

Botryoid Wilms Tumor: A Rare Diagnosis With an Atypical Presentation



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Wilms tumor commonly presents as an asymptomatic abdominal mass. In some cases, it can be accompanied with hypertension, constitutional symptoms, and hematuria when involving the collecting system. Below, we review the case of a child diagnosed with botryoid Wilms tumor involving the upper calyces and renal pelvis in which the presenting symptom was a concern for a foreign body in her left ear, and the only abnormality during initial history and physical examination was stage II hypertension. *UROLOGY* 126: 192–194, 2019. © 2019 Elsevier Inc.

Wilms tumor is the most common malignant primary renal tumor of childhood, affecting approximately 1 in 10,000 children per year.¹ An incidentally discovered asymptomatic abdominal mass is the most common initial sign, occurring in over 90% of cases.

Associated symptoms can include hypertension (25%), hematuria (20%), and constitutional symptoms (10%).² Botryoid tumors, an uncommon subclass of Wilms tumors, are intrapelvic polyploid masses that either extend from the renal parenchyma or are primarily pelvis based tumors. It is exceptionally rare for an isolated finding of hypertension to be the impetus for a new diagnosis of botryoid Wilms tumor.

CASE PRESENTATION

An otherwise healthy 3-year-old African-American female presented to our emergency department with concern for a foreign body in her left ear. During triage she had a negative otoscopic exam but was noted to be hypertensive with systolic and diastolic blood pressures between 153 and 174, and 89 and 108, respectively. She was afebrile with no palpable abdominal mass or costovertebral angle tenderness. Her creatinine was 0.4 mg/dL and urinalysis revealed 22 white blood cells but was otherwise unremarkable. Aldosterone, metanephrine, and renin levels were normal and she was started on amlodipine.

A renal ultrasound (Fig. 1) demonstrated mildly echogenic avascular-appearing material filling the left upper pole collecting system accompanied with lower calyceal

dilation consistent with an infectious vs neoplastic process. Therefore, an abdominal CT was performed (Fig. 2) demonstrating left-sided hydronephrosis with poorly perfused renal parenchyma and heterogenous debris within the upper calyces and renal pelvis, suggesting infectious etiology over a neoplastic process.

She then underwent cystoscopy, left renal pelvis washings and left retrograde pyelogram (Fig. 3a) which was remarkable for a filling defect in the upper pole extending towards the renal pelvis and creating middle and lower calyceal hydronephrosis. Ureteroscopy was attempted in order to visually confirm a solid component but was unsuccessful, as the ureteral orifice could not be safely traversed.

Ultimately, her urine and renal pelvis cultures returned negative. However, her pelvic washing cytology demonstrated numerous small dark cells and spindle-shaped cells. These cells stained positive with immunoperoxidase for Wilms tumor while the spindle cell areas were consistent with desmin and smooth muscle actin. Although non-diagnostic, Wilms tumor was suggested. We elected to proceed with left nephrectomy, partial ureterectomy, and hilar lymph node dissection. Pathology (Fig. 3b) returned as a 5.6 cm triphasic Wilms tumor with favorable histology, negative margins, and negative lymph nodes, consistent with local stage I disease. Postoperatively the patient had an uncomplicated course.

She subsequently enrolled in the NWTS-5 treatment protocol at St. Jude Children's Research Hospital for 18 weeks of vincristine and actinomycin D. She recently completed chemotherapy, has been normotensive without amlodipine, and has no signs of recurrence or metastatic disease 6 months postoperatively.

DISCUSSION

Entirely intrapelvic botryoid Wilms tumors are uncommonly reported, and this case is exceedingly atypical for several reasons. First, it is the only reported botryoid Wilms tumor to present without gross or microscopic

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Figure 1. Renal US demonstrating mildly echogenic debris filling the left upper pole with layering debris and hydronephrosis of the lower pole.



Figure 2. CT with contrast demonstrating poor perfusion of the left kidney and mildly hypoattenuating material filling the left upper pole renal collecting system and extending into the renal pelvis. (Color version available online.)

hematuria. Second, it is the only reported botryoid Wilms tumor to present not only with isolated hypertension but also a normal renin level.

With timely and appropriate work up, botryoid Wilms tumors themselves do not portend a worse prognosis.³ However, the variable presentation of Wilms tumor is important to fully comprehend as improper work up and testing can alter the patient's staging and ultimate prognosis. When imaging demonstrates any pathology that could be a solid renal mass, whether parenchymal or intrarenal, a high suspicion for Wilms tumor is necessary. Retrograde pyelogram and ureteroscopy should play an essential diagnostic role. These interventions not only allow for visual confirmation of a solid mass but can also delineate the distal extent of the tumor for surgical planning and complete

excision. Avoidance of leaving residual tumor, either by ureteroscopic or percutaneous biopsy, or by transecting the ureter proximal to the intraluminal tumor margin, is critical as this immediately and unnecessarily upstages the patient's disease and escalates their treatment plan.

Due to the location of botryoid tumors within the collecting system, it is unsurprising that all cases in the literature reported microscopic or gross hematuria on initial presentation. Although our patient's mass clearly encompasses the collecting system (Fig. 3), it is unique that she had no history of microscopic or gross hematuria which had not been previously reported. Many Wilms tumors

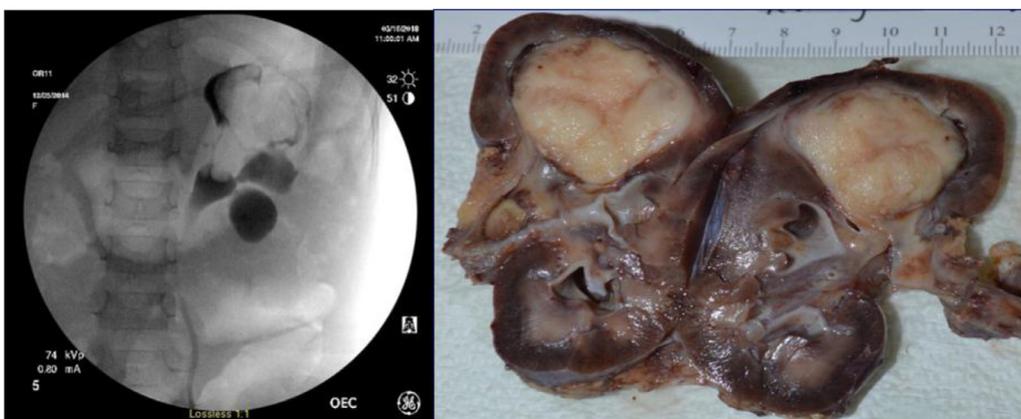


Figure 3. (A) Left retrograde pyelogram demonstrates a large upper pole filling defect extending into the renal pelvis with near complete separation of the defect from the parenchyma by contrast. (B) 5.6 × 4.0 × 3.2 cm circumscribed white-tan firm tumor focally protruding into the dilated upper major calyx and pelvis. No vascular, capsular, or soft tissue invasion identified. (Color version available online.)

have hemorrhage and necrosis noted on gross surgical pathology, however, this was absent in our specimen. Perhaps necrosis and hemorrhage are late developments in the natural course of Wilms tumor progression and our patient presented prior to this stage of her disease process. Conceivably, these histopathological findings, more so than collecting system involvement, determine the likelihood of hematuria.

Her newly discovered hypertension allowed us to discover her botryoid Wilms tumor. As previously reported, hypertension is associated with Wilms tumor in 25% to 55% of cases.⁴ The majority of patients with Wilms tumor and hypertension have increased blood pressure related to renin elevation.^{5,6} In our patient, the renal parenchyma appeared poorly perfused on CT imaging. Typically, renal hypoperfusion would activate the renin—angiotensin—aldosterone mechanism to attempt to return renal blood flow back to equilibrium. However, in our patient this was not the case.

CONCLUSION

This case of a botryoid Wilms tumor that presented with a solitary abnormality of hypertension has never been

reported in the literature. As with many single patient case reports, we present more questions than answers. Given the rarity of botryoid Wilms tumors, further research into the pathophysiology of these masses will help to determine their etiology and mechanisms in which they present. Although an isolated discovery of hypertension is rare in these children, we suggest considering Wilms tumor as a potential cause of hypertension in children with abnormal renal imaging.

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