

# A Case of Wilms' Tumor in an Adult

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## Introduction

Nephroblastoma, or Wilms' tumor, is the most common primary renal tumor in children, and the fourth most common malignancy in children. Almost all cases occur before the age of ten, but there are rare cases found in an adult. Proper sectioning by the pathologists' assistant is needed to confirm the stage of such cases, which directly relates to the correct treatment and prognosis for the patient. This poster presents a woman in her 40's with a 9.0 x 6.0 x 5.4 cm stage 3 Wilms' tumor in her right kidney.

## Clinical History

A woman in her 40's presented to her physician with complaints of painless hematuria and a 20-pound weight loss over the course of the year. She underwent a magnetic resonance imaging (MRI) scan that revealed a multilobulated partially cystic lesion in the superior right kidney. A renal biopsy was obtained, and histological analysis and immunohistochemical stains showed that the preliminary diagnosis was adult Wilms' tumor. The patient underwent six weeks of chemotherapy, after which a right nephrectomy was performed (**Fig. 1**).

## Gross Description

Container label: Name, medical record number, and "Right kidney Wilms tumor".

Received: Fresh and placed in formalin

Specimen: Right nephrectomy with attached adipose tissue

Size: 416.0 gm, 14.0 x 6.5 x 5.5 cm

- Kidney: 13.8 x 6.4 x 5.3 cm

- Ureter: 0.4 cm in length, 0.2 cm in diameter

- Adrenal gland: 3.0 x 1.1 x 0.4 cm

Orientation: None provided.

Mass

- Location: Upper pole kidney directly underlying adrenal, with extension into renal sinus.

- Size: 9.0 x 6.0 x 5.4 cm, involving the renal pelvis

- Description: There are two discrete components to the mass. Component one comprises approximately 60-70% of the total tumor burden, and is a yellow-golden, lobulated, rubbery mass with multiple fibrous septae intervening the lobules. Within this component, there is focal hemorrhage. The second component comprises approximately 20-30% of the total tumor burden and is well-circumscribed, ovoid, tan-pink to focally white-tan, firm to rubbery in texture, and fleshy in appearance. Fresh tissue from both components is frozen for future diagnostic purposes.

- Gerota's fascia: Mass directly abuts Gerota's fascia, but does not penetrate.

- Renal artery: Grossly negative.

- Renal vein: Grossly negative, tumor thrombus absent.

- Ureter: Grossly negative.

- Sinus adipose tissue: Grossly involved by mass.

- Adrenal: Grossly negative.

Gross photographs: Yes, including dissection map. (**Fig 2**)

Cassette Summary

A1-19: Sections of tumor (see dissection map).

- Note: A12 and A14 have no ink as the fat/capsule was disrupted during sectioning.

A20-22: Uninvolved renal parenchyma (PAS ordered)

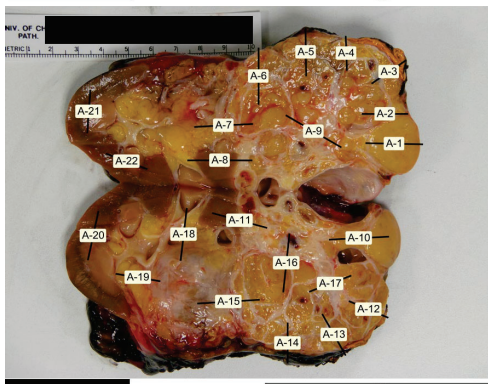
A23: Ureteral margin

A24: Vascular margins

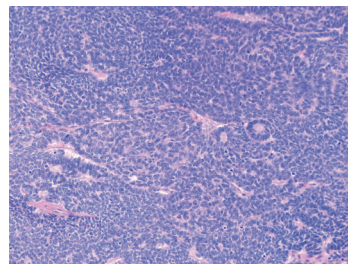
A25: Renal sinus adipose tissue margin, shaved and submitted en face



**Figure 1:**  
Specimen received in the anatomic pathology lab after the patient's right nephrectomy procedure



**Figure 2:** Image of the patient's mass in the bisected right kidney, as well as an overlain diagram representing sections taken by the pathologists' assistant



**Figure 3:**  
Photomicrograph of the patient's mass showing the triphasic appearance of Wilms' tumor: blastemal, epithelial, and stromal components

## Final Diagnosis

Stage III adult Wilms' tumor with favorable histology, focally necrotic and extensively collagenized. Tumor extended into the renal sinus and perinephric adipose tissue; however, no tumor was present at the margins of resection (soft tissue, ureter, renal artery, renal vein), adrenal gland, and all 21 lymph nodes. Immunohistochemical stains showed that the tumor was positive for Cam5.2, PAX8, WT-1 and focally for CK7 and GATA3. The tumor was negative for OCT3/4, ER, PR, TTF-1 and CDX2.

## Discussion

Wilms' tumor has a peak incidence between the ages of two and five years. Adult Wilms' tumor is rare, occurring at a rate of one per five million adults per year. Symptoms differ between pediatric and adult patients; adults typically present with sudden weight loss, hematuria, and/or flank pain, and children typically present with palpable abdominal masses, high blood pressure, and/or nausea/vomiting. It can be difficult to diagnose adult Wilms' tumor due to the rarity of the disease, leading to a poorer prognosis compared to pediatric patients. The staging and treatment for Wilms' tumor is the same for children and adults, since the adult form is so rare. The staging system that is used was developed by the Children's Oncology Group and relies upon extent of invasion of the tumor. Because of this, the pathologists' assistant role is vital. For this patient, sections A1-A6, A12-A14, and the ureteral and vascular margins were key in determining if there was microscopic invasion out of the kidney. The patient was diagnosed as a stage III because of her biopsy before the nephrectomy, due to the risk of local spillage, however all the lymph nodes being negative ensured she was not a stage IV. Histologically, children and adult Wilms' tumors are the same. They are composed of three phases in varying percentages: blastemal, stromal, and epithelial components (**Fig. 3**). Adult Wilms' tumors that are predominantly blastemal tend to be more aggressive than the other two, and luckily this patient's tumor had stromal predominance. Another important feature is the degree of anaplasia present. Focal anaplasia, indicated as favorable histology, represents a lower risk tumor that is more susceptible to treatment. Diffuse anaplasia, or unfavorable histology, represents a much higher risk and requires more aggressive treatment. This patient's tumor had favorable histology. Despite the histological similarities between adults and pediatric Wilms' tumors, there is some evidence that the pathogenesis of Wilms' tumor is different in adults compared to children, due to different alterations in genes. Interestingly, this could indicate that adult Wilms' tumors are a completely distinct specimen from pediatric Wilms' tumors.

## References

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