# Can Wilms' tumor recur in the ureteric stump?

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**ABSTRACT**

Wilms' tumor recurrences are notably encountered in the tumor bed, less commonly in the lungs. Metastatic involvement ureter markedly uncommon, though extension of the tumor at the presentation of primary tumor is known. We are documenting the metastasis to the ureteric stump which occurred in our patient nearly 2 years after completion of treatment for Wilms' tumor.

**Keywords:** Hematuria; recurrent Wilms' tumor; ureteric metastasis.

## Introduction

Wilms' tumor is the commonest renal tumor in children. It is associated with a good prognosis if it is detected at a early stage and good response to chemo/radiotherapy is obtained. However, survival rates in recurrent Wilms' tumor are reportedly not as good as those of primary tumor. Several prognostic factors such as tumor spillage, unfavourable histology, early recurrence within 12 months of initial diagnosis have been identified with tumor recurrence.[1]

We report an interesting case of late recurrence of Wilms' tumor seen in ipsilateral ureteric stump.

## Case presentation

A 7-year-old-boy, known case of Stage III Wilms' tumor of the right kidney with favourable histology presented with painless gross hematuria persisted for one day. Having completed chemotherapy and radiotherapy 2 years earlier, as per National Wilms Tumor Study (NWTS-5) protocol, he was on regular follow-up. On sonography, a suspicious lesion of 4x1 cm was identified at the right vesicoureteric junction. Contrast-enhanced CT confirmed the lesion to be at the right ureteric stump, without any locoregional metastases (Figure 1). Cystoscopy revealed an extramucosal lesion abutting the right vesicoureteric junction (Figure 2). On discussion with tumor board, administration of second-line chemotherapy [ifosfamide Cyclophosphamide Etoposide (ICE) regimen] was planned. After one cycle of chemotherapy, the lesion on re-assessment was found to be without any significant reduction in size. Surgery was undertaken after written informed consent was obtained from his parents. Complete excision including a cuff of bladder was performed via extravesical approach (Figure 3). Targeted radiotherapy of the pelvic region, as per the tumor board recommendation completed the treatment. Histology of the lesion (Figure 4) demonstrated a well-differentiated Wilms' tumor, infiltrating the ureteric wall. Based on follow-up evaluation, he is doing well 2 years after completion of multimodality treatment. His current renal function as evaluated according to serum creatinine levels and glomerular filtration rate by nuclear scintigraphy is within normal limits.

## Discussion

Wilms' tumour involves ureters in only 2% of the cases. Wilms' tumor is known to grow into the pelvis and ureter with consequent hematuria. Also, intraurethral passage of clots or tissue fragments, flank pain and intraurethral mass are additional features indicating ureteric involvement. Imaging may not be accurate in the identification

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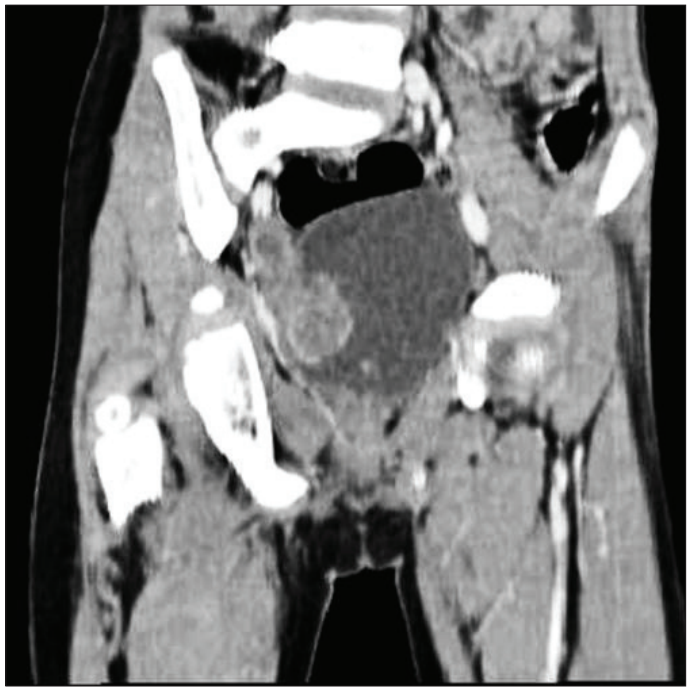

Figure 1. CT of pelvis: (coronal section) Lesion at the right vesico-ureteric junction

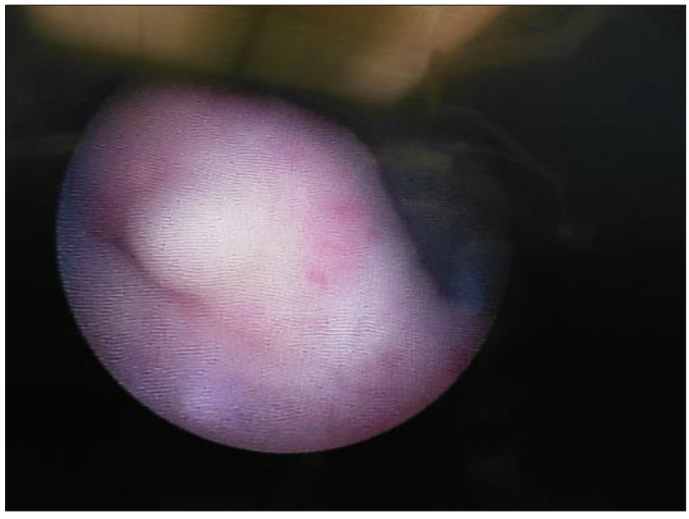

Figure 2. Cystoscopic view of extramucosal tumor abutting the right vesico-ureteric junction

of ureteric involvement, as only 30% of the cases are detected pre-operatively. However, presence of hydroureteronephrosis should alert the physician to the possibility of ureteric involvement.[2]

Wilms' tumor recurs in about 16-40% the cases. The tumor recurs locally in tumor bed, and distally to the lung. They usually relapse within 2 years of tumor diagnosis.[3] More commonly anaplastic type Wilms' tumor recurs. Notably, the ability of the tumor to

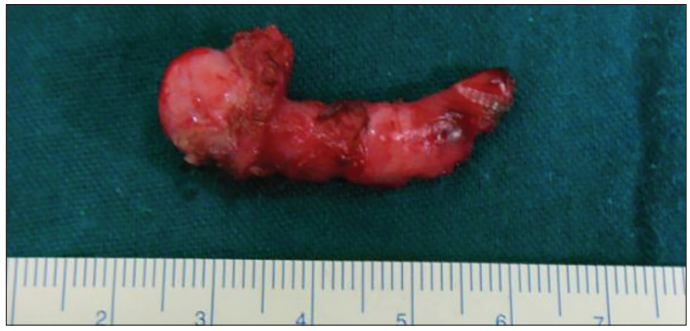

Figure 3. Excised specimen of the ureteric stump together with the bladder cuff

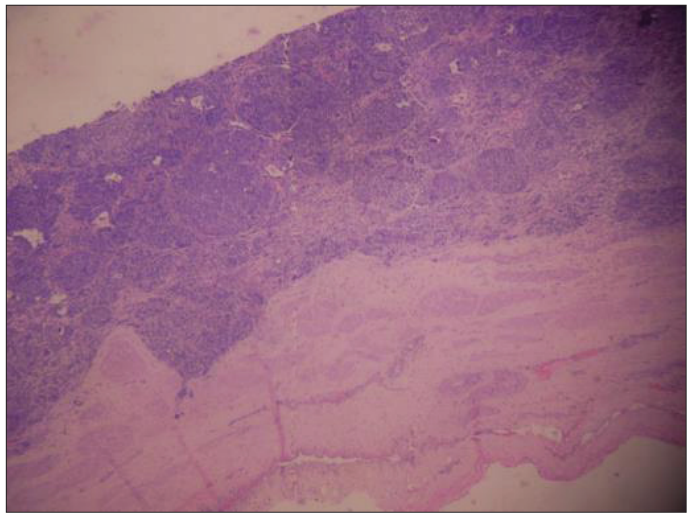

Figure 4. Histopathology: Ureteric wall infiltrated by tumour X100, H/E

invade lymphovascular structures and responsiveness of the remnant tumor to adjuvant therapy are causes of recurrence. Impact of genetic aberrations on tumor recurrence are well known. Specifically, gene expression signatures such as LOH 1p and 16q indicate poor prognosis due to their increased association with tumor relapse in stage III tumors with favourable histology.[4]

Microscopic tumor deposits, which get seeded in the ureter and lie dormant escape from host immune surveillance to manifest as late recurrence is one of the explanations for late tumor relapse in Wilms' tumor. Radiotherapy has been blamed as the culprit for the delay in resumption of the activity of the tumor cells, inducing them to be in a dormant state of variable time. This possibly can explain the unusual finding in the case described.[5]

Administration of second-line chemotherapy has profound effects on the solitary kidney. Hence careful assessment is necessary to identify renal damage during the follow-up period.

In conclusion, isolated ureteric metastasis is a rare occurrence in Wilms' tumor, more so when the recurrence happens long after the tumor nephrectomy. Although the burden of morbidity is likely to be more with additional chemotherapy on the solitary kidney, the prognosis appears to be good after surgical excision and consolidation with multi-modality treatment. Regular follow-up is vital to detect such unfortunate and uncommon recurrences.

**Informed Consent:** Written informed consent was obtained from patient's parents who participated in this case.

**Peer-review:** Externally peer-reviewed.

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