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Case report

# Synchronous bilateral Wilms tumour: A case report with review of literature

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ABSTRACT

Wilms tumour is the most common malignant renal tumour in childhood. Approximately 5–7% of Wilms tumour patients present with bilateral disease, either synchronously or metachronously. However, its association with hypospadias is seen in only 1.8% cases. Synchronous bilateral Wilms tumour poses the special challenge of establishing local tumour control while preserving renal function. We describe the case of synchronous bilateral Wilms tumour associated with hypospadias in a 1-year-old male child. A review of the aetiology, demographics, diagnosis and imaging, staging and treatment of Wilms tumour with emphasis on bilateral disease will be presented.

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## 1. Introduction

Wilms tumour (WT) is the most common paediatric renal mass accounting for 87% of cases, with peak incidence at 3–4 years of the age and more commonly in girls.[1] Involvement of both kidneys either at presentation or subsequently was seen in 5–7% of cases, but the majority of bilateral Wilms tumour (BWT) with synchronous disease at diagnosis.[2] The great majority of tumours occur at mean age of 3 and half year for unilateral (sporadic) cases and 2 and half year for bilateral or heritable cases.[1–3] Bilateral Wilms tumour has an increased incidence of congenital anomalies such as crytorchidism (in 2.8%), hemihypertrophy (in 2.5%), hypospadias (in 1.8%) and sporadic aniridia.[3]

## 2. Case report

A 1-year-old previously well boy presented to our institution with abdominal distension and palpable lump in abdomen on either side. There was no hematuria and no abdominal pain. The clinical examination revealed hypospadias in malnourished child. No hemihypertrophy, aniridia or other congenital anomalies were identified. X-ray abdomen and ultrasonography of abdomen were performed in our Radio diagnosis department. Plain X-ray abdomen revealed ill defined soft tissue haziness in either side of abdomen with regional displacement of pro-peritoneal fat plane without any calcification or bony lesions (Fig. 1). Abdominal ultrasound revealed about 10 cm × 8.5 cm × 7.6 cm homogenous mass lesion involving lower pole of right kidney and similar mass lesion of about 10.1 cm × 8.0 cm × 6.3 cm involving almost whole of left kidney. No hydronephrosis was noted in either of kidneys. Plain and contrast-enhanced computed tomography (CT) of abdomen and chest was performed under short general anaesthesia. CT findings were similar to that of ultrasound without evidence of inferior vena cava and renal vein involvement. No evidence of retroperitoneal lymphadenopathy was noted (Fig. 2). Chest CT scan did not show evidence of metastasis.

A synchronous BWT diagnosis was put on the basis of typical radiological findings. Then biopsies from masses of both kidneys were performed. The histopathology shows triphasic pattern with favourable histology of Wilms tumour. Diagnosis was consistent with stage V favourable-histology WT and treatment was initiated with chemotherapeutic agents. During the course of chemotherapy, the child died.

## 3. Discussion

### 3.1. Demographics

About 95% cases are diagnosed before age 10, with a peak incidence at age of 3. A child with a sibling or parent with BWT has a 30% risk of developing the tumour.[4] Two-thirds of bilateral cases present synchronously; the remainder are typically diagnosed from 2 years to as long as 19 years after the initial tumour.

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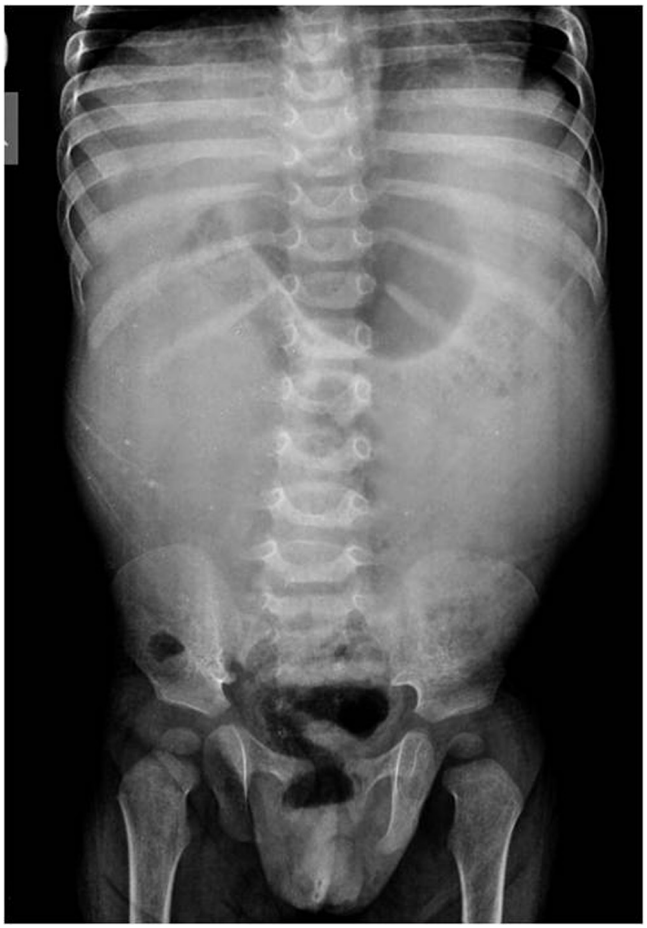

**Fig. 1.** Plain X-ray abdomen showing huge opacity in abdomen on either side of spine displacing bowel loops with obscuration of renal margin.

Synchronous bilateral disease tends to present at a younger age than does unilateral disease.[5,6] The risk for developing Wilms tumour is increased in certain congenital syndrome like Beckwith–Wiedemann syndrome, Isolated hemihypertrophy, Perlman syndrome, Sotos syndrome, Simpson–Golabi–Behmel syndrome, WAGR syndrome, Bloom syndrome and Denys–Drash syndrome. According to some authors, the frequency of these anomalies may be up to ten times higher in cases of bilateral diseases, especially with regard to hemihypertrophy and hypospadias,[6,7] while some other authors described that, with respect to unilateral versus bilateral Wilms tumour, there is not much difference in terms of prevailing genetic factors and associated syndrome.[4]

### 3.2. Aetiology

The tumour typically arises from mesodermal precursors of renal parenchyma (metanephros). Hereditary cases may account for 20–38% of Wilms tumour cases. Increasingly, gene foci are being implicated on chromosome 11 (WT1; 11 p13 and WT2: 11p15) as well as other loci on chromosomes 1, 8 and 12.[8] It is speculated that WT is associated with nephroblastomatosis, which represents the persistence of immature renal tissue beyond the time of completion of nephrogensis.[9] According to Machin et al., all cases of his study show evidence of nephroblastomatosis supporting two hypothesis that this multicentric abnormality important in occurrence of multifocal neoplasm and BWT represents multiple primary tumours rather than metastasis from the contralateral kidney.[8] But our case had no evidence of nephroblastomatosis changes in biopsy.

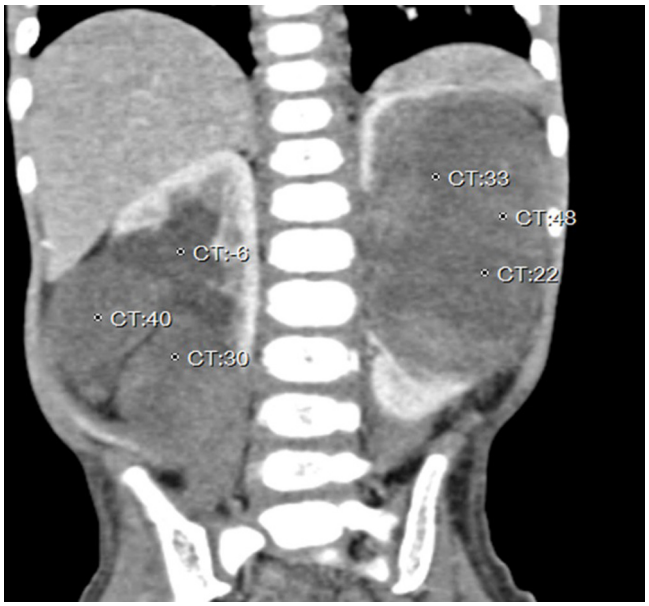


**Fig. 2.** Coronal image of venous phase of CT scan shows heterogeneous contrast enhancement of mass lesions.

### 3.3. Diagnosis and imaging

The most common presenting symptoms in patients with WT are abdominal mass, pain, hematuria and hypertension. Later, two have been noted to be more frequent in cases of bilateral disease.[10]

In the past, the intravenous pyleography has been the mainstay of evaluation of patients with WT, typically revealing calyceal distortion and displacement. On ultrasound, WT appears heterogeneous in echogenicity due to presence of haemorrhage, fat, necrosis or calcification.[7] Contrast-enhanced CT scan reveals the extent of tumour mass, renal functional status, Inferior Vena Cava (IVC) patency, hepatic and nodal metastasis and associated nephrogenic rests.[11] In cases of bialterality, it has been shown to be quite sensitive in preoperative assessment of feasibility of partial nephrectomy. On magnetic resonance imaging (MRI), WT demonstrates low signal intensity on T1-weighted and high signal intensity on T2-weighted images. MRI is reported to be the most sensitive modality for determination of IVC patency.[11] In the study by Stickles et al., of three patients with WT (one with bilateral tumour), there was a good correlation between the flurodeoxyglucose uptake and tumour/metastatic chest disease, which is crucial in the staging and surveillance of this tumour.[12] While in earlier days surgical exploration of contralateral kidney was the only method to detect a contralateral tumour, the necessity of this way of assessment has been questioned with the introduction of modern imaging techniques particularly, CT and MRI. But even these modalities miss up to 50% lesions below 1 cm in its greatest dimension. Paya et al. reported in their study that bilaterality could not be determined despite the use of all available imaging methods prior to surgery.[13] The differential diagnoses of BWT include neuroblastoma, Non-Hodgkin's lymphoma, rhabdomyosarcoma and mesoblasticnephroma, cystic nephroma and clear cell sarcoma. They all are differentiated from BWT according to its age of presentation, its characteristic features and its association with other diseases.

### 3.4. Staging

Staging is an important step that can help doctors to determine the most effective treatment approach. National Wilms' Tumour Study Group (NWTS) staging system is the most widely used system (Table 1).[14] The NWTS staging system places all patients