Renal cell carcinoma in a crossed ectopic kidney

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Introduction

Congenital malformations of the urinary system are present in 10% of all births (1). The ectopic kidney in the pre-sacral position can occur in 1 of 800 subjects (2). A renal cell carcinoma arising in a renal ectopia is extremely rare. Its atypical symptomatology causes a diagnostic as well as therapeutic dilemma.

Case report

A 63-year old male presented with anintra abdominal umbilical region lump for 10 years and haematuria for two months which was ignored because there was no significant increase in size of the lump with absence of any other symptom. In the past he had been diagnosed to have an uncomplicated crossed ectopic left kidney causing a palpable abdominal mass.

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Figure 1. Left crossed ectopic pelvic kidney in the lower lumbar and pre-sacral region with a neoplastic lesion.

The ultrasound scan of the abdomen showed a large solid mass of 8.5x7centimeter in the upper pole of the

pre-sacral, crossed ectopic kidney. The CT urography revealed a radiologically neoplastic mass in the left-to-right renal ectopia which was found in the lower lumbar and pre-sacral region (Figure 1). Well excreting right kidney was found in the normal position not attached to the ectopic left kidney.

A hypoechoic solitary lesion in the right lobe of the liver was confirmed to be a benign liver cyst.

During exploratory laparotomy the tumour was found in the pre-sacral and lower lumbar region, in the retroperitoneum, deep to the lower half of the root of the mesentery (Figure 2). Both the sigmoid colon and the caecum were mobilized to approach the retroperitoneal plane anterior to the tumour. It was well separate from the normal right kidney.

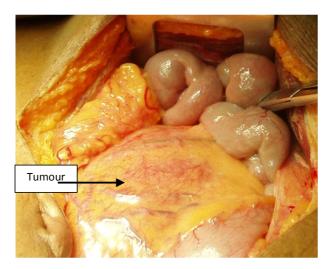


Figure 2. The tumour forming a bulge at the lower part of the root of the mesentry.

The left renal vein was identified draining in to the inferior vena cava just above its confluence. The left ureter was dissected free from its course parallel to

the left iliac vessels. The arterial supply was mainly from the left common iliac. There were smaller arteries supplying from the right common iliac and directly from the aorta which were divided and ligated. After dividing the vein and the ureter the left kidney with the tumour was delivered. A thorough inspection was carried out to find para-caval, para-aortic and iliac lymph nodes which were not present.



Figure 3. The specimen of the neoplastic kidney with the ureter following radical nephrectomy.

The right kidney was normal in position and appearance. The patient had an uncomplicated recovery.

Macroscopically the tumour had the typical variegated appearance confined to peri-renal fat and the Gerota's fascia. The histology showed an organ confined clear cell renal cell carcinoma staged pT2N0M0.

Discussion

Incidence of ectopic kidney reported in literature is 1:500 to 1:110. Incidence of one normal and one pelvic kidney is 1:800 to 1:3000 (3,4). There is no increased risk of malignancywithin an ectopic kidney (5). But the diagnostic difficulty and unusual presentation can lead to diagnostic dilemmas. The incidence of neoplasms in the horseshoe kidneys is estimated to be 1% to 12% and more than 50% of them are renal cell carcinomas (6).

In a study with 61 patients with ectopic kidneys the commonest presentations were palpable abdominal lump (31%) and abdominal pain (27%). Only 18%

had macroscopic haematuria. Renal stones and pelviureteric junction obstruction were the commonest pathologies encountered (7). With the onset of macroscopic haematuria diagnostic workup should be on standard guidelines. The USS and the CT scan would confirm the mass to be a renal tumour as in this case without a biopsy.

Various arterial supplies from common iliac, internal iliac, common inferior mesenteric trunk and directly from the aorta have been described in the literature. And in the present case it was mainly from the left common iliac artery. Venous drainage is usually to the common iliac, IVC and it was a single renal vein directly to the IVC on the ventral aspect in this patient. Pedicle dissection was tedious and we strongly felt that a CT angiogram would have been helpful during dissection to anticipate the difficulties and also to give more details about the extent of fusion of the two kidneys.

The ureter was found parallel to the left common iliac artery and emptied into the bladder at the normal position on the left as documented in the other cases.

The present case emphasize the need of adherence to common haematuria guidelines in all congenitally abnormal kidneys even with a long term diagnosis and also highlights the need of a pre operative CT angiography to study the abnormal vasculature.

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