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Botryoid Wilms tumor and renal cell carcinoma: A case-based approach

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Abstract

Pediatric renal tumors are uncommon, with Wilms tumor (WT) being the most frequently diagnosed. Among its subtypes, Botryoid Wilms tumor (BWT) is a rare variant distinguished by its multilobular architecture. Although more typical in adults, renal cell carcinoma (RCC) may also develop in children, most often originating in the renal pelvis or calyceal system. Differentiating botryoid Wilms tumor (BWT) from renal cell carcinoma (RCC) in pediatric patients is particularly challenging due to overlapping clinical and radiologic features, including renal masses, hematuria, and hydronephrosis.

We present two pediatric cases that highlight the complexities in distinguishing these tumors. The first, a 3-year-old child, presented with hematuria and an abdominal mass. Imaging revealed a lobulated renal tumor with both cystic and solid areas, consistent with BWT, which was later confirmed through histopathology following nephrectomy. The child remained recurrence-free after two years of follow-up. The second case involved an 11-year-old presenting with flank pain and gross hematuria. Imaging raised suspicion for Wilms tumor; however, histopathological examination confirmed RCC. The patient underwent partial nephrectomy and adjuvant therapy and remained stable at 18-month follow-up.

These cases emphasize the diagnostic overlap between BWT and RCC and the importance of histological confirmation when imaging is inconclusive. Early, accurate identification is essential, as management strategies and long-term outcomes differ considerably. While WT generally responds well to multimodal therapy, RCC may necessitate long-term monitoring due to a higher risk of metastasis. Advancements in imaging techniques and molecular profiling may enhance diagnostic accuracy in future cases.

Introduction

Pediatric kidney tumors, although rare, are of significant concern due to their clinical presentation and the need for accurate diagnosis. Among these tumors, Wilms tumor (WT) is the most common, particularly in children under the age of 5. Botryoid Wilms tumor (BWT) is a rare variant of WT, characterized by its multilobular, grape-like appearance. Renal cell carcinoma (RCC), although more common in adults, can also occur in children, typically originating from the renal pelvis or calyces. Both BWT and RCC can present similarly with renal mass formation and hydronephrosis. However, these two rare tumors have distinct radiological and clinical features that help differentiate them. This article reviews the radiological findings of two pediatric cases—one of botryoid Wilms tumor and the other of renal cell carcinoma—and compares their imaging characteristics. **Citation:** Yammouri Z, Mouhcine Y, Ouazzani H, Chaouche S, Akammar A, et.al. Botryoid Wilms tumor and renal cell carcinoma: A case-based approach. J Clin Images Med Case Rep. 2025; 6(7): 3660.

Case presentations

Case 1

A 3-year-old Caucasian female presented with an abdominal mass and mild abdominal distension. Ultrasound and contrastenhanced CT revealed a large, heterogeneous right renal mass with a multilobular configuration, extending into the superior and middle calyceal groups as well as the ipsilateral ureter. Surgical exploration and subsequent histopathological analysis confirmed the diagnosis of a botryoid Wilms tumor (BWT). The patient underwent radical nephrectomy, which included resection of the tumor involving the renal pelvis and ureter, dissection of the renal pedicle, and ligation of the ureter. The inferior vena cava was inadvertently involved during surgery but managed appropriately. Postoperatively, the patient received chemotherapy according to standard Wilms tumor protocols [1,2]. At two-year follow-up, the patient remained recurrence-free, with no evidence of disease progression (Figures 1, 2 and 3).

Case 2

An 11-year-old Caucasian female presented with flank pain and gross hematuria. Ultrasound and contrast-enhanced CT revealed a large, irregular renal pelvic mass with features suggestive of malignancy, including areas of necrosis, hemorrhage, and increased vascularity. The mass demonstrated apparent invasion of the renal capsule and perinephric fat, raising suspicion for renal cell carcinoma (RCC). A radiologically guided biopsy was performed, and histopathological analysis confirmed clear cell RCC originating from the renal pelvis. The patient underwent a partial nephrectomy, followed by adjuvant chemotherapy and radiotherapy to address any potential residual disease [3,4]. At



Figures 1, 2 and 3: Axial (1), coronal (2) and sagittal (3) scans showing large, heterogeneous, multilobulated mass originating from the upper pole of the right kidney, extending approximately 3.5 cm into the superior and middle calyces and down into the homolateral ureter, blue arrows. The mass contains mixed solid and cystic components, with no calcifications.



Figure 4, 5, 6 , 7 and 8: CT imaging reveals a large, irregular, and heterogeneous mass with ill-defined borders, capsular breach, and perinephric fat infiltration centered in the renal pelvis of the right kidney, measuring approximately $9.0 \times 6.0 \times 5.5$ cm. The blue arrows mark areas of central necrosis and hemorrhage, reflecting the tumor's aggressive and vascular nature. The orange arrows identify calcific foci within the tumor. The white arrows indicate the tumor's extension into the renal pelvis.

18-month follow-up, the patient remained disease-free, with preserved renal function and no evidence of recurrence (Figures 4, 5 and 6).

Discussion

The occurrence of Botryoid Wilms tumor (BWT) and renal cell carcinoma (RCC) within the renal excretory system in pediatric patients is exceptionally rare and poses a considerable diagnostic dilemma. While these tumors are histologically distinct, their clinical presentations often overlap, with symptoms such as hematuria, abdominal pain, and palpable masses being common to both. These shared manifestations, along with similar radiological features, can complicate early differentiation and delay definitive diagnosis [3,4,5].

Radiologically, both BWT and RCC may appear as heterogeneous renal masses exhibiting both cystic and solid components. Additionally, each can lead to hydronephrosis due to obstruction of the collecting system. However, certain imaging characteristics can assist in distinguishing between the two.

Table 1: Summary of radiological differences [3,4,5,6,7,8,9]

Feature	Botryoid Wilms Tumor (BWT)	Renal Cell Carcinoma (RCC)
Age of occurrence	Younger children (under 5 years)	Older children or adolescents
Ultrasound	Hypoechoic, lobulated, may involve renal pelvis	Solid, irregular mass, often hypoechoic
CT scan	Heterogeneous, lobulated mass, renal pelvis extension, grape-like (botryoid) appearance	Solid, irregular mass, may have necrosis or hemorrhage
MRI T1- weighted	Hypointense	Hypointense
MRI T2- weighted	Hyperintense (due to cystic/hemorrhagic areas)	Heterogeneous signal, may show necrosis
Renal pelvis involvement	Common, causes dilation and extension into calyces	Rare, may cause distortion
Tumor margins	Well-defined, with botryoid pattern	Irregular, infiltrative
Vascular involvement	Rare	May show renal vein or IVC thrombus in advanced stages
Calcifications	Uncommon	More frequent, especially when originating in the renal pelvis or calyces

BWT frequently presents as a well-circumscribed, lobulated mass that extends into the renal pelvis and ureter, giving it a botryoid or "grape-like" morphology. It typically exhibits heterogeneous but less aggressive enhancement and lacks features such as capsular breach or perinephric invasion [5,6]. In contrast, RCC is more likely to appear as an irregular, solid mass with necrosis, hemorrhage, and calcifications. It often demonstrates invasive features, including involvement of the renal capsule and surrounding fat, and shows strong early contrast enhancement due to its vascularity [3,4,7].

Because of these overlapping and sometimes ambiguous imaging findings, histopathological confirmation remains essential for accurate diagnosis. Timely differentiation is crucial, as the management and prognosis for BWT and RCC differ significantly. BWT is typically treated with nephrectomy followed by chemotherapy and has a generally favorable outcome when managed according to pediatric oncology protocols [1,2,8]. On the other hand, RCC requires surgical resection and, depending on disease extent, may also necessitate adjuvant therapy and long-term monitoring due to its potential for metastasis and recurrence [4,9,10].

These two cases underscore the importance of comprehensive imaging assessment and multidisciplinary collaboration in the diagnostic process. As diagnostic imaging continues to advance, particularly with the use of diffusion-weighted MRI, functional imaging, and radiomics, greater accuracy in tumor characterization may be achieved [5,6]. Molecular and genetic profiling could further enhance differentiation, reducing the need for invasive interventions. Ultimately, early and accurate identification of these tumors enables personalized treatment planning, reduces unnecessary therapeutic exposures, and improves prognostic outcomes in pediatric patients [8,10].

Conclusion

While both Botryoid Wilms tumor (BWT) and renal cell carcinoma (RCC) share overlapping radiological features, they can be differentiated by their characteristic imaging findings. BWT typically presents as a multilobular, cystic and solid mass with a more encapsulated growth pattern, while RCC is often more irregular, necrotic, and exhibits local invasion and vascularity. Recognizing these distinct radiological features early in the diagnostic process is crucial for determining the appropriate treatment strategy and improving outcomes in pediatric patients.

Declarations

Ethical statement: Written informed consent was obtained from the parents of both patients for publication of this case and associated images.

Conflict of interest: The authors declare no conflict of interest.

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