



Pediatrics

An unusual radiologic appearance of Wilms tumor

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ARTICLE INFO

Keywords:

Nephroblastoma

Wilms tumor

Nephrectomy

Introduction

Wilms tumor is the most common primary malignant renal tumor in children, accounting for 7% of all childhood cancers. The tumor is equally common in girls and boys, with a mean age at diagnosis of 3.5 years.¹ The most common manifestation of Wilms tumor (90%) is an incidental detection of an asymptomatic abdominal mass, often by a parent or a physician. Abdominal pain, fever, anorexia, weight loss and microscopic hematuria are other common findings at diagnosis, while gross hematuria is rare. On imaging, Wilms tumor characteristically appears as a large (mean diameter, 11 cm), spherical and at least partially intra-renal mass. It may be formed by homogeneous tissue, but oftentimes is heterogeneous, containing areas that represent necrosis, hemorrhage or cystic degeneration. One of the well-known radiological signs of the tumor is the “claw sign” – referring to the sharp angles on either side of the mass, which the surrounding normal parenchyma forms when the mass arises from it. Herein, we present a case report of a toddler with an atypical renal mass mimicking a space-occupying lesion in the upper urinary collecting system, a finding that later proved to be a Wilms tumor.

Case report

A 3.5 years old healthy male infant presented with intermittent abdominal pain without fever, gastrointestinal symptoms or hematuria. Physical examination revealed a prominent mass in the right upper abdominal quadrant.

Abdominal ultrasound (Fig. 1) demonstrated an enlarged,

hydronephrotic right kidney (approximately 11 cm) with a 5 × 7.8 cm upper pole mass. The mass extended into the superior calyces and the renal pelvis.

Computerized tomography (CT) and magnetic resonance imaging (MRI) (Figs. 2 and 3) demonstrated similar findings, namely, a distinct upper pole renal mass measuring 3.9 × 4.8 cm. The mass measured 85 HU, with no enhancement. The collecting system was not well visualized and there were some filling defects in the upper calices and in the renal pelvis. Yet, there was no evidence of extra renal extension, contralateral pathology or distant metastasis.

A diagnostic ureteroscopy combined with retrograde intra-renal surgery was performed, demonstrating edematous urothelium with signs of external pressure causing constriction of the lumen. Selective urine cytology and biopsies from the urothelium of the superior collecting system were obtained. No malignant cells were identified on the cytological studies, while the histological examination was compatible with metanephric adenoma, even though the possibility of Wilms tumor could not be histologically ruled out.

In view of the extent of a potentially malignant tumor which does not enable nephron sparing surgery, an open right radical nephrectomy was performed. The final pathology was consistent with favorable histology Wilms tumor, mainly showing blastemal components and wide areas of necrosis. The tumor was confined to the kidney, with free surgical margins and no involvement of the ureter or the lympho-vascular structures.

Postoperative course was uneventful, and the child was subsequently treated by Vincristine and Dactinomycin (EE4A protocol), successfully accomplished 28 weeks postoperatively, with no

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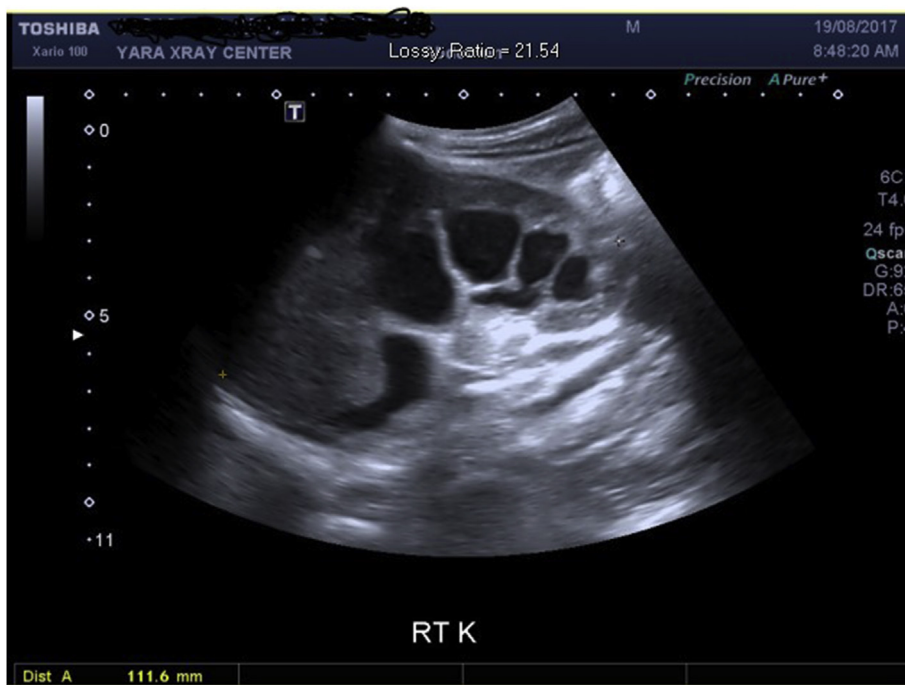


Fig. 1. Abdominal US showing an enlarged right kidney (about 11 cm) with an upper pole mass (5 × 7.8 cm) extending into the superior calyces and renal pelvis and hydronephrosis.



Fig. 2. CT shows a distinct upper pole renal mass with extension in the upper calyces and renal pelvis.

complications. A CT scan performed 6 months postoperatively, showed no evidence of recurrence.

Discussion

Wilms tumor, which accounts for 95% of all kidney cancers in children under the age of 15 years, usually has a typical characteristic radiological appearance. Nevertheless, it is still stated in the textbook that “a precise histological diagnosis cannot be obtained on the pre-operative imaging study”.² Moreover, we should always bear in mind

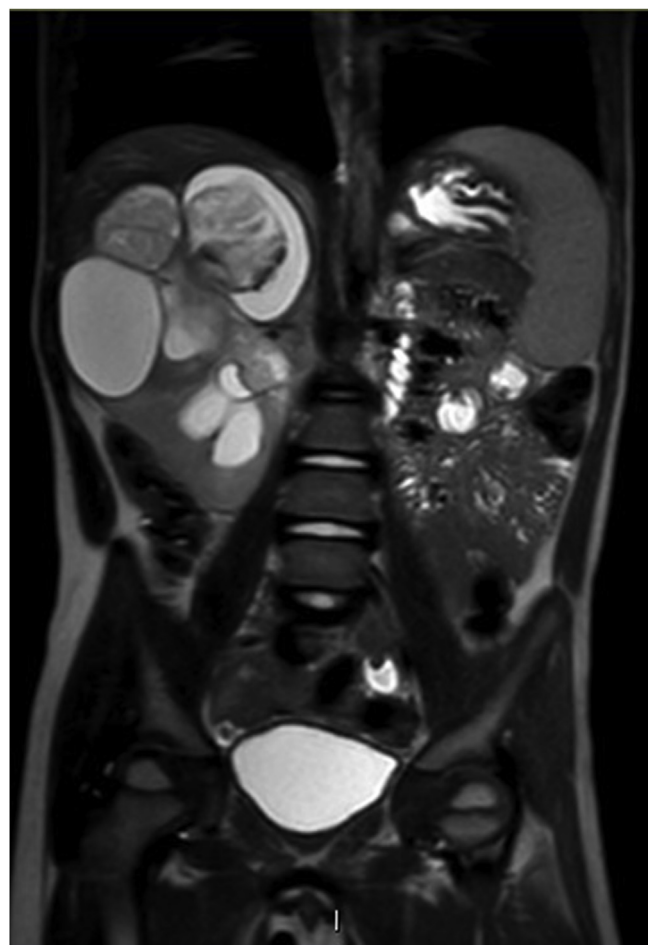


Fig. 3. MRI shows a distinct upper pole renal mass with extension in the upper calyces and renal pelvis.

that “all the solid renal tumors of childhood have some common radiographic features”.² In fact, 5.4% of the suspected Wilms patients treated in the SIOP-9 study with preoperative chemotherapy have been found on nephrectomy to have either other renal malignancies, or even benign renal pathology.² Therefore, medical teams involved in the treatment of these patients should not be strict and dogmatic, as a differential diagnosis does exist including clear cell sarcoma of the kidney, rhabdoid tumor, metanephric adenofibroma, angiomyolipoma and renal cell carcinoma (RCC).

Regarding the present case, which showed a renal parenchymal mass associated with suspected filling defects in the upper urinary collecting system, other options such as infectious process, blood clots due to, ignored traumatic blunt renal injury, as well as upper tract transitional cell carcinoma (TCC) were also considered. Nevertheless, the possibility of an unusual appearance of Wilms tumor has never been neglected.

Literature review reveals few case reports describing cases where strictly looking renal intra-pelvic masses, even extending to the ureter or bladder, which have eventually been found to be Wilms tumor.^{3,4} In such cases, when the tumor expands into the renal collecting system and produces a polypoid shape resembling botryoid sarcoma, it is called “botryoid Wilms tumor”,⁴ and overall 26 such cases have been reported in the literature till 2017, mainly in the right kidney and in males.⁵

Another noteworthy message from the current case management is regarding the approach by which the renal biopsy should be taken. Understandably, whenever a non-typical renal mass is encountered, a biopsy is usually performed, oftentimes through a percutaneous

approach, either ultrasound or CT guided with the disadvantage of upstaging to stage 3 due to associated needle track seeding. In the current case, the mass protruded into the renal calyces and pelvis and therefore a retrograde endoscopic biopsy was performed prior to surgery. Thus, upon complete removal, the tumor could still be defined as Stage 1 disease, sparing the patient adjuvant abdominal radiotherapy and a more aggressive chemotherapy treatment.

Conclusion

Wilms tumor may rarely present as a space-occupying lesion of the upper urinary collecting system. Endoscopic sampling should then be preferably considered whenever technically feasible, as it avoids tumor spread, consequent disease upstaging and resultant either adjuvant or potentially more aggressive chemo/radiotherapy.

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