RADIOLOGICAL CASE

CLINICS IN DIAGNOSTIC IMAGING (4)

W C G Peh, J H C Li

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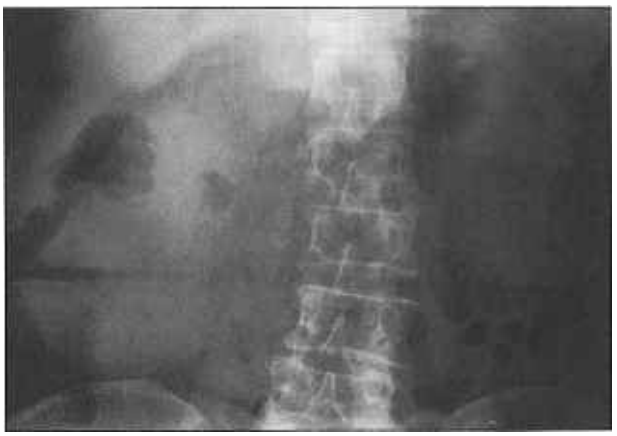
CASE REPORT

A 69-year-old Chinese woman was admitted for a one-day episode of colicky pain over the right upper quadrant of her abdomen. The pain was of moderate-to-severe intensity, with radiation to the back. There was no associated chills, rigors or tea-coloured urine, nor was there a history of recent trauma. On further questioning, she had occasional bouts of mild right-sided abdominal pain over the past 20 years. She suffered from diabetes mellitus and was currently taking oral hypoglycaemics.

On physical examination, the patient was afebrile and haemodynamically stable, with no jaundice or pallor. Abdominal examination revealed only mild tenderness over the right upper quadrant. Her blood biochemistry, including urea, electrolytes, liver and renal function tests, was unremarkable. Haemoglobin level was 11.7 g/dL.

Plain radiographs and ultrasound of the abdomen were performed for suspected biliary colic. However, no gallstone, focal liver lesion or dilated intrahepatic duct was detected sonographically. What do the plain abdominal radiograph (Fig 1) and ultrasound of the right kidney (Fig 2) show? What does the intravenous urogram (IVU) (Fig 3) demonstrate? What further investigation would be useful in confirming the diagnosis?

Fig 1 – Supine anteroposterior radiograph of the upper abdomen.



Department of Diagnostic Radiology
The University of Hong Kong
Queen Mary Hospital
Hong Kong

W C G Peh, FRCR, FHKAM, FAMS
Senior Lecturer and Consultant

J H C Li, MBBS, DMRD, FRCR
Senior Medical Officer

Correspondence to: Dr W C G Peh

Fig 2 – Ultrasound of the right kidney. (a) Longitudinal scan of the upper kidney. (b) Transverse scan of the lower renal pole.

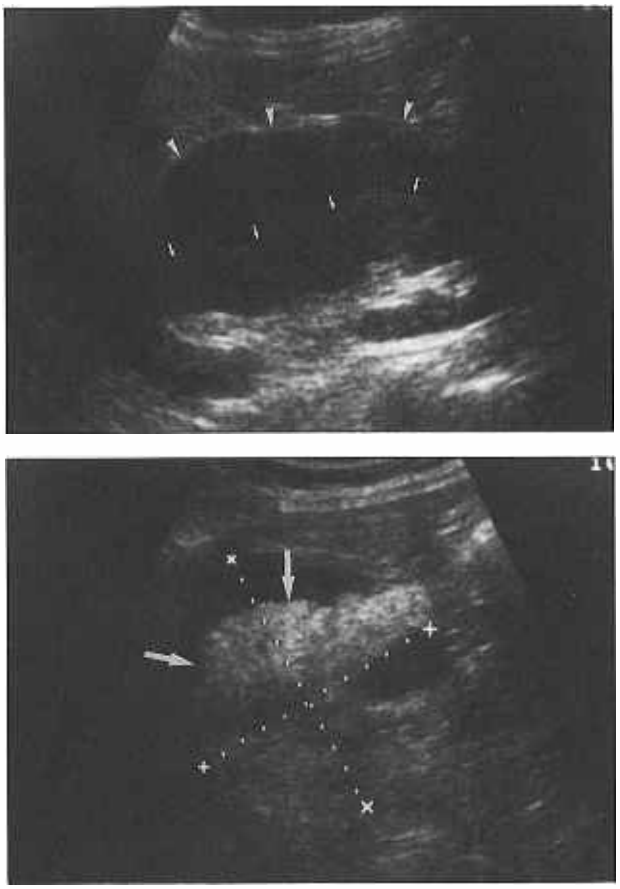


Fig 3 – Urogram of the kidney (15-minute IVU film).

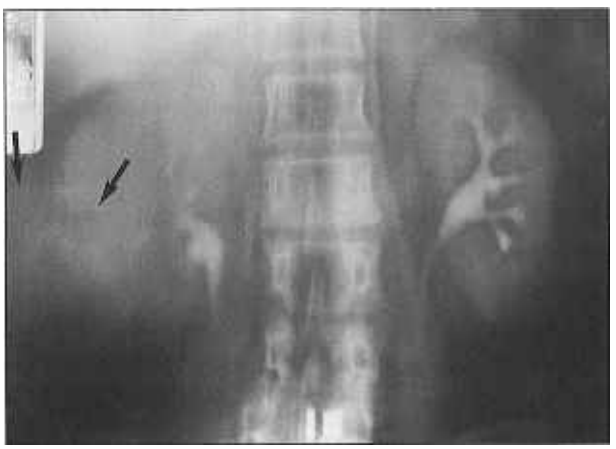


IMAGE INTERPRETATION

On the abdominal radiograph, there was a large ovoid mass in the region of the right kidney (Fig 1). Ultrasound demonstrated a hypoechoic area (arrowheads) adjacent to and slightly indenting the lateral cortex of the right kidney (small arrows), consistent with a perinephric fluid collection (Fig 2a). There was a large hyperechoic mass (arrowed) arising from the lower renal pole (Fig 2b). IVU showed a large rounded mass at the lateral aspect of the lower right kidney (arrowed). It was of mixed density, containing lucent areas (Fig 3). The right pelvicalyceal system was under-filled, consistent with extrinsic compression by a surrounding mass. The left kidney had a normal appearance. The combination of radiographic lucency together with sonographic hyperechogenicity suggested a fatty component within the renal mass. Subsequent computerised tomography (CT) of the kidneys showed a well-defined, largely fatty, lower pole mass; as well as a lateral perinephric fluid collection. The mass contained markedly enhanced linear areas, representative of blood vessels (Fig 4).

DIAGNOSIS

Renal angiomyolipoma with spontaneous perinephric haemorrhage

Clinical Course

The patient's painful symptoms settled with analgesics and she was discharged three days after initial presentation. At last follow-up nine months later, she remained asymptomatic, with no further episodes of abdominal pain. Repeat ultrasound showed no change in appearance of the angiomyolipoma (Fig 5). The perinephric collection had resolved completely. The plan of management was to continue monitoring the patient clinically and radiologically.

DISCUSSION

Spontaneous perinephric or retroperitoneal haemorrhage from renal tumour rupture is a rare but recognised surgical emergency, the commonest cause of which is renal carcinoma[1]. The aetiology in patients of Chinese origin, however, tends to be renal angiomyolipoma rather than carcinoma[2]. Angiomyolipoma is an uncommon benign tumour of the kidney with an incidence in the general population of about 0.3%[3]. Angiomyolipomas are generally regarded as hamartomas, being composed of mature fat cells, smooth muscle and thick-walled blood vessels. The degree of tumour vascularity, together with structural rigidity, inelasticity and tortuosity of the blood vessels in these tumours make them prone to haemorrhage[4].

Fig 4 – CT kidneys. Non-enhanced scans at the levels of the (a) mid and (b) lower poles show a fatty mass (arrows) arising from the lateral aspect of the inferior right kidney. After contrast injection, scans at similar levels, (c) and (d), demonstrate the lateral perinephric fluid collection (arrowheads), as well as enhancement of vessels within the fatty mass (arrows).

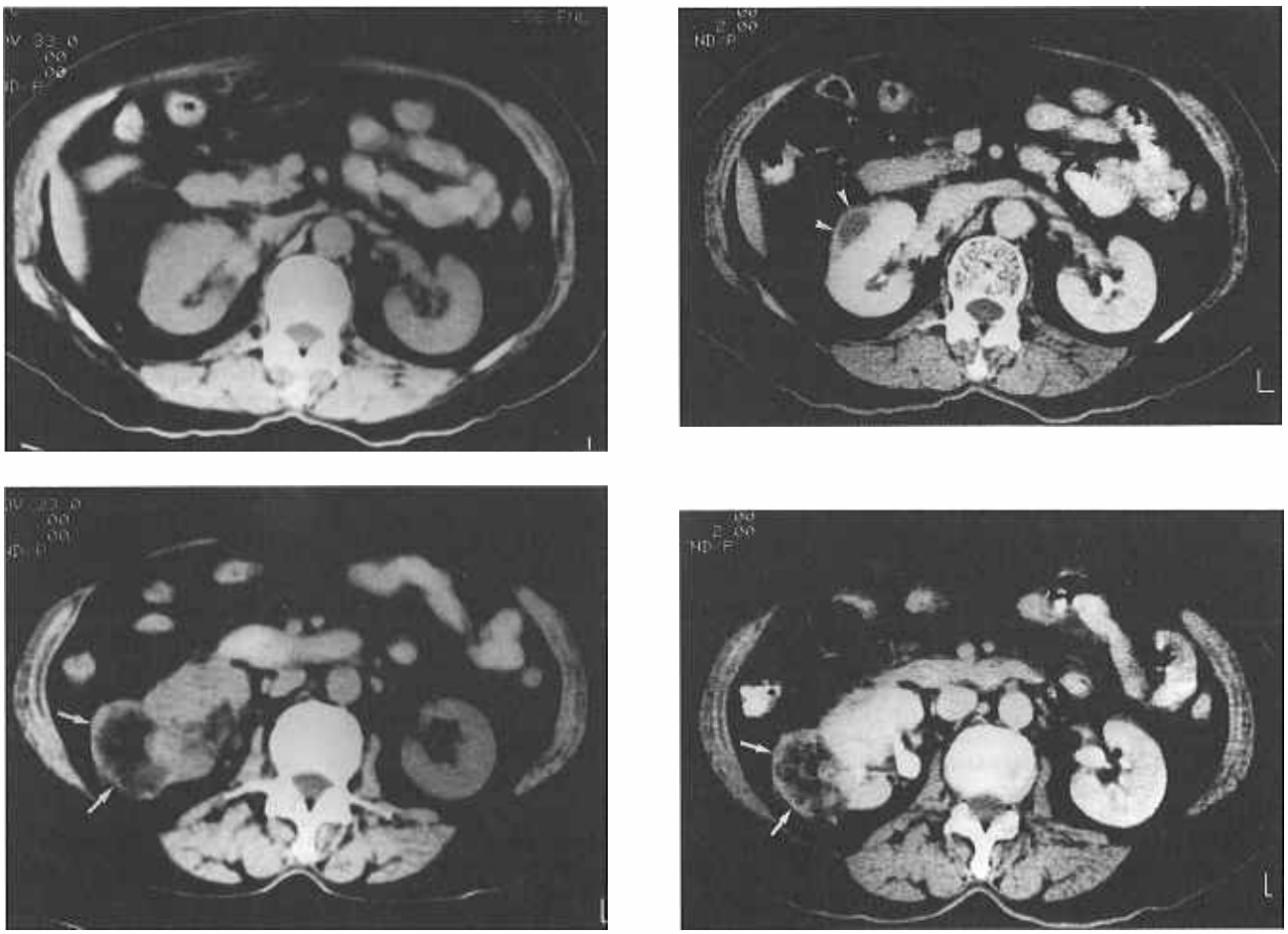
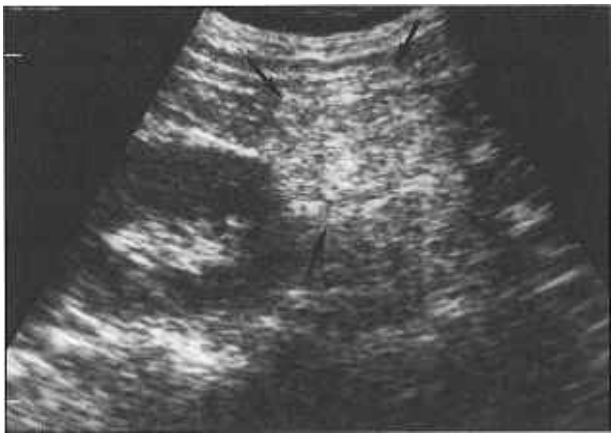


Fig 5 – Repeat ultrasound shows the angiomyolipoma as a rounded well-defined echogenic mass (arrows) arising from the lower renal pole.



Renal angiomyolipomas have been classified into two types based on clinical features. The more common type is associated with tuberous sclerosis and is typically bilateral, small, multiple and asymptomatic. It is estimated that 40% to 80% of patients with complete expression of tuberous sclerosis have renal angiomyolipomas, while about 50% of all renal angiomyolipomas occur in patients with tuberous sclerosis[5]. The other type not associated with tuberous sclerosis tends to be unilateral, large and symptomatic. This isolated type is usually found in middle-aged women, and classically present with a triad of loin pain(87%), haematuria(40%) and a palpable mass(47%). In a review of the literature, about 25% of these patients presented with severe abdominal pain and shock, necessitating emergency laparotomy[6].

It is obviously important to be able to distinguish a bleeding angiomyolipoma from ruptured renal cell carcinoma. Plain radiograph, intravenous urogram and angiography are generally unhelpful in this differentiation. The angiographic features of hypervascularity, tortuous vessels and microaneurysms seen in angiomyolipomas are non-specific as malignant tumours can produce similiar appearances. These older methods have been superceded by modern techniques, such as ultrasound and CT, which can generally provide an accurate diagnosis in a non-invasive manner.

Angiomyolipoma produces a characteristic highly echogenic focal pattern sonographically. Even though this appearance is not specific, its presence should suggest the possibility of angiomyolipoma in most cases. Detection of fat using CT confirms the diagnosis and is said to be the only radiological finding which can differentiate angiomyolipoma from renal cell carcinoma. The use of thin (5 mm) sections improves the accuracy of CT[7]. Where the diagnosis remains uncertain despite ultrasound and CT, magnetic resonance imaging (MRI) has been suggested[8]. The diagnosis is again dependent on detecting the fatty component which is seen as areas of high signal intensity in T1-weighted, and low signal intensity in T2-weighted images. The presence of haemorrhage, however, complicates the imaging appearance of this tumour. This is especially so if the tumour contains only small amounts of fat, compounding the difficulty of pre-operative diagnosis. Fine needle aspiration has been advocated in these doubtful situations[9].

The ability to make a confident diagnosis and to monitor the progress of angiomyolipomas radiologically in most cases has led to the current trend of managing these tumours conservatively. This has been attributed to the increasing detection, by ultrasound and CT, of small asymptomatic angiomyolipomas in patients without tuberous sclerosis, thus altering the traditional concept of the two clinical types mentioned earlier. The scheme of management depends on the tumour size and patient symptomatology. In general, symptomatic tumours larger than 4 cm in size should be studied angiographically and considered for selective arterial embolisation. Smaller and/or asymptomatic lesions should be monitored at regular intervals using ultrasound or CT, particularly for surveillance of haemorrhage and synchronous malignancy. If surgery is required, a renal-preserving procedure should be performed wherever possible. Radical nephrectomy, however, is necessary if malignancy is suspected[10,11].

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ABSTRACT

A 69-year-old Chinese woman presented with acute right-sided abdominal pain. Intravenous urogram and ultrasound demonstrated a fatty lower renal pole mass with a perinephric collection. The diagnosis of renal angiomyolipoma was confirmed by computerised tomography. The perinephric haemorrhage resolved with conservative treatment. The classification, clinical features and management of this entity is discussed. The role of imaging in diagnosis and follow-up of renal angiomyolipoma is emphasised.

Keywords: *angiomyolipoma, computerised tomography(CT), hamartoma, kidney neoplasms, spontaneous haemorrhage*