Clinics in Diagnostic Imaging (46)

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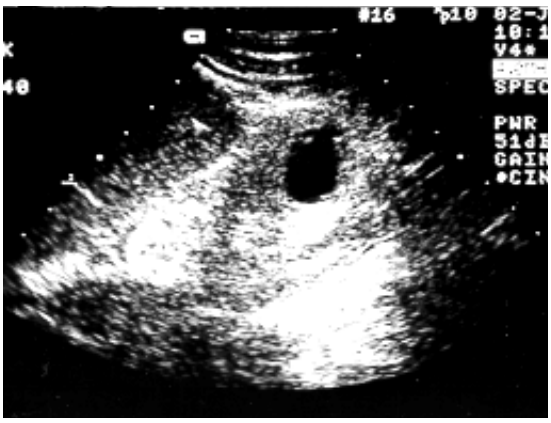

Fig 1a

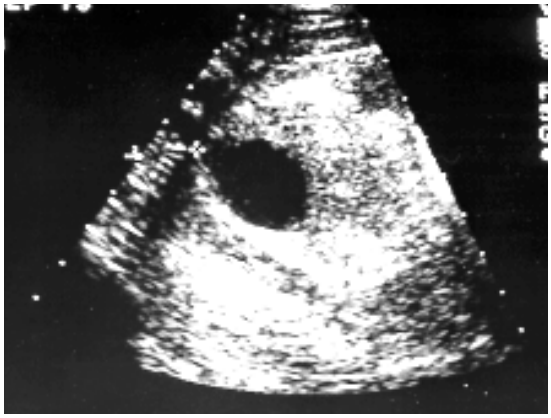

Fig 1b

Fig 1 - (a) Longitudinal and (b) transverse US scans of the right kidney.

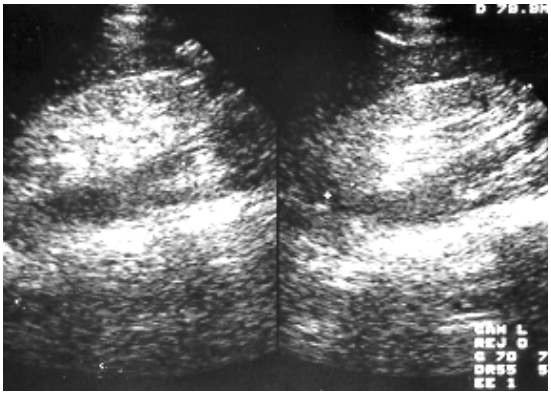

Fig 2 - Longitudinal US scans of the right kidney taken 6 years previously. The left kidney has a similar appearance.

CASE PRESENTATION

A 40-year-old Chinese man underwent surveillance ultrasonography (US) of the kidneys (Figs 1a & 1b). He was suffering from chronic renal failure and was on regular haemodialysis for the past six years. The patient was otherwise asymptomatic and his dialysis program was unremarkable. Previous US scans (Fig 2) performed at the time of the first diagnosis of renal failure were available for comparison. What do the current US scans show? How do these differ from the previous study? What other investigation will be useful?

IMAGE INTERPRETATION
US scans showed a largely cystic lesion at the lower pole of the right kidney (Figs 1a & 1b). However, there was a suspicious solid component at the rim of the lesion. Previous US scan performed 6 years ago revealed bilateral small and atrophic kidneys with loss of corticomedullary differentiation, compatible with renal failure (Fig 2). No cystic changes were noted at that time. Unenhanced and enhanced computed tomography (CT) scans of the kidneys were performed. Unenhanced CT scan showed a low-density lesion at the lower pole of the right kidney, with a small spike of calcification at its margin (Fig 3a). After contrast injection, the lesion was found to be predominantly cystic, but an enhancing solid component was noted at the rim of the cystic wall (Fig 3b). A slightly more cranial CT section showed the multiseptated portion of the lesion which comprised cystic and solid components (Fig 4). These findings were highly suspicious of a cystic renal cell carcinoma. Other CT sections confirmed the presence of bilateral atrophic kidneys with multiple cystic changes, compatible with acquired cystic disease of the kidney (Fig 5).

DIAGNOSIS
Renal cell carcinoma in acquired cystic kidney disease.

CLINICAL COURSE
The patient underwent radical nephrectomy and he made an uneventful recovery. Histopathological examination confirmed a T2 G2 renal cell carcinoma in a background of acquired cystic disease of the kidney.

DISCUSSION
In 1977, Dunnill et al[1] first described the development of acquired cystic kidney disease (ACKD) and renal cell carcinoma in patients undergoing haemodialysis. ACKD is defined as the presence of more than 3 cysts or when more than 25% of the renal parenchyma is involved by cystic change[2]. Chronic renal failure (CRF) is the key to the development of ACKD that begins even before the initiation of dialysis[3]. Its pathogenesis is still unclear. However, there has been some speculation of possible stimulation by a renal cell growth factor[4] and renotropic factors[5] in the presence of insufficient renal function causing hyperplastic changes and formation of RCC.

The incidence of acquired cystic disease ranges from 10% to 90%, depending on the period of dialysis[2]. It has also been found that patients on haemodialysis and continuous ambulatory peritoneal dialysis (CAPD) exhibit a similar prevalence of ACKD[3]. There is some suggestion that successful renal transplantation (and hence cessation of dialysis) may lead to regression of ACKD[5], but this remains controversial[6]. On the other hand, the rate of malignant transformation of ACKD to RCC varies throughout the literature, ranging from 1% to 8%[1,2,6-9]. Several reports have suggested that the prevalence of RCC is greater in haemodialysis patients[3,7] and in the native kidneys of renal transplant recipients[8,9], compared to the general population. Doublet et al[10] recently estimated the risk of RCC to be as high as 100 times greater in asymptomatic renal transplant recipients versus those in the general population. Radiological surveillance for CRF patients have therefore been advocated by some authors[7,10,11].

Terasawa et al[11] reported a 100% diagnostic rate by US. They proposed that US was the most useful method to diagnose RCC in haemodialysis patients, with CT and angiography being only adjuncts. Cogny-Van Weydevelt et al[12] also suggested that US was accurate for the detection of RCC in renal transplant recipients. Jamis-Dow et al[13] studied the detection of small renal masses by CT versus US in 21 patients with von Hippel-Lindau syndrome, a condition also characterised by multiple cyst formation and possible neoplastic changes. Two hundred and five renal masses were removed surgically and pathological correlation with the pre-operative imaging was made. Among lesions 10 - 35 mm in size, 90% and 82% were correctly characterised with CT and US respectively. When CT and US were combined, 95% of the lesions were correctly categorised.

In our opinion, US is useful as the initial screening modality in this group of patients and when suspicious, CT is the next imaging investigation of choice. CT also provides more information about tumour invasion and lymph node status that US may be unable to provide. The anatomical details of locally- advanced RCC and changes of renal failure are well-depicted on CT scans (Fig 6). Our local experience suggested that most of these lesions were of low grade and low stage at the time of diagnosis[14].

While there are numerous reports of increased incidental detection of renal tumours due to the availability of US and CT[15-17], it remains debatable as to whether it is cost effective to screen all chronic renal failure and post-renal transplant patients radiologically. However, such screening and routine cytology examinations are prudent in patients at high risk for development of urinary malignancies, including patients with analgesic nephropathy as their cause of CRF[5]. For the subgroup of patients who are undergoing surveillance programs for their hepatitis or post-transplant graft status, it is natural to extend the assessment to the native kidneys. In patients presenting with haematuria, imaging studies by means of US or CT are definitely indicated, and comparision with previous imaging studies often helps to identify new or suspicious lesions.

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ABSTRACT

Acquired cystic disease of the kidney and renal cell carcinoma are associated with chronic renal failure. In recent years, there has been increased incidental detection of renal tumours through the liberal use of ultrasonography (US) and computed tomography (CT). A 40-year-old man suffering from chronic renal failure and who was being treated with haemodialysis for six years, was found to have a complex cystic lesion on US and CT. Nephrectomy was performed and the lesion was confirmed to be renal cell carcinoma associated with acquired cystic kidney disease. The current role of imaging in the management of affected patients are discussed.

Keywords: renal cell carcinoma, acquired cystic kidney disease (ACKD), chronic renal failure, dialysis, computed tomography (CT), ultrasonography (US)

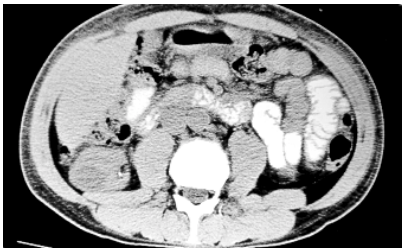

Fig 3a

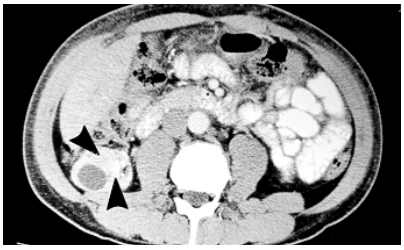

Fig 3b

Fig 3 - (a) Unenhanced CT scan shows a hypodense lesion at the lower pole of the right kidney, and an adjacent small focus of calcification. (b) Enhanced CT scan shows a solid component (arrow heads) at the wall of the cyst.

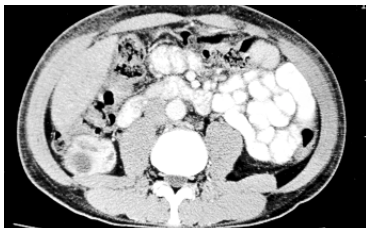

Fig 4 - Enhanced CT scan of the lower right kidney shows a multiseptated lesion with cystic components.

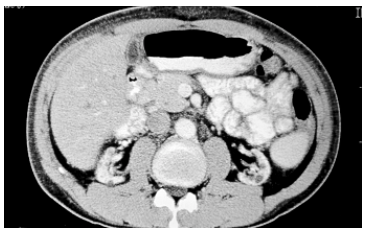

Fig 5 - Enhanced CT scans show small atrophic kidneys bilaterally, with multiple small cysts.

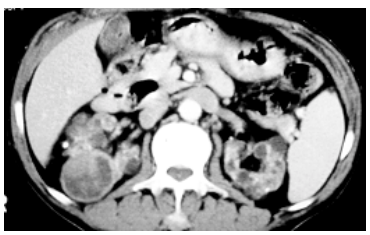

Fig 6 - Enhanced CT scan of a 59-year-old man shows a large solid tumour arising from a background of acquired cystic disease of the right kidney. This patient underwent bilateral nephrectomies and interestingly, was also found to have microscopic foci of RCC in the contralateral kidney.