RENAL ANGIIOMYOLIPOMA – A CASE REPORT

F W K Yip, S H Lee

ABSTRACT
Renal angiomyolipoma is a benign tumour of the kidney which is often mistaken for a renal cell carcinoma resulting in an unnecessary nephrectomy. It can be diagnosed preoperatively and managed conservatively without surgery. Large symptomatic renal angiomyolipomas can often be treated by renal-sparing surgery. We report a case of a large renal angiomyolipoma diagnosed preoperatively and excised completely with preservation of the kidney.

Keywords: Renal angiomyolipoma, benign tumour, characteristic radiological features, conservative management.

INTRODUCTION
Renal angiomyolipoma is an uncommon benign mesenchymal tumour of the kidney. It contains fat, smooth muscle and tortuous blood vessels in varying proportions. First described by Bourneville in 1880,[1] it is often considered to be a hamartoma rather than a malignant neoplasm. Two forms of this tumour are recognized clinically: the first is associated with tuberous sclerosis, where the tumour is usually small, multiple, bilateral and has an equal sex distribution. The second form occurs more often in women and is not associated with tuberous sclerosis, the tumour is often large, single and unilateral.

CASE REPORT
A 40-year-old woman presented with a large abdominal mass of 3 years duration. Three years prior to admission she had noticed progressive abdominal swelling over the right flank. She was otherwise well with no symptoms in the gastrointestinal or urinary tract.

Physical examination revealed a healthy woman. There was no pallor, the blood pressure was normal. A large abdominal mass was palpable in the right flank extending across the midline and extending inferiorly into the right iliac fossa. It was non-tender and easily ballotable. Neurological examination revealed no evidence of tuberous sclerosis.

The serum biochemistry was normal. Urine microscopy revealed no haematuria. Ultrasonography revealed a large hypoechoic mass originating from the lower pole of the right kidney. An intravenous urogram (Fig 1) revealed a soft tissue mass arising from the mid and lower pole of the right kidney causing distortion to the lower and midpole calyces and displacing the ureter to the left. A CT-scan demonstrated a large hypodense lesion (HU-39U) with septation arising from the lower pole of the right kidney (Fig 2). A pre-operative diagnosis of a large solitary right renal angiomyolipoma was made.

At operation, an irregularly shaped tumour consisting mainly of fat was found adherent to the inferior pole of the right kidney. The right ureter was stretched over the upper surface of this tumour. The mass measuring 20x17x7 cm and weighing 1.1 kg was excised together with a small rim of normal renal tissue (Fig 3).

Histopathology confirmed the diagnosis of a renal angiomyolipoma with the characteristic picture of a tumour composed of mature fat cells, irregular, thick-walled blood vessels and sheets of smooth muscle cells.

Post-operative recovery was uneventful and the patient was discharged on the eighth post-operative day. On follow-up five months later she was well. A repeat intravenous urogram showed no evidence of obstruction.

DISCUSSION
Before the advent of ultrasonography and the CT scanner, renal angiomyolipomas were often misdiagnosed as renal cell

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Fig 1 – Intravenous urogram showing mass in the right kidney with calyceal distortion and displacement of the ureter.
carcinoma, with the result of nephrectomy being the only treatment. The histological diagnosis often come as a surprise after surgery.

In recent years, the characteristic radiological findings of hypoechoic areas on the renal ultrasonography together with the characteristic pattern of fat density on the CT scan has made the diagnosis of renal angiomyolipoma almost certain in virtually all cases. As a result of this, small asymptomatic lesions can be accurately diagnosed and managed conservatively with frequent follow-up. In larger lesions, symptoms of pain and mass tend to dictate some form of invasive intervention. Oesterling et al. found that 82% of tumours greater than 4cm in diameter tend to be symptomatic. The common symptoms include acute flank and abdominal pain, a palpable mass and sometimes spontaneous haemorrhage. Further, large tumours are at a high risk of spontaneous rupture or rupture after minor trauma.

Although the advent of selective arterial embolization has rendered many of these tumours amenable to less aggressive therapy, the sheer size of this tumour in this patient which was causing partial obstruction to the ureteric drainage necessitated operative excision.

At surgery, total excision of the tumour was possible because of the very narrow pedicle attachment to the kidney. A small rim of normal renal tissue was excised together with the tumour without the need for a nephrectomy.

When the tumour attachment to the kidney is more extensive, or where there are multiple tumours located in one pole of the kidney, a partial nephrectomy with pedicle control is usually an easy procedure.

It is important to recognize the benign nature of these tumours and their characteristic radiological features. Most of these tumours are amenable to conservative management with frequent follow-up. Even when surgical excision is indicated, the kidney itself can often be spared without jeopardizing the patient. Only when the tumour has replaced the whole kidney or when there is a concomitant renal carcinoma present is a nephrectomy necessary. Such cases are extremely rare.

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REFERENCES