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Case reports

# The use of preoperative chemotherapy in Wilms' tumor with contained retroperitoneal rupture

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#### Index words:

Wilms' tumor; Neoadjuvant chemotherapy; Retroperitoneal; Rupture

#### **Abstract**

**Purpose:** The National Wilms Tumor Study currently describes 3 indications for the use of preoperative chemotherapy: extensive caval involvement, bilateral tumors, and patients who only have a single kidney. However, the management of patients who present with a contained retroperitoneal rupture is not specifically addressed. This is relevant because of the strong possibility of peritoneal contamination when performing a primary resection and the resultant requirement for total abdominal radiation. The use of neoadjuvant chemotherapy in this subgroup of patients may be warranted.

**Methods:** We retrospectively reviewed our experience with Wilms' tumor and identified 3 cases with contained rupture at presentation. Details of their initial evaluation and therapy, resection and pathologic findings, and follow-up constitute this report. Institutional review board waiver was obtained for the purposes of this review.

**Results:** Two male patients, aged 2.9 years, and 1 female patient, aged 9.3 years, were identified. All patients received preoperative chemotherapy with vincristine and dactinomycin (n = 1) plus doxorubicin (n = 2) for 4 to 6 weeks before surgical resection. One patient underwent pretreatment computed tomography—guided biopsy of the kidney mass for diagnostic purposes. Presurgical computed tomographic scans showed resolution of perinephric blood and fluid with tumor shrinkage. Histopathologic analyses showed all tumors were resected with negative margins, and there was no intraoperative tumor spillage. All patients received 1050 to 1080 cGy of flank radiation postoperatively. All patients are currently alive at follow-up without evidence of local recurrence or distant disease.

**Conclusions:** Neoadjuvant chemotherapy allowed for complete resection and avoidance of total abdominal radiation in 3 patients with ruptured Wilms' tumor and hematoma within the retroperitoneum. These data support the use of initial chemotherapy in children with retroperitoneal rupture and hematoma of Wilms' tumor at diagnosis.

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Current Wilms' tumor treatment is dependent on stage and histologic type with a recommendation for initial nephrectomy and lymph node sampling followed by chemotherapy [1-3]. There are 3 instances where preoperative treatment with chemotherapy is recommended: bilateral

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renal involvement, Wilms' tumor in a solitary kidney, and large tumors that will require resection of adjacent organs to remove and/or vena caval extension above the hepatic veins [4-6]. However, the management of a Wilms' tumor with a contained retroperitoneal rupture at presentation has not been specifically addressed. This is relevant because of the strong possibility of peritoneal contamination owing to fluid leak during resection. This would then require total abdominal radiation rather than localized flank radiation. We describe our experience with neoadjuvant chemotherapy used in 3 consecutive patients with Wilms' tumor presenting with contained retroperitoneal rupture.

### 1. Methods

This is a retrospective review of 3 patients with Wilms' tumor who presented with a contained retroperitoneal rupture seen on preoperative imaging studies. Contained retroperitoneal rupture was identified by clear areas of fluid collection within Gerota fascia. Approval from the institutional review board at Memorial Sloan Kettering Cancer Center (MSKCC; New York, NY) was obtained for the initiation of the study. All Health Insurance Portability and Accountability Act (HIPAA) guidelines were followed to ensure patient privacy. The patients' inpatient and outpatient charts were reviewed along with all available imaging studies, operative reports, and pathologic reports. The workup, preoperative treatment, and outcomes are discussed below.

### 2. Results

Two male patients, both with an age of 2.9 years, and 1 female patient aged 9.3 years were identified. Patient data, treatment, and follow-up data are listed in Table 1. Diagnoses





**Fig. 1** A, Computed tomographic scan of patient 1 before treatment. B, Computed tomographic scan of patient 1 after neoadjuvant chemotherapy.

were suspected after imaging studies were performed at outside institutions. Patients were then referred to MSKCC for further testing and treatment evaluations. All 3 patients

|             | Sex/age<br>at diagnosis | Stage at presentation/side                | Neoadjuvant chemotherapy  | Interval<br>till<br>surgery | Histopathologic subtype/margins | Follow-up/<br>disease<br>status |
|-------------|-------------------------|---|---|-----------------------------|---------------------------------|---------------------------------|
| Patient 1 a | Male/2.9 y              | Stage IV/left<br>+pulmonary<br>metastasis | Vincristine 0.05 mg/(kg·wk) × 5;<br>dactinomycin<br>45 µg/kg × 1; doxorubicin<br>1.53 mg/kg × 1 | 44 d                        | Favorable/negative              | 66 mo/NED                       |
| Patient 2   | Male/2.9 y              | Stage III/left                            | Vincristine 0.05 mg/(kg·wk) × 5;<br>dactinomycin<br>45 µg/kg × 1; doxorubicin<br>1.53 mg/kg × 1 | 30 d                        | Favorable/negative              | 38 mo/NED                       |
| Patient 3   | Female/9.3 y            | Stage III/left                            | Vincristine 1.5 mg/(m <sup>2</sup> ·wk) × 5;<br>dactinomycin<br>45 $\mu$ g/kg × 3               | 42 d                        | Unfavorable/negative            | 12 mo/NED                       |

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