A trio of multiple Wilms' tumours

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Abstract

Wilms' tumour is the commonest malignant abdominal tumour in children. Unilateral multicentric, and bilateral Wilms' tumours are, however, less common, occurring in 7% and 5% of cases respectively. These are often associated with sporadic aniridia, genitourinary anomalies, hemihypertrophy and nephroblastomatosis. Nephroblastomatosis is a separate entity that may act as a precursor of Wilms' tumours. We present three cases of multiple Wilms' tumours. Two cases also had nephroblastomatosis which was not seen on pre-operative CT imaging but was identified in one case at MRI. In cases of multiple Wilms' tumours, MRI provides better delineation of the tumours and may, therefore, affect management.

Key words
Wilms' tumour, nephroblastomatosis, magnetic resonance imaging (MRI), computed tomography (CT), horseshoe kidney.

Introduction

Wilms' tumour (WT) is the commonest malignant abdominal tumour in children.[1] Unilateral multicentric, and bilateral WT are less common. We present three cases of multiple WT and their imaging features.

Case 1

A 2-year-old boy presented with a month's history of weight loss and a palpable right-sided abdominal mass. Ultrasound (US) and computed tomography (CT) showed large upper pole and smaller lower pole masses in the right kidney (Figures 1a and b). A right nephrectomy was performed and histology demonstrated nephroblastomatosis and three WT of favourable histology.

Case 2

A 4-year-old boy with multiple congenital anomalies, including a horseshoe kidney, presented with macroscopic haematuria. A palpable abdominal mass was sonographically shown to arise from the left side of the horseshoe kidney. On CT, bilateral renal masses were seen in keeping with WT (Figures 2a and b). In

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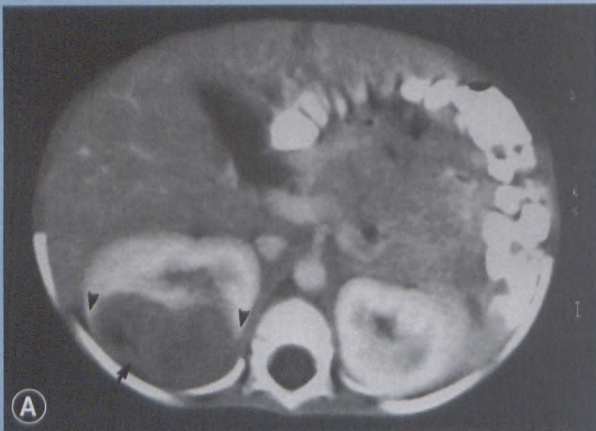- **low-osmolar**
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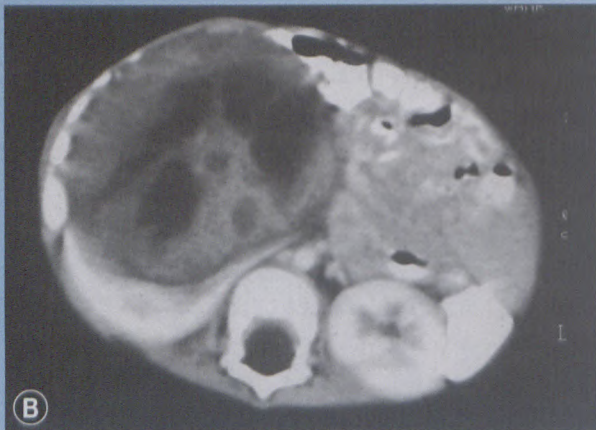**Schering
Diagnostics**

From seeing to understanding

A trio of multiple Wilms' tumours

*Figures 1a and b: Two tumours in a single kidney*

(A)

a. Post-contrast CT shows a low density mass (arrow) in the posterola-teral upper pole of the right kidney with a typical claw sign (arrowheads).

(B)

b. Post-contrast CT of the same patient shows another mass with cystic components in the anteromedial lower pole of the right kidney.

view of the clinical setting, the child was managed conservatively until his death.

Figures 2a and b: Bilateral tumours in a horseshoe kidney

(A)
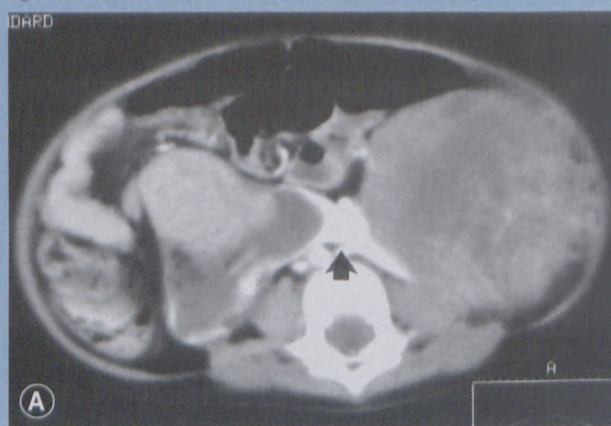
a. Post-contrast CT shows masses in both kidneys with a contrast-enhancing isthmus (arrow) of the horseshoe kidney.

Case 3

A 4-year-old boy presented with a 10-day history of anorexia, abdominal pain and intermittent fever. An US and CT scan (Figure 3a) demonstrated bilateral renal masses (two in the right kidney and one on the left). After chemotherapy, the left WT was removed by partial nephrectomy. A DMSA scan demonstrated a differential function of 68% on the right and 32% on the left. Repeat CT showed more than one mass in the right kidney. In view of possible salvage surgery in the better functioning right kidney, an MRI was performed to delineate the extent of the masses. At least five masses were demonstrated (Figures 3b,c,d), three of which were histologically proven nephroblastomatosis. A repeat course of chemotherapy was instituted.

Discussion

Wilms' tumour is the commonest malignant abdominal tu-

(B)
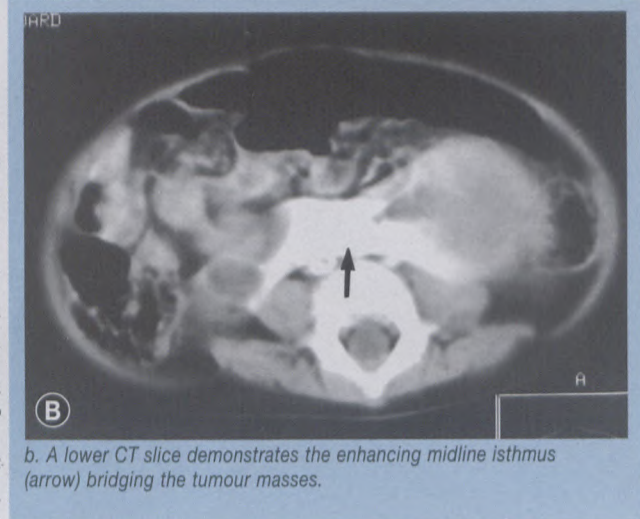
b. A lower CT slice demonstrates the enhancing midline isthmus (arrow) bridging the tumour masses.

mour of children between one and eight years of age and is the third commonest cause for a renal mass in childhood after hydronephrosis and multicystic dysplastic kidney.[1] The commonest presentation is with an asymptomatic abdominal mass. Uncommon presentations include abdominal pain, fever, anorexia, haematuria and hypertension. Although mostly sporadic, WT are associated with several conditions, including horseshoe kidneys, Beckwith-Wiedemann syndrome and sporadic aniridia.[2] Bilateral tumours occur in 5% of cases and unilateral multicentric tumours in 7%. Bilateral tumours are often associated with sporadic aniridia, genitourinary anomalies, hemihypertrophy and nephroblastomatosis (persistent metanephric blastemal rests found after 34 weeks' gestation). Nephroblastomatosis is considered an intermediate between a malformation and neoplasm.[3,4] These nephrogenic rests are seen in 1% of autopsies of children under three months of age.[2] They are associated with synchronous bilateral WT in 99%, and with metachronous bilateral WT in 94% of cases. Nephroblastomatosis therefore has
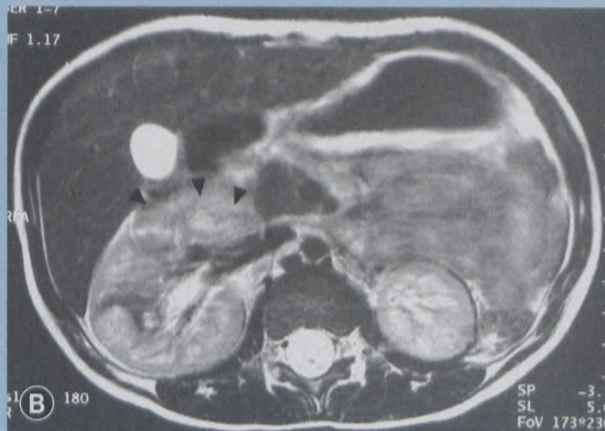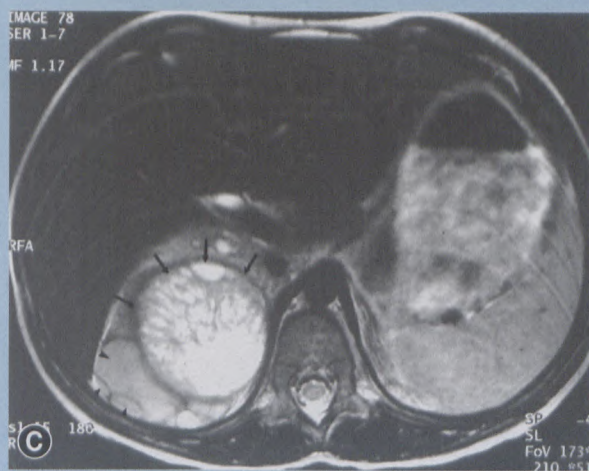
Figures 3a-d: Bilateral tumours
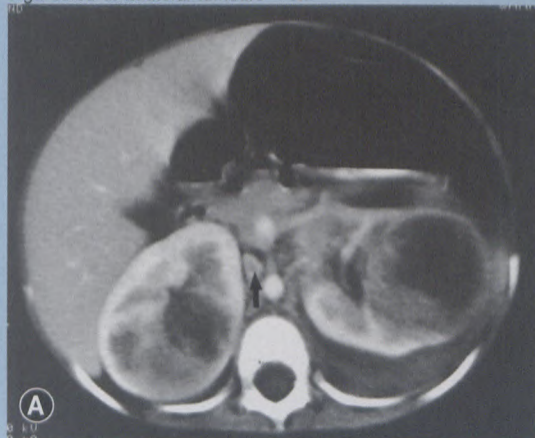
a. Post-contrast CT shows bilateral low density renal masses with cystic components. Note the thrombus in the IVC (arrow).

b. Axial T2-weighted MRI post left partial nephrectomy shows an anteromedial upper pole renal mass (arrowheads) that is iso-intense to the rest of the kidney.

c. Axial T2-weighted MRI shows clear differentiation between the inhomogeneous Wilms' mass (arrow) with increased central T2 signal intensity, and the peripheral, flatter nodules of nephroblastomatosis (arrowhead) that are iso-intense to cortex.

d. T2-weighted MRI in the coronal plane allows evaluation of multiple masses (open arrows) with greater ease.

malignant potential and is hence a precursor of WT.[3] While ultrasound is often the initial investigation and provides good detail of IVC tumour thrombus, CT scanning provides better detail of the tumours. Post-treatment imaging is necessary to detect recurrence and metachronous tumours – especially with associations known to develop multiple WT.[2] On CT, the tumour is typically of inhomogeneous low-density with increased inhomogeneity post-contrast. A 'claw sign' confirms the renal origin of the mass. The mass is usually solid but haemorrhage and necrosis may cause cystic areas. Calcification

is seen in up to 15% of cases. MRI is helpful in difficult cases as coronal imaging allows better tumour delineation. Generally WT are heterogeneous with low signal intensity on T1-weighting, high signal on T2-weighting and show inhomogeneous contrast enhancement. Haemorrhage and necrosis may alter these appearances.[3] Differential diagnosis includes neuroblastoma, nephroblastomatosis, mesoblastic nephroma, multilocular cystic renal tumours, renal cell carcinoma, renal lymphoma, renal leukaemia, clear-cell sarcoma and rhabdoid tumours of the kidney. Nephrogenic rests may either be perilobar (periph-

eral) or intralobar (central). The intralobar rests are associated with more frequent development of WT and at a younger age.[4] On CT, the nephrogenic rests are typically well-defined plaque-like or nodular subcapsular masses that are homogenously isodense to renal cortex. They enhance poorly and hence appear hypo-dense relative to renal cortex. On MRI they are iso- to hypo-intense to renal cortex on both T1- and T2-weighted sequences. They enhance poorly with gadolinium and are hypo-intense relative to renal cortex. Both nephroblastomatosis and WT are best seen on post-gadolinium images. The

most reliable differentiation between them is based on their homogeneity.[3] In practice, the imaging[4] and surgery[5] for WT often differs from the National Wilms' Tumour Study Group (NWTSG) protocol. Controversial points include radiological or surgical staging, timing of chemotherapy, the adequacy of partial nephrectomy, and the optimum follow-up surveillance.[4,5] This is especially so for multiple WT where the surgical approach and need for accurate follow-up surveillance is complicated by the presence of multiple potentially malignant nephrogenic rests. Cases one and three had nephroblastomatosis which was not seen on pre-operative CT imaging. In case three it was, however, identified on the pre-operative MRI scan. In view of these problems, it may be advisable to consider MRI in complicated cases with multiple WT.

Conclusion

In cases of multiple WT, MRI provides better delineation of the tumours and increased sensitivity for detecting nephroblastomatosis. It should be considered in the tumour work-up as it may affect management.

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