A malignant ‘incidentaloma’ in a patient with autosomal dominant polycystic kidney disease

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Case report

A 72-year-old woman with known, stable, renal insufficiency was admitted because of fever, malaise, and dysuria, which began 48 h prior to admission. She had known autosomal dominant polycystic kidney disease (ADPKD) and several family members had developed end-stage renal disease. She had previously had urinary tract infections which had responded to antibiotics. Her creatinine had exceeded 200 µmol/l a year prior to admission and was gradually increasing. On physical examination she was moderately febrile (38°C), but not in acute distress. She had mild back and abdominal tenderness. The kidneys and the liver were easily palpable and exhibited gross, nodular structures consistent with ADPKD. The urinalysis showed many leukocytes and bacteria. Her serum creatinine concentration was 351 µmol/l. A urine culture yielded Klebsiella sp. and ciprofloxacin was administered with good response.

An ultrasound examination revealed cystic changes in both kidneys and the liver, as well as a denser structure in the upper pole of the left kidney, prompting the computerized tomographic study shown in Figure 1. A large, dense structure with a single cavity can be seen in the upper pole of the left kidney. A T1-weighted, magnetic resonance imaging examination followed, as shown in Figure 1. This examination revealed similar changes. A selective renal arteriogram is shown in Figure 2, which shows several abnormal polar vessels supplying the upper pole mass. Involvement of the renal vein by tumour could not be shown. An ultrasound-guided biopsy specimen revealed typical changes of a clear-cell renal-cell carcinoma (RCC). The right kidney was carefully visualized with the imaging studies, but revealed no suspicious lesions. Additional work-up failed to show any evidence of distant metastasis and the patient recovered from her urinary tract infection uneventfully, although her creatinine concentration remained at 348 µmol/l.

Discussion

Modern imaging techniques not uncommonly confront physicians with disturbing findings in asymptomatic patients. The term ‘incidentaloma’ is generally applied to accidentally located adrenal masses; however, the term appears appropriate here. Our patient with ADPKD had developed RCC, which we carefully and zealously documented. Her actual reason for coming to the hospital had resolved and she wanted to go home. We were confronted with serious medical decisions in this case of an elderly woman with ADPKD-induced chronic renal failure, who was otherwise doing relatively well.

The association of RCC and ADPKD is controversial; an increased incidence has not been clearly shown. Zeier et al. [1] could find no evidence for an increase and indicated that patients with von Hippel–Lindau syndrome may have skewed the evidence. The absence of a higher risk was confirmed by Ritz and colleagues in an analysis based on the EDTA data [2]. Keith...
et al. [3] described three cases from the Mayo clinic and reviewed the literature since 1955. They examined 22 case reports, which included sufficient clinical information for analysis in addition to their own cases. They reported that RCC was more often concurrently bilateral, sarcomatoid in type, and occurred at a younger age than in the general population. Their own three patients had unilateral tumours and two had clear-cell RCC, similar to our patient.

Since the report of Keith et al. [3], Soderdahl et al. [4] described a 63-year-old man with end-stage renal disease from ADPKD, who had bilateral suspicious-looking renal masses. He proved to have bilateral RCC of papillary and clear cell varieties. The patient survived a bilateral nephrectomy uneventfully and appears to support the authors’ call for an aggressive work-up including multiple imaging modalities, renal biopsy, surgical exploration of the contralateral side, and even bilateral nephrectomy.

The appropriate management for our patient was not entirely clear. Whether or not RCC in the face of ADPKD is more or less aggressive than is generally the case for this unpredictable tumour, is not known. Surgical excision of the tumour usually includes nephrectomy and could have resulted in loss of renal function sufficient to make dialysis mandatory in our patient. The conservative senior clinician in this case advised against operation and argued that the survival as a dialysis patient might be less than expected without treatment. The life expectancy of elderly chronic dialysis patients has recently been reviewed and compared to the survival of patients with colon cancer with sobering results [5]. The options were discussed with the patient and an excision of the tumour with preservation of remaining renal tissue was advised. This procedure was successfully performed and the patient left the hospital, rid of her urinary tract infection and free of her RCC, with a serum creatinine concentration unchanged from the preoperative values.

**Teaching point**

ADPKD makes renal masses more difficult to evaluate technically; is not permissive in the development of renal cell carcinoma; but then again, does not protect from cancer either. As always, vigilance is warranted.

**References**