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Traumatic Rupture of Wilm's Tumour in Children, Case Report of 3.5 Years Girl

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Abstract

Background: We report a case of a 3.5-year-old girl presenting with hemorrhagic shock and gross heamaturia. Emergency evaluation via ultrasound and CT revealed a large perinephric hematoma and a non-secretory left kidney. Due to hemodynamic instability, an emergent laparotomy and radical nephrectomy was performed. **Results:** Histopathology confirmed a Stage III mixed-type Wilms tumor. The upgrade to Stage III was necessitated by intraoperative tumor rupture and local spillage. Despite a postoperative complication of intestinal obstruction, the patient successfully completed the SIOP-based chemotherapy and radiotherapy protocol and remains recurrence-free at one-year follow-up. **Conclusion:** Traumatic rupture of a Wilms tumor is a surgical emergency. Immediate intervention and upstaging to Stage III are critical for survival and long-term oncological control.

Keywords

Wilms Tumor, Blunt Renal Trauma, Hemorrhagic Shock, Radical Nephrectomy, SIOP Staging

1. Introduction

Wilms tumor, or nephroblastoma, stands as the most prevalent primary renal malignancy in the pediatric population, accounting for approximately 6% to 7% of all childhood cancers [1]. Historically, the prognosis for Wilms tumor has been one of the greatest success stories in modern oncology, with overall survival rates now exceeding 90% due to highly refined multi-modal treatment protocols [1]. Most cases

present as an incidental, painless abdominal mass discovered by a parent or during a routine pediatric examination. However, a small but clinically challenging subset of patients presents with acute abdominal crisis due to tumor rupture.

The occurrence of traumatic rupture in an occult Wilms tumor is a rare clinical entity that places the patient at immediate life-threatening risk. From a physiological standpoint, Wilms tumors are often hypervascular and possess a fragile, thin capsule [2]. This makes the tumor tissue significantly more susceptible to parenchymal disruption than the elastic, healthy renal tissue found in children. Consequently, even low-energy blunt abdominal trauma—such as a fall from a moderate height or a minor sports injury—can lead to "disproportionate injury," where the degree of renal hemorrhage and capsular breach is far greater than what would be expected from the mechanism of injury alone.

When a rupture occurs, it initiates a complex "dual-emergency" scenario:

1) Hemorrhagic Shock: The rapid extravasation of blood into the perinephric space or the peritoneal cavity can lead to sudden hemodynamic collapse, requiring urgent surgical hemostasis.

2) Oncological Seeding: The breach of the tumor capsule allows for the dissemination of malignant blastemal cells. This "spill" transforms a potentially localized Stage I or II tumor into a Stage III disease, fundamentally altering the patient's long-term treatment trajectory [3].

The management of such cases remains a point of divergence between the world's two primary pediatric oncology groups. The Children's Oncology Group (COG) in North America generally advocates for primary nephrectomy, whereas the International Society of Paediatric Oncology (SIOP) in Europe and elsewhere typically favors preoperative chemotherapy [3]. In the setting of an acute traumatic rupture, the SIOP "chemotherapy-first" paradigm must be abandoned in favor of emergency surgery.

This case report details the presentation of a 3.5-year-old girl where the intersection of trauma and oncology demanded immediate surgical intervention. We highlight the critical importance of recognizing the staging implications of a traumatic spill and the necessity of a multidisciplinary approach—combining emergency urological surgery with intensified adjuvant chemoradiotherapy—to ensure a favorable oncological outcome despite a catastrophic initial presentation.

2. Case Presentation

2.1. Emergency Presentation and Resuscitation

A 3.5-year-old girl was brought to the emergency department following a fall from a height. On arrival, she was in a state of compensated hemorrhagic shock: pale, tachycardic with a weak thready pulse, and a blood pressure of 60/40 mmHg. Initial laboratory investigations revealed a hemoglobin level of 9 g/dL, a white blood cell count of 12×10^3 cells /mm^3, and a serum creatinine of 0.6 mg.

Physical examination revealed a palpable, tender mass in the left upper quadrant and localized abdominal distention. Gross hematuria was evident upon uri-

nary catheterization, suggesting that the traumatic forces had disrupted both the renal parenchyma and the collecting system.

Upon arrival, the patient's clinical status dictated immediate activation of the massive transfusion protocol. Given her profound hypotension (60/40 mmHg) and tachycardia (165 bpm), resuscitation began with an initial bolus of 20 mL/kg of warmed isotonic crystalloids.

Due to the lack of hemodynamic response and a drop in hemoglobin to 9 g/dL, the team transitioned to blood products. The patient received:

- **Packed Red Blood Cells (PRBCs):** 10 mL/kg of O-negative blood, followed by cross-matched units.
- **Fresh Frozen Plasma (FFP):** Administered in a 1:1 ratio with PRBCs to address potential trauma-induced coagulopathy.
- **Tranexamic Acid (TXA):** A loading dose was administered to stabilize clot formation.

Justification for Immediate Nephrectomy

While blunt renal trauma in children is often managed nonoperatively (NOM), several critical factors made conservative management inappropriate in this case:

1) Hemodynamic Instability: The patient remained persistently hypotensive despite aggressive fluid and blood resuscitation. In pediatric trauma, "refractory shock" is an absolute indication for surgical exploration.

2) CT Grade and Extravasation: Contrast-enhanced CT demonstrated a Grade V renal injury equivalent, characterized by complete anatomical distortion and active contrast extravasation ("blush"). This indicated an arterial-grade bleed that was unlikely to tamponade spontaneously.

3) Presence of an Underlying Mass: The identification of a suspected Wilms tumor altered the management priority. Unlike a standard kidney injury where the goal is "renal salvage," a ruptured malignant mass carries the risk of ongoing hemorrhage from abnormal neoplastic vasculature and the threat of extensive peritoneal seeding.

4) Inefficacy of Angioembolization: Given the patient's unstable state, transferring her to an interventional radiology suite for Transcatheter Arterial Embolization (TAE) was deemed too high-risk. Surgical control was the fastest route to definitive hemostasis.

So due to the patient's persistent hemodynamic instability and the high clinical suspicion of active hemorrhage, non-operative management was deemed inappropriate.

2.2. Diagnostic Imaging

A Focused Assessment with Sonography for Trauma (FAST) was performed bedside, which identified a large heterogeneous mass in the left renal bed and free fluid. Subsequent contrast-enhanced CT (CTU) confirmed a large left perinephric hematoma with active extravasation (**Figure 1(a)**). The left kidney was non-secretory and its architecture was totally distorted by a suspected underlying tumor.

Given the patient's refractory hypotension and the CT evidence of active hemorrhage from a ruptured renal mass, the medical team opted for urgent surgical intervention.

2.3. Surgical Intervention

The patient was taken for emergent exploration via a midline trans-peritoneal incision. Upon entering the abdomen, a large retroperitoneal hematoma was identified. The posterior peritoneum was opened, and the mass was addressed within Gerota's fascia. Intraoperative findings revealed a macroscopic rupture of the renal capsule with tumor involvement and significant local extravasation of blood (**Figure 1(b)-(c)**). A left radical nephrectomy was performed, successfully removing the kidney and the ruptured tumor *en bloc* within Gerota's fascia to ensure oncological safety. The procedure lasted 2 hours and 40 minutes, and the patient was stabilized postoperatively.

2.4. Pathology and Final Diagnosis

Gross and microscopic examination of the nephrectomy specimen confirmed a Wilms tumor (nephroblastoma) of mixed type with favorable histology and no evidence of focal or diffuse anaplasia. The tumor exhibited invasion of the renal sinus fat and positive lymphovascular invasion, which are markers of intermediate-risk biology. All surgical margins—including the hilar vascular structures and the ureteric margin—were free of neoplastic cells. Four hilar lymph nodes were identified and were negative for malignancy (pN0) (**Figure 2**). However, consistent with the intraoperative findings and preoperative imaging, there was a definitive rupture of the renal capsule resulting in a localized tumor spill into the retroperitoneal space.

Protocol Linkage and Treatment Rationale

Under the SIOP Umbrella Protocol, the combination of mixed-type histology and the absence of anaplasia classifies this as an Intermediate-Risk tumor. While the clear margins and negative lymph nodes would typically suggest Stage II, the traumatic capsular rupture (spill) necessitates an upstage to Stage III. This classification directly dictated the intensification of the adjuvant regimen:

- **Chemotherapy:** The patient was transitioned from a standard two-drug regimen to a three-drug "Regimen AF" (Vincristine, Actinomycin-D, and Doxorubicin) to address the high risk of recurrence associated with tumor seeding.
- **Radiotherapy:** Due to the local spill, the patient received flank irradiation (10.8 - 15 Gy). This is a mandatory component of Stage III management to sterilize the retroperitoneal space and prevent local abdominal relapse, a complication that is significantly more frequent in cases of preoperative rupture managed without radiation.

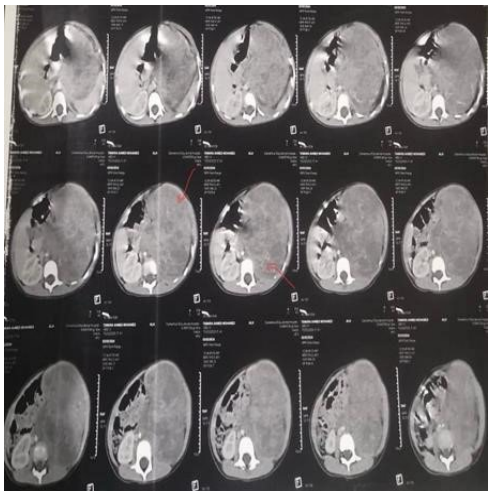
2.5. Postoperative Course and Follow-Up

Following the emergency radical nephrectomy, the patient was referred to the pediatric oncology center. Despite the absence of nodal involvement, the child was

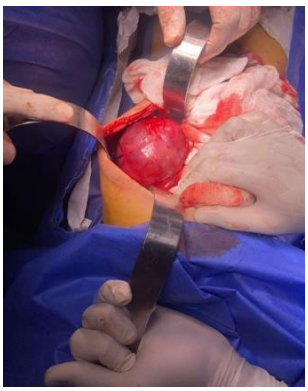
managed under the SIOP Umbrella protocol for Stage III intermediate-risk disease. This decision was dictated by the traumatic spill, requiring a three-drug chemotherapy regimen (Vincristine, Actinomycin-D, and Doxorubicin) and localized radiotherapy to the tumor bed to minimize the risk of local abdominal recurrence). The course was briefly interrupted by an intestinal obstruction requiring surgical adhesiolysis; findings confirmed non-neoplastic adhesions. Following recovery, she completed her oncology regimen. At the 12-month follow-up, there is no clinical or radiological evidence of recurrence (Table 1).

Table 1. Summary of results.

Feature	Finding	Staging impact
Histology	Mixed type (blastemal, stromal, epithelial)	Intermediate risk
Lymph nodes	0/4 (Negative for malignancy)	Would be stage II
Margins	Hilar, vascual, and ureteric (Free)	Would be stage II
Trauma status	Preoperative capsular rupture	Upstaged to stage III
Final stage	SIOP stage III	Final clinical/pathological stage



(a)



(b)

(c)

Figure 1. (a) (lt): axial cuts of CTU showing large Lt perinephric hematoma with extravasation; (b) (Rt): intraoperative view; (c): Lt nephrectomy showing ruptured tumor.

Nature of Specimen: **Left nephrectomy**

Gross Pathology:

Received is a left nephrectomy specimen 13x10.5X5.5cm.showing a large mass 8X6.5 cm The mass is homogenous tan coloured with few hemorrhagic and necrotic areas. The hilar structures are grossly free.

Histopathology Report:

Microscopic examination reveals a cellular triphasic malignant neoplastic growth predominantly formed of primitive (blastema cells) 50% of the tumor tissue formed of sheets and cords of cells having large dark nuclei with basophilic cytoplasm and mitosis .Blastema cells are separated by fibrotic stroma (10%) of tumor with primitive glomeruli and tubules (40%) of the tumor. Focal necrosis and many karyorrhectic debris are noted. The renal hilum is free as well as the peri-nephric fat. Tumor invades the renal sinus. No nephrogenic rests detected and no tumor anaplasia. Margins are free.

Conclusion:

- WILM'S TUMOUR [NEPHROBLASTOMA] (MIXED), INTERMEDIATE RISK (SIOP), WITH FAVOURABLE HISTOLOGY (NO ANAPLASIA).
- TUMOR INVADES THE SINUS FAT.
- POSITIVE VASCULAR INVASION.
- FREE PERI-NEPHRIC FAT, AND URETERIC MARGIN .
- NO NEPHROGENIC RESTS OR ANAPLASIA.
- REACTIVE FOUR HILAR LYMPH NODES (NEGATIVE FOR MALIGNANCY).
- STAGE II ACCORDING TO SIOP (INTERNATIONAL SOCIETY OF PEDIATRIC ONCOLOGY).
- FREE HILAR MARGIN.

Figure 2. Histopathology report of the case.

3. Discussion

The Management Dilemma: Surgery vs. Nonoperative Management (NOM).

In the modern management of pediatric blunt renal trauma, the standard of care—as outlined by the American Association for the Surgery of Trauma (AAST)—is nonoperative management (NOM) for the majority of high-grade (Grades III - V)

injuries, provided the patient remains hemodynamically stable. However, the presence of an occult malignancy fundamentally alters this algorithm.

In this case, several factors rendered NOM inappropriate. First, the patient exhibited refractory hemorrhagic shock, an absolute indication for surgical exploration. Second, the pathophysiology of a ruptured Wilms tumor differs significantly from that of a healthy kidney. While a normal kidney may benefit from the "tamponade effect" of an intact Gerota's fascia, Wilms tumor tissue is exceptionally friable and hypervascular. This prevents normal physiological hemostasis [4] and clot stabilization, making emergent radical nephrectomy the only viable life-saving intervention to achieve definitive surgical hemostasis.

The Impact of Trauma on Oncological Staging

Accurate staging following trauma is the most critical factor in determining long-term survival [5]. A primary challenge in this case was reconciling the localized nature of the tumor with the catastrophic clinical presentation. While the histopathological absence of nodal involvement or distant metastasis initially suggested a lower stage, the traumatic rupture of the renal capsule became the defining feature for staging [6].

Under the SIOP Umbrella protocol, any preoperative or intraoperative rupture—whether spontaneous or traumatic—is categorized as a local tumor spill [7]. This breach of capsule integrity carries an inherent risk of seeding malignant blastemal cells into the retroperitoneal or peritoneal space. Consequently, the patient was upstaged from Stage II to Stage III. This transition is vital for oncological safety, as Stage III protocols mandate a more aggressive therapeutic approach to mitigate the high risk of local abdominal recurrence.

Clinical Indicators and Diagnostic Correlation

The presence of gross hematuria in this patient served as an essential clinical marker, indicating that the traumatic force was sufficient to disrupt the deep renal parenchyma [8] and extend into the renal collecting system or pelvis. Intraoperatively, this correlated with a macroscopic rupture at the lower pole of the mass.

The decision to proceed with an immediate midline transperitoneal radical nephrectomy allowed for rapid vascular control and complete extirpation of the ruptured mass. This approach addresses the dual crisis of hemodynamic instability and potential oncological dissemination.

Adjuvant Therapy Rationale

The classification of this case as Stage III Intermediate Risk directly dictated the postoperative regimen. To treat the "spill" area, the patient required:

- **Intensified Chemotherapy:** A three-drug regimen (Vincristine, Actinomycin-D, and Doxorubicin) rather than the standard two-drug protocol used for Stage II.
- **Localized Radiotherapy:** Flank irradiation (10.8 - 15 Gy) to "sterilize" the retroperitoneal space [9].

The successful one-year follow-up, despite the initial rupture and a postoperative complication of adhesive intestinal obstruction, validates this aggressive mul-

tidisciplinary management strategy. It underscores that while trauma complicates the initial presentation, adherence to standardized upstaging protocols remains the cornerstone of successful outcomes in ruptured pediatric renal tumors.

4. Conclusion

Traumatic rupture of a Wilms tumor should be suspected in any child presenting with shock and a renal mass following minor trauma. Immediate surgical intervention is required for unstable patients [9]. Pathological staging must account for the rupture, ensuring patients receive the necessary Stage III adjuvant therapy to mitigate the risk of local recurrence.

Declarations

Consent for Publication: Written informed consent was obtained from the patient's parents for the publication of this case report and any accompanying images. **Ethics Approval:** This case report was conducted in accordance with the ethical standards of the Damanhour Medical National Institute.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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