

RADIOLOGICAL CASE

CLINICS IN DIAGNOSTIC IMAGING (18)

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CASE REPORT

A one-year three-month old boy was incidentally noted to have a left loin mass. Antenatal history was unremarkable. He had no dysmorphic features. His blood pressure and pulse rate measured 107/71 mmHg and 80/min respectively. On examination, the left loin mass was ovoid-shaped, ballotable, smooth in outline and measured about 9 cm in length. Full blood count, renal and liver function tests, and urine microscopy were normal. The urine catecholamine metabolites, VMA and HVA, were within normal limits. What do the plain abdominal radiograph (Fig 1), ultrasound of the left kidney (Fig 2), and computed tomography (CT) scans (Fig 3) show? What is the diagnosis?

Fig 1 – Frontal radiograph of the abdomen, taken at admission.

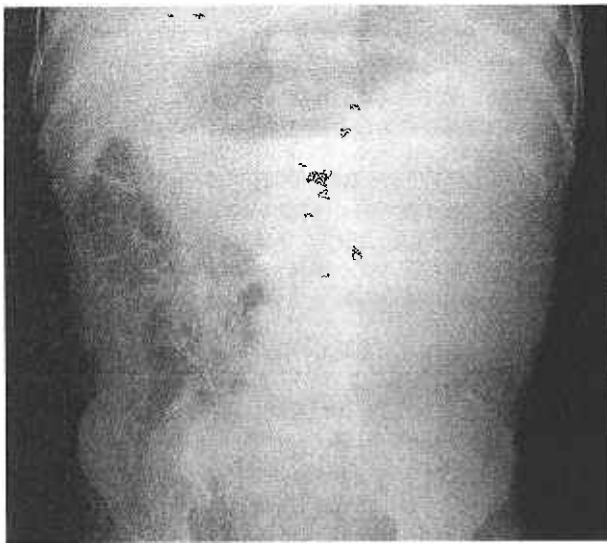
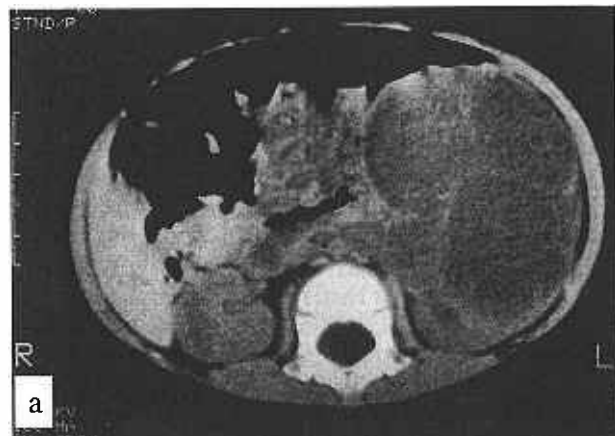


Fig 2 – Ultrasound of the loin mass (lower pole of the left kidney [LK] is labelled).



Fig 3 – (a) Unenhanced and (b) Enhanced CT scans at the level of the mid-right kidney.



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IMAGING INTERPRETATION

Plain radiograph (Fig 1) showed a large soft tissue shadow in the left side of the abdomen, with displacement of the gas-filled bowel loops to the right. The left psoas shadow was obscured. No soft tissue calcification or bony abnormality was demonstrated. Ultrasound (Fig 2) showed a cystic mass, with multiple thick and highly-echogenic septations, arising from the upper pole of the left kidney. No solid component nor internal calcification was seen within this multicystic mass.

The CT scan (Fig 3) demonstrated a well-defined septated mass originating from the anterior aspect of the upper left kidney. The septae were of variable thicknesses and enhanced with intravenous contrast. The cystic spaces were generally rounded in shape, differed in size and had CT numbers of near-water density. No abnormal calcification was demonstrated.

DIAGNOSIS

Multilocular cystic nephroma

CLINICAL COURSE

During the operation, a large tumour arising from the upper pole of the left kidney was found. There was no surrounding infiltration nor enlarged lymph nodes. Incidentally, the ureter was noted to be bifid in its proximal 6 cm. Left nephrectomy was performed. The tumour was spherical, well-circumscribed, and measured 7.5 cm in diameter. Bisection of the specimen demonstrated multiple cystic spaces, lined by smooth septae, containing clear light yellow fluid. Multiple cysts were found herniated from the tumour into the renal pelvis. The renal pelvic mucosa appeared normal (Fig 4).

Microscopic examination showed that the renal tumour was composed of cysts lined variably by flattened columnar and stratified transitional epithelia. The fibrous septae were oedematous and focally infiltrated by inflammatory cells. No immature blastematos tissue could be identified. The diagnosis was multilocular cystic nephroma (Fig 5). The patient recovered uneventfully from surgery and has remained well on follow-up.

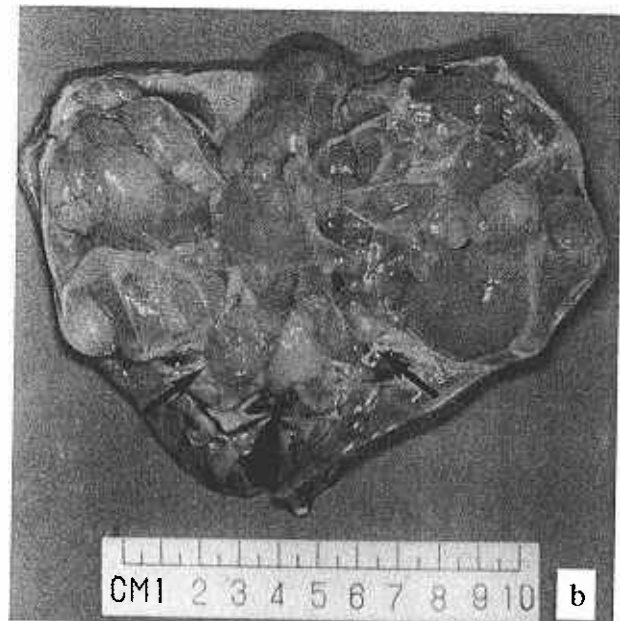
DISCUSSION

A renal mass in an infant can be caused by a variety of lesions such as hydronephrosis, cystic disease, tumour, renal vein thrombosis, haematoma and inflammation (Table 1). Ultrasound is the initial technique of choice for imaging an infant with an abdominal or suspected renal mass. Besides the avoidance of ionizing radiation hazard, ultrasound has the advantages of multiplanar imaging and the ability to characterise masses as cystic, solid or mixed.

Ultrasound is an excellent antenatal screening tool for obstructive renal lesions (Fig 6). Furthermore, ultrasound guides utilisation of other imaging modalities⁽¹⁾. Besides diagnosing hydronephrosis, ultrasound aids in assessing the level of obstruction (Fig 7), detecting underlying anomalies and directing further investigations. For example, in PUJ obstruction, renal scintigraphy is complementary in providing physiological information. Micturating cystourethrography can be performed to assess the degree of vesico-ureteric reflux (Fig 8).

Multicystic renal dysplasia is easily evaluated by sonography, being seen as multiple renal cysts of variable sizes with no normal intervening renal parenchyma or sinus echoes. On CT scan, multiple cysts which are present replace the renal tissue almost entirely. In a small number of patients, remnant renal tissue may persist and be visualised on renal scintigraphy⁽²⁾. Infantile polycystic kidney disease produces bilateral renal enlargement. Sonographically, the kidneys are diffusely echogenic, due to the multiplicity of tiny cysts, with poor corticomedullary differentiation (Fig 9). The severity of liver involvement is

Fig 4 – (a) Nephrectomy specimen showing the large tumour mass, incidentally found bifid ureter (arrows) and remnant lower pole of the left kidney (arrowheads). (b) Cut section of the tumour showing the multiple cysts of varying sizes, as well as internal septae. Herniation into the renal pelvis is arrowed.



recognised to be inversely proportionate to the renal abnormality⁽²⁾.

Medullary sponge kidney is best diagnosed by intravenous urography (IVU). Plain films demonstrate the characteristic pattern of medullary nephrocalcinosis while the classical "paintbrush" appearance of dilated pyramidal collecting tubules are seen on IVU (Fig 10). Renal vein thrombosis is a condition that typically occurs in neonates who have suffered dehydration or asphyxia. Associated adrenal haemorrhage may also be present. The initial diagnosis of renal vein thrombosis is best made with colour Doppler sonography. The kidney is enlarged

Fig 5 – Histological microphotograph of the tumour shows cystic spaces lined by simple flattened and columnar epithelium which have a hobnail appearance in places. The septae are composed of bland fibrovascular tissue. (Haematoxylin and eosin stain magnification x 100).

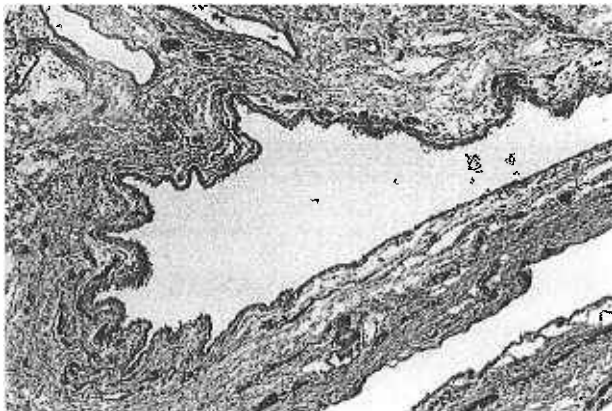


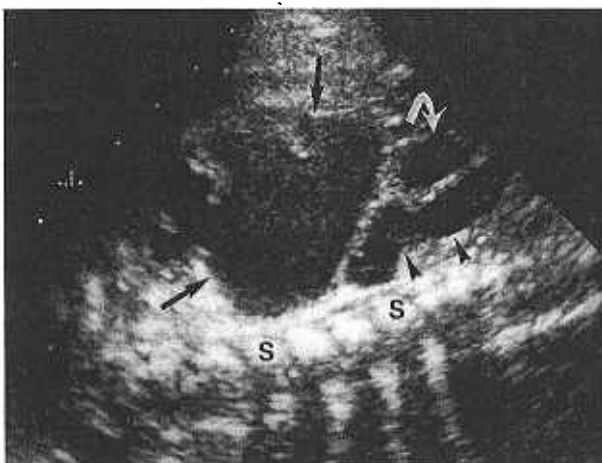
Fig 7 – Ultrasound of an ureterocolic, showing the classical “sitting duck” sign. The head of the duck represents the ureterocolic (arrowheads) and the body, the dilated lower end of ureter (arrows). Bladder is labelled “B”.



Table 1 – Differential diagnoses of a renal mass in infancy

- | |
|---|
| 1. Hydronephrosis |
| - pelvi-ureteric junction (PUJ) obstruction |
| - obstructive megaureter |
| - posterior urethral valves |
| - vesico-ureteric reflux |
| - ureterocolic |
| 2. Cystic disease |
| - infantile polycystic disease |
| - multicystic renal dysplasia |
| - medullary sponge kidney |
| 3. Renal tumour |
| - multilocular cystic nephroma |
| - Wilms' tumour |
| - mesoblastic nephroma |
| 4. Renal vein thrombosis |
| 5. Haematoma |
| 6. Inflammatory disease and abscess |
| 7. Renal ectopia |

Fig 6 – Antenatal ultrasound of a foetus at 32-week gestation showing hydronephrosis in a double-moiety kidney. The dilated lower moiety (arrows), obstructed upper moiety ureter (arrowheads), and part of the upper calyces (curved arrow) are clearly seen anterior to the foetal spine [S]. This was an incidental finding in a patient being investigated for antepartum haemorrhage.



with a disorganised heterogeneous echo pattern.

Wilms' tumour is seen sonographically as an echogenic mass replacing normal renal parenchyma. Pre-contrast CT may show variable patterns of calcification not evident on plain radiographs. Wilms' tumour however has a peak incidence in children at 3 years, unlike mesoblastic nephroma which commonly presents in the first 3 months of life. The usual sonographic appearance of mesoblastic nephroma is that of a huge solid echogenic mass with central hypoechoic areas caused by haemorrhage or necrosis. It is not encapsulated but may penetrate the renal capsule, extending into the perinephric tissue (Fig 11). CT demonstrates a heterogeneous mass. In extensive atypical mesoblastic nephroma, pre-operative chemotherapy is needed to render the tumour operable⁽³⁾.

Multilocular cystic nephroma, also known as multilocular cyst and cystic hamartoma, is an uncommon, benign lesion of uncertain aetiology⁽⁴⁾. It usually presents incidentally, as in our patient. Sometimes, it may manifest with haematuria, pain or even a rapid growing mass⁽⁵⁾. Most patients present below two years of age with an abdominal mass.

Powell et al in 1951 proposed a set of eight criteria for the diagnosis of multilocular cystic nephroma: (a) unilateral involvement, (b) solitary lesion, (c) multilocular, (d) non-communication of the cysts with one another, (e) non-communication with the renal pelvis, (f) loculi lined by epithelium, (g) interocular septa devoid of renal parenchyma, and (h) residual renal tissue, if present, is normal. Joshi et al⁽⁶⁾ added two more criteria to distinguish multilocular cystic nephroma from cystic partially-differentiated Wilms' tumour, namely: the tissue in the cyst must be entirely differentiated tissue and there must not be any blastematosus or embryonal element.

Madewell et al⁽⁷⁾ reviewed the radiographs and pathology of 58 patients with multilocular cystic nephroma, indicating that the diagnosis can be strongly suspected pre-operatively if there is pelvic herniation or if a multilocular pattern is demonstrated on imaging studies. This pelvic herniation is clearly demonstrated in the bisected specimen of our patient (Fig 4b).

Banner et al⁽⁸⁾ suggested partial nephrectomy for this tumour, based on a review of the literature confirming its benign nature. However, it is only after careful histological examination that malignant elements in a cystic renal lesion can be identified. Beckwith⁽⁹⁾ emphasized the difficulty in distinguishing multilocular cystic nephroma from cystic Wilms' tumour and cystic mesoblastic nephroma pre-operatively. Partial nephrectomy alone would not be adequate for the latter two

Fig 8 – Micturating cystourethrogram showing a dilated right pelvicalyceal system secondary to vesico-ureteric reflux (B=bladder).

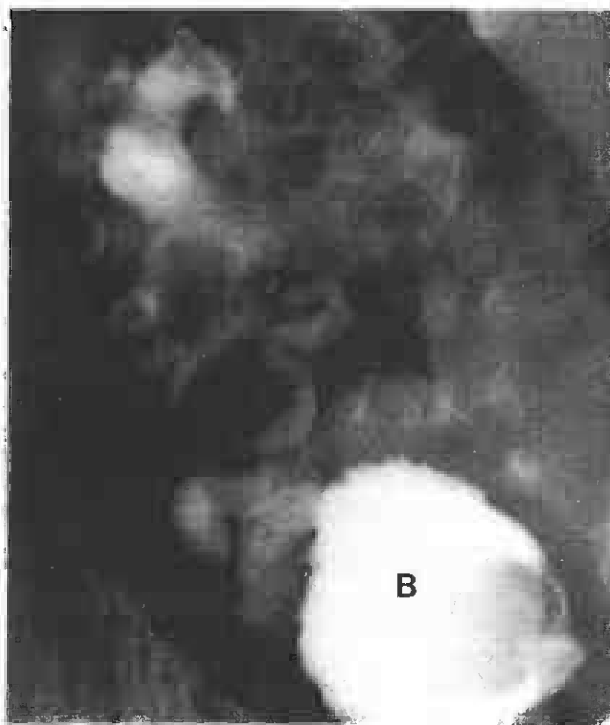


Fig 9 – Right renal ultrasound of infantile polycystic kidney disease shows typical findings of diffuse echogenicity, loss of corticomedullary differentiation and some tiny cysts. The left kidney had a similar appearance.



lesions. Furthermore, ultrasound-guided aspiration cytology has not been reliable for diagnosis as atypical cells have been identified in the aspirate⁽¹⁰⁾. A multilocular cystic nephroma, without any evidence of neoplastic foci, can still cause complications such as hydronephrosis, hypertension and external compression of the renal pelvis. Thus, nephrectomy is recommended as the standard treatment for this lesion.

ACKNOWLEDGEMENT

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Fig 10 – Medullary sponge kidneys, (a) Radiograph shows bilateral nephrocalcinosis (arrows). (b) IVU shows streaky appearance of dilated collecting tubules in the renal medulla.

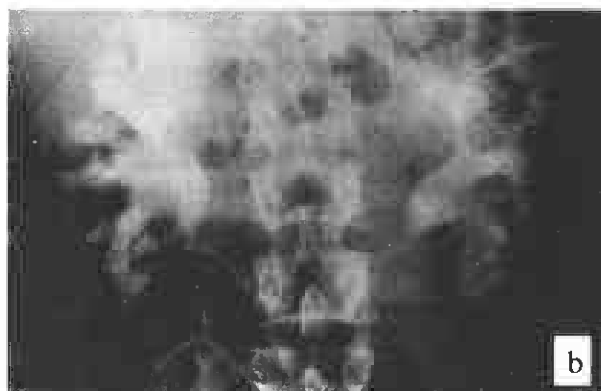
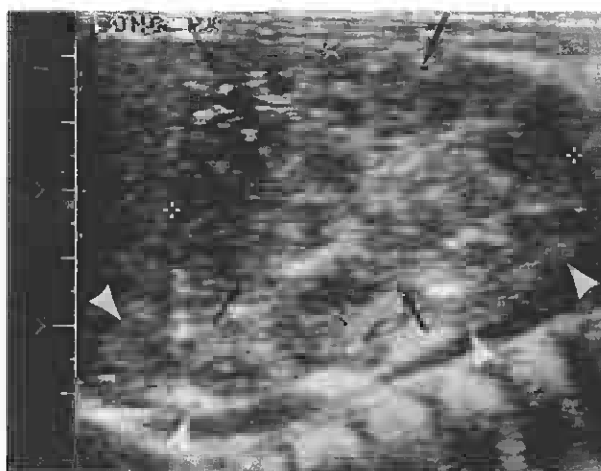


Fig 11 – Mesoblastic nephroblastoma of the right kidney. Ultrasound shows a complex solid mass (arrows) arising from the cortex of the kidney (arrowheads).



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ABSTRACT

A male Chinese infant presented with the incidental finding of a left loin mass. Plain abdominal radiograph showed a large left-sided soft tissue shadow. Ultrasound and computed tomography scans demonstrated a multiloculated cystic lesion arising from the upper pole of the left kidney. Histological examination of the nephrectomy specimen confirmed the diagnosis of multilocular cystic nephroma. The differential diagnoses of infantile renal masses, the role of imaging in assessment of these masses and the surgical management of multilocular cystic nephroma are discussed.

Keywords: multilocular cystic nephroma, renal cyst, renal mass, ultrasound.