatomical location and risk stratification. Low-risk tumours can be treated with renal-sparing surgery such as endoscopic ablation and segmental ureteral resection, while high-risk tumours require radical nephroureterectomy with cuff of bladder excision and possible adjuvant chemotherapy.[4] Our patient had metastases on follow-up imaging and was started on palliative chemotherapy. The POUT trial demonstrated that gemcitabine-platinum combination chemotherapy initiated within 90 days after nephroureterectomy

significantly improved disease-free survival in patients with locally advanced UTUC.[3]

This case highlights the importance of including UTUC as part of the differential diagnosis of a renal mass. Furthermore, correct workup and staging of a renal tumour, especially in the setting of such an atypical presentation, can expedite the correct surgical management and subsequent follow-up of such cases.

Conflict of interest

The authors declare no conflict of interest.

Funding source

The authors declare that they have no relevant financial interests.

Ethical approval

Ethics committee approval was obtained from the Stellenbosch University Human Research Ethics Committee (C20/04/008).

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