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## CASE REPORT

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# Bilateral Wilms' Tumour

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### ABSTRACT

*We present a case of bilateral Wilms' tumours in a three-year-old girl. In addition, we briefly discuss the differential diagnoses of bilateral Wilms' tumours and their further characterisation from unilateral tumours. Younger age at presentation, association of renal vein and inferior vena cava thrombosis and lung metastases, may help making a diagnosis of the Wilms' tumour over other renal tumours.*

*Key Words:* Child; Kidney neoplasms; Neoplasm metastasis; Wilms tumor

## 中文摘要

### 雙側腎母細胞瘤

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本文報告一宗發生在一名三歲女孩的雙側腎母細胞瘤病例，並簡短討論此病的鑒別診斷及其與單側腫瘤的不同之處。與其他類型的腎腫瘤不同，腎母細胞瘤通常在年少時發病，並可併發腎靜脈及下腔靜脈血栓和肺轉移。

### INTRODUCTION

Wilms' tumours account for 87% of paediatric renal masses, with peak incidence at 3 to 4 years of age.<sup>1</sup> Unilateral Wilms' tumour present at an earlier age in males (mean age, 42 months) than in females (mean age, 47 months), while the mean age of presentation for bilateral Wilms' tumours (BWTs) is lower, at 30 months and 33 months, respectively. BWTs account for 4 to 13% of children with Wilms' tumours and have an increased incidence of congenital anomalies such as cryptorchidism, hemihypertrophy, hypospadias, and sporadic aniridia.<sup>2</sup>

### CASE REPORT

A three-year-old previously well girl presented with abdominal distension and haematuria in April 2004. On general examination, the child was anaemic, and the abdomen was tender and distended. Plain radiography revealed increased opacity of the abdomen with obscuration of renal and psoas muscle margins (Figure 1), raising the suspicion of retroperitoneal mass, but abnormal calcifications or bony lesions were not detected. Urgent ultrasound examination confirmed the presence of bilateral renal masses. Computed tomography (CT) of her abdomen showed

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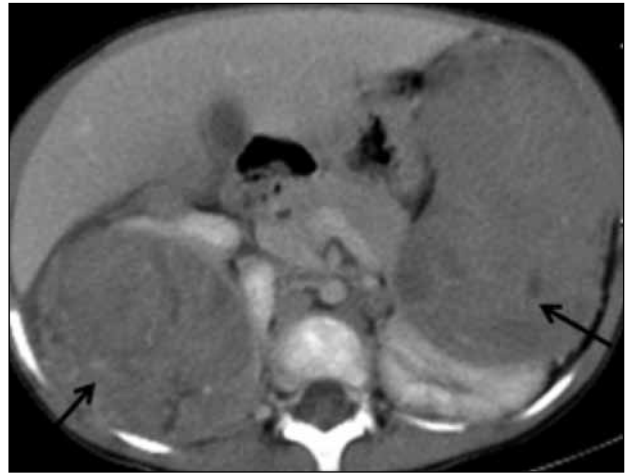
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multiple heterogeneously enhancing solid-cystic renal masses (Figure 2). The renal veins and inferior vena cava (IVC) were patent (Figure 3). There was bilateral hydronephrosis and mild ascites (Figures 4 and 5). No significant retroperitoneal lymphadenopathy or distant metastases were detected, and renal vessels were unremarkable bilaterally. Considering patient's age, a working diagnosis of BWT was made, while lymphoma and metastases were considered to be rare possibilities. Biopsy of the renal masses confirmed the diagnosis of BWT. The patient received a 21-week course of neoadjuvant chemotherapy consisting of vincristine, actinomycin, and doxorubicin. There was partial response to chemotherapy, with shrinkage of the bilateral renal masses. Bilateral partial nephrectomy was then performed. Histology showed viable tumour with no anaplastic features. Whole abdominal radiation therapy (1080 cGy) was given postoperatively, followed by a further chemotherapy course with vincristine



**Figure 1.** A plain radiograph, which shows increased opacity of the abdomen with scanty bowel gas. The psoas muscles are not identified, which is suspicious of a retroperitoneal mass. There is no abnormal calcification or bony lesion.



**Figure 2.** A contrast-enhanced axial computed tomographic image of the abdomen showing multiple heterogeneous renal masses arising from both kidneys (black arrows). Both renal arteries are intact. There is no evidence of liver metastases or enlarged para-aortic lymph glands.



**Figure 3.** A contrast-enhanced axial computed tomographic image of the abdomen showing the left renal vein (arrow) and inferior vena cava (arrowhead) are patent.

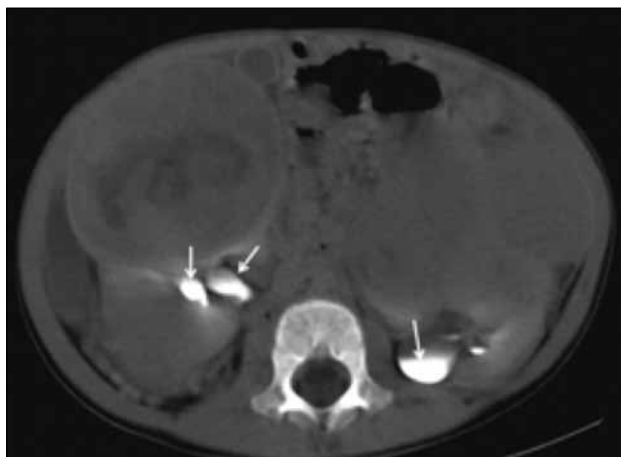
and actinomycin for 54 weeks. To date, the girl is in remission with no evidence of recurrence at the age of 10 years.

## DISCUSSION

BWT tends to have a more indolent clinical course than unilateral case. A child with a sibling or parent with BWT has a 30% risk of developing the tumour. These patients may present with an abdominal mass, haematuria and / or abdominal pain. Raised blood pressure due to renin production by the tumour may ensue.



**Figure 4.** A contrast-enhanced axial computed tomographic image of the abdomen showing ascites (white arrowhead) and bilateral hydronephrosis (black arrowheads). Bilateral renal masses similar to those in Figure 1 (black arrows) are evident. There is no suspicion of para-aortic lymphadenopathy.



**Figure 5.** An axial computed tomographic image during the excretory phase of both kidneys. Obstructive hydronephrosis is clearly demonstrated by contrast in the dilated renal pelvis (arrows) of each kidney.

Suppression of the *WT1* gene at the 11p13 locus, and the *WT2* gene at the 11p15 locus, which are known to play an important role in the normal development of the urogenital tract are considered responsible genetic factors. The risk for developing Wilms' tumour is increased in certain congenital syndromes, namely: Beckwith-Wiedemann syndrome, isolated hemihypertrophy, Perlman syndrome, Sotos syndrome, Simpson-Golabi-Behmel syndrome, isolated aniridia, trisomy 18, the WAGR syndrome (i.e., Wilms' tumour, aniridia, genitourinary abnormalities, and mental retardation), the Bloom syndrome, and the Denys-Drash

syndrome. However, with respect to unilateral versus bilateral Wilms' tumours, there is not much difference in terms of prevailing genetic factors and associated syndromes.

The National Wilms' tumour study staging comprises: stage I, tumour limited to the kidney and is completely resectable; stage II, tumour extending beyond the kidney but still completely resectable; stage III, residual tumour confined to the abdomen without distant metastasis; stage IV, haematogeneous metastases to lung, liver, bone, brain; and finally stage V, bilateral renal involvement appearing initially or manifesting during treatment. Thus, BWTs are all considered stage V disease. Generally, both kidneys are asymmetrically involved, one kidney being more severely affected without any salvageable renal substance while the opposite side has multiple cortical lesions. Most patients present with synchronous disease. Metachronous disease (two or more cancers appearing at different points in time) has an ominous prognosis. In our patient, the tumours were synchronous, with right kidney being more severely affected than the left.

On imaging, BWT commonly presents as solid, often heterogeneous, intrarenal masses, with internal haemorrhage, fat, necrosis, or calcification.<sup>2</sup> Wilms' tumours typically spread by direct extension and displace adjacent structures. There may be invasion of the renal vein and IVC, with occasional extension into the right atrium. Metastases to lung are more common to bone or liver.<sup>3</sup>

On ultrasound, Wilms' tumours appear heterogeneous in echogenicity due to the presence of haemorrhage, fat, necrosis, or calcification.<sup>2</sup> On CT, they also appear heterogeneous. Contrast-enhanced CT is better for demonstrating tumour extension into the renal vein or IVC, hepatic and nodal metastases, and associated nephrogenic rests.<sup>3</sup> On magnetic resonance imaging (MRI), Wilms' tumours demonstrate low signal intensity on T1-weighted and high signal intensity on T2-weighted images. IVC patency, hepatic metastases, and multifocal disease can also be well depicted by MRI. Renal vein and IVC involvement necessitate modification of the surgical approach. MRI is reported to be the most sensitive modality for determination of IVC patency.<sup>3</sup> However, the longer sedation required for the examination has limited its use in young children. Nevertheless, surgical planning can be achieved based on meticulous ultrasound and contrast-enhanced CT

examination.<sup>3</sup> In the study by Stickles et al<sup>4</sup> of three patients with Wilms' tumour (one with BWT), there was a good correlation between the fluorodeoxyglucose uptake and tumour / metastatic chest disease, which is crucial in the staging and surveillance of this tumour.<sup>4</sup>

Differential diagnoses for BTWs include: bilateral renal cell carcinomas, metastases, lymphoma, and neuroblastoma. Other differentials include: mesoblastic nephroma, multilocular cystic renal tumour, clear cell sarcoma, rhabdoid tumour, angiomyolipoma, and renal medullary carcinoma.<sup>3</sup> Although renal cell carcinoma tends to be smaller than Wilms' tumour, its gross morphology is similar, and preoperatively the two can be indistinguishable. They both give rise to infiltrative solid masses with variable necrosis, haemorrhage, calcification, and cystic degeneration. Compared to Wilms' tumour, renal cell carcinoma is more likely to manifest bilaterally and metastasise to bone. There is a higher frequency of calcification (25%) than in the former (9%).<sup>2</sup> Compared to Wilms' tumour, metastases and lymphoma are relatively rare at this age. Unilateral Wilms' tumours need to be differentiated from neuroblastoma, another common paediatric abdominal tumour. Neuroblastoma commonly arises from an adrenal gland, is usually calcified, crosses the midline, and is more commonly associated with bone than lung metastases. In a neonate with a solid renal mass, mesoblastic nephroma remains a major concern and characteristically manifests as a multilocular cystic renal tumour with little solid tissue.<sup>3</sup> Clear cell sarcoma and rhabdoid tumour should be considered in the presence of concurrent skeletal metastases and brain neoplasms, respectively.<sup>3</sup> Angiomyolipoma is characterised by fat and is associated with tuberous sclerosis. Renal

medullary carcinoma manifests as an infiltrative mass with metastases, and is frequently reported in patients with sickle-cell trait or haemoglobin sickle-cell disease.

Treatment of BWT aims at maximal conservation of uninvolved renal parenchyma, in order to salvage renal function. Preoperative chemotherapy is particularly important because each kidney is staged separately, and complete resolution of disease in one kidney may allow surgery on the contralateral kidney to achieve a definitive cure. A larger tumour is usually dealt with by radical nephrectomy, otherwise the surgical approach mainly entails tumour resection with sparing of normal parenchyma. In contrast, unilateral Wilms' tumour is generally treated by nephrectomy, followed by adjuvant chemotherapy. Pre-surgical treatment with chemotherapy may be used to promote shrinkage of the tumour, so as to improve outcome.<sup>5</sup>

In conclusion, this report describes the radiological appearance of BWTs. The differential diagnosis of bilateral renal masses and their differentiating features are discussed, and may help radiologists reach a correct diagnosis.

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